

2021 EAU Paediatric Urology Guidelines Search Strategy – Congenital Penile Curvature

Database: Embase <1974 to 2020 June 05>, OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present, EBM Reviews - Cochrane Database of Systematic Reviews <2005 to June 03, 2020>

Search Strategy:

-
- 1 ((penile or penis or penil) adj3 (curvature* or curved or bend or bent)).tw,kw. (2824)
 - 2 exp *hypospadias/ (8916)
 - 3 (Hypospadia* or Epispadia*).tw,kw. (16238)
 - 4 exp *epispadias/ (1269)
 - 5 or/1-4 (19544)
 - 6 exp Child/ or exp Infant/ or exp Minors/ or exp Adolescent/ or exp adolescence/ or exp Pediatrics/ or exp newborn/ or exp Puberty/ or exp Schools/ or high school/ or kindergarten/ or middle school/ or nursery school/ or primary school/ (7310904)
 - 7 (baby or babies or child or children or pediatric* or paediatric* or peadiatric* or infan* or infancy or neonat* or newborn* or new born* or kid or kids or adolescen* or preschool or pre-school or toddler*).tw,kw. (4750428)
 - 8 (postmatur* or prematur* or preterm* or perinat* or boy* or girl* or teen* or minors or prepubescen* or prepuberty* or pubescen* or puber*).tw,kw. (1290517)
 - 9 (elementary school* or high school* or highschool* or kindergar* or nursery school* or primary school* or secondary school* or youth* or young or student* or juvenil* or underage* or (under* adj age*) or "under 16" or "under 18").tw,kw. (2068343)
 - 10 or/6-9 (9986491)
 - 11 5 and 10 (12016)
 - 12 (exp animals/ or exp animal/ or exp nonhuman/ or exp animal experiment/ or animal model/ or animal tissue/ or non human/ or (rat or rats or mice or mouse or swine or porcine or murine or sheep or lambs or pigs or piglets or rabbit or rabbits or cat or cats or dog or dogs or cattle or bovine or monkey or monkeys or trout or marmoset\$1 or basic research or cell lines or in vitro or animal model or canine).tw.) not (humans/ or human/ or (human* or patients or subjects).tw.) (10875069)
 - 13 11 not 12 (11885)
 - 14 conference abstract.pt. or Congresses as Topic/ or Conference Review.pt. or "Journal: Conference Abstract".pt. (3939033)
 - 15 case report/ or case reports/ or case report.ti. (4648524)
 - 16 note/ or editorial/ or letter/ or Comment/ or news/ or (note or editorial or letter or Comment or news).pt. (4646379)

17 or/14-16 (12520958)
18 13 not 17 (8212)
19 limit 18 to yr="2016 -current" (1446)
20 limit 19 to english language [Limit not valid in CDSR; records were retained] (1360)
21 remove duplicates from 20 (789)

1.

Ambiguous Genitalia And Disorders of Sexual Differentiation. [Review]

Mehmood KT; Rentea RM.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid

MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

StatPearls Publishing. 2020 01.

[Review]

UI: 32491367

The birth of an infant with ambiguous genitalia generates difficult multiple medical, surgical, ethical, psychosocial, and physical issues for patients and their parents. Phenotypic sex results from the differentiation of internal ducts and external genitalia under the influence of hormones and other additional factors. When discordance occurs among three processes (chromosomal, gonadal, phenotypic sex determination), a DSD is the result. Terminology such as hermaphrodite, pseudo-hermaphrodite, and intersex, are considered to be pejorative and dated. These terms have been replaced by the term disorders of sexual development (DSD) by the consensus statement on management of intersex disorders.[1][2] Disorders of sexual development are defined as congenital conditions characterized by atypical development of chromosomal, gonadal, or anatomic sex.[3] Normal sexual development in utero is dependent upon a precise and coordinated spatiotemporal sequence of various activating and repressing factors.[4] Any deviations from the usual pattern of differentiation can present as DSDs. Two distinct processes occur in normal sexual development. The first of which is sex determination in which the bi-potential gonads are induced to form either the male testes or the female ovaries. Secondly, the newly formed gonads secrete hormones to modulate the formation of internal and external

genitalia.[5] The phenotypic manifestation of DSDs are diverse and can include; bilateral undescended testes, severe hypospadias (scrotal or perineal), clitoromegaly, a fusion of posterior labial folds, female external genitalia with palpable gonad, discordant genitalia and sex chromosomes. The inclusion of disorders in which there is no genital/gonadal discordance like Turner syndrome, Klinefelter syndrome, simple hypospadias remains controversial. Regardless of presentation or severity, individuals require a multidisciplinary approach that is warranted to improve the quality of life and achieve the best possible outcomes.

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Book Title

StatPearls

Version ID

1

Authors Full Name

Mehmood, Khawar T.; Rentea, Rebecca M..

Institution

Mehmood, Khawar T.. Hameed Latif Hospital Rentea, Rebecca M.. Children's Mercy

Publisher

StatPearls Publishing

Year of Publication

2020

2.

Ingrafts in hypospadias surgery: Longer-term outcomes.

Loloi J; Harrington S; Boltz S; Decter RM.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid

MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Journal of pediatric urology. 2020 May 07.

[Journal Article]

UI: 32471708

BACKGROUND: The technique of hypospadias repair with a dorsal inlay graft (ingraft) was initially reserved for boys with an unfavorable glans configuration or in previously failed repairs. Although the ingraft technique has been used for some time, there is scarce literature describing

its longer term outcomes. Additionally, there is minimal data comparing inner preputial skin and buccal mucosal graft outcomes in primary and reoperative surgery.

OBJECTIVE: To describe longer-term outcomes in the use of buccal mucosa and inner preputial skin ingrafts for primary and revisional hypospadias repairs.

STUDY DESIGN: We conducted a single-center retrospective review of our hypospadias repairs employing ingrafts along with a review of the literature.

RESULTS: A total of 47 patients met inclusion criteria. Primary repair was performed in 38 patients, all with unfavorable glans characteristics at a mean age of 16 months and redo repairs in 9 boys at a mean age of 110 months. We used a buccal mucosal graft (BMG) in 19 boys and the remaining 28 were grafted with inner preputial skin. The complication rate was 32% in primary repairs and amongst those, 6 of 28 patients (21%) with skin grafts and 6 of 10 patients (60%) with BMGs suffered a complication. Only BMGs were used in reoperative situations and complications occurred in 2 of 9 cases (22%) of those cases. Overall, 8 patients (42%) with BMG and 6 patients (21%) with preputial skin ingrafts experienced a complication, at an average time of 17 months (range: 0.4-66 months) and 24 months (range: 1.1-113 months), respectively. Surgical correction of the complications resulted in resolution of symptoms in a majority of patients.

DISCUSSION: In our experience, BMGs used as salvage therapy in revisional hypospadias surgery, had lower observed complication rates when compared to its use in primary repair. Inner preputial skin ingrafts for primary repair yielded an acceptable complication rate. Our study describes some of the longest follow-up times in the literature with complications observed even up to 10 years postoperatively. This reinforces the need for active long-term follow-up in reporting outcomes in hypospadias surgery.

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Version ID

1

Status

Publisher

Authors Full Name

Loloi, Justin; Harrington, Stephen; Boltz, Suzanne; Decter, Ross M.

Institution

Loloi, Justin. The Pennsylvania State University, College of Medicine, Hershey, PA, USA.

Harrington, Stephen. The Pennsylvania State University, College of Medicine, Hershey, PA, USA.

Boltz, Suzanne. Department of Surgery, Division of Urology, Penn State Milton S. Hershey Medical Center, Hershey, PA, USA.

Decter, Ross M. Department of Surgery, Division of Urology, Penn State Milton S. Hershey Medical Center, Hershey, PA, USA. Electronic address: rdecter@pennstatehealth.psu.edu.

Year of Publication
2020

3.

Urethral duplication in male children: A study of 12 cases.

Kang SK; Kim J; Lee YS; Han SW; Kim SW.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid
MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Journal of Pediatric Surgery. 2020 Jan 29.

[Journal Article]

UI: 32063365

PURPOSE: To present our experience in urethral duplication focusing on detailed surgical management.

METHODS: We retrospectively reviewed the records of 12 male patients treated for urethral duplication between 2005 and 2017. Evaluations included ultrasound, retrograde urethrography, cystoscopy, and voiding cystourethrography.

RESULTS: The age at presentation ranged from birth to 11years. All 12 cases were classified using the Effmann classification. Case 1-4 patients with type I underwent excision of the dorsal accessory urethra by stripping technique. In case 5 patient(type IA) with two adjacent apical urethras, the septum was opened to form a single channel. Case 6 patient with type IB underwent visual internal urethrotomy near bulbous urethra to combine urethra into one channel. Five patients classified as type II (one with a type IIA1, and four with type IIA2 urethras). Urethral duplication was incidentally found during epispadias repair in case 7 patient with type IIA1, which was corrected by ventral plication, and excision of the dorsal epispadial urethra with stripping technique just below pubic bone. Case 8 patient with type IIA2 also required dorsal urethral excision with stripping technique. The two Y-type patients (case 10, 11) underwent urethrourethrostomy with a single-stage buccal mucosa tube graft, followed by repetitive surgeries owing to urethral stricture. One type III patient presented with penile inflammation and suprapubic pain, and underwent excision of both the dorsal urethra and nonfunctional anterior bladder.

CONCLUSIONS: Urethral duplication requires individualized surgical approaches based on the anatomical and functional characteristics. Because prognosis is variable depending on type and

accompanied anomalies, these should be taken into account when planning a comprehensive workup and surgical management.

LEVEL OF EVIDENCE: Level IV.

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Version ID

1

Status

Publisher

Authors Full Name

Kang, Sung Ku; Kim, Jinu; Lee, Yong Seung; Han, Sang Won; Kim, Sang Woon.

Institution

Kang, Sung Ku. Department of Urology, Yonsei University College of Medicine, Seoul, Republic of Korea. Kim, Jinu. Department of Urology, Yonsei University College of Medicine, Seoul, Republic of Korea.

Lee, Yong Seung. Department of Urology, Yonsei University College of Medicine, Seoul, Republic of Korea.

Han, Sang Won. Department of Urology, Yonsei University College of Medicine, Seoul, Republic of Korea.

Kim, Sang Woon. Department of Urology, Yonsei University College of Medicine, Seoul, Republic of Korea. Electronic address: khisin@yuhs.ac.

Year of Publication

2020

4.

Outcomes of preputioplasty in patients undergoing TIP urethroplasty (tubularization of incised urethral plate) for distal and mid penile hypospadias.

Shoor G; Sugandhi N; Acharya SK; Chakraborty G; Teckchandani N; Dixit A; Kour H; Bagga D.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid

MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Journal of pediatric urology. 2020 Apr 21.

[Journal Article]

UI: 32376290

BACKGROUND: Preputioplasty as a part of hypospadias repair restores the normal appearance of phallus, which is especially important in distal and mid penile hypospadias. However possibility of its inherent complications such as iatrogenic phimosis or preputial breakdown are the cause of controversy and reluctance regarding this procedure. This study evaluates the results of preputial reconstruction with TIP urethroplasty in distal and mid penile hypospadias repair and analyses if preputioplasty may be offered to these patients.

MATERIALS & METHODS: In this prospective observational study, 48 cases of distal and mid penile hypospadias underwent TIP urethroplasty and preputioplasty and results were assessed at 2 weeks, 3 months and 6 months. Major complications included preputial dehiscence, tight prepuce (iatrogenic phimosis) and minor complications included ventral tethering, persistent dorsal whorls and redundant prepuce. Data was analysed with Microsoft Excel spreadsheet where descriptive statistics were obtained.

RESULTS: Preputioplasty was performed in 48 children with a mean age of 5.1 years. Preputioplasty dehiscence was seen in three (6%) patients, which gave an appearance of irregular prepuce on 6 m follow up. Two patients (4%) were confirmed to have preputial tightness at 3 months but this resolved conservatively in one patient and only one patient (2%) required circumcision for a tight prepuce. Minor complications included ventral tethering, persistence of dorsal whorls and redundant prepuce. Ventral tethering was present in 3 patients (6.25%). Redundant prepuce was observed in 2 patients (4.16%). Additionally, unsightly dorsal whorls were found to be persistent in 2 children (4.16%). None of these patients opted for circumcision. The rest of the children had a cosmetically and functionally normal prepuce. Two patients (4%) developed urethrocutaneous fistula at 3 months' follow-up.

CONCLUSION: Preputial reconstruction is feasible with a good cosmetic outcome and minimal complications in patients of distal and mid penile hypospadias undergoing TIP urethroplasty. Mild preputial tightness evolves over time and resolves with conservative measures. In patients with very prominent dorsal whorls and underlying bulky tissues the preputioplasty does not appear to be of satisfactory cosmesis. To help the patient and parents take a well informed decision, it would be useful to explain all possible major and minor foreskin complications, and their rectification.

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Version ID

1

Status

Publisher

Authors Full Name

Shoor, Gunjan; Sugandhi, Nidhi; Acharya, Samir Kant; Chakraborty, Goutam; Teckchandani, Narinder; Dixit, Anand; Kour, Harshita; Bagga, Deepak.

Institution

Shoor, Gunjan. Department of Paediatric Surgery, VMMC and Safdarjung Hospital, New Delhi, India. Sugandhi, Nidhi. Department of Paediatric Surgery, VMMC and Safdarjung Hospital, New Delhi, India. Electronic address: drnidhisugandhi@gmail.com.

Acharya, Samir Kant. Department of Paediatric Surgery, VMMC and Safdarjung Hospital, New Delhi, India.

Chakraborty, Goutam. Department of Paediatric Surgery, VMMC and Safdarjung Hospital, New Delhi, India.

Teckchandani, Narinder. Department of Paediatric Surgery, VMMC and Safdarjung Hospital, New Delhi, India.

Dixit, Anand. Department of Paediatric Surgery, VMMC and Safdarjung Hospital, New Delhi, India.

Kour, Harshita. Department of Paediatric Surgery, VMMC and Safdarjung Hospital, New Delhi, India.

Bagga, Deepak. Department of Paediatric Surgery, VMMC and Safdarjung Hospital, New Delhi, India.

Year of Publication

2020

5.

Urinary flow measurement in hypospadias correlated to surgical procedure and risk of development of urethra-cutaneous fistula.

Winberg H; Anderberg M; Arnbjornsson E; Stenstrom P.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Journal of pediatric urology. 2020 Mar 29.

[Journal Article]

UI: 32295743

AIM: To explore the correlation between fistula development and urinary flow measurements after hypospadias repair with emphasis on patients with urethrocuteaneous fistula complications and to identify risk factors for fistula development.

METHODS: Urinary flow was examined in boys operated on for hypospadias. Outcome of maximum urinary flow (ml/s) (Qmax), voided volume for age (ml) (Volume), and pathological flow pattern (n) (Curve) was compared between the Byars, Mathieu, and Tubularized Incised Plate (TIP) surgical repair methods and between the groups of those who had and had not developed a fistula. Logistic regression analysis was performed for age at operation, genetics, comorbidity, or urinary flow measurements regarding the development of urethrocuteaneous fistula.

RESULTS: Seventy-three boys underwent hypospadias repair. Overall, the urinary flow measurements differed significantly between the three reconstructive methods, being favorable for the Mathieu procedure regarding Qmax ($p < 0.01$), volume ($p = 0.04$), and frequency of pathological voiding curve ($p < 0.01$; Table). The frequency of urethrocuteaneous fistula was 18% (13/73) and did not differ significantly between the three different reconstructive surgery methods (Byar 33%, Mathieu 32%, and TIP 12%; $p = 0.22$). Urinary flow measurements did not differ between patients with and without fistula complications regarding Qmax 10 ml/s (4-16) vs. 8 ml/s (2-18), voided volume 74 ml (35-171) vs. 71 ml (9-270), or abnormal urinary flow pattern (23% vs. 30%). On logistic regression analysis, age at operation, genetics, comorbidity, and urinary flow measurement parameters did not turn out to be independent risk factors for development of urethrocuteaneous fistula after hypospadias repair.

DISCUSSIONS: The study demonstrated significant differences between the urinary flow measurement results between the three different repair methods, favoring the Mathieu procedure. A low Qmax was a common postoperative finding. Urinary flow measurements did not differ between boys developing fistula and those who did not. No risk factors for fistula development were identified. The study did not support that it would be possible, at an early postoperative stage, to identify those with an upcoming postoperative fistula neither with urinary flow measurements nor through risk factors. No similar reports have studied the possibility of using postoperative urinary flow measurements to determine patients at risk of fistula development after hypospadias repair.

CONCLUSIONS: Urinary flow measurements were favorable after hypospadias reconstruction with Mathieu compared with Byars and TIP. Furthermore, urinary flow measurements did not differ between reconstructed with and without a fistula complication. No risk factor for fistula development was identified.

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Version ID

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Publisher

Authors Full Name

Winberg, Hans; Anderberg, Magnus; Arnbjornsson, Einar; Stenstrom, Pernilla.

Institution

Winberg, Hans. Department of Pediatric Surgery, Skane University Hospital, 221 85, Lund, Sweden; Department of Clinical Sciences, Pediatrics, Lund University, Lund, Sweden. Electronic address: winberghans@gmail.com. Anderberg, Magnus. Department of Pediatric Surgery, Skane University Hospital, 221 85, Lund, Sweden; Department of Clinical Sciences, Pediatrics, Lund University, Lund, Sweden. Electronic address: magnus.anderberg@med.lu.se.

Arnbjornsson, Einar. Department of Pediatric Surgery, Skane University Hospital, 221 85, Lund, Sweden; Department of Clinical Sciences, Pediatrics, Lund University, Lund, Sweden. Electronic address: einar.arnbjornsson@telia.com.

Stenstrom, Pernilla. Department of Pediatric Surgery, Skane University Hospital, 221 85, Lund, Sweden; Department of Clinical Sciences, Pediatrics, Lund University, Lund, Sweden. Electronic address: pernilla.stenstrom@med.lu.se.

Year of Publication

2020

6.

A case of a parthenogenetic 46,XX/46,XY chimera presenting ambiguous genitalia.

Kawamura R; Kato T; Miyai S; Suzuki F; Naru Y; Kato M; Tanaka K; Nagasaka M; Tsutsumi M; Inagaki H; Ioroi T; Yoshida M; Nao T; Conlin LK; Iijima K; Kurahashi H; Taniguchi-Ikeda M.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Journal of Human Genetics. 2020 Apr 10.

[Journal Article]

UI: 32277176

Sex-chromosome discordant chimerism (XX/XY chimerism) is a rare chromosomal disorder in humans. We report a boy with ambiguous genitalia and hypospadias, showing 46,XY[26]/46,XX[4] in peripheral blood cells. To clarify the mechanism of how this chimerism took place, we carried out whole-genome genotyping using a SNP array and microsatellite analysis.

The B-allele frequency of the SNP array showed a mixture of three and five allele combinations, which excluded mosaicism but not chimerism, and suggested the fusion of two embryos or a shared parental haplotype between the two parental cells. All microsatellite markers showed a single maternal allele. From these results, we concluded that this XX/XY chimera is composed of two different paternal alleles and a single duplicated maternal genome. This XX/XY chimera likely arose from a diploid maternal cell that was formed via endoduplication of the maternal genome just before fertilization, being fertilized with both X and Y sperm.

Version ID

1

Status

Publisher

Authors Full Name

Kawamura, Rie; Kato, Takema; Miyai, Shunsuke; Suzuki, Fumihiko; Naru, Yuki; Kato, Maki; Tanaka, Keiko; Nagasaka, Miwako; Tsutsumi, Makiko; Inagaki, Hidehito; Ioroi, Tomoaki; Yoshida, Makiko; Nao, Tomoya; Conlin, Laura K; Iijima, Kazumoto; Kurahashi, Hiroki; Taniguchi-Ikeda, Mariko.

Institution

Kawamura, Rie. Department of Molecular Genetics, Fujita Health University, Toyoake, Japan.

Kato, Takema. Department of Molecular Genetics, Fujita Health University, Toyoake, Japan.

Miyai, Shunsuke. Department of Molecular Genetics, Fujita Health University, Toyoake, Japan.

Suzuki, Fumihiko. Department of Molecular Genetics, Fujita Health University, Toyoake, Japan.

Naru, Yuki. Department of Molecular Genetics, Fujita Health University, Toyoake, Japan.

Kato, Maki. Department of Molecular Genetics, Fujita Health University, Toyoake, Japan.

Tanaka, Keiko. Division of Genetic Counseling, Kobe University Hospital, Kobe, Japan.

Nagasaka, Miwako. Division of Genetic Counseling, Kobe University Hospital, Kobe, Japan.

Nagasaka, Miwako. Department of Pediatrics, Kobe University Graduate School of Medicine, Kobe, Japan.

Tsutsumi, Makiko. Department of Molecular Genetics, Fujita Health University, Toyoake, Japan.

Inagaki, Hidehito. Department of Molecular Genetics, Fujita Health University, Toyoake, Japan.

Ioroi, Tomoaki. Department of Pediatrics, Comprehensive Perinatal Maternal and Child Medical Center, Himeji Red Cross Hospital, Himeji, Japan.

Yoshida, Makiko. Department of Pathology, Hyogo Prefectural Kobe Children's Hospital, Kobe, Japan.

Nao, Tomoya. Department of Urology, Hyogo Prefectural Kobe Children's Hospital, Kobe, Japan.

Conlin, Laura K. Division of Genomic Diagnostics, Department of Pathology and Laboratory Medicine, Children's Hospital of Philadelphia, Philadelphia, PA, USA.

Iijima, Kazumoto. Division of Genetic Counseling, Kobe University Hospital, Kobe, Japan.

Iijima, Kazumoto. Department of Pediatrics, Kobe University Graduate School of Medicine, Kobe, Japan.

Kurahashi, Hiroki. Department of Molecular Genetics, Fujita Health University, Toyoake, Japan.

Taniguchi-Ikeda, Mariko. Department of Molecular Genetics, Fujita Health University, Toyoake, Japan. mtani@fujita-hu.ac.jp.

Taniguchi-Ikeda, Mariko. Department of Clinical Genetics, Fujita Health University Hospital, Toyoake, Japan. mtani@fujita-hu.ac.jp.

Year of Publication

2020

7.

Re: The Biomechanical Properties of the Urethra in Boys with Hypospadias: A Preliminary Study.

Canning DA.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid

MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Journal of Urology. 101097JU000000000000104903, 2020 Apr 09.

[Journal Article]

UI: 32272051

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1

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Authors Full Name

Canning, Douglas A.

Year of Publication

2020

8.

Save the foreskin: Outcomes of preputioplasty in the treatment of childhood phimosis.

Hotonu S; Mohamed A; Rajimwale A; Gopal M.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid

MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Surgeon Journal of the Royal Colleges of Surgeons of Edinburgh & Ireland. 18(3):150-153, 2020 Jun.

[Journal Article]

UI: 31548120

BACKGROUND: Symptomatic phimosis is a common childhood urology complaint. Circumcision was traditionally the treatment of choice, but its popularity in cases of non-scarred phimosis has been superseded by more conservative methods like preputioplasty. We sought to examine outcomes of preputioplasty for the treatment of non-scarred pathological phimosis in two UK paediatric surgery tertiary centres.

METHODS: Retrospective case series selecting cases performed in both departments over a 4 year period (January 2012-December 2015).

INCLUSION CRITERIA: non-scarred pathological phimosis treated with preputioplasty.

EXCLUSION CRITERIA: diffuse scarring of foreskin or presence of balanitis xerotica obliterans (BXO), preputioplasty performed as part of hypospadias repair. Outcome measure was treatment success as evidenced by fully retractile prepuce at follow up. Follow up occurred between 3 and 24 months.

RESULTS: We identified 126 patients, 6 were excluded due to the above criteria. Median age was 13.4 years (range 10 months-18 years). Median follow up was 13 months (range 3-24 months). 115 patients (96%) had successful treatment as evidenced by satisfactory post-operative cosmesis and complete resolution of phimosis at follow up. Recurrence of phimosis occurred in 5 patients (4%). Mean time of recurrence was 6 months, with a median age of recurrence of 15.3 years (range 10.7-16.7 years). All patients with recurrence were successfully treated with circumcision.

CONCLUSION: Foreskin conserving methods like preputioplasty are a valid option in the treatment of non-scarred pathological phimosis.

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Version ID

1

Status

In-Process

Authors Full Name

Hotonu, Sesi; Mohamed, Ahmed; Rajimwale, Ashok; Gopal, Milan.

Institution

Hotonu, Sesi. The Great North Children's Hospital Queen Victoria Road, Newcastle upon Tyne, NE1 4LP, UK. Electronic address: sesihotonu@doctors.org.uk. Mohamed, Ahmed. The Great North Children's Hospital Queen Victoria Road, Newcastle upon Tyne, NE1 4LP, UK. Electronic address: dr.ahmed.osama.salama@gmail.com.

Rajimwale, Ashok. Leicester Children's Hospital Infirmary Square, Leicester, LE1 5WW, UK. Electronic address: ashok.rajimwale@nhs.net.

Gopal, Milan. The Great North Children's Hospital Queen Victoria Road, Newcastle upon Tyne, NE1 4LP, UK. Electronic address: milan.gopal@nuth.nhs.uk.

Year of Publication

2020

9.

Wilms' Tumor and Benign Renal Tumor Combined With Hypospadias and Incomplete Orchicatabasis Appearing Simultaneously in a 10 Months Old Boy.

Li C; Li WS.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present
Urology Journal. 17(3):321-323, 2020 May 16.

[Journal Article]

UI: 31134609

We herein report a case of left renal Wilms' tumor and right renal hamartoma combined with hypospadias and incomplete orchicatabasis in a 10-month-old boy. In the literature to date, no case has been reported. The preoperative abdominal computerized tomography (CT) scan was suggestive of bilateral nephroblastomas, and clinical diagnosis was bilateral renal tumors with external genitals malformation, a syndrome? Finally, this case was used by B-ultrasonic guided percutaneous biopsy to help determine the nature of bilateral renal tumors. Afterwards, the boy underwent preoperative chemotherapy, surgery (a left radical nephrectomy and right wedge excision of the renal tumor) and postoperative chemotherapy. After 3 years of follow-up, there was no evidence of tumor recurrence, the renal function was normal, and the boy's height, weight and intelligence were also within normal range. Owing to no similar cases as a reference, we

discussed the preoperative imaging diagnosis, final etiological diagnosis and appropriate treatment of this disease. Long-term follow-up with a sufficient number of cases may be needed to optimize methods of diagnosis and define optimal treatment options for patients with this extremely rare disease.

Version ID

1

Status

In-Process

Authors Full Name

Li, Chun; Li, Wei-Song.

Institution

Li, Chun. The First Affiliated Hospital of Anhui Medical University, pediatric surgery.

leiforget1970@hotmail.com.

Year of Publication

2020

10.

In cases of familial primary ovarian insufficiency and disorders of gonadal development, consider NR5A1/SF-1 sequence variants.

Bertrand-Delepine J; Manouvrier-Hanu S; Cartigny M; Paris F; Mallet D; Philibert P; Morel Y; Lefevre C; Dewailly D; Catteau-Jonard S.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid

MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Reproductive Biomedicine Online. 40(1):151-159, 2020 Jan.

[Journal Article]

UI: 31831369

RESEARCH QUESTION: Primary ovarian insufficiency (POI) is defined as the early exhaustion of ovarian function, before the age of 40 years. Its origin is genetic in 20-25% of cases. In rare cases, sequence variants of the NR5A1/SF-1 gene may result in POI, or in various disorders of gonadal development (DGD) or adrenal insufficiency.

DESIGN: This study describes the cases of two families in which the association of DGD and POI enabled a diagnosis of NR5A1 deleterious variations. Their clinical, hormonal, ultrasound and genetic characteristics are reported.

RESULTS: The mothers of the affected children were 21 and 29 years when POI was diagnosed. Each nonetheless had two spontaneous pregnancies. The children have different phenotypes and different forms of DGD. None of the affected family members had adrenal insufficiency. A new sequence variant of the NR5A1 gene was identified in one family: p.Cys283Phe (c.848G>T), and the NR5A1 sequence variant c.86G>C was found in the other family.

CONCLUSION: Sequence variation of the NR5A1 gene is a possibility that must be considered when a woman with POI or a diminished ovarian reserve has a family member or child with DGD. If a variant is identified, genetic counselling is essential for the patient and his/her family.

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Version ID

1

Status

In-Data-Review

Authors Full Name

Bertrand-Delepine, Juliette; Manouvrier-Hanu, Sylvie; Cartigny, Maryse; Paris, Francoise; Mallet, Delphine; Philibert, Pascal; Morel, Yves; Lefevre, Christine; Dewailly, Didier; Catteau-Jonard, Sophie.

Institution

Bertrand-Delepine, Juliette. CH Calais, Department of Obstetrics and Gynecology, Calais F-62100, France. Manouvrier-Hanu, Sylvie. Universite de Lille. Lille, CHU Lille, Clinique de Genetique, EA 7364-RADEME, Lille F-59000, France.

Cartigny, Maryse. CHU Lille, Centre de Reference DEV-GEN, Lille F-59000, France.

Paris, Francoise. Departement d'Endocrinologie et de Gynecologie Pediatrique CHU Arnaud de Villeneuve, 34090 Montpellier, Departement de Genetique, IURC, Equipe DEV-GEN, Montpellier 34090, France.

Mallet, Delphine. CHU Lyon, Laboratoire de Biochimie et Biologie Moleculaire Grand Est, UM Pathologies Endocriniennes Renales Musculaires et Mucoviscidose - Centre de Reference DEV-GEN, Bron F-69677, France.

Philibert, Pascal. Departement d'Endocrinologie et de Gynecologie Pediatrique CHU Arnaud de Villeneuve, 34090 Montpellier, Departement de Genetique, IURC, Equipe DEV-GEN, Montpellier 34090, France.

Morel, Yves. CHU Lyon, Laboratoire de Biochimie et Biologie Moleculaire Grand Est, UM Pathologies Endocriniennes Renales Musculaires et Mucoviscidose - Centre de Reference DEV-

GEN, Bron F-69677, France; Universite de Lyon, Universite Claude Bernard Lyon 1, Lyon, France.

Lefevre, Christine. CHU Lille, Centre de Reference DEV-GEN, Lille F-59000, France.

Dewailly, Didier. Universite de Lille, CHU Lille, INSERM U1172, Lille F-59000, France.

Catteau-Jonard, Sophie. Universite de Lille, CHU Lille, INSERM U1172, Lille F-59000, France; Universite de Lille, CHU Lille, Department of Reproductive Medicine, Lille F-59000, France.

Electronic address: sophie.jonard@chru-lille.fr.

Year of Publication

2020

11.

Psychosexual Functioning Outcome Testing after Hypospadias Repair.

Majstorovic M; Bizic M; Nikolic D; Stojanovic B; Bencic M; Joksic I; Djordjevic M.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid

MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Healthcare. 8(1), 2020 Feb 05.

[Journal Article]

UI: 32033376

Self-confidence plays an important role in both genders' sexual functioning. Lack of genital self-esteem may have negative effects on psychosexual development, especially in males, where deviations from a standardized normal penile appearance can lead to inhibitions in entering into sexual relationships. The aim of our study was to evaluate the informativeness of studied domains of the Global Sexual Functioning (GSF) questionnaire and sexual functioning of patients surgically treated in childhood for different types of hypospadias. We evaluated 63 males with hypospadias and 60 healthy age- and gender-matched controls. The GSF questionnaire was used to estimate psychosexual function as a long-term follow-up after the surgical correction of hypospadias in the patient and control groups. Sexual activity ($p = 0.017$), arousal ($p = 0.033$) and orgasmic abilities ($p = 0.002$) values were significantly increased in patients. Strong correlation was noticed between sexual activity and sexual desire ($R = 0.872$); arousal and sexual desire ($R = 0.753$), as well as orgasmic and erectile abilities ($R = 0.769$). Different domains of psychosexual functioning in the patient group correlated with each other to various degrees, resulting in a

heterogeneous expression of psychosexual dysfunctions, implicating the necessity of a personalized treatment approach.

Version ID

1

Status

PubMed-not-MEDLINE

Author Initials

Nikolic, Dejan; ORCID: <https://orcid.org/0000-0002-0609-9916>

Authors Full Name

Majstorovic, Marko; Bizic, Marta; Nikolic, Dejan; Stojanovic, Borko; Bencic, Marko; Joksic, Ivana; Djordjevic, Miroslav.

Institution

Majstorovic, Marko. University Children's Hospital, Tirsova 10, 11000 Belgrade, Serbia.

Majstorovic, Marko. Faculty of Medicine, University of Belgrade, 11000 Belgrade, Serbia.

Bizic, Marta. University Children's Hospital, Tirsova 10, 11000 Belgrade, Serbia.

Bizic, Marta. Faculty of Medicine, University of Belgrade, 11000 Belgrade, Serbia.

Nikolic, Dejan. University Children's Hospital, Tirsova 10, 11000 Belgrade, Serbia.

Nikolic, Dejan. Faculty of Medicine, University of Belgrade, 11000 Belgrade, Serbia.

Stojanovic, Borko. University Children's Hospital, Tirsova 10, 11000 Belgrade, Serbia.

Stojanovic, Borko. Faculty of Medicine, University of Belgrade, 11000 Belgrade, Serbia.

Bencic, Marko. University Children's Hospital, Tirsova 10, 11000 Belgrade, Serbia.

Joksic, Ivana. Faculty of Medicine, University of Belgrade, 11000 Belgrade, Serbia.

Joksic, Ivana. Clinic for Gynecology and Obstetrics "Narodni front", 11000 Belgrade, Serbia.

Djordjevic, Miroslav. University Children's Hospital, Tirsova 10, 11000 Belgrade, Serbia.

Djordjevic, Miroslav. Faculty of Medicine, University of Belgrade, 11000 Belgrade, Serbia.

PMID

<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC7151077>

Year of Publication

2020

12.

Further delineation of putative ACTB loss-of-function variants: A 4-patient series.

Baumann M; Beaver EM; Palomares-Bralo M; Santos-Simarro F; Holzer P; Povysil G; Muller T; Valovka T; Janecke AR.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Human Mutation. 41(4):753-758, 2020 Apr.

[Journal Article]

UI: 31898838

ACTB encodes beta-cytoplasmic actin, an essential component of the cytoskeleton. Based on chromosome 7p22.1 deletions that include the ACTB locus and on rare truncating ACTB variants, a phenotype resulting from ACTB haploinsufficiency was recently proposed. We report putative ACTB loss-of-function variants in four patients. To the best of our knowledge, we report the first 7p22.1 microdeletion confined to ACTB and the second ACTB frameshifting mutation that predicts mRNA decay. A de-novo ACTB p.(Gly302Ala) mutation affects beta-cytoplasmic actin distribution. All four patients share a facial gestalt that is distinct from that of individuals with dominant-negative ACTB variants in Baraitser-Winter cerebrofrontofacial syndrome. Two of our patients had strikingly thin and sparse scalp hair. One patient had sagittal craniosynostosis and hypospadias. All three affected male children have attention deficits and mild global developmental delay. Mild intellectual disability was present in only one patient. Heterozygous ACTB deletion can allow for normal psychomotor function.

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1

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Author Initials

Janecke, Andreas R; ORCID: <http://orcid.org/0000-0001-7155-0315>

Authors Full Name

Baumann, Matthias; Beaver, Erin M; Palomares-Bralo, Maria; Santos-Simarro, Fernando; Holzer, Peter; Povysil, Gundula; Muller, Thomas; Valovka, Taras; Janecke, Andreas R.

Institution

Baumann, Matthias. Department of Pediatrics I, Medical University of Innsbruck, Innsbruck, Austria. Beaver, Erin M. Mercy Kids Genetics, Mercy Children's Hospital St. Louis, St. Louis, Missouri.

Palomares-Bralo, Maria. Institute of Medical and Molecular Genetics, University Hospital La Paz, Madrid, Spain.

Santos-Simarro, Fernando. Institute of Medical and Molecular Genetics, University Hospital La Paz, Madrid, Spain.

Holzer, Peter. Intelligent Predictive Networks GmbH, Vienna, Austria.
Povysil, Gundula. Institute of Bioinformatics, Johannes Kepler University, Linz, Austria.
Muller, Thomas. Department of Pediatrics I, Medical University of Innsbruck, Innsbruck, Austria.
Valovka, Taras. Department of Pediatrics I, Medical University of Innsbruck, Innsbruck, Austria.
Janecke, Andreas R. Department of Pediatrics I, Medical University of Innsbruck, Innsbruck, Austria.
Janecke, Andreas R. Division of Human Genetics, Medical University of Innsbruck, Innsbruck, Austria.
PMID
<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC7155001>
Year of Publication
2020

13.

Discrete Hedgehog Factor Expression and Action in the Developing Phallus.
Tarulli GA; Pask AJ; Renfree MB.
OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid
MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present
International Journal of Molecular Sciences. 21(4), 2020 Feb 12.
[Journal Article]
UI: 32059607
Hypospadias is a failure of urethral closure within the penis occurring in 1 in 125 boys at birth and is increasing in frequency. While paracrine hedgehog signalling is implicated in the process of urethral closure, how these factors act on a tissue level to execute closure itself is unknown. This study aimed to understand the role of different hedgehog signalling members in urethral closure. The tammar wallaby (*Macropus eugenii*) provides a unique system to understand urethral closure as it allows direct treatment of developing offspring because mothers give birth to young before urethral closure begins. Wallaby pouch young were treated with vehicle or oestradiol (known to induce hypospadias in males) and samples subjected to RNAseq for differential expression and gene ontology analyses. Localisation of Sonic Hedgehog (SHH) and Indian Hedgehog (IHH), as well as the transcription factor SOX9, were assessed in normal phallus tissue using immunofluorescence. Normal tissue culture explants were treated with SHH or IHH and analysed

for AR, ESR1, PTCH1, GLI2, SOX9, IHH and SHH expression by qPCR. Gene ontology analysis showed enrichment for bone differentiation terms in male samples compared with either female samples or males treated with oestradiol. Expression of SHH and IHH localised to specific tissue areas during development, akin to their compartmentalised expression in developing bone. Treatment of phallus explants with SHH or IHH induced factor-specific expression of genes associated with bone differentiation. This reveals a potential developmental interaction involved in urethral closure that mimics bone differentiation and incorporates discrete hedgehog activity within the developing phallus and phallic urethra.

Version ID

1

Status

In-Process

Authors Full Name

Tarulli, Gerard A; Pask, Andrew J; Renfree, Marilyn B.

Institution

Tarulli, Gerard A. School of BioSciences, The University of Melbourne, Parkville, VIC 3010, Australia. Pask, Andrew J. School of BioSciences, The University of Melbourne, Parkville, VIC 3010, Australia.

Renfree, Marilyn B. School of BioSciences, The University of Melbourne, Parkville, VIC 3010, Australia.

PMID

<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC7072906>

Year of Publication

2020

14.

Expanding the phenotype of thrombocytopenia absent radius syndrome with hypospadias.

[Review]

Miertus J; Maltese PE; Hyblova M; Tomkova E; Durovcikova D; Risova V; Bertelli M.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid

MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Journal of Biotechnology. 311:44-48, 2020 Mar 10.

[Journal Article. Review]

UI: 32109542

Rare genetic diseases and syndromes may appear with unique features in some patients. In genetically-solved cases, this situation indicates a phenotypic expansion of the syndrome with additional features (i.e. the disease-associated gene gives rise to unusual clinical presentation). However, this situation can also hide a multilocus pathogenic variation that cannot be solved genetically except by a massive sequencing approach, such as exome sequencing. Here we describe the case of a child with bilateral radial aplasia, transient thrombocytopenia and anemia, cow's milk intolerance, hypospadias, facial dysmorphism, mild hypothyroidism and umbilical and inguinal hernia. Bilaterally absent radius, presence of thumbs and low platelet count are pathognomonic of thrombocytopenia absent radius (TAR) syndrome, but the child also showed other features beyond those reported in the literature. Since various diseases resembling the proband's phenotype required differential diagnosis, clinical exome sequencing was performed. The results showed compound heterozygous mutations in the RBM8A gene, confirming the suspicion of TAR syndrome. A truncating heterozygous variant in the DUOX2 gene, known to be associated with transient thyroid dysmorphogenesis type 6 (TDH6), was also detected and may explain the proband's mild hypothyroidism.

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1

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Authors Full Name

Miertus, Jan; Maltese, Paolo Enrico; Hyblova, Michaela; Tomkova, Erika; Durovcikova, Darina; Risova, Vanda; Bertelli, Matteo.

Institution

Miertus, Jan. Genius n. o., Mestska Poliklinika, Starohajska 2, Trnava, Slovakia; MAGI's Lab, via delle Maioliche 57/D, Rovereto, TN, Italy. Electronic address: director@genius.care. Maltese, Paolo Enrico. MAGI's Lab, via delle Maioliche 57/D, Rovereto, TN, Italy. Electronic address: paolo.maltese@assomagi.org.

Hyblova, Michaela. Genetics Lab, Medirex a.s., Galvaniho 17C, Bratislava, Slovakia. Electronic address: michaela.hyblova@medirex.sk.

Tomkova, Erika. Genetics Lab, Medirex a.s., Galvaniho 17C, Bratislava, Slovakia. Electronic address: erika.tomkova@medirex.sk.

Durovcikova, Darina. Genetic Clinic, Slovak Healthcare University, Limbova 12, Bratislava, Slovakia. Electronic address: darina.durovcikova@szu.sk.

Risova, Vanda. Histology Dept., "Commeniana" University, Spitalska 24, Bratislava, Slovakia.
Electronic address: vanda.risova@gmail.com.

Bertelli, Matteo. MAGI's Lab, via delle Maioliche 57/D, Rovereto, TN, Italy. Electronic address:
matteo.bertelli@assomagi.org.

Year of Publication
2020

15.

Urinary Tract Infection in Pediatric Patients on Clean Intermittent Catheterization via a Mitrofanoff port with Reused Catheters - Any Association with Catheter Sterility?.

Sam CJ; Jagadeesan CT; Sen S; Arunachalam P; Appalaraju B; Das PT.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid

MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Journal of Indian Association of Pediatric Surgeons. 25(2):91-95, 2020 Mar-Apr.

[Journal Article]

UI: 32139987

Objective: The objective of this study is to find out whether the reused catheters for clean intermittent catheterization (CIC) are colonized before insertion and its association with urinary tract infection (UTI).

Materials and Methods: This is a study conducted on 28 pediatric surgery patients who are on CIC via a Mitrofanoff port and who were reusing catheters, in a tertiary care private medical college hospital for 6-month period. Catheters to be used for the next catheterization were sent for culture along with urine culture. A questionnaire was utilized to assess CIC practice and UTI.

Results: Diseases of patients were: neurogenic bladder and exstrophy-epispadias and posterior urethral valve. Twenty-one of them had an augmented bladder. Hydronephrosis was present in ten and vesicoureteral reflux (VUR) in five. Their mean duration of CIC was 5.3 years. Of 28 catheter tip samples, 16 catheters were colonized with organism. Of the 28 urine culture samples, 17 cultures were positive and all were asymptomatic except one. Of the 16 positive catheter samples, only 9 had positive urine culture; four of them had grown different organisms and five of them had the same organism, and even in these five, single organism was seen only in three. Urine culture grew Gram-negative organism in 85%, but catheter grew Gram-positive organism in

46%. No difference was found in the variables between both groups such as hydronephrosis, VUR, and augmented bladder.

Conclusion: Reused catheters were colonized in 57% of the study patients, but reused catheter may not be the cause of culture positivity or UTI in the study population.

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Version ID

1

Status

PubMed-not-MEDLINE

Authors Full Name

Sam, Cenita James; Jagadeesan, Cindrel T; Sen, Sudipta; Arunachalam, Pavai; Appalaraju, B; Das, Praseetha T.

Institution

Sam, Cenita James. Department of Paediatric Surgery, PSG Institute of Medical Sciences and Research, Coimbatore, Tamil Nadu, India. Jagadeesan, Cindrel T. Department of Paediatric Surgery, PSG Institute of Medical Sciences and Research, Coimbatore, Tamil Nadu, India.

Sen, Sudipta. Department of Paediatric Surgery, PSG Institute of Medical Sciences and Research, Coimbatore, Tamil Nadu, India.

Arunachalam, Pavai. Department of Paediatric Surgery, PSG Institute of Medical Sciences and Research, Coimbatore, Tamil Nadu, India.

Appalaraju, B. Department of Microbiology, PSG Institute of Medical Sciences and Research, Coimbatore, Tamil Nadu, India.

Das, Praseetha T. Department of Paediatric Surgery, PSG Institute of Medical Sciences and Research, Coimbatore, Tamil Nadu, India.

PMID

<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC7020673>

Year of Publication

2020

16.

Androgen Receptor Expression in Hypospadias.

Balaji DR; Reddy G; Babu R; Paramaswamy B; Ramasundaram M; Agarwal P; Joseph LD; D'Cruze L; Sundaram S.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Journal of Indian Association of Pediatric Surgeons. 25(1):6-9, 2020 Jan-Feb.

[Journal Article]

UI: 31896892

Introduction: The exact mechanism behind the development of hypospadias is unclear. Research studies on androgen receptor (AR) expression are controversial with results stating all possible outcomes - AR elevated, similar, or reduced when compared to normal.

Aims: The aim is to study the AR expression and hormone levels in hypospadias patients and compare them with children having normal genitalia.

Methods: Group 1 (controls) involved patients who underwent circumcision for phimosis while Group 2 involved hypospadias patients who did not receive any preoperative testosterone.

Preoperative hormonal assay included luteinizing hormone, follicle-stimulating hormone, and free testosterone levels in all the patients. The foreskin specimen was analyzed for AR expression using immunohistochemistry (anti-AR antibody PathnSitu, clone R441, 1/100 dilution). AR staining was expressed as H score. The H score was calculated by multiplying the intensity of staining and the percentage of stained cells showing cytoplasmic positivity at high power (x40).

Results: There were 27 patients in Group 1 while 16 in Group 2 (distal 10; proximal 6). There was no significant difference in the age distribution. The mean H score was significantly higher (189.5) in hypospadias patients compared to controls (97.5) and was significantly higher in proximal (220) compared to distal (159) hypospadias. There was no significant difference in hormone levels between groups.

Conclusion: AR expression was significantly elevated in hypospadias patients. It was higher in proximal compared to distal hypospadias, probably due to end-organ overexpression. Further larger trials are likely to throw light into this controversial subject.

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Version ID

1

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PubMed-not-MEDLINE

Authors Full Name

Balaji, Dhanvanth Rajesh; Reddy, Goutham; Babu, Ramesh; Paramaswamy, Balamourougane; Ramasundaram, Madhu; Agarwal, Prakash; Joseph, Leena Dennis; D'Cruze, Lawrence; Sundaram, Sandhya.

Institution

Balaji, Dhanvanth Rajesh. Department of Pediatric Urology, Sri Ramachandra Medical College, Chennai, Tamil Nadu, India. Reddy, Goutham. Department of Pediatric Urology, Sri Ramachandra Medical College, Chennai, Tamil Nadu, India.

Babu, Ramesh. Department of Pediatric Urology, Sri Ramachandra Medical College, Chennai, Tamil Nadu, India.

Paramaswamy, Balamourougane. Department of Pediatric Surgery, Sri Ramachandra Medical College, Chennai, Tamil Nadu, India.

Ramasundaram, Madhu. Department of Pediatric Surgery, Sri Ramachandra Medical College, Chennai, Tamil Nadu, India.

Agarwal, Prakash. Department of Pediatric Surgery, Sri Ramachandra Medical College, Chennai, Tamil Nadu, India.

Joseph, Leena Dennis. Department of Pathology, Sri Ramachandra Medical College, Chennai, Tamil Nadu, India.

D'Cruze, Lawrence. Department of Pathology, Sri Ramachandra Medical College, Chennai, Tamil Nadu, India.

Sundaram, Sandhya. Department of Pathology, Sri Ramachandra Medical College, Chennai, Tamil Nadu, India.

PMID

<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC6910049>

Year of Publication

2020

17.

Association of Maternal First Trimester Serum Levels of Free Beta Human Chorionic Gonadotropin and Hypospadias: A Population Based Study.

Peycelon M; Lelong N; Carlier L; Monn MF; De Chalus A; Bonnard A; Rachid M; Houang M; Paye-Jaouen A; Ali L; Lecourbe A; Grapin C; Audry G; Legendre M; Muller F; Dreux S; El Ghoneimi A; Benachi A; Khoshnood B; Siffroi JP.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Journal of Urology. 203(5):1017-1023, 2020 05.

[Journal Article. Multicenter Study]

UI: 31855125

PURPOSE: Human chorionic gonadotropin stimulates fetal testosterone production and contributes to normal development of male genitalia. Using population based data we hypothesized that differences in maternal free beta human chorionic gonadotropin may be associated with hypospadias.

MATERIALS AND METHODS: Data were obtained from the Paris Registry of Congenital Malformations (REMAPAR) (2011 to 2016). The initial study population included 3,172 pregnant women who gave birth to a singleton live born male infant with a congenital malformation. After exclusion of cases with unknown beta human chorionic gonadotropin and those with chromosomal or genetic abnormalities, the study population included 194 boys with isolated hypospadias and 1,075 controls. For cases with operative notes (125) we obtained data on type (proximal/distal) of hypospadias. Using quantile regression we compared median values of multiple of median beta human chorionic gonadotropin measured for first trimester Down syndrome screening (10th to 13th gestational weeks) for overall as well as by type of hypospadias vs controls. We also considered possible effects of placental dysfunction (maternal age, intrauterine growth retardation and preterm births) as potential confounding factors.

RESULTS: Overall the median beta human chorionic gonadotropin multiple of median was comparable for women who had an infant with hypospadias vs controls (0.99 vs 0.95, $p=0.3$). However, proximal hypospadias was associated with a statistically significant higher median multiple of median than distal hypospadias or unspecified (1.49 vs 0.92 vs 1.05, $p=0.02$). The estimates were comparable after adjustment for placental dysfunction.

CONCLUSIONS: Our findings support the hypothesis that an alteration in maternal beta human chorionic gonadotropin levels is associated with hypospadias. However, this association appears to be limited to proximal hypospadias.

Version ID

1

Status

MEDLINE

Authors Full Name

Peycelon, Matthieu; Lelong, Nathalie; Carlier, Lea; Monn, M Francesca; De Chalus, Alienor; Bonnard, Adeline; Rachid, Myriam; Houang, Muriel; Paye-Jaouen, Annabel; Ali, Liza; Lecourbe, Alexandra; Grapin, Christine; Audry, Georges; Legendre, Marie; Muller, Francoise; Dreux, Sophie; El Ghoneimi, Alaa; Benachi, Alexandra; Khoshnood, Babak; Siffroi, Jean-Pierre.

Institution

Peycelon, Matthieu. Department of Pediatric Surgery and Urology, Robert-Debre University Hospital, APHP, Paris, France. Peycelon, Matthieu. Universite Paris Diderot, Sorbonne Paris Cite, Paris, France.

Peycelon, Matthieu. Centres de Reference Maladies Rares, Maladies Endocriniennes Rares de la Croissance (CRMERC), Paris, France.

Peycelon, Matthieu. Sorbonne Universite, INSERM, Maladies genetiques d'expression pediatrique, Hopital d'Enfants Armand Trousseau, APHP, Paris, France.

Peycelon, Matthieu. Department of Pediatric Urology, Riley Children's Hospital at Indiana University Health, Indianapolis, Indiana.

Peycelon, Matthieu. Department of Urology, Indiana University School of Medicine, Indianapolis, Indiana.

Lelong, Nathalie. Universite de Paris, CRESS Obstetrical, Perinatal and Pediatric Epidemiology Research Team (EPOPe), INSERM, INRA, Paris, France.

Carlier, Lea. Sorbonne Universite, INSERM, Maladies genetiques d'expression pediatrique, Hopital d'Enfants Armand Trousseau, APHP, Paris, France.

Monn, M Francesca. Department of Pediatric Urology, Riley Children's Hospital at Indiana University Health, Indianapolis, Indiana.

Monn, M Francesca. Department of Urology, Indiana University School of Medicine, Indianapolis, Indiana.

De Chalus, Alienor. Sorbonne Universite, INSERM, Maladies genetiques d'expression pediatrique, Hopital d'Enfants Armand Trousseau, APHP, Paris, France.

Bonnard, Adeline. Sorbonne Universite, INSERM, Maladies genetiques d'expression pediatrique, Hopital d'Enfants Armand Trousseau, APHP, Paris, France.

Rachid, Myriam. Sorbonne Universite, INSERM, Maladies genetiques d'expression pediatrique, Hopital d'Enfants Armand Trousseau, APHP, Paris, France.

Houang, Muriel. Department of Pediatric Endocrinology, Hopital d'Enfants Armand Trousseau, APHP, Paris, France.

Paye-Jaouen, Annabel. Department of Pediatric Surgery and Urology, Robert-Debre University Hospital, APHP, Paris, France.

Paye-Jaouen, Annabel. Centres de Reference Maladies Rares, Maladies Endocriniennes Rares de la Croissance (CRMERC), Paris, France.

Ali, Liza. Department of Pediatric Surgery and Urology, Robert-Debre University Hospital, APHP, Paris, France.

Ali, Liza. Centres de Reference Maladies Rares, Maladies Endocriniennes Rares de la Croissance (CRMERC), Paris, France.

Lecourbe, Alexandra. Universite de Paris, CRESS Obstetrical, Perinatal and Pediatric Epidemiology Research Team (EPOPe), INSERM, INRA, Paris, France.

Grabin, Christine. Department of Pediatric Surgery and Urology, Robert-Debre University Hospital, APHP, Paris, France.

Grabin, Christine. Universite Paris 13, Sorbonne Paris Cite, Paris, France.

Grapin, Christine. Centres de Reference Maladies Rares, Maladies Endocriniennes Rares de la Croissance (CRMERC), Paris, France.

Audry, Georges. Department of Pediatric Surgery, Hopital d'Enfants Armand Trousseau, APHP, Paris, France.

Legendre, Marie. Sorbonne Universite, INSERM, Maladies genetiques d'expression pediatrique, Hopital d'Enfants Armand Trousseau, APHP, Paris, France.

Muller, Francoise. Department of Biochemistry, Robert-Debre University Hospital, APHP, Paris, France.

Dreux, Sophie. Department of Biochemistry, Robert-Debre University Hospital, APHP, Paris, France.

El Ghoneimi, Alaa. Department of Pediatric Surgery and Urology, Robert-Debre University Hospital, APHP, Paris, France.

El Ghoneimi, Alaa. Universite Paris Diderot, Sorbonne Paris Cite, Paris, France.

El Ghoneimi, Alaa. Centres de Reference Maladies Rares, Maladies Endocriniennes Rares de la Croissance (CRMERC), Paris, France.

Benachi, Alexandra. Department of Gynecology-Obstetrics, Antoine Beclere Hospital, APHP and Universite Paris Sud, Clamart, France.

Khoshnood, Babak. Universite de Paris, CRESS Obstetrical, Perinatal and Pediatric Epidemiology Research Team (EPOPe), INSERM, INRA, Paris, France.

Siffroi, Jean-Pierre. Sorbonne Universite, INSERM, Maladies genetiques d'expression pediatrique, Hopital d'Enfants Armand Trousseau, APHP, Paris, France.

Comments

Comment in (CIN)

Year of Publication

2020

18.

Expression of androgen receptor and estrogen receptor-beta in foreskin tissue of hypospadiac and healthy boys. Expresion de receptores androgenicos y estrogenicos beta en la piel prepucial de hipospadias y ninos sanos. <Expresion de receptores androgenicos y estrogenicos beta en la piel prepucial de hipospadias y ninos sanos.>

Kocaturk H; Ozkaya F; Adanur S; Yapanoglu T; Aksoy Y.

OID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid
MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Archivos Espanoles de Urologia. 73(2):119-125, 2020 Mar.

[Journal Article]

UI: 32124842

OBJECTIVES: This prospective study aimed to investigate the expression of the androgen receptor(AR) and the estrogen receptor-beta (ER-beta) in foreskin tissues in boys with and without distal hypospadias.

METHODS: Thirty boys with distal hypospadias were evaluated. Fifteen boys who underwent elective circumcision over a period of 18 months served as the control group. The presence of AR and ER-beta in foreskin tissues was investigated immunohistochemically.

RESULTS: The percentages of AR in epithelial cells were 18.9 +/- 27.3% in the hypospadias group and 3.3 +/- 5.3% in the circumcision group, and the difference between the groups was significant ($p=0.034$). Of the stromal cells, 19.5 +/- 26.8% in the hypospadias group and 2.6 +/- 4.4% in the circumcision group were positive for AR ($p=0.004$). In the hypospadias group, significantly higher stromal cell percentage of ER-beta was found compared to that in the control group (24 +/- 24.5% and 1.3 +/- 1.1%, respectively, $p<0.001$). Moreover, epithelial cell percentage of ER-beta was higher in the hypospadias group than that in the control group, and the respective values were 6.8 +/- 10.1% and 0.9 +/- 1.3% ($p<0.0001$). **CONCLUSION:** The percentage of AR and ER-beta expression were higher in hypospadias-affected foreskin than in the normal foreskin. Whether the normal function of these receptors reveals, there is a need for more detailed studies.

Version ID

1

Status

MEDLINE

Authors Full Name

Kocaturk, Huseyin; Ozkaya, Fatih; Adanur, Senol; Yapanoglu, Turgut; Aksoy, Yilmaz.

Institution

Kocaturk, Huseyin. Department of Urology. Health Sciences University. Erzurum Regional Training and Research Hospital. Erzurum. Turkey. Ozkaya, Fatih. Department of Urology. Health Sciences University. Erzurum Regional Training and Research Hospital. Erzurum. Turkey. Adanur, Senol. Department of Urology. School of Medicine. Ataturk University. Erzurum. Turkey. Yapanoglu, Turgut. Department of Urology. School of Medicine. Ataturk University. Erzurum. Turkey.

Aksoy, Yilmaz. Department of Urology. School of Medicine. Ataturk University. Erzurum. Turkey.

Collaborator Alias

Publisher: Este estudio prospectivo que pretende investigar la expresion del receptor androgenico y estrogenico en la piel prepucial en ninos con y sin hipospadias distal. METODOS: Treinta ninos con hipospadias distal fueron evaluados. 15 ninos recibieron una circuncision electiva en un periodo de 18 meses y sirvieron de grupo control. La presencia de RA y RE-beta en la piel prepucial se investigo por inmunohistoquimica. RESULTADOS: El porcentaje de expresion del receptor androgenico en celulas epiteliales fue de 18,9+/-27,3% en el grupo hipospadias y 3,3+/- 5,3% en el grupo de circuncision. La diferencia entre ambos grupos fue significativo ($p=0,034$). En las celulas estromales, 19,5+/- 26,8% en el grupo hipospadias y 2,6+/- 4,4% en el grupo circuncision fueron positivos para el RA ($p=0,004$). En el grupo de hipospadias, un porcentaje mas elevado de expresion de RE-beta se evidencio en comparacion al grupo control (24+/-24,5% y 1,3+/-1,1, respectivamente, el porcentaje de celulas epiteliales con RE-beta fue superior en el grupo hipospadias que en el grupo control; los valores respectivos fueron 6,8+/-10,1% y 0,9+/-1,3% ($p<0,0001$)). CONCLUSION: En este estudio se sugiere que la expresion de RA y RE fueron superiores en el grupo con hipospadias que en piel prepucial normal. Se requieren mas estudios para determinar el significado de esta expresion.; Language: Spanish

Year of Publication

2020

19.

Update on Pregnancy after Heart Transplantation.

Punnoose L., Coscia L.A., Kliniewski D., Constantinescu S., Moritz M.J.

Embase

The Journal of heart and lung transplantation : the official publication of the International Society for Heart Transplantation. S. 39 (4) (pp S142-S143), 2020. Date of Publication: 01 Apr 2020.

[Article]

AN: 631927856

PURPOSE: The purpose of this study was to describe 171 pregnancies in 100 heart transplant recipients.

METHOD(S): Data regarding pregnancies occurring between 1987 and 2018 were collected by the Transplant Pregnancy Registry International (TPR) via questionnaires, telephone interviews, and medical records.

RESULT(S): The mean age at first transplant was 20+/-8.5 yrs. The transplant to conception interval was 6.8+/-10 yrs (range 0.15-26 yrs) and 37% of the pregnancies were unplanned. Immunosuppression was calcineurin inhibitor-based with 20% exposed to a mycophenolic acid product (MPA). Comorbid conditions during pregnancy included: hypertension 46%, preeclampsia 29%, and diabetes requiring insulin 7%. Rejection occurred during 14 pregnancies (8%) and within 3 months post-partum in 11 pregnancies. Graft loss within 2 years of delivery occurred in 4 recipients; 1 recipient was successfully re-transplanted. Pregnancy outcomes (n=177 includes multiple births) included: live births 67%, miscarriages 25% (46% with MPA exposure), terminations 5%, ectopic 1% and stillbirths 1%. Of the 119 newborn, mean gestational age was 36.1+/-3.4 wks and mean birth weight was 2568+/-701 g. Birth defects were reported in 10 children and included: duodenal atresia, AV canal defect, Tetralogy of Fallot (MPA exposure); facial deformities (MPA exposure), laryngomalacia (MPA exposure), cystic hygroma, vermian hypoplasia of the cerebellum, hypospadias, undescended testicle, pectus excavatum, hydronephrosis, and tongue-tie. Seven additional children inherited their mother's cardiac disease; 4 children have received a heart transplant. At last follow-up, mean 8.2 +/- 6.3 yrs, 33 recipients had died (average age of their 36 children at time of maternal death was 10.2 +/- 6.1 yrs), 7 had reduced cardiac function, 8 unknown, and 52 recipients reported adequate transplant function.

CONCLUSION(S): This is the largest reported series of pregnancies in heart transplant recipients to date. The majority of pregnancies reported are successful. MPA exposure presents significant concerns. Pre-pregnancy counseling should include discussion of inheritable cardiac conditions, MPA avoidance, risk of rejection/graft dysfunction, and long-term maternal survival.

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Institution

(Punnoose) Division of Cardiovascular Medicine, Vanderbilt University School of Medicine, TN, Nashville, United States (Coscia, Kliniewski) Transplant Pregnancy Registry International, Philadelphia, United States

(Constantinescu) Medicine, Lewis Katz School of Medicine, at Temple University, Philadelphia, United States

(Moritz) Surgery, Lehigh Valley Health Network, Allentown, United States

Publisher

NLM (Medline)

Year of Publication

2020

20.

Outcomes of urinary tract abnormalities diagnosed by the routine third trimester scan.

Bakalis S., Cao K., Graham R., Cuckow P., Johal N., Winyard P., Pandya P., Desai D.

Embase

European Journal of Obstetrics and Gynecology and Reproductive Biology. 250 (pp 150-154), 2020. Date of Publication: July 2020.

[Article]

AN: 2005916577

Objective: The aim of this study is to determine the incidence of congenital abnormalities of the kidneys and urinary tract (CAKUT) detected for the first time in an unselected population undergoing a routine third trimester scan between 30-34 week's gestation.

Method(s): This was a retrospective analysis of 8562 routine third trimester ultrasound scans during which the fetal anatomy was evaluated, and, any structural abnormalities detected, recorded onto a fetal database which was subsequently analysed for CAKUT. All postnatal records of antenatally diagnosed CAKUT were obtained and analysed for diagnosis and management.

Result(s): There were 26 cases of urological abnormalities detected for the first time in the third trimester. The most frequent abnormality was unilateral renal pelvis dilatation (73%). Postnatal ultrasound confirmed abnormalities in 19 (73%) newborns, with two (8%) resolving antenatally and four (15%) postnatally. The overall incidence of new CAKUT detected by the third trimester scan was 0.22% (19/8562) with a male to female ratio of 1:1.6. Four patients required surgery, two received cystoscopic injection of Deflux with circumcision, one received cystoscopic valve ablation and one patient received a staged hypospadias repair.

Conclusion(s): Routine third trimester scanning is already performed in many countries with proposed benefits primarily directed towards the monitoring of fetal growth and late pregnancy malpresentation. For healthcare systems that still utilize two routine scans, debate is ongoing as to the value of introducing a routine third trimester scan. The ability to detect additional and potentially missed CAKUT is a further benefit, which in isolation is likely of too small an impact to merit implementation. However, the combination of fetal structural assessment, growth velocity monitoring and fetal presentation evaluation presents a strong case for inclusion in an antenatal screening program. The findings of this study highlight the importance of a detailed fetal structural evaluation at each antenatal ultrasound scan.

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Status

Embase

Author NameID

Bakalis, Spyros; ORCID: <http://orcid.org/0000-0002-6285-3346>

Institution

(Bakalis) Department of Obstetrics, Fetal and Maternal Medicine, Guy's and St Thomas NHS Trust, London, United Kingdom (Cao, Cuckow, Johal, Desai) Department of Urology, Great

Ormond Street Hospital for Children, London, United Kingdom

(Graham) Department of Obstetrics and Gynaecology, Homerton University Hospital, London, United Kingdom

(Winyard) Department of Nephrology, Great Ormond Street Hospital for Children, London, United Kingdom

(Pandya) Fetal Medicine Unit, University College London Hospital, London, United Kingdom

Publisher

Elsevier Ireland Ltd

Year of Publication

2020

21.

Evaluating the anxiety levels of parents of children undergoing hypospadias surgery.

Hipospadias cerrahisi geciren cocuklarin ebeveynlerinin kaygi duzeyinin degerlendirilmesi

<Hipospadias cerrahisi geciren cocuklarin ebeveynlerinin kaygi duzeyinin degerlendirilmesi.>

Erdem A.O., Dogan B., Memis C.O., Sevincok L., Yazici M., Ozkisacik S.

Embase

Duzce Medical Journal. 22 (1) (pp 23-28), 2020. Date of Publication: 30 Apr 2020.

[Article]

AN: 2004437509

Aim: Hypospadias is an anomaly in which urethral meatus is located more proximally. Its treatment is surgery. Children undergoing hypospadias surgery are much more prone to

emotional disorders than the other surgical procedures. In this study, it was aimed to evaluate the emotional and temperament characteristics of the parents of children undergoing hypospadias surgery and normally circumcised.

Material(s) and Method(s): All voluntary parents of children with hypospadias surgery and of children with normal circumcision were included in this study. A semi-structured sociodemographic data form, The Temperament Evaluation of Memphis, Pisa, Paris and San Diego Autoquestionnaire (TEMPS-A), State-Trait Anxiety Inventory (STAI) I-II, and Beck Depression Scale (BDS) were used to collect data from these parents.

Result(s): There was no significant difference between the parents in terms of sociodemographic and clinical characteristics. When the parents of children with hypospadias and normal circumcision were compared in terms of TEMPS-A temperament scale scores, STAI total and I-II subscale scores, and BDS scores, no statistically significant difference was found.

Conclusion(s): Hypospadias is a complex biological development defect. In men who underwent hypospadias surgery, sexual dysfunctions that develop in further period usually develop on psychological grounds. Parents' anxiety levels should be taken into consideration and everyone should act together in management process of hypospadias, operation of which is planned in early period and may have many effects in the future. We think that the study we have done with parents in this area may guide to other studies.,.

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Status

In-Process

Author NameID

Erdem, Ali Onur; ORCID: <http://orcid.org/0000-0002-9584-4200> Yazici, Mesut; ORCID: <http://orcid.org/0000-0003-1960-7825>

Ozkisacik, Sezen; ORCID: <http://orcid.org/0000-0003-3302-727X>

Dogan, Bilge; ORCID: <http://orcid.org/0000-0001-7895-9738>

Memis, Cagdas Oyku; ORCID: <http://orcid.org/0000-0001-6777-4172>

Sevincok, Levent; ORCID: <http://orcid.org/0000-0003-3770-8881>

Institution

(Erdem, Yazici, Ozkisacik) Adnan Menderes University, Faculty of Medicine, Department of Pediatric Surgery, Aydin, Turkey (Dogan, Memis, Sevincok) Adnan Menderes University, Faculty of Medicine, Department of Psychiatry, Aydin, Turkey

Publisher

Duzce University Medical School (E-mail: duzcetipdergisi@gmail.com)

Year of Publication

2020

22.

Bladder Herniation as an Auto-Augmentation Technique in Bladder Exstrophy: Initial Experience in Patients with Small Bladder Plate.

Sabetkish S., Kajbafzadeh A.-M.

Embase

Urology. 139 (pp 151-155), 2020. Date of Publication: May 2020.

[Article]

AN: 2005279598

Objective: To present our long-term experience of bladder plate herniation technique in patients with bladder exstrophy epispadias complex (BEEC) and inadequate bladder plate.

Method(s): Ten BEEC patients with inadequate bladder plates were referred. The bladder underlying fascia was opened and the exstrophic bladder was fixed above the peritoneal cavity to herniate the peritoneal contents beneath the bladder plate so that the abdominal pressure would be directly transferred to the posterior bladder wall; causing gradual bladder expansion and auto-augmentation. In 5 patients, the inguinal hernia was fixed to increase the pressure transferred to the exstrophic bladder. The bladder capacity was measured while the patient was crying and when the bladder was enlarged. Cystometry and voiding cystourethrogram were performed before the application of this technique and during the next 6 to 8 months, to determine the bladder capacity for further primary bladder closure.

Result(s): The bladder was enlarged during straining/crying without any complications. The average bladder capacity was increased about 2.5 to 3 times after 8 months of follow-up so that it was suitable for undergoing primary closure. None of the children needed bladder augmentation following the single-stage total BEEC reconstruction.

Conclusion(s): This technique seems to be safe, effective, and feasible in patients with small-sized bladder and may be performed before the primary closure to increase the success rate. This technique may be effective in increasing the bladder capacity for staged bladder closure and bladder neck reconstruction without further need for bladder augmentation.

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Status

Embase

Institution

(Sabetkish, Kajbafzadeh) Pediatric Urology and Regenerative Medicine Research Center,
Section of Tissue Engineering and Stem Cells Therapy, Children's Hospital Medical Center,
Tehran University of Medical Sciences, Tehran, Iran, Islamic Republic of

Publisher

Elsevier Inc. (E-mail: usjcs@elsevier.com)

Year of Publication

2020

23.

Psychosexual Outcome, Sexual Function, and Long-Term Satisfaction of Adolescent and Young Adult Men After Childhood Hypospadias Repair.

Tack L.J.W., Springer A., Riedl S., Tonnhofer U., Weninger J., Hiess M., Van Laecke E., Hoebeke P., Spinoit A.-F., Cools M., Van Hoecke E.

Embase

Journal of Sexual Medicine. (no pagination), 2020. Date of Publication: 2020.

[Article]

AN: 2005935018

Background: The psychosexual outcome in adolescents and young adults (AYA) men born with hypospadias is precarious. However, the factors responsible for impaired outcome in some AYA men have been understudied.

Aim(s): To explore the outcome after hypospadias repair in childhood of AYA men aged 16-21 years and examine their opinion and their parents' opinion about this type of surgery.

Method(s): Cross-sectional assessment of 193 AYA men born with hypospadias and 50 male controls was performed. Questionnaires such as the Decision Regret Scale, Pediatric Penile Perception Score, Sexual Quality of Life-Male, International Index of Erectile Function, and a custom-made questionnaire were used. The Decision Regret Scale and a custom-made questionnaire were also completed by the participants' parents. Physical examination including Hypospadias Objective Penile Evaluation and measuring stretched penile length was performed.

Outcome(s): This study reports the psychosexual functioning (ie, social, relational, and sexual), erectile and sexual function after childhood hypospadias repair, using ad hoc measures. In addition, the opinion about hypospadias repair of patients and their parents is represented.

Result(s): The number of surgeries and satisfaction regarding penile appearance were the most important factors associated with the opinion on hypospadias repair and the psychosexual outcome. Most AYA men were more satisfied with their penile appearance than the physician. 80% of men were satisfied with having had a childhood hypospadias repair, even though they had not been able to consent to surgery themselves. Erectile and ejaculation problems were mild and seen in approximately 10% of the population. Clinical Implications: Based on our data, deferring hypospadias repair until the patient can decide himself is not warranted. However, physicians who accept a suboptimal esthetic outcome and withdraw from repeated surgery may contribute importantly to the patient's well-being, especially in proximal forms of hypospadias. Strengths & Limitations: This is one of the rare studies addressing the AYA's psychosexual outcome after childhood hypospadias repair. Strengths include the combination of clinical and psychosexual data from a very large cohort of men and their parents to provide a more holistic view. By entering this study, participants might have a different comfort level regarding their sexuality or have a different body image than the overall population of young men. Conclusion(s): Uncomplicated hypospadias surgery results in equal psychosexual outcome as controls and in high satisfaction rates; multiple surgeries are a risk factor for poorer outcomes. 80% of men are satisfied with childhood hypospadias repair. Tack LJW, Springer A, Riedl S, et al. Psychosexual Outcome, Sexual Function, and Long-Term Satisfaction of Adolescent and Young Adult Men After Childhood Hypospadias Repair. J Sex Med 2020;XX:XXX-XXX.

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32444342 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=32444342>]

Status

Article-in-Press

Institution

(Tack, Cools) Division of Paediatric Endocrinology, Department of Internal Medicine and Paediatrics, Ghent University Hospital, Ghent University, Ghent, Belgium (Springer, Tonnhofer, Weninger) Department of Pediatric Surgery, Medical University of Vienna, Vienna, Austria (Riedl) Department of Pediatric Pulmonology, Allergology and Endocrinology, Medical University of Vienna, Vienna, Austria

(Riedl) Department of Pediatrics, St Anna Children's Hospital, Medical University of Vienna, Vienna, Austria

(Hiess) Department of Urology, Medical University of Vienna, Vienna, Austria

(Van Laecke, Hoebeke, Spinoit) Division of Paediatric Urology, Department of Urology, Ghent University Hospital, Ghent University, Ghent, Belgium

(Van Hoecke) Department of Pediatric Psychology, Ghent University Hospital, Ghent, Belgium

Publisher

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Year of Publication
2020

24.

Six novel Mutation analysis of the androgen receptor gene in 17 Chinese patients with androgen insensitivity syndrome.

Jiang X., Teng Y., Chen X., Liang N., Li Z., Liang D., Wu L.

Embase

Clinica Chimica Acta. 506 (pp 180-186), 2020. Date of Publication: July 2020.

[Article]

AN: 2005458291

Background: Androgen insensitivity syndrome (AIS) is the most common type of 46, XY disorders of sex development (DSD), with a wide range of clinical heterogeneity, from male infertility, hypospadias to completely normal female external genitalia. Mutation of the androgen receptor (AR) gene on the X chromosome (Xq11.2q12) is the main cause of AIS.

Method(s): By phenotype evaluation, hormone test, ultrasound scan and G-banding karyotype, 17 unrelated Chinese patients were clinical diagnosed with AIS. Sanger sequencing of the AR was performed in these 17 patients. Functional studies were carried out for the novel mutations.

Result(s): We identified 16 mutations in all patients, including six novel mutations (Q59*, F171Sfs*4, E204*, G209E, I870T, *921R). It is the first time that a stop codon mutation (*921R) in AR has been identified. Expression and nuclear localization analysis showed the *921R mutation caused an elongated abnormal polypeptide chain of the AR protein, and the abnormal protein could not be transported to the nucleus to stimulate the expression of downstream genes after androgenic treatment. Expression analysis showed the protein level of G209E mutation was obviously decreased.

Conclusion(s): Our study expands the spectrum of AR mutations and could provide evidence for the genetic and reproductive counseling of families with AIS. All of these findings broadened the mutation spectrum of AR, which were significantly valuable for patient gender assignment, genetic counseling and the clinical and psychological management.

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Status

Embase

Author NameID

Li, Zhuo; ORCID: <http://orcid.org/0000-0001-6010-6139> Wu, Lingqian; ORCID:
<http://orcid.org/0000-0002-7070-5896>

Institution

(Jiang, Chen, Liang, Li, Liang, Wu) Center for Medical Genetics & Hunan Key Laboratory of
Medical Genetics, School of Life Sciences, Central South University, Changsha, Hunan 410078,
China (Teng, Liang, Wu) Hunan Jiahui Genetics Hospital, Changsha, Hunan 410078, China

Publisher

Elsevier B.V.

Year of Publication

2020

25.

Female Authorship Publishing Trends and Forecasting in Pediatric Urology: Are We Closer to
Gender Equality?.

Rickard M., Hannick J.H., Blais A.-S., Wang J., Santos J.D., Lorenzo A.J.

Embase

Urology. 139 (pp 141-150), 2020. Date of Publication: May 2020.

[Article]

AN: 2005190021

OBJECTIVE: To review the literature of 5 pediatric urology topics and conduct gender based and
forecasting analyses of first and corresponding authors.

METHOD(S): A PubMed search was performed for hypospadias, hydronephrosis, vesicoureteral
reflux, bladder and bowel dysfunction, and cryptorchidism over 3 decades from 1990 to 2019.
The 50 most relevant "best match" papers from each decade were extracted by topic. Author
gender, specialty, and advanced degrees, along with journal and publication variables were
collected. Forecasting analyses were conducted through the Holt-Winters method.

RESULT(S): Among 750 papers analyzed, 78% of corresponding and 70% of first authors were
male. A significant upward trend was observed for female-authored publications in both first and

corresponding positions over time ($P < .01$). Forecasting analyses predicted a continuing upward trend for female corresponding (55%) and first authors (83%) by 2049. Most studies originated from pediatric urology (59%), followed by pediatric surgery (9%) and endocrinology/genetics (5%). Papers focused in The Journal of Urology (30%) with the majority originating from the United States (38%). Most were retrospective (44%) and discussed medical (54%) versus surgical management (20%).

CONCLUSION(S): The majority of pediatric urology literature has been generated by male authors. A persistent, rising trend in female authorship across all examined pediatric urology topics was noted. These encouraging findings are projected to continue to increase in the future, suggesting a movement toward equal and fair gender representation in authorship in pediatric urology.

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Embase

Institution

(Rickard, Hannick, Blais, Wang, Santos, Lorenzo) Division of Urology, The Hospital for Sick Children, Toronto, Ontario, Canada (Hannick) Division of Pediatric Urology, UH Rainbow Babies and Children's Hospital, Cleveland, OH, United States

(Blais) Division of Urology, Centre Hospitalier Universitaire de Quebec, Quebec City, Quebec, Canada

(Wang) Department of Statistical Sciences, University of Toronto, Toronto, Ontario, Canada

Publisher

Elsevier Inc. (E-mail: usjcs@elsevier.com)

Year of Publication

2020

26.

Ventral penile curvature estimation using an app.

Villanueva C.A.

Embase

Journal of Pediatric Urology. (no pagination), 2020. Date of Publication: 2020.

[Article]

AN: 2005899302

Introduction and Objective: The assessment of penile curvature is a key component of hypospadias surgery, as it often determines if a 1 or 2-stage procedure should be done. The objective of this study was to test the accuracy of penile curvature measurements using an App-based method among non-urologists.

Method(s): Lateral pictures of six plastic 3-dimensional penile models representing each decile of curvature from 20 to 70degree were obtained and stored on the research project iPad. Each picture was labeled with a name (i.e. Jim). Non-urology medical professionals were recruited to estimate the curvature of the six pictures with an App-based method consisting of the Photoblend Pro App and 9 provided graded 2-dimensional penile curvature images (representing each decile of curvature from 10 to 90degree). Curvature estimations were done by merging the picture of the 3-dimensional penile model with one of the 9 provided graded 2-dimensional penile curvature images inside the App in an iterative process until the penile model picture matched the graded image (see figure). A research associate taught the research subjects the App-based method and then the subjects were asked to estimate the ventral penile curvature of each of the six penile model pictures. Measurement error was calculated as the absolute difference in between the measured value and the true value for the 6 models. A comparison was then made with previous research where pediatric urologists used either a goniometer or unaided visual inspection (UVI) to measure the ventral curvature of the same plastic 3-dimensional penile models used for the pictures of this study.

Result(s): Twenty-one subjects were recruited, and all completed the study. Mean errors using the App ranged from 1.9degree to 7.1degree, compared to 6.5degree-15degree for UVI and 4.4degree-15.9degree for goniometry. The median error for the App was 0degree compared to 5-10degree for both UVI and goniometry. Mean errors were significantly lower ($p < 0.05$) when using the app compared to UVI/goniometry for all except the 30degree and 50degree models. Assuming patients with VC $\leq$ 30degree would have had a one stage repair versus a 2-stage repair if curvature was >30 degree, the number of measurements that could have resulted in the unintended operation was calculated. There was a statistically significant difference in number of potential unintended surgeries in between App (17%) versus UVI + Goniometer (37%) ($p = 0.0133$).

Conclusion(s): This pilot study demonstrated better penile curvature estimations using the App compared to the two most common methods currently used by pediatric urologists. Plastic models provide an avenue to test and compare penile curvature measurement techniques.[Formula presented]

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Institution

(Villanueva) Phoenix Children's Hospital, 1920 E. Cambridge Ave., Ste. 302, Phoenix, AZ 85006,
United States

Publisher

Elsevier Ltd

Year of Publication

2020

27.

Cosmetic and functional outcome for the use of stitch by stitch technique in hypospadias surgery; results of 235 patients.

Elmoghazy H., Saber M., Mamdoh A., Rashed E., Badawy A.A., Riayd A.M.

Embase

Journal of Pediatric Urology. (no pagination), 2020. Date of Publication: 2020.

[Article]

AN: 2005780936

Introduction: One of the essential goals of hypospadias surgery is to create a cosmetically and functionally accepted penis with a near-normal looking glans and external meatus. An objective: We developed our technique stitch by stitch to allow glans closure in a conical manner with centrally located external urethral meatus in hypospadias repair using onlay flap. Study design (patient and methods): A total of 235 hypospadias male patients (0.5-30 years old) were included in the study. All of them treated by flap urethroplasty techniques and meatoglanuloplasty using stitch by stitch technique. Operative technique (described in details by illustrations); starts with penile degloving, preparation of the flap (Site of hypospadiac meatus is determined after degloving). Creation of a narrow strip of the urethral plate with wide glanular wings is done using bilateral incisions which are extended to the tip of the glans. The width of the urethral plate is about 3 mm then we suture the flap to the narrow urethral plate in two layers in continuous manner using vicryl 6/0 over a suitable plastic catheter. Triangular (V shape) area of the distal

part of the flap is excised at the midline. After this we anastomose the flap to the tip of the glans. The glans appears opened in the midline as inverted V shape. Closure of the glans is finished in the midline using three mattress interrupted sutures with another three simple stitches are used in between.

Result(s): From February 2007 to December 2016; 235 hypospadias patients fulfilled criteria and were included in our prospective cohort study table (1).

Discussion(s): The meatoplasty technique and stitch-by-stitch glans closure were used in all patients. Excellent results were achieved with an overall degree of satisfaction of 87% in patients older than six years (based on the Pediatric Penile Perception Score), and 84% in patients younger than six years. The complication rate was low at 8.5% and was related to a failure of the flap technique. Many surgical techniques have been described for glans closure and meatoplasty with variable outcomes, but none of them fits all patients with hypospadias; each procedure has its limitations and demands specific selection criteria. Limitations of this study come from a relatively short follow-up, and a single surgeon experience that needs to be reproduced by other surgeons.

Conclusion(s): Meatoglanuloplasty using stitch-by-stitch technique provides satisfactory cosmetic and functional results in hypospadias patients. [Table presented]

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Status

Article-in-Press

Institution

(Elmoghazy, Saber, Mamdoh, Rashed, Badawy, Riayd) Department of Urology, Sohag University Hospital, Sohag, Egypt

Publisher

Elsevier Ltd

Year of Publication

2020

28.

Severity-Dependent Profile of the Metabolome in Hypospadias.

Pineyro-Ruiz C., Chorna N.E., Perez-Brayfield M.R., Jorge J.C.

Embase

Frontiers in Pediatrics. 8 (no pagination), 2020. Article Number: 202. Date of Publication: 24 Apr 2020.

[Article]

AN: 631679580

Background & Objective: Hypospadias, characterized by the displacement of the opening of the urethra at any point in the medial-ventral side of the penis, is classified upon severity as mild (Type I) and severe (Type II and Type III) hypospadias. Hypospadias' etiology is idiopathic in the majority of cases, and underlying causes seem of multifactorial origin. Studies regarding genetic variants support this notion. It is unknown whether downstream gene products fit this profile. This study evaluated the metabolome of hypospadias by using the emerging technology of metabolomics in the search for distinct cellular processes associated with hypospadias' etiology according to the severity of this congenital urogenital condition.

Method(s): Foreskin samples were collected during urethroplasty from boys with Type I, II, and III hypospadias or undergoing elective circumcision (N = 28) between 5 and 28 months of age.

Samples were processed and submitted to gas chromatography-mass spectrometry (GC/MS).

MetaboloAnalyst (<http://www.metaboanalyst.ca/>) online platform was used for bioinformatic analyses.

Result(s): Thirty-five metabolites across experimental groups were identified by GC/MS. Principal component analysis (PCA) and partial least squares-discriminant analysis (PLS-DA) showed that the metabolome of Type II and Type III hypospadias patients differs from the metabolome of Type I hypospadias and control patients. Of those 35, 10 amino acids were found in significantly low concentrations in severe hypospadias: aspartate, glutamate, glycine, isoleucine, leucine, lysine, methionine, phenylalanine, proline, and tyrosine. A high concentration of the amino acid lysine was detected in mild hypospadias.

Conclusion(s): The observed downregulation of specific amino acids in severe hypospadias provides alternative routes for future research aiming to identify disrupted networks and pathways while considering the severity of hypospadias.

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Status

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Institution

(Pineyro-Ruiz, Jorge) Department of Anatomy and Neurobiology, School of Medicine, University of Puerto Rico, San Juan, United States (Chorna) Department of Biochemistry, PR-INBRE Metabolomics Research Core, University of Puerto Rico, Medical Sciences Campus, San Juan, United States

(Perez-Brayfield) Department of Surgery, Urology Section, School of Medicine, University of Puerto Rico, San Juan, United States

Publisher

Frontiers Media S.A. (E-mail: info@frontiersin.org)

Year of Publication

2020

29.

Preoperative topical estrogen treatment versus placebo in 244 children with midshaft and posterior hypospadias.

Gorduza D., Plotton I., Remontet L., Gay C.-L., El Jani M., Cheikhelard A., Blanc T., Elghoneimi A., Leclair M.-D., Roy P., Pirot F., Mimouni Y., Gaillard S., Chatelain P., Morel Y., Kassai B., Mouriquand P.

Embase

The Journal of clinical endocrinology and metabolism. (no pagination), 2020. Date of Publication: 09 May 2020.

[Article]

AN: 631700305

PURPOSE: Urethral fistula and dehiscence are common after hypospadias surgery. Preoperative androgens have been considered to reduce these complications although not evidence-based. Dermatologists have reported the benefits of topical Estrogens on skin healing. We investigated whether the preoperative use of topical Promestriene could reduce healing complications in hypospadias surgery. Our primary objective was to demonstrate a reduction of healing complications with Promestriene versus placebo. Impact on reoperations and other complications, clinical tolerance, bone growth and biological systemic effects of the treatment were also considered.

METHOD(S): We conducted a prospective, randomized, placebo-controlled, double-blind, parallel group trial, between 2011 and 2015, in 4 French centers. One-stage Transverse Preputial Island Flap Urethroplasty (Onlay urethroplasty) was selected for severe hypospadias. Promestriene or placebo was applied on the penis for 2 months prior to surgery. The primary outcome was the presence of postoperative urethral fistula or dehiscence in the first year post-surgery. For safety reasons, hormonal and anatomical screenings were performed.

RESULT(S): Out of 241 patients who received surgery, 122 patients were randomized to receive placebo and 119 Promestriene. The primary outcome was unavailable for 11 patients. Healing complications were assessed at 16.4% (19/116) in the placebo versus 14.9% (17/114) in the Promestriene arm and the odds ratio adjusted on centre was 0.93 [0.45; 1.94] (p=0.86).

CONCLUSIONS AND RELEVANCE: Although we observed an overall lower risk of complications compared to previous publications, post-surgery complications were not different between promestriene and placebo, because of a lack of power of the study or the inefficacy of Promestriene.

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PMID

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Institution

(Gorduja, Plotton, Gay, Chatelain, Morel, Mouriquand) Centre de Reference Maladies Rares
Developpement Genital : du Foetus a l'Adulte (Gorduja, Mouriquand) Service de Chirurgie Uro-
Viscerale de l'Enfant-Hopital Mere Enfant, Hospices Civils de Lyon

(Plotton, Morel) Laboratoire d'Hormonologie d'Endocrinologie Moleculaire et des Maladies Rares,
INSERM, Groupement Hospitalier Est, Hospices Civils de Lyon, Bron, France

(Remontet, El Jani, Roy, Mimouni, Gaillard, Kassai) Universite de Lyon, F-69000, Lyon, France ;
Universite Lyon 1 ; CNRS, UMR, Laboratoire de Biometrie et Biologie Evolutive, Villeurbanne,
France

(Remontet, Roy) Hospices Civils de Lyon, Service de Biostatistiques, Lyon, France

(Gay, Chatelain) Service d'Endocrinologie Pediatrique, Hopital Mere-Enfant, Centre Hospitalo-
Universitaire de Lyon, Bron Cedex, France

(El Jani, Mimouni, Gaillard, Kassai) Hospices Civils de Lyon, EPICIME-CIC 1407 de Lyon,
Inserm, Department of clinical epidemiology, CHU-Lyon, Bron, France

(Cheikhelard, Blanc) Service de chirurgie viscerale et urologie pediatrique, APHP, Hopital
Necker, Paris, France ; Universite Sorbonne Paris cite, Paris, France

(Elghoneimi) Service de chirurgie viscerale et urologie pediatrique, Centre de Reference des
Maladies Endocriniennes Rares de la Croissance, Hopital Robert Debre, APHP, Universite Paris
Diderot, Paris, France

(Leclair) Service de chirurgie pediatrique, CHU de Nantes, 38 boulevard Jean Monnet, Nantes,
France

(Piot) Service pharmaceutique, Plateforme FRIPHARM, Groupement Hospitalier Edouard
Herriot, Lyon Cedex, Place d'Arsonval, France

(Piot) Laboratoire de Recherche et Developpement de Pharmacie Galenique Industrielle,
Plateforme FRIPHARM, Faculte de Pharmacie, Laboratoire de Biologie Tissulaire et Ingenierie

Therapeutique - UMR, Lyon Cedex, Universite Claude Bernard Lyon 1 ,8 Avenue Rockefeller,
France

(Mouriquand) Universite Claude-Bernard-Lyon 1-France

Publisher

NLM (Medline)

Year of Publication

2020

30.

Delaying reclosure of bladder exstrophy leads to gradual decline in bladder capacity.

Wu W.J., Maruf M., Harris K.T., Manyevitch R., Patel H.D., Di Carlo H.N., Gearhart J.P.

Embase

Journal of Pediatric Urology. (no pagination), 2020. Date of Publication: 2020.

[Article]

AN: 2005680779

Introduction: After unsuccessful repair of bladder exstrophy, when to repeat surgical intervention is unclear. One must balance time required for tissue healing with the damaging effects of an exposed urothelium to the environment.

Objective(s): The authors aim to study whether a relationship exists between bladder growth/capacity and time till eventual successful closure. Study design: An institutional database of exstrophy-epispadias complex patients was queried for failed exstrophy closure with successful repeat reconstruction, at least three consecutive bladder capacity measurements, and measurements obtained at least three months following successful closure. Patients closed successfully in the neonatal period were used as a comparative group. Linear mixed effects models were used to study the effect of time and age on bladder capacity.

Result(s): Forty-seven patients requiring reclosure and 117 who had successful neonatal closures were included. Two models were created. The first linear mixed effects model found that for a given age, the bladder capacity declined approximately 9.6 mL per year ($p = 0.016$). The second model found that when time to successful closure was grouped by quartiles, compared to neonates, those in the fourth quartile had significantly decreased bladder capacity of 28.8 cc ($p = 0.042$). An interaction model comparing neonates and those requiring reclosure did not

demonstrate a significant change in bladder growth rate ($p = 0.098$). A model stratified by quartiles similarly did not find any significant impact to bladder growth rate.

Discussion(s): From the general linear mixed effects models, the authors conclude when compared to neonates, (1) there was an approximate 9.6 cc loss of total bladder capacity per year taken until successful closure, and that (2) those who were delayed the longest had the most significant difference in bladder capacity. This study required stricter inclusion criteria compared to previous publications, and therefore the conclusions that can be drawn regarding bladder growth rates may be more reliable. Future studies will examine the effects of delayed closure on the bladder at the cellular level.

Conclusion(s): There is a demonstrable significant impact on overall bladder capacity with increasing delay to successful reclosure. One should be cautious when prolonging reconstruction of the bladder as these data demonstrate a time dependent decline in overall capacity.[Formula presented]

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Status

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Author NameID

Harris, Kelly T.; ORCID: <http://orcid.org/0000-0002-3868-4423> Patel, Hiten D.; ORCID: <http://orcid.org/0000-0003-4028-010X>

Institution

(Wu, Maruf, Harris, Manyevitch, Patel, Di Carlo, Gearhart) Robert D. Jeffs Division of Pediatric Urology, Brady Urological Institute, The Johns Hopkins Medical Institutions, Baltimore, MD, United States

Publisher

Elsevier Ltd

Year of Publication

2020

31.

Challenges in pediatric urologic practice: a lifelong view.

Wiener J.S., Huck N., Blais A.-S., Rickard M., Lorenzo A., Di Carlo H.N.M., Mueller M.G., Stein R.
Embase

World journal of urology. (no pagination), 2020. Date of Publication: 23 Apr 2020.

[Article]

AN: 631624576

The role of the pediatric urologic surgeon does not end with initial reconstructive surgery. Many of the congenital anomalies encountered require multiple staged operations while others may not involve further surgery but require a life-long follow-up and often revisions. Management of most of these disorders must extend into and through adolescence before transitioning these patients to adult colleagues. The primary goal of management of all congenital uropathies is protection and/or reversal of renal insult. For posterior urethral valves, in particular, avoidance of end-stage renal failure may not be possible in severe cases due to the congenital nephropathy but usually can be prolonged. Likewise, prevention or minimization of urinary tract infections is important for overall health and eventual renal function. Attainment of urinary continence is an important goal for most with a proven positive impact on quality of life; however, measures to achieve that goal can require significant efforts for those with neuropathic bladder dysfunction, obstructive uropathies, and bladder exstrophy. A particular challenge is maximizing future self-esteem, sexual function, and reproductive potential for those with genital anomalies such as hypospadias, the bladder exstrophy epispadias complex, prune belly syndrome, and Mullerian anomalies. Few endeavors are rewarding as working with children and their families throughout childhood and adolescence to help them attain these goals, and modern advances have enhanced our ability to get them to adulthood in better physical and mental health than ever before.

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Institution

(Wiener) Division of Urologic Surgery, Duke University Medical Center, Durham, United States

(Huck, Stein) Department of Pediatric, Adolescent and Reconstructive Urology, Medical Faculty

Mannheim, University Medical Center Mannheim, Heidelberg University, Mannheim, Germany

(Blais, Rickard, Lorenzo) Division of Pediatric Urology, Hospital for Sick Children, ON, Toronto, Canada

(Lorenzo) Department of Surgery, University of Toronto, ON, Toronto, Canada

(Di Carlo) James Buchanan Brady Urologic Institute, Johns Hopkins University School of Medicine, MD, Baltimore, United States

(Mueller) Division of Female Pelvic Medicine and Reconstructive Surgery, Departments of Obstetrics & Gynecology and Urology, Feinberg School of Medicine, Northwestern University, Chicago, United States

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Year of Publication
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32.

Stented versus Non-Stented Snodgrass Urethroplasty for Distal Hypospadia Repair.

Almusafer M., Abduljabbar O.H., Buchholz N.

Embase

Urologia Internationalis. 104 (1-2) (pp 156-159), 2020. Date of Publication: 01 Apr 2020.

[Article]

AN: 629904525

Background: Hypospadia is one of the most common congenital anomalies in children. Patients with distal hypospadias can be treated successfully with a tubularized incised plate (TIP) urethroplasty, usually with a postoperative urethral stent to divert urine into the diaper or a urine bag for approximately 1 week. However, these stents have their own morbidity and complications. We therefore tried to determine the safety of distal penile hypospadias repair without the use of a postoperative stent. Patients and Method: Fifty patients with distal penile hypospadias were prospectively assessed from May 2016 to August 2018. All patients underwent Snodgrass urethroplasty by the same surgeon. Half of the patients had a postoperative stent for 1 week. The other half had no stent. Clinical follow-up was over 6 months with an emphasis on possible stent-related complications.

Result(s): Fifty children underwent TIP urethroplasty for distal hypospadia repair. The mean age was 5.9 years (range 2-12). In 25 cases, a stent was removed within 1 week. In the other 25 cases, no postoperative stent was placed. The overall complication rate for the stented group was 48% (n = 12) and for the non-stented group 68% (n = 17), respectively. In the stented group, 1 patient (4%) developed a fistula, whilst there were 2 (8%) in the non-stented group. All fistulas were repaired after 6 months postoperatively. Neourethral stenosis and glans dehiscence occurred in each 1 case (4%) in both groups. Differences were not statistically significant. However, there were significantly more wound infections in the stented group. On the other hand, stents prevented temporary urinary retention which occurred in 2 patients in the non-stented group.

Conclusion(s): Despite the limited number of cases, our study suggests that, all in all, there is no significant difference in severe complication rates regardless whether a postoperative stent is used or not.

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Institution

(Almusafer, Buchholz) U-merge Ltd., London ME71GD, United Kingdom (Almusafer, Abduljabbar) Department of Urology, University of Basrah, Basrah, Iraq

Publisher

S. Karger AG

Year of Publication

2020

33.

Long-term outcomes in primary male epispadias.

Thomas J.S., Shenoy M., Mushtaq I., Wood D.

Embase

Journal of Pediatric Urology. 16 (1) (pp 80.e1-80.e6), 2020. Date of Publication: February 2020.

[Article]

AN: 2004127015

Purpose: The authors present a case series of 30 male patients who presented with primary epispadias between 1989 and 2002 and looked at their long-term outcomes.

Objective(s): (1) What procedures and operations did these patients require following their original surgery? (2) What were their outcomes as adults in terms of continence, cosmesis, and sexual function?

Material(s) and Method(s): Primary male epispadias patients who had gone through transition into Adolescent and Adult Urology services were identified retrospectively from electronic patient records.

Result(s): The authors identified 30 male patients with a median follow-up of 18.5 years. Twenty-four had penopubic epispadias, and six had penile epispadias. All initial surgery took place between 9 and 48 months. Twenty-eight patients needed further surgery over the follow-up period, 26 had surgery for continence, and 12 required revision surgery. At follow-up, 15 patients were continent voiding per urethra, nine patients reported stress leakage with volitional urethral voiding, six patients were using a Mitrofanoff to void, and four of these had an ileocystoplasty. Nineteen patients had documentation on their feelings toward cosmetic outcome; 17 expressed concern. Twenty-one patients had documentation about sexual function; 20 had normal erections with six reporting chordee and nine reporting retrograde ejaculation. No patients were recorded to have fathered any children.

Discussion(s): There are only a few published studies looking at long-term outcomes of genitourinary reconstruction in primary male epispadias and their sexual function in adulthood. The majority of this patient cohort required surgery to improve their continence and had more than one continence procedure. There is limited data on continence outcomes in the literature with small cohorts and rates varying between 40 and 100% at 10-year follow-up. Despite corrective surgery, nearly all the patients were concerned about their genital appearance. Other studies have shown similar outcomes in terms of patient satisfaction and sexual function. The limitations of this work are that the information was gathered retrospectively from the electronic patient record and validated instruments on outcomes were not used.

Conclusion(s): The study outcomes will be useful for clinicians who are counseling patients and parents regarding what to expect as adults. [Table presented]

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Status

Embase

Institution

(Thomas, Mushtaq) Great Ormond Street Hospital for Children NHS Foundation Trust, London, United Kingdom (Thomas, Wood) University College London Hospital NHS Foundation Trust, London, United Kingdom

(Shenoy) Nottingham Children's Hospital, Nottingham, United Kingdom

Publisher

Elsevier Ltd

Year of Publication

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34.

Distal hypospadias repair using the needle point bipolar cutting-coagulation forceps.

Bagnara V., Giammusso B., Castagnetti M., Esposito C., Bianchi A.

Embase

Journal of Pediatric Urology. 16 (1) (pp 69.e1-69.e5), 2020. Date of Publication: February 2020.

[Article]

AN: 2003794212

Introduction: To determine the outcome of distal hypospadias repairs performed using bipolar diathermy (BD) for all the dissection.

Method(s): Retrospective review of 310 patients undergoing distal hypospadias (264 subcoronal/distal penile and 46 coronal/glanular) over a 11-year period. Median age at surgery was 2.0 years (range 9 months-15 years). Caudal anesthesia was performed in all patients. All children underwent an in situ tubularization of the urethral plate, which was combined with midline incision of the plate in 30 (10%). Preputial reconstruction was performed in 303 (98%) patients. BD forceps was used for coagulation and for all the dissection including skin incision, elevation of glans wings, separation of the corpus spongiosum from the corpora cavernosa, and urethral plate incision, when deemed appropriate. The authors assessed surgical complications and cosmetic results. The latter using the hypospadias objective score (HOSE), with a score ≥ 14 considered as acceptable.

Result(s): The average operative time was 70 min. There was no postoperative bleeding or hematomas that required surgical intervention. There were no wound infections or necrosis. Complications occurred in 37 patients (11.9%). Urethral fistula formation was the commonest (n = 18). Postoperative persistent preputial swelling occurred in 3.5% of cases. Two hundred twelve patients (90.6%) had a HOSE score ≥ 14 , and no patient required revision surgery for skin problems after a median follow-up of 8.1 (range 1.2-13.1) years.

Discussion(s): The study is limited by its retrospective nature and by the fact that a number of other pre-operative, intra-operative, and postoperative variables can affect outcome.

Conclusion(s): Bipolar diathermy can be safely used for distal hypospadias repairs. It allowed careful control of intra-operative bleeding and also clear visualization of tissue planes.

Complication rate overall compares favorably with the literature, and cosmetic results were satisfactory. [Table presented]

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Status

Embase

Author NameID

Castagnetti M.; ORCID: <http://orcid.org/0000-0003-1282-1351>

Institution

(Bagnara, Giammusso) Paediatric Surgery Unit, Polyclinic G.B. Morgagni, Catania, Italy

(Castagnetti) Section of Paediatric Urology, Department of Surgical, Oncological and Gastrointestinal Sciences, University Hospital of Padova, Padua, Italy

(Esposito) Paediatric Surgery Unit, University Hospital of Naples, Naples, Italy

(Bianchi) Specialist Paediatric Reconstructive Surgeon, Royal Manchester Children's Hospital, United Kingdom

Publisher

Elsevier Ltd

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35.

Perioperative complications within 30 days of hypospadias surgery: Results from NSQIP-Pediatrics.

Jiang D.D., Chakiryan N.H., Gillis K.A., Acevedo A.M., Austin J.C., Seideman C.A.

Embase

Journal of Pediatric Urology. (no pagination), 2020. Date of Publication: 2020.

[Article]

AN: 2005613287

Introduction: There are no large multi-institutional studies reporting on perioperative complications of hypospadias repairs. We sought to determine perioperative complications of hypospadias repairs from the National Surgical Quality Improvement Program Pediatrics (NSQIP-P) to aid in patient counseling. Study design: This cohort study from 2012 to 2017 was conducted using NSQIP-P database. Pediatric patients undergoing hypospadias surgery were identified and compared based on 4 major categories: distal/midshaft repair, one-stage repair proximal, stage one repair, and stage two repair. Baseline demographics between the four groups and

perioperative parameters were compared. Multivariable logistic regression analysis models including type of repair was used to determine associations with overall complications, infectious complications, and dehiscence.

Discussion(s): There were 11,292 patients identified in the study population. Overall, 78% underwent distal/midshaft hypospadias repair, 12% underwent one-stage proximal repair, 1.4% underwent proximal first stage repair and 9% underwent proximal second stage repair.

Multivariable logistic regression analysis revealed that proximal first stage procedures had similar overall complications to distal/mid repairs but proximal one-stage and proximal second stage procedures were associated with significantly more overall complications, local infectious complications, and dehiscence. Age, race, operative time, prematurity were also independently associated with increased overall complications. As expected, complication rates are higher in those with proximal hypospadias. In staged hypospadias, first stage has a lower complication rate compared to second stage. All complications, especially of infectious and dehiscence are the highest in the one-stage proximal and proximal second stage repairs.

Conclusion(s): We report large multi-institutional analysis of 30-day peri-operative hypospadias repair complications; this information is useful for patient counseling and education. [Table presented]

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Status

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Author NameID

Jiang, Da David; ORCID: <http://orcid.org/0000-0003-3543-491X> Chakiryan, Nicholas H.; ORCID: <http://orcid.org/0000-0001-9263-8957>

Institution

(Jiang, Chakiryan, Gillis, Acevedo, Austin, Seideman) Department of Urology, Oregon Health & Sciences University, Portland, OR, United States (Jiang, Chakiryan, Gillis) O'Brien Research Group, Portland, OR, United States

Publisher

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36.

Risk factors for urethrocutaneous fistula following hypospadias repair surgery in Indonesia.

Duarsa G.W.K., Tirtayasa P.M.W., Daryanto B., Nurhadi P., Renaldo J., Tarmono, Utomo T., Yuri P., Siregar S., Wahyudi I., Situmorang G.R., Ansharullah Palinrungi M.A., Hutasoit Y.I., Hutahaeen A.Y.A., Zulfiqar Y., Sigumonrong Y.H., Mirza H., Rodjani A., Kloping Y.P.

Embase

Journal of Pediatric Urology. (no pagination), 2020. Date of Publication: 2020.

[Article]

AN: 2005727095

Introduction: Hypospadias is one of the most common congenital malformations with a worldwide increasing trend over the years. Despite advancements in hypospadias repair, complications still occur. One of the most common complications of hypospadias repair surgery is Urethrocutaneous fistula. Studies attempting to analyze the association between the complication and risk factors are always beneficial, especially for studies performed in different areas of the world. We hypothesize that several evaluated risk factors among Indonesian hypospadias patients could be associated with the occurrence of urethrocutaneous fistula after the repair procedure.

Objective(s): To determine the risk factors associated with urethrocutaneous fistula after hypospadias repair surgery by collecting and analyzing data obtained from multiple centers in Indonesia.

Material(s) and Method(s): A nationwide, retrospective study with 12 hospitals in Indonesia of children with a diagnosis of hypospadias was conducted. The collected data, taken from patients admitted in 2018, from each center's medical records consisted of patient identity, repair technique used, neourethra length, percutaneous cystostomy, and splint size as independent variables speculated to be possible risk factors correlated to the presence of urethrocutaneous fistulae. Binomial logistic regression analysis was performed using SPSS 21.0 to determine the relationship between urethrocutaneous fistulae as a post-repair complication and possible risk factors.

Result(s): We collected 591 hypospadias cases from 12 centers in 9 cities in Indonesia. Most patients came when they were already at the age of more than four years old (60.4%). The chordee-only and failed urethroplasty groups are excluded from the analysis as they are not classified as true hypospadias. Most repairs were performed by using the Tubular Incised Plate (TIP) with Thiersch-Duplay technique (44.16%). Most of the reconstructed neourethra are 2-3 cm in length (32.13%). The 8 Fr urethral splint (46.41%) was mostly used during the operation. Most surgeons decided not to perform cystostomy throughout the procedure (61.03%) based on

personal preferences. Urethrocuteaneous fistula was found in 80 patients (15.27%) out of the total patients who underwent the surgery. The binomial logistic regression analysis shows that age (OR = 1.398, $p = 0.015$), the decision to not perform cystostomy (OR = 2.963, $p = 0.014$), and splint size (OR = 1.243, $p = 0.023$) are significantly associated ($p < 0.05$) with the development of urethrocuteaneous fistula.

Conclusion(s): Age and splint size are significant risk factors for urethrocuteaneous fistula after hypospadias repair in Indonesia, whereas performing percutaneous cystostomy during the repair decreases the risk for urethrocuteaneous fistula occurrence.

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Status

Article-in-Press

Author NameID

Kloping, Yudhistira Pradnyan; ORCID: <http://orcid.org/0000-0002-7227-8349>

Institution

(Duarsa, Tirtayasa) Division of Urology, Department of Surgery, Faculty of Medicine Universitas

Udayana, Sanglah Hospital, Denpasar, Indonesia (Daryanto, Nurhadi) Department of Urology,

Faculty of Medicine Universitas Brawijaya, Saiful Anwar Hospital, Malang, Indonesia

(Renaldo, Tarmono) Department of Urology, Faculty of Medicine Universitas Airlangga, Soetomo Hospital, Surabaya, Indonesia

(Utomo, Yuri) Department of Urology, Faculty of Medicine Universitas Gadjah Mada, Sardjito Hospital, Yogyakarta, Indonesia

(Siregar) Department of Urology, Faculty of Medicine Universitas Padjajaran, Hasan Sadikin Hospital, Bandung, Indonesia

(Wahyudi, Situmorang, Hutasoit, Hutahaeen, Mirza, Rodjani) Department of Urology, Faculty of Medicine Universitas Indonesia, Cipto Mangunkusumo Hospital, Jakarta, Indonesia

(Ansharullah Palinrungi) Division of Urology, Department of Surgery, Faculty of Medicine Universitas Hasannudin, Wahidin Sudirohusodo Hospital, Makassar, Indonesia

(Zulfiqar) Division of Urology, Department of Surgery, Faculty of Medicine, Universitas Andalas, M.Djamil Hospital, Padang, Indonesia

(Sigumonrong) Department of Urology, Faculty of Medicine, Universitas Sumatera Utara, Adam Malik Hospital, Medan, Indonesia

(Kloping) Medical Doctor, General Practitioner, Medical Faculty of Airlangga University, Surabaya, East Java, Indonesia

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37.

Consulting "Dr. YouTube": an objective evaluation of hypospadias videos on a popular video-sharing website.

Salama A., Panoch J., Bandali E., Carroll A., Wiehe S., Downs S., Cain M.P., Frankel R., Chan K.H.

Embase

Journal of Pediatric Urology. 16 (1) (pp 70.e1-70.e9), 2020. Date of Publication: February 2020. [Article]

AN: 2004559329

Introduction: Parents who make decisions about hypospadias repair for their child may seek information from online platforms such as YouTube.

Objective(s): The purpose of this study is to evaluate the health literacy demand of hypospadias videos on YouTube using the Patient Education Materials Assessment Tool for Audiovisual Materials (PEMAT-A/V). Study design: We performed a YouTube search using the term "hypospadias," limiting results to the first 100 videos. We excluded videos that were <1 min or >20 min and videos that were not in English or did not include subtitles. Two evaluators independently examined videos and determined PEMAT-A/V scores for understandability and actionability (i.e., ability to identify actions the viewer can take). Videos with scores >70% are understandable or actionable. The inter-rater reliability (kappa) and intraclass correlation coefficient (ICC) of PEMAT scores were calculated. Bivariate and multivariable linear regression models assessed the association of video characteristics with respective scores.

Result(s): Of the 100 videos that were identified on YouTube, 47 (47%) were excluded leaving 53 for analysis: 14 were >20 min, 14 were <1 min, 9 had no audio or subtitles, 7 were not in English, 1 was a duplicate, 1 was unrelated to hypospadias, and 1 was deleted at the time of data analysis. Three (5.6%) were understandable (mean score 54.5%, standard deviation (SD) 14.9) and eight (15.1%) were actionable (mean score 21.8%, SD 16.6) (Extended Summary Figure). Kappa values ranged from 0.4 to 1. The ICC's were 0.55 and 0.33 for understandability and actionability, respectively. In the bivariate analysis, mean understandability scores were significantly higher for English language videos ($p = 0.04$), videos with animation ($p = 0.002$), and those produced by industry ($p = 0.02$). In the multivariable analysis, mean understandability scores were significantly higher for "expert testimonial" or "other" video types after adjusting for

graphics type and overall tone ($p = 0.04$). Mean understandability scores were also significantly higher for videos with animation after adjusting for video type and overall tone ($p = 0.01$). Mean actionability scores were significantly higher for videos with a negative tone ($p = 0.01$).

Discussion(s): The vast majority of hypospadias-related YouTube content is not appropriate for users with low health literacy although certain types of videos, such those with animation and expert testimonials, scored higher on understandability than other types.

Conclusion(s): Due to the lack of sufficient online informational content regarding hypospadias, we plan to engage parents of sons with hypospadias in the development of high-quality patient educational materials about hypospadias.[Formula presented]

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Status

Embase

Author NameID

Salama, Amr; ORCID: <http://orcid.org/0000-0003-4107-2919>

Institution

(Salama, Panoche, Bandali, Cain, Chan) Department of Urology, Indiana University School of Medicine, United States (Carroll, Chan) Department of Pediatrics, Center for Pediatric and Adolescent Comparative Effectiveness Research, United States

(Wiehe, Downs) Department of Pediatrics, Children's Health Services Research Center, United States

(Frankel) Department of Medicine, Indiana University School of Medicine, Indianapolis, IN, United States

(Frankel) Cleveland Clinic Learner Institute, Cleveland, OH, United States

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Music Interventions in Pediatric Surgery (The Music under Surgery in Children Study): A Randomized Clinical Trial.

Kuhlmann A.Y.R., Van Rosmalen J., Staals L.M., Keyzer-Dekker C.M.G., Dogger J., De Leeuw T.G., Van Der Toorn F., Jeekel J., Wijnen R.M.H., Van Dijk M.

Embase

Anesthesia and Analgesia. (pp 991-1001), 2020. Date of Publication: 2020.

[Article]

AN: 631587319

BACKGROUND: Perioperative music interventions have been shown to reduce anxiety and pain in adults. This inexpensive, easily applicable intervention could be of benefit to children as well. Our objective was to determine the effects of music interventions on distress, anxiety, and postoperative pain in infants undergoing surgery.

METHOD(S): The Music Under Surgery In Children study was designed as a parallel, single-blind, randomized controlled trial with an a priori formulated hypothesis. Data were collected between August 2015 and October 2016 in a single tertiary care children's hospital. There was a 24-hour follow-up with blind primary outcome assessment. A random sample of 432 eligible 0-3 years of age infants admitted for orchidopexy, hypospadias, or inguinal hernia repair receiving general anesthesia and caudal block were asked for participation. Subjects were assigned to a preoperative music intervention, pre- And intraoperative music intervention, or no music intervention (control) via random allocation using a computer-generated list with the use of opaque envelopes. The main outcome measure was the postoperative level of distress assessed with the COMFORT-Behavior scale, which is an observational scale; furthermore, preoperative level of distress, preoperative anxiety, and physiological measurements such as heart rate (HR) and blood pressure were measured. The trial was registered at the Dutch Trial Register, number NTR5402 (www.trialregister.nl).

RESULT(S): One hundred ninety-five infants with median age 6.9 months (interquartile range, 3.3-11.1) were randomized, 178 of whom were included in the primary analysis. A nonsignificant difference in COMFORT-Behavior scale scores between the pre- And intraoperative music intervention group and control group at 4 hours after surgery was found (mean difference, -1.22; 95% CI, 2.60-0.17; $P = .085$). Additional analysis showed weak nonsignificant evidence for an interaction effect between music exposure and COMFORT-Behavior score at baseline ($P = .027$ with a Bonferroni-adjusted significance level of .025). General linear modeling showed a statistically significantly reduced HR after the preoperative music intervention in the holding area in the combined preoperative music intervention and intraoperative music intervention group compared to the control group ($P = .003$). The differences in HR among the 3 study arms at all time points were not statistically significant ($P = .069$).

CONCLUSION(S): Music interventions do not seem to benefit all young infants undergoing surgery. The potential benefits of music interventions in the preoperative period and in more distressed children warrant further exploration.

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PMID

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Status

Article-in-Press

Institution

(Kuhlmann, Keyzer-Dekker, Wijnen, Van Dijk) Department of Pediatric Surgery, Erasmus University Medical Center, Sophia Children's Hospital, PO Box 2060, Room SK1264, Rotterdam 3000CA, Netherlands (Van Rosmalen) Department of Biostatistics, Erasmus University Medical Center, Rotterdam, Netherlands

(Staals, Dogger, De Leeuw) Departments of Anesthesiology, PO Box 2060, Room SK1264, Rotterdam 3000CA, Netherlands

(Van Der Toorn) Pediatric Urology, Erasmus University Medical Center, Sophia Children's Hospital, Rotterdam, Netherlands

(Jeekel) Departments of Neuroscience, PO Box 2060, Room SK1264, Rotterdam 3000CA, Netherlands

(Van Dijk) Internal Medicine, Erasmus University Medical Center, Rotterdam, Netherlands

Publisher

Lippincott Williams and Wilkins (E-mail: kathiest.clai@apta.org)

Year of Publication

2020

39.

Androgen receptor expression in preputial dartos tissue correlates with physiological androgen exposure in congenital malformations of the penis and in controls.

Tack L.J.W., Praet M., Van Dorpe J., Haid B., Buelens S., Hoebeke P., Van Laecke E., Cools M., Spinoit A.F.

Embase

Journal of Pediatric Urology. 16 (1) (pp 43.e1-43.e8), 2020. Date of Publication: February 2020.

[Article]

AN: 2004075023

Introduction: The androgen receptor (AR) plays an important role in the development of male genitalia, and impaired androgen signalling has been hypothesised to underlie congenital penile malformations (CPM) such as hypospadias. Previous studies exploring the role of AR expression in the development of CPM have yielded conflicting results.

Objective(s): To assess AR expression in human foreskin of boys/men born with hypospadias, buried penis versus controls. Study design: Foreskin samples of 428 boys and men undergoing primary penile surgery (198 controls, 197 hypospadias, and 33 buried penis) were collected between October 2013 and July 2018. AR staining was performed in all samples and semi-quantitatively scored by two researchers independently, using a modified quick score (mQuicks) that assesses the proportion and intensity of AR staining in smooth muscle fibres.

Result(s): The interobserver variability of the mQuicks had a high level of agreement for the total score, as well as for the subscores. Two phases of high AR expression were observed in all groups, the first following the postnatal gonadotropin surge (i.e., mini-puberty) and the second in (pre-) puberty. No differences in AR expression were found in hypospadias or buried penis cases as compared to controls matched for age at time of surgery.

Discussion(s): This study describes the physiological evolution in AR expression in the human foreskin of boys with CPM and explains the cause of the previously reported, conflicting results. Despite the very large cohort, the limitations of this study are the low number of cases younger than six months at the time of surgery and the lack of Tanner stages to correlate with the mQuicks in adolescents.

Conclusion(s): The mQuicks is a straightforward and informative tool to semi-quantitatively assess AR expression in the dartos tissue. In this study, AR expression in human foreskin shows a bimodal distribution in boys with CPM and controls, following physiological androgen exposure. No statistically significant difference in AR expression could be found between both groups. Whether other local mechanisms are affected by these physiological changes is currently unclear. However, strict age-matching should be considered when exploring the mechanisms underlying disturbed penile and urethral development in CPM.[Formula presented]

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Status

Embase

Author NameID

Tack L.J.W.; ORCID: <http://orcid.org/0000-0001-9352-5444> Buelens S.; ORCID:

<http://orcid.org/0000-0002-1752-6821>

Spinoit A.F.; ORCID: <http://orcid.org/0000-0002-9045-9127>

Institution

(Tack, Cools) Pediatric Endocrinology Service, Ghent University Hospital, Department of Internal Medicine and Pediatrics, Ghent University, Ghent, Belgium (Praet, Van Dorpe) Department of

Pathology, Ghent University Hospital, Ghent, Belgium

(Haid) Department of Pediatric Urology Ordensklinikum Linz, Hospital of the Sisters of Charity, Linz Austria & Department of Urology, Ludwig-Maximilian University of Munich, Germany

(Buelens, Hoebeke, Van Laecke, Spinoit) Department of Urology, Ghent University Hospital, Ghent, Belgium

Publisher

Elsevier Ltd

Year of Publication

2020

40.

FGF8, FGF10 and FGF receptor 2 in foreskin of children with hypospadias: an analysis of immunohistochemical expression patterns and gene transcription.

Haid B., Pechriggl E., Nagele F., Dudas J., Webersinke G., Rammer M., Fritsch H., Oswald J.

Embase

Journal of Pediatric Urology. 16 (1) (pp 41.e1-41.e10), 2020. Date of Publication: February 2020.

[Article]

AN: 2003710058

Introduction: Fibroblast growth factors (FGFs) play a crucial role in early embryogenesis of the genital tubercle and are involved in the development of hypospadias, affecting both endo- and ectodermally derived tissues. It was hypothesized that expression of FGFs could be qualitatively or quantitatively altered in skin of children with hypospadias.

Objective(s): The objective of the study was to investigate expression patterns and transcription levels of FGF8, FGF10, and FGF Receptor 2 (FGFR2) in patients with hypospadias compared to normal controls.

Patients and Methods: Skin samples from the ventro-lateral aspect of the foreskin of 32 patients with hypospadias (17 distal and 15 proximal, mean age 25 months) and 10 normal foreskin samples (mean age 77 months) were analyzed by immunohistochemistry. Staining, localization,

and distribution of positive cells in epidermis and dermis were categorized independently by two researchers. Complementary DNA (cDNA) samples prepared from messenger RNA (mRNA) isolates of the same samples were analyzed by quantitative polymerase chain reaction (qPCR), comparing expressions of FGF8, FGF10, and FGFR2 with loading controls.

Result(s): Patients with hypospadias consistently showed aberrant immunohistochemical staining patterns for FGF8/FGF10/FGFR2 in epidermis and dermis compared to patients without penile malformation ($p < 0.01$ for all markers). qPCR displayed no difference in expression levels on mRNA level (FGFR2 $p = 0.44$, FGF8 $p = 0.77$, and FGF10 $p = 0.17$) comparing normal foreskin with foreskin from patients with hypospadias. Figure.

Discussion(s): The results point at an impact of FGF signaling during embryological development of hypospadias on skin, as an ectodermally derived tissue. Similar to the urethral development, this might be a result of mesothelial-epithelial interactions. The differing expression patterns in immunohistochemistry are not matched by a quantitative difference in marker expression on the mRNA level, putatively caused by post-translational modifications or alterations of the downstream pathway. FGFs, particularly FGF10 and FGFR2, are critically involved in wound healing.

Conclusion(s): There are significant differences in localization and distribution of FGF8, FGF10, and FGFR2 in comparisons of normal foreskin to foreskin of patients with hypospadias, whereas there is no difference in the quantitative expression of these markers on the mRNA level. This confirms the notion that penile skin is affected as well by the embryological aberrations during the embryogenesis of hypospadias.[Formula presented]

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Status

Embase

Institution

(Haid, Oswald) Department of Pediatric Urology, Hospital of the Sisters of Charity, Linz, Austria

(Haid) Department of Urology, Ludwig Maximilians University, Munich, Germany

(Pechriggl) Department of Plastic, Reconstructive and Aesthetic Surgery, Medical University Innsbruck, Innsbruck, Austria

(Nagele) Department of Cardiac Surgery, Medical University Innsbruck, Innsbruck, Austria

(Dudas) Molecular Biology and Oncology Lab, Department of Otorhinolaryngology, Medical University Innsbruck, Innsbruck, Austria

(Webersinke, Rammer) Laboratory for Molecular Biology and Tumor Cytogenetics, Department of Internal Medicine I, Hospital of the Sisters of Charity, Linz, Austria

(Fritsch) Section for Clinical and Functional Anatomy, Medical University Innsbruck, Innsbruck, Austria

Publisher

Elsevier Ltd

Year of Publication

2020

41.

Provider perspectives on shared decision-making regarding hypospadias surgery.

Chan K.H., Misseri R., Cain M.P., Whittam B., Szymanski K., Kaefer M., Rink R., Cockrum B., Moore C., Wiehe S.

Embase

Journal of Pediatric Urology. (no pagination), 2020. Date of Publication: 2020.

[Article]

AN: 2005598403

Introduction: Many parents experience decisional conflict and decisional regret around hypospadias surgery. The utilization of a shared decision-making (SDM) process may mitigate these issues, however addressing the principal components of the SDM process is a complex task that requires the investment of providers.

Objective(s): The purpose of this study was to facilitate a discussion about SDM anchored on hypospadias with pediatric urology and general pediatric providers to explore perspectives, clinical applications and barriers to adopting SDM in clinical practice. Study design: We conducted two focus groups in order to engage pediatric urology and general pediatric providers in guided discussions about SDM anchored on hypospadias. All activities were audio recorded and professionally transcribed. The transcripts were analyzed by three coders using directed qualitative content analysis techniques to identify themes and relationships between themes to inform the development of an affinity diagram (Extended Summary Figure).

Result(s): Two focus groups were held; one with seven pediatric urology providers in November 2018 and one with ten general pediatric providers in January 2019 (median age 51 years, 88.2% Caucasian, 58.8% female, 70.6% physicians and 29.4% nurse practitioners). Both groups identified some of the key components of SDM including engaging families in decision-making, informing them about treatment options and clarifying values/preferences (Extended Summary

Figure). They thought that SDM was useful for discussing preference-sensitive conditions (e.g. hypospadias) and addressing parental compliance. General pediatric providers also suggested that SDM helped them avoid unnecessary referrals to specialists. Both groups identified parental, provider and systemic barriers to the adoption of SDM: a) desire for paternalism, b) misperceptions about medical evidence, c) completion of parental decision-making prior to the clinical visit, d) provider bias/lack of interest and e) time constraints/productivity pressures. Discussion(s): Providers who care for hypospadias patients are knowledgeable about SDM and its potential clinical applications. They identified several potentially modifiable barriers to the adoption of a SDM process about hypospadias surgery in a pediatric clinical setting. Conclusion(s): Based on feedback from providers, we plan to implement a hypospadias decision aid early in the parental decision-making process about hypospadias such as in the postpartum unit and at well-child visits in the newborn period and provide a provider training session about SDM to address the identified knowledge gaps. [Figure presented]

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Status

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Author NameID

Chan, Katherine H.; ORCID: <http://orcid.org/0000-0001-9124-1704> Moore, Courtney; ORCID: <http://orcid.org/0000-0003-3563-0259>

Institution

(Chan, Misseri, Whittam, Szymanski, Kaefer, Rink) Department of Urology, Indiana University School of Medicine, Indianapolis, IN, United States (Chan) Department of Pediatrics, Center for Pediatric and Adolescent Comparative Effectiveness Research, Indiana University School of Medicine, Indianapolis, IN, United States

(Cain) Department of Urology, University of Washington, Seattle, WA, United States

(Cockrum, Moore, Wiehe) Research Jam, Indiana Clinical and Translational Sciences Institute, Indiana University School of Medicine, Indianapolis, IN, United States

Publisher

Elsevier Ltd

Year of Publication

2020

42.

Diagnosis and Treatment of Hypospadias With Megameatus Intact Prepuce.

Duan S.X., Li J., Jiang X., Zhang X., Ou W., Fu M., Chen K., Zheng L., Ma S.H.

Embase

Frontiers in Pediatrics. 8 (no pagination), 2020. Article Number: 128. Date of Publication: 31 Mar 2020.

[Article]

AN: 631486804

Purpose: To evaluate the diagnosis and treatment methods of hypospadias with megameatus intact prepuce (MIP).

Material(s) and Method(s): A retrospective analysis was performed in 27 MIP children, 13 of whom underwent tubularized incised plate urethroplasty (TIP procedure), 7 underwent the Duplay procedure, 5 underwent the Mathieu procedure, 1 underwent meatal advancement and glanuloplasty (MAGPI procedure), and 1 underwent the glans approximation procedure (GAP). The patients were followed for 6-36 months to evaluate the surgical outcomes by the Pediatric Penile Perception Score (PPPS).

Result(s): A total of 27 patients with a mean age of 8.12 +/- 3.0 years were enrolled in this study, and 25 cases (25/27, 92.6%) were accidentally discovered during the first visit for phimosis. The patients had a formed urethra of 0.5 to 1.5 cm. Complications occurred in 4 of the 27 patients (14.81%): 2 patients with urethral fistula and 2 patients with meatal stenosis. One patient had a case of self-healed urethral fistula, and the remaining 3 patients underwent reoperation. The post-operative effect was satisfactory in all patients, and the urinary flow and stream during urination were normal. The overall average PPPS score of non-operative surgeons and parents was satisfactory. There were no significant differences in meatus appearance, glans appearance, skin appearance, and general appearance PPPS score among the Mathieu, TIP, and Duplay surgical procedures.

Conclusion(s): MIP clinical manifestations are concealed and usually noted when circumcision is attempted. The suitable procedure for each patient should be tailored according to the anatomic features, and several techniques can be used with good functional and cosmetic results.

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Status

Embase

Institution

(Duan, Zheng) Department of Pediatric Surgery, The First Affiliated Hospital of Shantou University Medical College, Shantou, China (Duan, Li, Ou, Fu, Chen) Department of Pediatric Surgery, The Second Affiliated Hospital of Shantou University Medical College, Shantou, China (Jiang, Zhang) Department of Pediatric Surgery, Shenzhen Pingshan District Woman's and Children's Hospital, Southern Medical University, Shenzhen, China (Ma) Department of Radiology, The First Affiliated Hospital of Shantou University Medical College, Shantou, China
 Publisher
 Frontiers Media S.A. (E-mail: info@frontiersin.org)
 Year of Publication
 2020

43.

Overview of paediatric urology practice in lagos state university teaching hospital, Ikeja, Lagos, Nigeria.

Abolarinwa A.A., Ojewuyi O.O., Solarin A.U.

Embase

The Nigerian postgraduate medical journal. 27 (2) (pp 132-135), 2020. Date of Publication: 01 Apr 2020.

[Article]

AN: 631534272

Introduction: Paediatric urology is one of the subspecialities of urology, and in most climes, it is practised by the urologists and paediatric surgeons, and likewise in the Lagos State University Teaching Hospital (LASUTH). The urologists see and manage most of these cases in LASUTH. There has been no formal training in this subspeciality. However, both the urologists and paediatric surgeons in LASUTH have acquired some measure of skill and experience over time by virtue of the relatively high volume of the cases seen. This study is aimed at reviewing the practice of paediatric urology in the urology division of LASUTH and to advocate for formal training in an otherwise rare but direly needed subspeciality.

Patients and Methods: The ports of entry of paediatric patients with urologic conditions were assessed retrospectively over a 5-year period (2014-2018). The paediatric age range based on the Lagos State Government policy for health care is from birth to 12 years old. The ports of entry

included the urologic outpatient department, paediatric and the adult surgical emergency units and the paediatric wards. Patients referred to and managed by the paediatric surgery division were excluded from this study.

Result(s): The total paediatric urology cases seen and managed by the urologist in LASUTH within the period of review were 421. A total of 363 paediatric urology cases were seen during the period under review, making up 7.96% of the urology cases seen at the surgical outpatient department. The most common cases managed were hypospadias, posterior urethral valves and hydronephrosis. A variety of other cases include priapism, circumcision and post-circumcision injuries, urethral prolapse, testicular torsion, cystic renal dysplasia, disorder of sexual differentiation and several others. Three hundred and seven surgical procedures were done in the period of review on 272 (64.6%) patients.

Conclusion(s): There is a need for subspecialisation in paediatric urology to harness more specialists with a specific focus, training and interest in children and their urological conditions.

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32295945 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=32295945>]

Institution

(Abolarinwa) Department of Surgery, College of Medicine, Lagos State University, Ikeja, Lagos State, Nigeria (Ojewuyi) Department of Surgery, Ladoke Akintola University Teaching Hospital, Osogbo, Osun State, Nigeria

(Solarin) Department of Surgery; Department of Paediatrics, College of Medicine, Lagos State University, Ikeja, Lagos State, Nigeria

Publisher

NLM (Medline)

Year of Publication

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44.

Recurrent NEDD4L Variant in Periventricular Nodular Heterotopia, Polymicrogyria and Syndactyly.

Stouffs K., Verloo P., Brock S., Regal L., Beysen D., Ceulemans B., Jansen A.C., Meuwissen M.E.C.

Embase

Frontiers in Genetics. 11 (no pagination), 2020. Article Number: 26. Date of Publication: 05 Feb 2020.

[Article]

AN: 630978493

NEDD4L encodes an ubiquitin ligase which is expressed in the cortex and ventricular zone of the fetal brain. Missense variants in NEDD4L have been reported in nine patients with periventricular nodular heterotopia (PNH), polymicrogyria, cleft palate, and syndactyly. All reported variants are located in the HECT domain, causing deregulation of signaling pathways, including the AKT/mTOR pathway. Here we describe a first familial case with four affected members with a high degree of intra-familial phenotypic variability. Phenotypic features in the proband consisted of severe neurodevelopmental delay, refractory seizures, bilateral PNH, and perisylvian polymicrogyria. The other family members were less severely affected with mild developmental delay and isolated bilateral PNH. All family members had syndactyly. An unrelated patient presented with severe neurodevelopmental delay, seizures, and hypospadias, expanding the phenotypic spectrum. MRI revealed bilateral PNH and perisylvian polymicrogyria. All tested patients carry the recurrent variant c.623G > A, p.(Arg208Gln) in the WW domain of NEDD4L. The variant in the unrelated patient occurred de novo. This is the first report of a NEDD4L variant located in the WW domain which is probably involved in the recognition of substrates for ligation suggesting a loss of function variant.

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Status

Embase

Institution

(Stouffs) Center for Medical Genetics, UZ Brussel, Brussels, Belgium (Stouffs, Brock, Jansen)

Neurogenetics Research Group, Reproduction Genetics and Regenerative Medicine Research Cluster, Vrije Universiteit Brussel, Brussels, Belgium

(Verloo) Department of Pediatric Neurology, Ghent University Hospital, Ghent, Belgium

(Brock) Department of Pathology, UZ Brussel, Brussels, Belgium

(Regal, Jansen) Pediatric Neurology Unit, Department of Pediatrics, UZ Brussel, Brussels, Belgium

(Beysen, Ceulemans) Department of Pediatric Neurology, Antwerp University Hospital, Edegem, Belgium

(Meuwissen) Center of Medical Genetics, Antwerp University Hospital, Edegem, Belgium

(Meuwissen) Center of Medical Genetics, University of Antwerp, Edegem, Belgium

Publisher

Frontiers Media S.A. (E-mail: info@frontiersin.org)

Year of Publication

2020

45.

Paediatric urology.

Undre S., Cherian A.

Embase

Surgery (United Kingdom). 38 (4) (pp 224-230), 2020. Date of Publication: April 2020.

[Review]

AN: 2005117568

Paediatric urology is a subject that covers the urological aspects of care in children, some of which are seen also in adults, but may have specific diagnostic methods and treatments that are quite different. Additionally, it covers a range of congenital anomalies either on their own or in combination with a spectrum of disorders that need more complex management available at only specialized centres. For the purposes of a broad and basic understanding of the subject, this article will cover relevant topics and up-to-date guidelines.

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Institution

(Undre, Cherian) Lister Hospital, Stevenage, United Kingdom (Undre, Cherian) Great Ormond Street Hospital, London, United Kingdom

Publisher

Elsevier Ltd

Year of Publication

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46.

Use of Acellular Dermal Matrix for Urethroplasty Coverage in Proximal Hypospadias Repair: a Pilot Study.

Lin D., Wang G., Song H., Qu Y., Liu P., Liang H., Xu S., Chen S., Zhang W., Zhao Y., Chen B., Sun N.

Embase

Advances in Therapy. 37 (4) (pp 1425-1435), 2020. Date of Publication: 01 Apr 2020.

[Article]

AN: 2004278492

Introduction: The complication rates of proximal hypospadias, especially fistula, are much higher than those of distal hypospadias. Urethral coverage is an effective method for reducing fistulas. Acellular dermal matrix (ADM) has been shown to exhibit structural compatibility and biocompatibility, both of which promote tissue healing.

Method(s): The present non-randomized study evaluated the efficiency, feasibility, and safety of using ADM for urethroplasty coverage in patients with proximal hypospadias. This prospective study enrolled 35 patients (age range 15-60 months) with proximal hypospadias who underwent operation between September 2018 and March 2019 at Beijing Children's Hospital (Beijing, China). Urethroplasties were performed by the transverse preputial island flap (TPIF) technique. ADM was applied and sutured over the urethroplasty as an additional covering layer. Patient outcomes were compared with those of 80 non-matched control patients with proximal hypospadias who underwent the same procedure, with dartos as a covering layer.

Result(s): During a median follow-up of 11.56 months (range 9-15 months), urethral fistula occurred in six patients (17.1%) in the ADM group and 28 patients (35%) in the dartos group. Superficial wound infection was observed in six patients (17.1%) in the ADM group and 10 patients (12.5%) in the dartos group. One patient in the ADM group had diverticulum, compared with five patients (6.25%) in the dartos group. Meatal stenosis and urethral stricture were observed in four patients (11.4%) in the ADM group and six patients (7.5%) in the dartos group; all of these complications were treated conservatively. No glans dehiscence was observed in either group.

Conclusion(s): Use of ADM may be a safe and efficient covering technique to provide an additional coverage layer for proximal hypospadias repair, thereby reducing the incidence of fistula formation, especially among patients who have poor-quality covering materials.

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Status

Embase

Author NameID

Sun, Ning; ORCID: <http://orcid.org/0000-0002-3443-6991>

Institution

(Lin, Wang, Song, Qu, Liu, Liang, Xu, Chen, Zhang, Sun) Department of Urology, Beijing Children's Hospital Affiliated to Capital Medical University, National Center for Children's Health, Beijing 100045, China (Zhao, Chen) State Key Laboratory of Molecular Developmental Biology, Institute of Genetics and Developmental Biology, Chinese Academy of Sciences, Beijing 100190, China

Publisher

Adis

Year of Publication

2020

47.

Evaluation of surgical outcomes in different hypospadias types by HOSE score.

Guner E., Arikan Y.

Embase

Journal of Urological Surgery. 7 (1) (pp 54-57), 2020. Date of Publication: 2020.

[Article]

AN: 2005104888

Objective: We aimed to present outcomes of hypospadias surgery with the help of the Hypospadias objective scoring evaluation (HOSE) system and to demonstrate the reliability of the HOSE.

Material(s) and Method(s): Data of patients, who were operated for hypospadias in our clinic between January 2017 and March 2019, were retrospectively analyzed. The parameters included demographic data, location of the meatus preoperative, postoperative complications, follow-up period, recurrence and postoperative HOSE score.

Result(s): A total of 46 patients were included in the study. The mean age of the patients was 8.5+/-5.4 years. Fifteen patients had distal hypospadias, 13- subcoronal, 9- penile , 6- proximal and 3 had glanular hypospadias. Tubularized incised plate urethroplasty was performed in 35 patients, Bracka 2- stage repair in 6 patients and meatal advancement and glanduloplasty in 3 and, the onlay flap technique was used in 2 patients. The mean postoperative HOSE score was 14.6+/-1.7. The lowest HOSE score was found in patients with proximal hypospadias and the

highest HOSE score was in those with glanular hypospadias (12.6 versus 15.6) ($p=0.26$). When HOSE scores of the patients were evaluated by a different physician, the mean HOSE score was found to be 14.5 ± 1.7 . There was no significant intraobserver variation (Kappa score: 0.698), ($p<0.0001$).

Conclusion(s): HOSE is an objective and reliable scoring system that can be used to evaluate the outcomes of hypospadias surgery.

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Status

Embase

Author NameID

Arikan, Yusuf; ORCID: <http://orcid.org/0000-0003-0823-7400>

Institution

(Guner, Arikan) Bakirkoy Sadi Konuk Training and Research Hospital, Clinic of Urology, Istanbul, Turkey

Publisher

Galenos Publishing House (E-mail: info@balkanmedicaljournal.org)

Year of Publication

2020

48.

Evaluation of double-folded meatal-based flap (modified Mathieu) in redo circumcised distal penile hypospadias: a prospective multicenter study.

Nagla S., Radwan M., Alhefnawy M., Ghaith A., Hagrass A.M., Shalaby M., Awad M., Negm M.A., Elgamal S.

Embase

African Journal of Urology. 26 (1) (no pagination), 2020. Article Number: 13. Date of Publication: 01 Dec 2020.

[Article]

AN: 2004603714

Background: Sometimes it becomes a dilemma to repair a failed circumcised distal penile hypospadias with partial or complete wound dehiscence. In many of these cases, redo operations

need a graft or a flap. The aim of this study is to evaluate the double-folded meatal-based flap in redo circumcised hypospadias with shallow urethral plate and a small glans penis.

Result(s): From September 2015 to August 2018, we prospectively studied the double-folded meatal-based flap in 56 failed circumcised distal hypospadias without penile curvature with shallow urethral plate and a small glans penis. Distally folded Mathieu flap was done. Forty-seven cases only had completed the study. We had 19 patients with distal penile hypospadias, 19 with sub-coronal hypospadias, and 9 with coronal hypospadias. Median operative time was 50 (range 40-80 min). The median length of the double-folded flap was 14 (range 10-18 mm). The median follow-up was 12 (range 1-33 months). The success of the operation means that there is an absence of any complications. Our success rate was 73%. We had 8 patients with meatal retraction, one fistula, and one patient with wound disruption. Only 3 patients (6.4%) needed re-operation: one case with meatal retraction, another with fistula, and a third with complete wound disruption.

Conclusion(s): Distally folded meatal-based flap is an additional option in redo circumcised hypospadias with shallow urethral plate, small glans penis, and pliable anterior penile skin with a considerable high percent of meatal recession.

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Status

Embase

Institution

(Nagla, Radwan, Ghaith, Hagrass, Elgamal) Urology Department, Tanta University, Tanta, Egypt

(Alhefnawy) Urology Department, Benha University, Benha, Egypt

(Shalaby, Awad) Pediatric Surgery Unit, Tanta University, Tanta, Egypt

(Negm) Pediatric Surgery Unit, Qena Faculty of Medicine, South Valley University, Qena, Egypt

Publisher

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49.

Bracka's Method of Proximal Hypospadias Repair: Preputial Skin or Buccal Mucosa?.

Manasheva D., Kozyrev G., Nikolaev V., Abdullaev F., Abdulkarimov G., Kushnir B., Gazimiev M.

Embase

Urology. 138 (pp 138-143), 2020. Date of Publication: April 2020.

[Article]

AN: 2004683249

OBJECTIVE: To analyze treatment results of staged surgical repair of proximal forms of hypospadias according to Bracka's technique using preputial vs buccal grafts. MATERIAL AND METHODS: We retrospectively reviewed 220 patients with proximal forms of hypospadias treated with Bracka's urethroplasty technique. They were divided into 2 groups: Group I-108 patients treated with preputial skin grafts in 2001-2013; Group II-112 patients who underwent urethroplasty with buccal mucosa grafts in 2013-2016. RESULTS AND DISCUSSION: Of the Group I patients with preputial skin grafts, complications were obtained in 33 (31%) cases; in Group II-23 (20%) cases. Complications include fistulas, defects of urethra, and scar contraction of grafts. The cosmetic results according to Hypospadias Objective Penile Evaluation scale were more satisfactory when buccal mucosa grafts were used. Further studies are needed to analyze the long-term changes post-urethroplasty with both preputial skin and buccal mucosa grafts. CONCLUSION(S): This is one of the only studies to compare complications and histology of the 2 free grafts: preputial skin and buccal mucosa. This study affirms that a staged surgical method with the use of free grafts according to Bracka's technique is a successful method of treatment of proximal forms of hypospadias in children achieving good functional and cosmetic results with a relatively low rate of complications.

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Status

Embase

Institution

(Manasheva) Moscow State University, Faculty of Fundamental Medicine, Moscow, Russian Federation (Kozyrev, Nikolaev, Abdullaev, Abdulkarimov) Uroandrology Department of the Russian Children's Clinical Hospital of the Russian Ministry of Health, Moscow, Russian Federation

(Kushnir) Department of Anatomical Pathology of the Russian Children's Clinical Hospital of the Russian Ministry of Health, Moscow, Russian Federation

(Gazimiev) Urology Department of Sechenov First Moscow State Medical University, Moscow, Russian Federation

Publisher

Elsevier Inc. (E-mail: usjcs@elsevier.com)

Year of Publication

2020

50.

Associations between macrolide antibiotics prescribing during pregnancy and adverse child outcomes in the UK: Population based cohort study.

Fan H., Gilbert R., O'Callaghan F., Li L.

Embase

The BMJ. 368 (no pagination), 2020. Article Number: m331. Date of Publication: February 2020.

[Article]

AN: 631074298

Objective To assess the association between macrolide antibiotics prescribing during pregnancy and major malformations, cerebral palsy, epilepsy, attention deficit hyperactivity disorder, and autism spectrum disorder in children. **Design** Population based cohort study. **Setting** The UK Clinical Practice Research Datalink. **Participants** The study cohort included 104 605 children born from 1990 to 2016 whose mothers were prescribed one macrolide monotherapy (erythromycin, clarithromycin, or azithromycin) or one penicillin monotherapy from the fourth gestational week to delivery. Two negative control cohorts consisted of 82 314 children whose mothers were prescribed macrolides or penicillins before conception, and 53 735 children who were siblings of the children in the study cohort. **Main outcome measures** Risks of any major malformations and system specific major malformations (nervous, cardiovascular, gastrointestinal, genital, and urinary) after macrolide or penicillin prescribing during the first trimester (four to 13 gestational weeks), second to third trimester (14 gestational weeks to birth), or any trimester of pregnancy. Additionally, risks of cerebral palsy, epilepsy, attention deficit hyperactivity disorder, and autism spectrum disorder. **Results** Major malformations were recorded in 186 of 8632 children (21.55 per 1000) whose mothers were prescribed macrolides and 1666 of 95 973 children (17.36 per 1000) whose mothers were prescribed penicillins during pregnancy. Macrolide prescribing during the first trimester was associated with an increased risk of any major malformation compared with penicillin (27.65 v 17.65 per 1000, adjusted risk ratio 1.55, 95% confidence interval 1.19 to 2.03) and specifically cardiovascular malformations (10.60 v 6.61 per 1000, 1.62, 1.05 to 2.51). Macrolide prescribing in any trimester was associated with an increased risk of genital

malformations (4.75 v 3.07 per 1000, 1.58, 1.14 to 2.19, mainly hypospadias). Erythromycin in the first trimester was associated with an increased risk of any major malformation (27.39 v 17.65 per 1000, 1.50, 1.13 to 1.99). No statistically significant associations were found for other system specific malformations or for neurodevelopmental disorders. Findings were robust to sensitivity analyses. Conclusions Prescribing macrolide antibiotics during the first trimester of pregnancy was associated with an increased risk of any major malformation and specifically cardiovascular malformations compared with penicillin antibiotics. Macrolide prescribing in any trimester was associated with an increased risk of genital malformations. These findings show that macrolides should be used with caution during pregnancy and if feasible alternative antibiotics should be prescribed until further research is available. Trial registration ClinicalTrials.gov NCT03948620 Copyright © Published by the BMJ Publishing Group Limited. For permission to use (where not already granted under a licence) please go to.

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Embase

Institution

(Fan, Gilbert, Li) Population, Policy and Practice Programme, Great Ormond Street Institute of Child Health, University College London, London WC1N 1EH, United Kingdom (O'Callaghan) Developmental Neurosciences Programme, Great Ormond Street Institute of Child Health, University College London, London, United Kingdom

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Year of Publication

2020

51.

Comparison of snodgrass and bracka methods in terms of urethral fistula and meatal stenosis in hypospadias repair.

Kocak O.F., Avci V., Ayengin K.

Embase

Eastern Journal of Medicine. 25 (1) (pp 146-150), 2020. Date of Publication: 2020.

[Article]

AN: 2003713067

Hypospadias is one of the most frequent congenital anomalies in birth. Many factors may have part in etiology. Its treatment is possible only through surgical intervention, and there are more than 300 methods in literature identified for hypospadias repair. The purpose of this study is to compare Snodgrass and Bracka methods, the methods most frequently used for hypospadias repair, in terms of urethral fistula and meatal stenosis. In this study, the data of 40 patients operated between 2014 and 2019 years have been used. 20 of the patients have been operated with Snodgrass, and the others have been operated with Bracka method. The average ages of the patients operated with Snodgrass method and Bracka method were 4 years 3 months and 2 years 9 months respectively. Average length for follow-up for the patients operated with Snodgrass method was 18 months 9 days, and it was 35 months 1 day for the patients operated with Bracka method. Urethral fistula was observed to develop in %30 of the patients operated with Snodgrass method and %25 of those operated with Bracka method totally. In both methods, rates of meatal stenosis were more frequent compared to fistula (Snodgrass: %35, Bracka %40). Fistula development is found numerically more in the patients operated with Snodgrass method while meatal stenosis development is observed numerically more in those operated with Bracka method.

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Status

Embase

Institution

(Kocak) Department of Plastic, Reconstructive and Aesthetic Surgery, Van Yuzuncu Yil University, Faculty of Medicine, Van, Turkey (Avci, Ayengin) Department of Pediatric Surgery, Van Yuzuncu Yil University, Faculty of Medicine, Van, Turkey

Publisher

Yuzuncu Yil Universitesi Tip Fakultesi (Eastern Journal of Medicine, Van 65100, Turkey)

Year of Publication

2020

52.

PUVs are more common in boys with hypospadias.

Lee B., Driver C.P., Flett M.E., Steven L., Steven M., O'Toole S.

Embase

Journal of Pediatric Urology. (no pagination), 2020. Date of Publication: 2020.

[Article]

AN: 2005439593

Introduction: The association between posterior urethral valves (PUVs) and hypospadias has previously been reported in case reports. After the identification of this twin pathology in a number of patients, a national retrospective review of all patients with this dual diagnosis was performed.

Patients and Methods: All patients were identified in each centre from surgical databases of prospectively collated information on all surgical procedures. The medical notes were reviewed to ascertain demographics, the type of hypospadias, the mode of presentation of the valves and the outcome.

Result(s): Twenty-eight patients who had the dual diagnosis of hypospadias and PUV between 2002 and 2017 in the four tertiary paediatric centres where specialist paediatric urology is undertaken in our country were identified. Most patients (n = 24) had the valves diagnosed after hypospadias surgery. The median age at the time of hypospadias surgery was 1.4 years (range 1-4 years). There were 12 proximal and 16 mid or distal hypospadias. The commonest presentation was with problems voiding after surgery in 14 cases with a further seven boys who had urinary tract infections. Four patients had a urethro-cutaneous fistula after repair that initiated further assessment. Two boys had distal dehiscence of their repair. There was one boy presented with new onset daytime incontinence. The median time of follow-up after valve incision surgery was 4.9 years (range 0.1-12.3 years). Twenty-two patients (three pre toilet training) had no ongoing urinary symptoms. Twenty-one boys have normal renal function with one patient in stage 3b chronic kidney disease. The incidence of this dual diagnosis in Scotland is estimated at one in 100 cases of hypospadias in the paediatric population.

Conclusion(s): The incidence of PUV in boys with hypospadias is estimated at 1% patients.

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Status

Article-in-Press

Institution

(Lee, Flett, Steven, O'Toole) Department of Paediatric Surgery and Urology, Royal Hospital for Children, 1345 Govan Road, Glasgow G51 4TF, United Kingdom (Driver) Department of Paediatric Surgery and Urology, Royal Aberdeen Children's Hospital, Aberdeen AB25 2ZN, United Kingdom

(Steven) Department of Paediatric Surgery and Urology, Royal Hospital for Sick Children, 9
Sciennes Road, Edinburgh EH9 1LF, United Kingdom

Publisher

Elsevier Ltd

Year of Publication

2020

53.

5alpha-Reductase type 2 deficiency in families from an isolated Andean population in Venezuela.
Avendano A., Gonzalez-Coira M., Paradisi I., Rojas A., Da Silva G., Gomez-Perez R., Ceballos
J.O.

Embase

Annals of Human Genetics. 84 (2) (pp 151-160), 2020. Date of Publication: 01 Mar 2020.

[Article]

AN: 2003479473

5alpha-Reductase type 2 deficiency causes a 46,XY disorder of sex development (DSD) characterized by ambiguous external genitalia, rudimentary prostate, and normal internal genitalia. The disease prevalence worldwide is low, but in a small and isolated village of the Venezuelan Andes, a higher incidence has been found. DNA analysis of the SRD5A2 gene was performed in three inbred affected individuals clinically diagnosed with DSD. The entire coding regions, the p.L89V polymorphism (rs523349) and five intragenic SNPs (rs2300702, rs2268797, rs2268796, rs4952220, rs12470196) used to construct haplotypes were analyzed by Sanger sequencing. To assess the probable ethnic origin of the mutation in this geographic isolate, a population structure analysis was performed. Homozygosis for the p.N193S mutation was found in all patients, with a mutation carrier frequency of 1:80 chromosomes (0.0125) in the geographic focus, suggesting a founder phenomenon. The results of the population structure analysis suggested a mutation origin closer to the Spanish populations, according to the clusters grouping. The genotype-phenotype correlation in the patients was not absolute, being hypospadias and cryptorchidism the main traits that differentiate affected individuals.

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Author NameID

Paradisi, Irene; ORCID: <http://orcid.org/0000-0001-9039-8945>

Institution

(Avendano, Gonzalez-Coira, Da Silva) Medical Genetics Unit, Medicine Faculty, Los Andes University, Merida, Venezuela (Paradisi) Venezuelan Institute for Scientific Research (IVIC), Laboratory of Human Genetics, Caracas, Venezuela

(Rojas) National Center for Scientific Calculation (CeCalcULA), Los Andes University, Merida, Venezuela

(Gomez-Perez) Endocrinology Service, Los Andes University Hospital (I.A.H.U.L.A.), Merida, Venezuela

(Ceballos) Andrology Laboratory, Los Andes University, Electronic Microscopic Center, Merida, Venezuela

Publisher

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Year of Publication

2020

54.

Discrepant Rates of Hypospadias Surgical Complications: A Comparison of U.S. News & World Report and Pediatric Health Information System Data and Published Literature.

Pohl H.G., Rana S., Sprague B.M., Beamer M., Rushton H.G.

Embase

The Journal of urology. 203 (3) (pp 616-623), 2020. Date of Publication: 01 Mar 2020.

[Review]

AN: 629348713

PURPOSE: Complication rates of 5% to 10% and 12% to 23% have been observed following distal and proximal hypospadias repair, respectively. However, these rates may be overreported. We hypothesized that data from the Pediatric Health Information System would corroborate the complication rates reported in the literature and refute the rates established by U.S. News & World Report as quality metrics. MATERIALS AND METHODS: The Pediatric Health Information

System database was interrogated for hypospadias and revision CPT codes. To evaluate the appropriateness of the U.S. News & World Report code list to identify revisions, 3 CPT groups were defined. Group A included codes sought by U.S. News & World Report, group B included group A plus codes for acquired urethrocutaneous fistula in males and group C included group B plus any revision codes associated with the index procedures. To evaluate the appropriateness of U.S. News & World Report followup interval, we assessed revision rates with ever increasing followup intervals out to 7 years. Yearly revision rates were summarized by median and quantile to correlate whether median revision rates changed significantly depending on whether increased followup or enhanced code lists were used.

RESULT(S): Average complication rates for group A were 3.32% (range 0.48% to 7.36%) and 12.29% (3.48% to 36.36%) for distal and proximal repairs, respectively. Revision rates increased significantly from group A (median 3.32%) by inclusion of a more expansive list of CPT codes associated with revision procedures (group B, 4.26%, $p < 0.001$ and group C, 6.37%, $p < 0.001$) in distal hypospadias. Among proximal hypospadias cases this difference was not significant when comparing group A (12.29%) to group B (12.53%, $p = 0.813$), but was significant when comparing group A to group C (22.14%, $p < 0.001$). Median revision rates for distal and proximal hypospadias increased with longer followup for all 3 groups, although the upward trend was not statistically significant.

CONCLUSION(S): Depending on how one defines hypospadias revision, no hospital within the Pediatric Health Information System data set meets the U.S. News & World Report definition of perfection, a finding that is supported by recent reports from some of the largest, most prolific and most highly regarded pediatric urology programs. We posit that the U.S. News & World Report quality metrics do not accurately reflect the nature of hypospadias surgery complication rates.

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Institution

(Pohl, Rana, Sprague, Beamer, Rushton) Division of Urology, Children's National Health System, George Washington University School of Medicine and Health Sciences, D.C, WA

Publisher

NLM (Medline)

Year of Publication

2020

55.

The pediatric patient and future fertility: optimizing long-term male reproductive health outcomes.

Nassau D.E., Chu K.Y., Blachman-Braun R., Castellan M., Ramasamy R.

Embase

Fertility and Sterility. 113 (3) (pp 489-499), 2020. Date of Publication: March 2020.

[Review]

AN: 2005226288

Globally, male factor infertility accounts for 20%-70% of couples struggling to conceive. Certain male pediatric developmental conditions, such as cryptorchidism, hypospadias, testicular and other childhood cancers, infections, and pediatric varicocele have been associated with future infertility. Early fertility preservation, especially in those with pending chemotherapy or genetic conditions such as Klinefelter syndrome, should be strongly considered in patients expected to experience testicular loss. Although optimal treatment timing may be unknown owing to a paucity of long-term prospective studies, early diagnosis and targeted treatment may optimize fertility potential in adulthood.

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Institution

(Nassau) Department of Urology, Lenox Hill Hospital, Donald and Barbra Zucker School of

Medicine at Hofstra/Northwell, New York, NY, United States (Chu, Blachman-Braun,

Ramasamy) Department of Urology, University of Miami Miller School of Medicine, Miami, FL,

United States

(Castellan) Division of Pediatric Urology, Nicklaus Children's Hospital, Miami, FL, United States

Publisher

Elsevier Inc. (E-mail: usjcs@elsevier.com)

Year of Publication

2020

56.

The male genital system.

Wu W.J., Gitlin J.S.

Embase

Pediatrics in Review. 41 (3) (pp 101-111), 2020. Date of Publication: 01 Mar 2020.

[Article]

AN: 2005208079

* Male differentiation relies on the presence of the Y chromosome and the action of testosterone on bipotent genital structures. * Hypospadias is a commonly encountered disorder of the penis. Further evaluation is prudent in cases associated with undescended testis based on an observational study. (12) * By consensus, micropenis is defined as 2.5 SD or more below the mean stretched length for a patient's age. Endocrinologic evaluation should be instituted once this diagnosis is made. More benign entities, including webbed penis and buried penis, should be ruled out by proper measurement of the penis. Treatment with testosterone supplementation is effective based on observational studies. (17)(18)(19) * Physiologic phimosis resolves over time, with the prepuce becoming retractile in most patients by 4 years of age. Paraphimosis is an emergency condition that requires immediate medical attention. Complications from the prepuce may be treated medically or surgically. * Male circumcision has medical benefits, such as decreased risk of urinary tract infection, penile cancer, and contracting human immunodeficiency virus based on strong data from meta-analysis of the present literature. (24) * Penile trauma is an uncommon clinical entity. A thorough history should be obtained to ensure that the mechanism of injury is consistent with the physical injury. * Cryptorchidism is a common condition that requires close followup. Referral to a surgical specialist is recommended should a testis fail to descend by 6 months of age. There is strong evidence based on a literature review that surgical correction helps lower the risk of testicular malignancy in cryptorchid testes. (35) * By consensus, testicular torsion is an emergency condition that requires rapid diagnosis and management. This condition should be considered in all patients with acute scrotal pain. * Varicoceles are of clinical concern because of possible future infertility. Its management in the adolescent population is still a subject of great debate. * Epididymo-orchitis may mimic testicular torsion because patients present with similar symptoms of acute scrotal pain and swelling. Duplex ultrasonography is helpful in making the proper diagnosis. * Based on clinical studies, cancer of the testis is uncommon in the pediatric population. A slow-growing, nontender mass should raise concern for an intrascrotal malignancy. Referral to a urologist is indicated for surgical resection once the diagnosis is made. * Trauma to the testis may lead to testicular rupture. Early surgery is indicated to prevent testicular atrophy when rupture is suspected or when there is penetrating trauma. Conservative management is acceptable in select populations based on an observational study. (68).

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Status

Embase

Institution

(Wu) Smith Institute for Urology, Zucker School of Medicine, Lake Success, NY, United States

(Gitlin) Cohen Children's Medical Center, Division of Pediatric Urology, Zucker School of Medicine, New Hyde Park, NY, United States

Publisher

American Academy of Pediatrics (141 Northwest Point Blvd, P.O. Box 927, Elk Grove Village IL 60007-1098, United States)

Year of Publication

2020

57.

Hyperbaric oxygen therapy for pediatric "hypospadias cripple"-evaluating the advantages regarding graft take.

Neheman A., Rappaport Y.H., Verhovsky G., Bush N., Snodgrass W., Lang E., Zisman A., Efrati S.

Embase

Journal of Pediatric Urology. (no pagination), 2020. Date of Publication: 2020.

[Article]

AN: 2005199637

Introduction: Hypospadias cripple patients pose a major surgical challenge with high complication rates attributed mainly to graft contraction. Hyperbaric oxygen therapy (HBOT) is an established treatment for compromised grafts and used extensively as a salvage therapy for compromised grafts and ischemic non-healing wounds.

Objective(s): We evaluated the graft-take rates in hypospadias cripple cases undergoing a staged tubularized autograft repair (STAG) and compared between patients treated with or without preemptive HBOT.

Material(s) and Method(s): All patients underwent a STAG. Patients receiving preemptive HBOT were compared with patients receiving the standard surgical procedure without HBOT. The HBOT

protocol included a daily session, 5 days per week for four weeks before the surgery and 10 additional daily sessions immediately after first-stage surgery. Each HBOT session included 90 min exposure to 100% O₂ at 2 atmospheres absolute with 5 min air breaks every 20 min. The primary endpoint was graft take. Sequential tubularization without tension at second stage was defined as success.

Result(s): Seven boys received HBOT and 14 boys comprised the control group. All patients in the HBOT group had good graft take with no graft contraction. In the control group, 57% had good graft take and could proceed to the second-stage surgery and 43% had graft contraction (Table). Except for one patient who had claustrophobia while entering the chamber, no significant side-effects developed during the HBOT.

Discussion(s): The basic pathophysiology of compromised flaps includes both ischemia and reperfusion injury, which can be attenuated by HBOT. The beneficial effects of HBOT relates to several mechanisms, including hyperoxygenation, fibroblast proliferation, collagen deposition, angiogenesis, and vasculogenesis. Graft contraction is a well-known complication in hypospadias cripple population with reported failure rate of 39-63%. The HBOT procedure was found to be very effective and the entire HBOT group had a good graft take. Accordingly, all patients in the HBOT group proceeded to a successful second-stage tubularization. In addition, HBOT was found to be safe and generally well tolerated by this pediatric population. Study limitations were a relative small, non-homogenous sample size and lack of prospective randomization. Success was defined as sufficient graft elasticity sufficing for tubularization of the neourethra, and exact graft measurements are lacking in this study.

Conclusion(s): Preemptive HBOT can be used safely in the hypospadias cripple pediatric population and can potentially reduce the expected high surgical failure secondary to graft contraction. [Table presented]

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Status

Article-in-Press

Institution

(Neheman, Rappaport, Verhovsky, Zisman) Departments of Urology, Shamir (Assaf-Harofeh) Medical Center, Zerifin, Israel (Bush, Snodgrass) Parc Urology, Frisco, TX, United States (Lang, Efrati) Sagol Center for Hyperbaric Medicine and Research, Shamir (Assaf-Harofeh) Medical Center, Zerifin, Israel

(Neheman, Rappaport, Lang, Zisman, Efrati) Sackler School of Medicine, Tel Aviv University, Tel Aviv, Israel

Publisher

58.

Long-term sexual outcomes in patients with exstrophy-epispadias complex.

Sinatti C., Waterschoot M., Roth J., Van Laecke E., Hoebeke P., Spinoit A.-F.

Embase

International Journal of Impotence Research. (no pagination), 2020. Date of Publication: 2020.

[Article]

AN: 2004448229

Exstrophy-epispadias complex (EEC) is a spectrum of genitourinary malformations that ranges in severity and affects external genitalia and the lower urinary tract. The aim of this study was to determine the long-term sexual outcomes of patients with EEC. Sexual outcomes were hypothesized to be related to those of urinary ones. A retrospective database including all patients with EEC who had surgery at a tertiary referral institution from 1990 to 2019 was created. Data based on patient's charts were collected: demographics, surgeries, sexual outcomes, urinary outcomes. Fifty-eight patients with EEC had surgery at tertiary referral institution and entered our database. For this analysis of sexual outcomes, a sub-set of the whole population was selected: patients being 14 years old and older, having at least one surgery at our institution and having at least 12 months of follow-up. Applying this selection criteria to our database resulted in a series of 29 patients. High rates of sexual activity were observed in pubertal and post-pubertal men (96%) and women (75%). Seventy-nine percent of men and 67% of women reported sexual satisfaction; 63% of men reported normal ejaculation. To achieve these rates, 96% of men required surgery (84% penoplasty, 52% phalloplasty), and 25% of women required introitoplasty. Fertility was achieved in 67% of men and 100% of women. Assisted reproductive technology was needed in one man. Continence rates were high (diurnal continence in 83% and nocturnal continence in 93%). However, 76% required multiple continence procedures. Men and women with EEC can have good long-term sexual and urinary outcomes, but this may require multiple surgeries. Good sexual outcomes seem to be related to good urinary and continence outcome.

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Status

Article-in-Press

Author NameID

Spinoit, Anne-Francoise; ORCID: <http://orcid.org/0000-0002-9045-9127>

Institution

(Sinatti, Waterschoot, Roth, Van Laecke, Hoebeke, Spinoit) Department of Urology, ERN eUROGEN accredited centre, Ghent University Hospital, Ghent, Belgium (Roth) Division of Pediatric Urology, Riley Hospital for Children, Indiana University School of Medicine, 705 Riley Hospital Drive, Suite 4230, Indianapolis, IN 46202, United States

Publisher

Springer Nature

Year of Publication

2020

59.

Parental decisional satisfaction after hypospadias repair in the United Kingdom.

Bethell G.S., Chhabra S., Shalaby M.S., Corbett H., Kenny S.E., Godse A., Lall A., Taghizadeh A., Lee B., Driver C., Keene D., Marshall D., Murphy F., McAndrew F., Nicholls G., Chandran H., Steinbrecher H., Evans K., McCarthy L., Steven M., Shenoy M., Farrugia M.-K., Woodward M., Flett M., Gopal M., Godbole P., Daniel R., Romero R.M., Wragg R., Manoharan S., Griffin S., O'Toole S., Abbas T., Kalidasan V.

Embase

Journal of Pediatric Urology. (no pagination), 2020. Date of Publication: 2020.

[Article]

AN: 2005148453

Background: In hypospadias, the aim of surgical treatment is to achieve both desirable functional and cosmetic outcomes; however, complications following surgery are common and 18% of boys require re-operation. In mild degrees of hypospadias, repair may be offered entirely to improve cosmesis, meaning parents should be fully informed of this and the potential for complications, during the consent process. Parents' decision-making may be aided by making them aware of

how others in a similar position have felt about the decision that they made for their child. One method of measuring parental satisfaction is decisional regret (DR).

Objective(s): To assess parental satisfaction following hypospadias surgery in the United Kingdom by assessing DR and to determine the feasibility of obtaining meaningful data via a mobile phone survey. Study design: The National Outcomes Audit in Hypospadias database was commissioned by the British Association of Paediatric Surgeons to capture clinical information from hypospadias repairs. Following ethical approval (16/NW/0819), a text message was sent to mobile numbers in the database inviting participation in a questionnaire incorporating the validated DR scale (DRS). The primary outcome measure was mean DRS score, which was correlated with clinical information, a score of zero indicated no regret and 100 indicated maximum regret.

Result(s): There were 340 (37%) responses. The median age at the primary procedure was 16 (interquartile range 13-20) months. No DR (score = 0) was detected in 186 (55% [95%CI 49-60]) respondents; however, moderate-to-severe DR (score = 26-100) was seen in 21 (6.2% [95%CI 3.6-8.7]) respondents. On multivariate analysis, a distal meatus, a small glans and developing complications requiring repeat surgery were all associated with increased levels of regret (Table). There was no association between DR and cases performed per surgeon.

Discussion(s): Around half of respondents demonstrated no DR and postoperative complications requiring surgery were associated with the highest levels of DR, which is similar to a Canadian study. Lorenzo et al. however found that DR was associated with circumcision, which was undertaken in all boys; however, in this UK study, around a third of boys were circumcised and regret levels between those circumcised and those not circumcised were similar. The limitations of this work include the following: surgeons submitting their own data on complications and there is potential of selection bias between respondents and non-respondents as with any survey.

Conclusion(s): Data from this study can be used to improve pre-operative counselling during the consent process. Smart mobile phone technology can be used successfully to distribute and collect parent-reported outcomes. [Table presented]

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Institution

(Bethell, Chhabra, Corbett, Kenny) Alder Hey Children's Hospital, Liverpool L14 5AB, United Kingdom (Bethell, Chhabra, Kenny) University of Liverpool, Liverpool, Merseyside L69 3BX, United Kingdom

(Shalaby) Bristol Royal Hospital for Children, Bristol BS2 8BJ, United Kingdom

(Godse, Lall, Gopal) Royal Victoria Infirmary, Newcastle, United Kingdom
 (Taghizadeh) Evelina London Children's Hospital, London, United Kingdom
 (Lee, Steven, Flett, O'Toole) Royal Hospital for Sick Children, Glasgow, United Kingdom
 (Driver) Royal Aberdeen Children's Hospital, Aberdeen, United Kingdom
 (Keene) Manchester Children's Hospital, Manchester, United Kingdom
 (Marshall) Royal Belfast Hospital for Sick Children, Belfast, United Kingdom
 (Murphy, Evans) St George's Hospital, London, United Kingdom
 (McAndrew) Alder Hey Children's NHS Foundation Trust, Liverpool, United Kingdom
 (Nicholls, Woodward) Bristol Royal Hospital for Children, Bristol, United Kingdom
 (Chandran, McCarthy, Wragg, Abbas) Birmingham Children's Hospital, Birmingham, United Kingdom
 (Steinbrecher, Manoharan, Griffin) Southampton General Hospital, Southampton, United Kingdom
 (Shenoy) Queen's Medical Centre, Nottingham, United Kingdom
 (Farrugia) Chelsea & Westminster Hospital, London, United Kingdom
 (Godbole) Sheffield Children's Hospital, Sheffield, United Kingdom
 (Daniel) Hull Royal Infirmary, Hull, United Kingdom
 (Romero) Oxford University Hospital, Oxford, United Kingdom
 (Kalidasan) Royal Alexandra Hospital, Brighton, United Kingdom
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 2020

60.

Complete primary repair of bladder exstrophy: a systematic review.

Pathak P., Ring J.D., Delfino K.R., Dynda D.I., Mathews R.I.

Embase

Journal of Pediatric Urology. (no pagination), 2020. Date of Publication: 2020.

[Review]

AN: 2005127818

Objectives: Complete primary repair of exstrophy (CPRE) was established as a method to reduce numbers of procedures for the reconstruction of bladder exstrophy (BE). Performed since 1989, some suggest it as a replacement for the staged reconstructive procedure, the gold standard. Does CPRE reduce the numbers of procedures for reconstruction of BE? **Methods:** Literature was reviewed from 1989 to 2016, and articles evaluating outcomes of patients undergoing CPRE, extracted. Effort was made to obtain final data from each reporting institution/group. Eleven articles meeting criteria were evaluated for qualitative systematic review. Age at initial closure, complications, additional procedures, and outcomes were evaluated to provide an overview of CPRE.

Result(s): Ten groups reported BE management using the CPRE technique. 236 patients (153 boys; 72 girls; 11 unknown sex) had primary closure ranging from birth to 5.6 years. Osteotomy was favored by most in infants closed beyond the first 72 h of life along with spica cast immobilization. Three groups recommended concomitant augmentation for infants with small bladder capacities. Ureteral reimplantation was required in 58 patients with recurrent urinary tract infections resistant to prophylaxis. Hypospadias repair was required for most boys having complete penile disassembly, and most children eventually required bladder neck reconstruction (BNR) for continence. Overall, voiding without BNR was noted in 16-37% of children in the reported series.

Conclusion(s): Complete primary repair of exstrophy has been suggested as a single procedure for the management of BE. Literature review suggests most patients require multiple procedures to complete reconstruction and attain continence.

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Institution

(Pathak, Ring, Delfino, Dynda, Mathews) Southern Illinois University School of Medicine,
Springfield, IL, United States

Publisher

Elsevier Ltd

Year of Publication

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61.

Snodgrass vs Snodgraft operation to repair the distal hypospadias in the narrow urethral plate.

Eldeeb M., Nagla S., Abou-Farha M., Hassan A.

Embase

Journal of Pediatric Urology. (no pagination), 2020. Date of Publication: 2020.

[Article]

AN: 2005127673

Background: Using the Snodgraft technique in patients with urethral plate less than 8 mm to repair distal hypospadias is still debatable. Some authors assume that augmentation may be beneficial. We aimed to compare the outcomes of the Snodgrass vs Snodgraft procedure in patients with a narrow urethral plate less than 8 mm. Methodology: This prospective randomized study included 60 children who had been treated by the Snodgrass or Snodgraft procedure for repair of distal penile hypospadias with narrow urethral plate from March 2017 to September 2018. They were randomized into two subgroups. Group 1 (30 patients) underwent tubularized incised plate urethroplasty, whereas the second group (30 patients) underwent the Snodgraft procedure by using the inner prepuce. Operative details, postoperative period, and complications were reported and statistically analyzed using IBM SPSS software package version 20.0.

Result(s): The operative time was longer for patients who underwent the Snodgraft procedure: 78 (55-95) and 110 (80-140) minutes in groups 1 and 2, respectively. In group 1, there was one case of meatal stenosis which was resolved by urethral dilation using the local anesthetic cream in the outpatient clinic. In addition, there was another case of distal penile fistula. In group 2, there was a case of complete wound disruption and another of distal penile fistula. There was no significant difference in the complication rate in any group.

Conclusion(s): The operative time was longer in group 2 than in group 1 but with comparable outcomes. The Snodgraft procedure is not superior to the Snodgrass operation in the narrow healthy urethral plate. [Table presented]

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Institution

(Eldeeb, Nagla, Abou-Farha, Hassan) Urology Department, Tanta University, Egypt

Publisher

Elsevier Ltd

Year of Publication
2020

62.

Disorder of sex development with germ cell tumors: Which is uncovered first?.

Faure-Contier C., Orbach D., Fresneau B., Verite C., Bonneau J., Thebaud E., Poiree M.,
Thouvenin S., Pluchart C., Mure P.Y., Dijoud F., Morel Y.

Embase

Pediatric Blood and Cancer. 67 (4) (no pagination), 2020. Article Number: e28169. Date of
Publication: 01 Apr 2020.

[Article]

AN: 2004202394

Background: Disorders of sex development (DSD) are rare conditions. Although they are known to predispose to germ cell tumors (GCT), there is a paucity of information regarding the circumstances of DSD discovery. Design/methods: All patients with DSD registered in two French pediatric GCT protocols (TGM95 and 13) were analyzed.

Result(s): Sixteen patients were identified among 276 ovarian, 160 testicular, and 24 mediastinal GCT. Eleven phenotypic females (median age 15 years) exhibited gonadal GCT, including 10 with a 46,XY karyotype and gonadal dysgenesis and one with 46XX,45X0 mosaicism. None had genital anomalies, seven had spontaneous pubertal changes, and one had spontaneous menarche. The tumors were bilateral in four cases. DSD was diagnosed after the GCT diagnosis in seven cases. The reasons for karyotyping were bilateral tumors (3), gonadoblastoma/streak gonad/absence of egg follicles (3), or systematic for GCT (1). The karyotyping was performed before the GCT diagnosis in four cases: for polymalformative syndrome (2) or primary amenorrhea (2). Four males (median age 14 years) exhibited mediastinal GCT (metastatic in two cases) indicative of Klinefelter syndrome, despite typical phenotypes in all cases. The remaining patient had severe hypospadias, leading to the discovery of 46,XY/45,X0 mosaicism before the diagnosis of testicular nonseminomatous GCT at 16 years of age.

Conclusion(s): DSD are often uncovered at the time of GCT diagnosis (11/16 cases). This should prompt oncologists to rule out a DSD in patients with GCT, even in case of pubertal development. Earlier recognition of Klinefelter syndrome could potentially lead to GCT detection at an earlier stage.

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Status

Embase

Author NameID

Faure-Contier, Cecile; ORCID: <http://orcid.org/0000-0002-4717-4943> Orbach, Daniel; ORCID:

<http://orcid.org/0000-0002-2520-139X>

Fresneau, Brice; ORCID: <http://orcid.org/0000-0001-7603-7828>

Institution

(Faure-Contier) Institut d'Hematologie et d'Oncologie Pediatrique, Lyon, France (Orbach) Institut Curie, SIREDO Oncology Center, PSL University, Paris, France

(Fresneau) Gustave Roussy Cancer Campus, Pediatric Oncology Unit, Villejuif, France

(Verite) Department of Pediatric Oncology, Centre hospitalier universitaire, Bordeaux, France

(Bonneau) Department of Pediatric Oncology, Centre hospitalier universitaire, Rennes, France

(Thebaud) Department of Pediatric Oncology, Centre hospitalier universitaire, Nantes, France

(Poiree) Department of Pediatric Oncology, Centre hospitalier universitaire, Nice, France

(Thouvenin) Department of Pediatric Oncology, Centre hospitalier universitaire, Saint Etienne, France

(Pluchart) Department of Pediatric Oncology, Centre hospitalier universitaire, Reims, France

(Mure) Department of Pediatric Surgery, Hospices Civils de Lyon, Bron, France

(Dijoud, Morel) Institut de Pathologie Multisite, Groupement hospitalier Est, Hospices Civils de Lyon, UCBL Lyon 1 University, Lyon, France

Publisher

John Wiley and Sons Inc. (P.O.Box 18667, Newark NJ 07191-8667, United States)

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63.

Fertility in adult men born with hypospadias: A nationwide register-based cohort study on birthrates, the use of assisted reproductive technologies and infertility.

Skarin Nordenvall A., Chen Q., Norrby C., Lundholm C., Frisen L., Nordenstrom A., Almqvist C., Nordenskjold A.

Embase

Andrology. 8 (2) (pp 372-380), 2020. Date of Publication: 01 Mar 2020.

[Article]

AN: 2003653100

Background: Fertility in men with hypospadias may be affected due to anatomical, surgical, or etiological factors and associated conditions. Fertility is further influenced by psychosocial and genetic factors, often shared within families.

Objective(s): To evaluate fertility in men born with hypospadias and assess confounding by familial factors.

Material(s) and Method(s): A population-based cohort of 1.2 million men born in Sweden 1964-1998, identified through national demographic and healthcare registers. Associations between hypospadias and (a) being a biological father, (b) conceiving through ART, and (c) diagnosis of male infertility were investigated in the full cohort with logistic regression models and Cox proportional hazard models, expressed as odds ratios (ORs) and hazard ratios (HRs), respectively, with 95% confidence intervals (CIs). A stratified proportional hazard model, conditional on sibling group, was used to control for shared familial confounding.

Result(s): Men with hypospadias, as a whole group, had a lower probability of having biological children (adjusted HR 0.87, 95% CI 0.83-0.92). A significant association was present in both distal (adjusted HR 0.90, 95% CI 0.85-0.96) and proximal hypospadias (HR 0.59, 95% CI 0.42-0.81). Men with hypospadias more often became fathers through ART, regardless of concomitant cryptorchidism. The initial association between hypospadias and the diagnosis of infertility disappeared in sensitivity analyses excluding cryptorchidism.

Discussion(s): Men with hypospadias displayed lower birthrates as compared to their brothers and the general population. Mere birthrates may, however, be a questionable measure of fertility in a population using family planning. However, men with hypospadias were also at higher risk of reproducing through ART and did more often receive a diagnosis of male infertility. Altogether, these findings indicate impaired fertility in men with hypospadias.

Conclusion(s): Fertility in men with hypospadias is impaired, as shown by lower birthrates, increased use of ART and higher risk of receiving a diagnosis of male infertility.

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Status

Embase

Author NameID

Skarin Nordenvall, Anna; ORCID: <http://orcid.org/0000-0003-3034-9222>

Institution

(Skarin Nordenvall, Nordenstrom, Nordenskjold) Department of Women's and Children's Health, Karolinska Institutet, Stockholm, Sweden (Skarin Nordenvall, Chen, Norrby, Lundholm, Almqvist) Department of Medical Epidemiology and Biostatistics, Karolinska Institutet, Stockholm, Sweden

(Frisen) Department of Clinical Neuroscience, Karolinska Institutet, Stockholm, Sweden

(Frisen) Child and Adolescent Psychiatry Research Center, Stockholm, Sweden

(Nordenstrom) Department of Pediatric Endocrinology, Astrid Lindgren Children's Hospital, Karolinska University Hospital, Stockholm, Sweden

(Almqvist) Pediatric Allergy and Respiratory Medicine, Astrid Lindgren Children's Hospital, Karolinska University Hospital, Stockholm, Sweden

(Nordenskjold) Center for Molecular Medicine, Karolinska Institutet, Stockholm, Sweden

(Nordenskjold) Department of Pediatric Surgery, Astrid Lindgren Children's Hospital, Karolinska University Hospital, Stockholm, Sweden

Publisher

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64.

Penile Disassembly in Complete Primary Repair of Bladder Exstrophy: Time for Re-evaluation?.

Kasprinski M., Maruf M., Davis R., Jayman J., Benz K., Michaud J., Di Carlo H., Dunn E.A., Gearhart J.P.

Embase

Urology. 137 (pp 146-151), 2020. Date of Publication: March 2020.

[Article]

AN: 2004743838

OBJECTIVE: To explore a series of classic bladder exstrophy (CBE) cases referred to the authors' institution where primary closure with penile disassembly epispadias repair was complicated by penile injury. The penile disassembly technique is frequently combined with

bladder closure in patients with CBE undergoing the complete primary repair of exstrophy (CPRE). Penile disassembly has been posited as a risk for penile injury by ischemic mechanisms.

METHOD(S): A prospectively-maintained institutional database of 1337 exstrophy-epispadias complex patients was reviewed for CPRE cases referred to the authors' institution, and those with injury to the penis were identified. The location, extent of injury, and subsequent management is reported.

RESULT(S): One hundred and thirteen male CBE patients were referred after prior CPRE. Twenty-six (20%) were identified with penile loss and reviewed. Eighty-one percent were closed in the neonatal period, and 54% had a pelvic osteotomy. Median follow-up time was 9.9 years (range 0.6-21.3). Of 26 patients with penile loss, 77% had unilateral loss and in 23% had bilateral loss involving the glans and/or one or both corpora cavernosa. Three patients were successfully managed with myocutaneous neophalloplasty.

CONCLUSION(S): Complete penile disassembly during bladder exstrophy closure may lead to penile injury. This major complication questions the continued application of complete penile disassembly in the reconstruction of bladder exstrophy.

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Status

Embase

Institution

(Kasprenski, Maruf, Davis, Jayman, Benz, Michaud, Di Carlo, Gearhart) Division of Pediatric Urology, James Buchanan Brady Urological Institutions, Johns Hopkins Hospital, Johns Hopkins Medical Institutions, Charlotte Bloomberg Children's Hospital, Baltimore, MD, United States (Dunn) Division of Pediatric Radiology, Russell H. Morgan Department of Radiology and Radiological Science, Johns Hopkins Hospital, Baltimore, MD, United States

Publisher

Elsevier Inc. (E-mail: usjcs@elsevier.com)

Year of Publication

2020

Structural Analysis of the Bulbospongiosus Muscle in Patients With Bulbar Urethral Strictures.

Alves E.F., Gallo C.M., Costa W.S., Sampaio F.J., Favorito L.A.

Embase

Urology. 137 (pp 183-189), 2020. Date of Publication: March 2020.

[Article]

AN: 2004678391

OBJECTIVE: To characterize the bulbospongiosus muscle (BSM) in patients with bulbar urethral strictures. MATERIALS AND METHODS: We studied 21 patients divided into 2 groups: Stricture Group (n = 14; mean age = 62.00 years) with bulbar stricture submitted to open urethroplasty; and Control Group (n = 7; mean age = 60.14 years) with penile strictures (hypospadias cripples, penile cancer and/or penile infection) who were submitted to perineal urethrostomy. Samples of the BSM were dissected and histologic sections were stained by histochemical and immunohistochemical techniques. Histomorphometric analyzes were performed on photomicrographs. Means were statistically compared using the unpaired Student t test and the Mann-Whitney test (P <.05).

RESULT(S): The etiology of bulbar urethral stricture was idiopathic in 2 cases (14.29%), post-TURP in 6 (42.86%), post open radical prostatectomy in 5 (35.71%) and post open prostatectomy in 1 case (7.14%). The average length of the stricture was 2.08 cm. The only parameter analyzed with significant difference between the groups was the vessels (significant difference between the control group: 5.11 +/- 1.98% and stricture group: 3.57 +/- 1.32%, P =.0460). The quantitative analysis of collagen (Control Group: 10.63 +/- 5.37% and Stricture Group: 10.83 +/- 4.55%, P =.9296); diameter of BSM muscle fibers (Control Group: 41.71 +/- 14.63 micro m and Stricture Group: 40.11 +/- 8.59 micro m, P =.76 and elastic system fibers (Control Group; 3.83 +/- 1.54% and Stricture Group: 5.43 +/- 2.90%, P =.2601) showed no significant difference.

CONCLUSION(S): Histologic analysis showed a significant decrease of the BSM vessels in urethral stricture, without changes in elastic fibers, collagen, nerves, and muscle fiber diameter. These findings show that the bulbar urethral stricture causes minimal alterations in the structure of the BSM.

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Institution

(Alves, Gallo, Costa, Sampaio) Universidade do Estado do, Rio de Janeiro, Faculdade de Ciencias Medicas, Rio de janeiro, Brazil (Favorito) State University Rio de janerio, Rio de janeiro, Brazil

Publisher

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Year of Publication

2020

66.

Modern Management of the Failed Bladder Exstrophy Closure: A 50-yr Experience.

Kasprenski M., Benz K., Maruf M., Jayman J., Di Carlo H., Gearhart J.

Embase

European Urology Focus. 6 (2) (pp 383-389), 2020. Date of Publication: 15 March 2020.

[Article]

AN: 2001152705

Background: A failed closure of classic bladder exstrophy (CBE) has a negative long-term impact on the patient and the health care system.

Objective(s): To investigate the outcomes of CBE patients with failed primary bladder closure.

Design, setting, and participants: A database of 1317 exstrophy-epispadias complex patients was retrospectively reviewed for CBE patients with failed primary bladder closure from 1965 to 2017 with subsequent repeat closure.

Intervention(s): Repeat bladder exstrophy closure and subsequent continence procedure.

Outcome measurements and statistical analysis: Failed exstrophy closures are defined as occurrence of bladder prolapse, dehiscence, vesicocutaneous fistula, outlet obstruction, or combination of these factors. Successful repeat closures are defined as closures that require no further operative intervention as a consequence of these factors. Kaplan-Meier to determine time to successful repeat closure and receiver operator characteristic curve to determine the optimal time for secondary closure were determined. Results and limitations: In total, 170 CBE patients had at least one repeat closure following a failed primary closure (115 male/55 female). With continued closure attempts, 166/170 (97.6%) patients were successfully closed. The median time to successful closure from birth was 12.9 mo (95% confidence interval: 11.7-15.7). Furthermore, 52/153 (34%) patients had more than one osteotomy. Of 215 total osteotomies, 50 (29.4%) were performed during the 170 failed primary closures, 128 (75.3%) during the 170 second closures, and 27 (64.3%) during the 42 third closures. Of 96 patients with available continence data, 74 (77.1%) achieved urinary continence.

Conclusion(s): A successful repeat closure is possible, especially when used in conjunction with a pelvic osteotomy. Continent urinary diversion yielded the highest continence rate in this cohort.

Patient Summary: We looked at outcomes of classic bladder exstrophy closure in a large population. Successful repeat closure is possible in the majority of cases when used with pelvic osteotomy. A majority of patients achieved urinary continence using a continent diversion. Especially when performed in conjunction with osteotomy, a successful repeat bladder exstrophy closure is possible following a prior failed attempt. The majority of patients are able to achieve urinary continence after a prior failed closure using a continent diversion.

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Author NameID

Gearhart, John; ORCID: <http://orcid.org/0000-0002-0177-6121>

Institution

(Kasprenski, Benz, Maruf, Jayman, Di Carlo, Gearhart) Robert D. Jeffs Division of Pediatric Urology, James Buchanan Brady Urological Institutions, Johns Hopkins Hospital, Johns Hopkins Medical Institutions, Charlotte Bloomberg Children's Hospital, Baltimore, MD, United States

Publisher

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Year of Publication

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67.

Epidemiology of birth defects based on a birth defect surveillance system in Southern Jiangsu, China, 2014-2018.

Zhou Y., Mao X., Zhou H., Qin Z., Wang L., Cai Z., Yu B.

Embase

Journal of Maternal-Fetal and Neonatal Medicine. (no pagination), 2020. Date of Publication: 2020.

[Article]

AN: 2004337280

Introduction: Changzhou has been confronted with great challenges in birth defects (BDs) prevention, as the prevalence rates of BDs in Changzhou increased rapidly. The aims of this study were to describe the epidemiology of BDs in perinatal infants (PIs, including dead fetus, stillbirth, or live birth between 28 weeks of gestation and 7 days after birth) in Changzhou during the period from 2014 to 2018.

Method(s): The BD surveillance data of PIs were collected from 56 hospitals of Changzhou. The prevalence rate of BDs with 95% confidence interval (CI) were calculated by Poisson distribution. Univariate and multivariate Poisson regression was performed to identify the changing trends of prevalence rates of BDs by year and the association of regarding BD characteristics including year, infant gender, maternal age, and season with BDs successively.

Result(s): From 2014 to 2018, there were a total of 238,712 PIs of which 1707 had BDs, with the average prevalence of 71.509 per 10,000 PIs, showing a remarkable uptrend (aPRR = 1.133, 95%CI: 1.094-1.173). The ten leading BDs were polydactyly, congenital heart defects (CHD), syndactyly, microtia, cleft lip and palate (CLP), hypospadias, cleft palate, other malformation of external ear (OMEE), congenital atresia of rectum and anus, and congenital talipes equinovarus (CTE). During the study period, the prevalence rates of polydactyly, CHD and syndactyly increased significantly (PRR = 1.195, 95%CI: 1.109-1.288, PRR = 1.194, 95%CI: 1.105-1.291, and PRR = 1.143, 95%CI: 1.007-1.297, respectively); the prevalence rates of congenital esophageal atresia decreased significantly (PRR = 0.571, 95%CI: 0.395-0.826). The risk of BDs was higher in male PIs versus female PIs (aPRR = 1.235, 95%CI: 1.123-1.358).

Conclusion(s): A significant increase in the prevalence of BDs was detected from 2014 to 2018 in Changzhou. CHD, polydactyly, and syndactyly increased much and congenital esophageal atresia declined much. Male PIs was risk factor for occurrence of BDs. Collecting information on factors associated with BDs, setting the report time of BDs system at smaller gestational age so as to get an exact prevalence and make better prevention strategy, strengthening the publicity and education, improving the ability of monitoring, and wider use of new diagnosis technology are important to reduce the prevalence of BDs in PIs.

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Status

Article-in-Press

Institution

(Zhou, Mao, Zhou, Qin, Wang, Yu) Changzhou Maternity and Child Health Care Hospital affiliated to Nanjing Medical University, Changzhou, China (Cai) Changzhou Commission of Health, Changzhou, China

Publisher
Taylor and Francis Ltd
Year of Publication
2020

68.

Proximal Hypospadias: Isolated Genital Condition or Marker of More?.

Johnson E.K., Jacobson D.L., Finlayson C., Yerkes E.B., Goetsch A.L., Leeth E.A., Cheng E.Y.

Embase

The Journal of urology. (pp 101097JU0000000000000810), 2020. Date of Publication: 24 Feb 2020.

[Article]

AN: 631044348

PURPOSE: The prevalence of endocrine/genetic anomalies among boys with proximal hypospadias is unknown. This study aims to describe the endocrine/genetic evaluation for boys with proximal hypospadias to inform who may have a difference/disorder of sex development and/or benefit from additional testing.

METHOD(S): Boys with hypospadias seen at our hospital from 1/2013-10/2018 were retrospectively reviewed. Those with proximal (penoscrotal, scrotal, perineal) hypospadias who presented at <6 months old and underwent endocrine/genetic testing were included.

Demographics, test results, testicular exam at presentation, comorbidities, and diagnoses were abstracted.

RESULT(S): 1789 boys with hypospadias were identified. Of 131 boys with proximal hypospadias, all 60 who underwent endocrine/genetic evaluation were included. Most had bilateral palpable testes (52/60, 86%) that were fully descended (41/60, 68%). Associated anatomic anomalies were found in 53%. All boys underwent endocrine testing, which was completely typical for an infant male in most (41/60, 68%). Common genetic tests included karyotyping (100%), Microarray (38%), and multi-gene panel (13%). Genetic anomalies were found in 17 boys (28%): 7/41 (17%) with bilateral descended testes and 10/19 (53%) with ≥ 1 undescended testis ($p=0.01$). Most boys (6/8) with ≥ 1 non-palpable testis had a genetic anomaly (vs. 11/52 with bilateral palpable testes; $p=0.005$). Differences/disorders of sex development were found in 9 (15%).

CONCLUSION(S): Among 60 boys with proximal hypospadias, 53% had non-genital anomalies, 28% had genetic anomalies, and 15% had a difference/disorder of sex development. Although endocrine testing was clinically useful, genetic testing was most diagnostically revealing. Endocrine/genetic evaluation should be considered for boys with proximal hypospadias.

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Institution

(Johnson, Jacobson, Yerkes, Cheng) Department of Urology, Northwestern University, Evanston, IL, United States (Johnson, Jacobson, Yerkes, Cheng) Division of Pediatric Urology, Ann & Robert H. Lurie Children's Hospital of Chicago, Chicago, IL, Mexico

(Finlayson, Goetsch, Leeth) Department of Pediatrics, Northwestern University, Evanston, IL, United States

(Finlayson) Division of Pediatric Endocrinology, Ann & Robert H. Lurie Children's Hospital of Chicago, Chicago, IL, Mexico

(Goetsch) Division of Genetics, Birth Defects & Metabolism, Ann & Robert H. Lurie Children's Hospital of Chicago, Chicago, IL, Mexico

(Leeth) Department of Pathology and Laboratory Medicine, Ann & Robert H. Lurie Children's Hospital of Chicago, Chicago, IL, Mexico

Publisher

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2020

69.

Fertility and sexuality issues in congenital lifelong urology patients: male aspects.

Spinoit A.-F., Waterschoot M., Sinatti C., Abbas T., Callens N., Cools M., Hamid R., Hanna M.K., Joshi P., Misseri R., Salle J.L.P., Roth J., Tack L.J.W., De Win G.

Embase

World journal of urology. (no pagination), 2020. Date of Publication: 17 Feb 2020.

[Article]

AN: 630991198

PURPOSE: To review existing literature about fertility and sexuality of boys born with complex congenital genitourinary anomalies.

METHOD(S): A Pubmed review was performed in December 2018 to identify the most relevant original manuscripts regarding male complex congenital conditions affecting the urogenital system in male patients including spina bifida (SB), bladder exstrophy-epispadias complex (BEEC) and hypospadias. A comprehensive review was drafted exploring sexual dysfunction from a medical, psychosexual, surgical and reproductive point of view during transition from childhood (or adolescence) to adulthood.

RESULT(S): About 75% of men with SB have erectile dysfunction (ED) (Game et al. in Urology 67(3):566-570, 2006; Diamond et al. in 58(4):434-435, 1986). Most SB patients have impaired sexual development mainly due to diminished self-esteem, dependence on caregivers and lack of privacy (Blum et al. in Pediatrics 88(2):280-285, 1991). Men with BEEC have fewer intimate relationships than women because of the greater difficulties with issues regarding their genitalia and sexual activities (Deans et al. in Am J Obstet Gynecol 206(6):496.e1-496.e6, 2012).

However, a good quality of life is achievable with the effective use of coping strategies (Deng et al. in Transl Androl Urol 7:941, 2018; Rikken et al. in BMC Womens Health 18(1):163, 2018; Friedler et al. in Reprod Biomed Online 32(1):54-61, 2016). Chordee occurs in 25% of all hypospadias patients. More severe hypospadias is related to a greater risk for complications. The long-term sexual quality of life (QoL) in men who underwent hypospadias surgery is influenced by a lot of factors. Therefore, an interactive and dynamic biopsychosocial model of sexual QoL was proposed.

CONCLUSION(S): The care of patients with congenital urologic conditions becomes a challenge especially in the period of 'transition'. The goal of follow-up is a holistic management viewed from a medical, psychosexual, surgical end reproductive point. All patients should be asked for specific urinary, fecal or sexual concerns.

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Author NameID

Spinoit, Anne-Francoise; ORCID: <http://orcid.org/0000-0002-9045-9127>

Institution

(Spinoit, Waterschoot, Sinatti) Department of Urology, Ghent University Hospital, Ghent, Belgium

(Abbas) Pediatric Urology, Sidra Hospital, Doha, Qatar

(Callens) Centre for Research on Culture and Gender, Department of Languages and Cultures, Ghent University, Ghent, Belgium

(Cools) Department of Paediatric and Adolescent Endocrinology and Diabetology, Ghent University Hospital, Ghent, Belgium

(Hamid) Department of Neuro-Urology, Royal National Orthopaedic Hospital, Stanmore, Middlesex, UK

(Hamid) University College London Hospitals, London, United Kingdom

(Hanna) New York Presbyterian/Weill Cornell Medical Center, NY, United States

(Joshi) Kulkarni Reconstructive Urology Centre, Pune, India

(Misseri) Pediatric Urology, Riley Hospital for Children at Indiana University Health, Indiana University School of Medicine, IN, Indianapolis, United States

(Salle) Division of Urology, Department of Surgery, Sidra Medical and Research Center, Doha, Qatar

(Roth) Paediatric Urology, Riley Hospital for Children, Indiana University Health, IN, Indianapolis, United States

(Tack) Division of Paediatric Endocrinology, Department of Internal Medicine and Paediatrics, Ghent University Hospital, Ghent, Belgium

(De Win) Faculty of Medicine and Health Science, University of Antwerp, University Hospital Antwerp, Antwerp, Belgium

(De Win) Department of Urology, University of College London Hospitals, London, United Kingdom

Publisher

NLM (Medline)

Year of Publication

2020

70.

Comparing the outcomes of tubularized incised plate urethroplasty and dorsal inlay graft urethroplasty in children with hypospadias: a systematic review and meta-analysis.

Alshafei A., Cascio S., Boland F., O'Shea N., Hickey A., Quinn F.

Embase

Journal of Pediatric Urology. (no pagination), 2020. Date of Publication: 2020.

[Review]

AN: 2004913485

Background: Excellent outcomes have been reported following the widely accepted tubularized incised plate urethroplasty (TIPU) and its relatively recent modification, the dorsal inlay graft

urethroplasty (DIGU). However, there is a lack of consensus on which technique offers more favorable postoperative outcomes.

Aim(s): To systematically compare the reported outcomes of the TIPU and DIGU techniques in children undergoing primary hypospadias repair.

Design(s): A systematic review and meta-analysis of randomized and observational studies.

Method(s): An electronic database search was conducted up to May 2018. Sources included Medline, Embase, Cochrane library, CINAHL, Web of Science, and Google Scholar as well as trial registries and grey literature sources. Studies were selected if they compared the postoperative complications of TIPU and DIGU in children. Secondary outcomes included standardized cosmetic scores and urinary flow studies. A meta-analysis of reported complications was performed using a random-effects model.

Result(s): Two randomized, two prospective, and two retrospective studies met the inclusion criteria. TIPU and DIGU were performed in 350 and 267 patients, respectively. Pooled analysis did not demonstrate a significant difference regarding postoperative urethrocuteaneous fistula, meatal/urethral stenosis, wound dehiscence, or total complications. Subgroup analysis according to hypospadias severity did not alter initial findings. Statistical analysis of secondary outcomes was not feasible due to insufficient data. Most studies were of low methodological quality with a high risk of bias.

Conclusion(s): There is no strong evidence to suggest that either technique offers more favorable outcomes. Until more robust randomized trials exist, decisions regarding the appropriate repair should be based on the surgeon's experience and outcomes.

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Status

Article-in-Press

Institution

(Alshafei, Cascio, O'Shea, Quinn) Our Lady's Children's Hospital, Crumlin, Cooley Road, Dublin 12, Ireland (Boland, Hickey) Division of Population Health Science, Royal College of Surgeons in Ireland, Dublin 2, Ireland

Publisher

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2020

71.

Knowledge gaps and information seeking by parents about hypospadias.

Chan K.H., Panoch J., Carroll A., Wiehe S., Cain M.P., Frankel R.

Embase

Journal of Pediatric Urology. (no pagination), 2020. Date of Publication: 2020.

[Article]

AN: 2004913421

Introduction: Parents making complex decisions about hypospadias surgery may experience anxiety and uncertainty related to multiple sources of information with questionable reliability and limited relevance to their concerns.

Objective(s): The purpose of this study was to identify knowledge gaps, information-seeking behaviors, and informational needs of parents making decisions about hypospadias surgery as an initial step in the development of a hypospadias decision aid. Study design: We conducted semi-structured interviews with English-speaking parents (≥ 18 years of age) of children with hypospadias, inquiring about gaps in their knowledge, information-seeking behaviors, and perceived informational needs (Extended Summary Table). We conducted interviews until no new themes were identified, analyzing them iteratively using open, axial, and selective coding. We used grounded theory methods to develop an explanation of the information-seeking process about hypospadias surgery.

Result(s): Of the 43 eligible parents, 16 mothers and 1 father (39.5%) of 16 patients participated: 7 preoperative and 9 postoperative with distal (8) and proximal (8) meatal locations. Parents were aged 21-43: 15 Caucasians and 2 African-Americans. Educational backgrounds and marital status varied across subjects. We identified five categories of knowledge gaps relating to hypospadias surgery: 1) epidemiology, 2) timing/technique, 3) perioperative experience, 4) long-term cosmetic outcome, and 5) long-term risk of complications. Information-seeking behaviors included searching the internet, discussing hypospadias with the child's pediatrician and/or urologist, and obtaining information from their social network. Most parents sought information online prior to and/or after consultation with the urologist, from parent blogs/forums, medical school/hospital websites, journal articles, and medical databases. Perceived informational needs included clear and reliable information online, images of mild degrees of hypospadias, and images of repaired hypospadias cases. According to the parents, video testimonials from other parents would help them relate to others in their social network and build confidence about the surgical process.

Discussion(s): The findings of this study contribute to our understanding of parental decision-making about hypospadias surgery by highlighting specific knowledge gaps and informational needs for inclusion in a decision aid. Study limitations include a small sample size that is typical and expected for qualitative research studies and the underrepresentation of fathers, minorities, and same-sex couples.

Conclusion(s): The Internet is the primary source of information most parents use to address knowledge gaps about hypospadias. Parents expressed concerns about the reliability and clarity of information and identified informational needs including parent testimonials and a wide variety of hypospadias images for inclusion in a decision aid. [Table presented]

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Author NameID

Chan, Katherine H.; ORCID: <http://orcid.org/0000-0001-9124-1704>

Institution

(Chan, Panoch, Cain) Department of Urology, Indiana University School of Medicine, United States (Chan, Carroll) Department of Pediatrics, Center for Pediatric and Adolescent

Comparative Effectiveness Research, United States

(Wiehe) Department of Pediatrics, Children's Health Services Research, United States

(Frankel) Department of Medicine, Indiana University School of Medicine, Indianapolis, IN, United States

(Frankel) Cleveland Clinic Learner Institute, Cleveland, OH, United States

Publisher

Elsevier Ltd

Year of Publication

2020

72.

Delayed presentation of urethrocutaneous fistulae after hypospadias repair.

Johnston A.W., Jibara G.A., Purves J.T., Routh J.C., Wiener J.S.

Embase

Journal of Pediatric Surgery. (no pagination), 2020. Date of Publication: 2020.

[Article]

AN: 2004912323

Background: Delayed urethrocutaneous fistula (UCF) presentation after hypospadias repair is rarely reported. The aim of this study is to report our experience with delayed UCF presenting more than 5 years after hypospadias repair.

Method(s): We conducted a retrospective review of patients who underwent UCF repair (CPT codes 54,340 and 54,344) at our institution between 1997 and 2017. Delayed UCF presentation was defined as a single normal urinary stream after initial hypospadias repair and subsequent presentation of a UCF/s urinary stream more than 5 years after initial hypospadias or UCF repair. Demographic and clinical data were reviewed after approval from our institutional review committee.

Result(s): We identified 12 patients with delayed UCF. The mean age at hypospadias repair was 12.3 months (Range 6-32). The mean time to delayed UCF presentation was 11.5 years (Range 7.1-15.8). Four patients with delayed UCF (33.3%) required additional surgery for UCF recurrence with a mean time to recurrence of 2.2 years (Range < 1-5.6).

Conclusion(s): Delayed UCF presentation can occur more than 15 years after initial repair. Pubertal penile skin changes and increased genital awareness in older children may be contributing factors as all but one presented at age 10 years or older.

Level of Evidence: III

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Institution

(Johnston, Jibara, Purves, Routh, Wiener) Division of Urologic Surgery, Duke University Medical Center, Durham, NC, United States

Publisher

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Year of Publication

2020

73.

Increased hand digit length ratio (2D:4D) is associated with increased severity of hypospadias in pre-pubertal boys.

O'Kelly F., DeCotiis K., Zu'bi F., Farhat W.A., Koyle M.A.

Embase

Pediatric Surgery International. 36 (2) (pp 247-253), 2020. Date of Publication: 01 Feb 2020.

[Article]

AN: 2003653165

Introduction: Hypospadias is a common congenital male disorder, with much research focusing on prenatal androgen exposure as a causative factor. Whilst digit length ratios were apparent in sexual dimorphism since the nineteenth century, their role in hypospadias remains unknown. The objective of our study was to determine the correlation between digit length (2D:4D) ratio, hypospadias severity, and anogenital distance.

Method(s): Pre-pubertal boys (<3 years old) seen intra/postoperatively following hypospadias repair (June 2018-January 2019 inc.) were included. These were age-matched to non-hypospadias controls. Anthropomorphic measurements of digit lengths, penile/glans width, and anogenital distance were measured using digital calipers.

Result(s): Data measurements were collected for 105 boys with hypospadias (60 distal; 45 proximal) and 55 controls. There were significant differences in 2D:4D ratios in each hand ($p < 0.001$), as well as individual digits ($p < 0.001$), and a reduced anogenital distance ($p < 0.001$), when comparing the proximal group with distal or control groups. There were no significant differences in glans width, or between term- and preterm births.

Conclusion(s): This study is the first to demonstrate increased 2D:4D ratios with proximal hypospadias, which also correlate with a shortened anogenital distance. This may provide a non-invasive, potentially antenatal, anthropomorphic measurement, as an indirect indicator of aberrant urogenital development.

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Institution

(O'Kelly, DeCotiis, Zu'bi, Farhat, Koyle) Division of Urology, The Hospital for Sick Children, Toronto, ON M5G1X8, Canada

Publisher

Springer
Year of Publication
2020

74.

Utility of Skin Grafting and Tissue Expansion in Penile Reconstruction for the Exstrophy-Epispadias Complex.

Harris T.G.W., Maruf M., Barone A.A.L., Redett R.J., Gearhart J.P.

Embase

Urology. 136 (pp 231-237), 2020. Date of Publication: February 2020.

[Article]

AN: 2004207375

Objective: To describe the use of additional tissue recruited for coverage after penile lengthening in male exstrophy-epispadias complex patients using either local skin from tissue expansion (TE) or extragenital skin with a skin graft (SG) and report their respective outcomes.

Method(s): An institutionally approved database of exstrophy-epispadias complex patients was retrospectively reviewed for male patients who received penile reconstruction. This included a penile lengthening procedure and the subsequent use of TE and/or a full thickness skin graft to provide cutaneous coverage of gained corporal length.

Result(s): A total of 50 patients (mean age 18.1 years) underwent penile reconstruction. TE was used in 27 patients, SG in 19, and 4 received a combination of TE and SG. The mean number of previous penile operations was 2.7 for patients that received TE and 3.1 for SG. A successful outcome from primary reconstruction was achieved in 35 patients (70%) and overall successful reconstruction was achieved by 48 patients (96%).

Conclusion(s): TE and SG are useful techniques in providing soft tissue coverage following penile lengthening. TE is the preferred technique for primary reconstruction in a lengthening procedure. When genital skin is not expandable or coverage from TE is insufficient after lengthening, extragenital skin (SG) is recruited.

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Institution

(Harris, Barone, Redett) Department of Plastic and Reconstructive Surgery, Johns Hopkins University School of Medicine, Baltimore, MD, United States (Maruf, Gearhart) Jeffs Division of Pediatric Urology, Department of Urology, Brady Urological Institute, Johns Hopkins School of Medicine, Baltimore, MD, United States

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75.

Periconceptional exposure to air pollution and congenital hypospadias among full-term infants.

Huang C.-C., Pan S.-C., Chen B.-Y., Guo Y.L.

Embase

Environmental Research. 183 (no pagination), 2020. Article Number: 109151. Date of Publication: April 2020.

[Article]

AN: 2004729618

Background: Evidence regarding whether prenatal exposure to air pollution increases the risk of hypospadias remains limited. The aim of the study is to evaluate the association between exposure to ambient air pollution during early pregnancy and the incidence of hypospadias.

Method(s): We conducted a 1:10 case-control study using the Taiwanese Birth Registry database. Male full-term infants reported to have hypospadias were defined as cases, and controls were randomly selected from male full-term infants without any congenital anomaly. The monthly average of ambient air pollutants, including PM₁₀, PM_{2.5}, PM_{2.5-10}, NO₂, NO_x, O₃, and O₃ 8-h maximum, from 3 months before conception to 6 months post conception was retrieved from air quality monitoring stations and interpolated to the level of township using the kriging method. Multivariable logistic regression models were utilized to evaluate the associations.

Result(s): A total of 200 hypospadias cases, with 2000 healthy controls sampled, were reported during 2007-2014. The results revealed that PM_{2.5} exposure during the first 3 months after

conception (odds ratio [OR] = 1.29, 95% confidence interval [CI]: 1.01-1.65, per interquartile range [IQR] = 15.6 mug/m³) and O₃ exposure during the first month after conception (OR = 1.40, 95% CI: 1.08-1.82, per IQR = 8.0 ppb) were associated with a higher incidence of hypospadias.

Conclusion(s): The results of the study suggest that early gestational exposure to ambient air pollution increases the risk of hypospadias among full-term infants.

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Embase

Institution

(Huang, Guo) Department of Environmental and Occupational Medicine, National Taiwan University College of Medicine and Hospital, Taipei, Taiwan (Republic of China) (Pan, Guo) Institute of Environmental and Occupational Health Sciences, College of Public Health, National Taiwan University, Taipei, Taiwan (Republic of China)

(Chen) Department of Medical Research and Development, Chang Gung Memorial Hospital, Keelung, Taiwan (Republic of China)

(Guo) National Institute of Environmental Health Sciences, National Health Research Institutes, Miaoli, Taiwan (Republic of China)

Publisher

Academic Press Inc. (E-mail: apjcs@harcourt.com)

Year of Publication

2020

76.

Smith-Lemli-Opitz syndrome - Fetal phenotypes with special reference to the syndrome-specific internal malformation pattern.

Schoner K., Witsch-Baumgartner M., Behunova J., Petrovic R., Bald R., Kircher S.G., Ramaswamy A., Kluge B., Meyer-Wittkopf M., Schmitz R., Fritz B., Zschocke J., Laccone F., Rehder H.

Embase

Birth Defects Research. 112 (2) (pp 175-185), 2020. Date of Publication: 15 Jan 2020.

[Article]

AN: 2003909279

Background: Autosomal-recessive SLOS is caused by mutations in the DHCR7 gene. It is defined as a highly variable complex of microcephaly with intellectual disability, characteristic facies, hypospadias, and polysyndactyly. Syndrome diagnosis is often missed at prenatal ultrasound and fetal autopsy.

Method(s): We performed autopsies and DHCR7 gene analyses in eight fetuses suspected of having SLOS and measured cholesterol values in long-term formalin-fixed tissues of an additional museum exhibit.

Result(s): Five of the nine fetuses presented classical features of SLOS, including four cases with atrial/atrioventricular septal defects and renal anomalies, and one with additional bilateral renal agenesis and a Dandy-Walker cyst. These cases allowed for diagnosis at autopsy and subsequent SLOS diagnosis in two siblings. Two fetuses were mildly affected and two fetuses showed additional holoprosencephaly. These four cases and the exhibit had escaped diagnosis at autopsy. The case with bilateral renal agenesis presented a novel combination of a null allele and a putative C-terminus missense mutation in the DHCR7 gene.

Conclusion(s): In view of the discrepancy between the prevalence of SLOS among newborns and the carrier frequency of a heterozygous DHCR7 gene mutation, the syndrome-specific internal malformation pattern may be helpful not to miss SLOS diagnosis in fetuses at prenatal ultrasound and fetal autopsy.

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Author NameID

Rehder, Helga; ORCID: <http://orcid.org/0000-0002-5215-623X>

Institution

(Schoner, Ramaswamy, Rehder) Institute of Pathology, Philipps-University Marburg, Marburg, Germany (Witsch-Baumgartner, Zschocke) Institute of Human Genetics, Medical University Innsbruck, Innsbruck, Austria

(Behunova, Kircher, Kluge, Laccone, Rehder) Institute of Medical Genetics, Medical University Vienna, Vienna, Austria

(Petrovic) Institute of Medical Biology, Comenius University Bratislava, Bratislava, Slovakia

(Bald) Clinic of Gynecology and Obstetrics, Klinikum Leverkusen, Leverkusen, Germany

(Meyer-Wittkopf) Clinic of Gynecology and Obstetrics, University Clinic Oldenburg, Oldenburg, Germany

(Schmitz) Clinic of Gynecology and Obstetrics, University Clinic Muenster, Munster, Germany
(Fritz) Institute of Human Genetics, Philipps-University Marburg, Marburg, Germany

Publisher

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Year of Publication

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77.

Recurrent chordee in 59 adolescents and young adults following childhood hypospadias repair.

Abosena W., Talab S.S., Hanna M.K.

Embase

Journal of Pediatric Urology. (no pagination), 2020. Date of Publication: 2020.

[Article]

AN: 2004658517

Background: As children transition to adolescence, penile curvature may recur several years, sometimes, decades later. Herein we review our experience with a group of symptomatic patients, their surgical repairs and outcome.

Material(s) and Method(s): Reviews were done on the charts of 59 symptomatic adolescents aged 14-21 years who presented with recurrent penile curvature, causing either sexual dysfunction or significant deformity and had undergone surgical correction between 2000 and 2017. Their initial hypospadias repairs were: TIP & dorsal midline plication (28), Tiersch-Duplay (T-D) urethroplasty and Nesbit dorsal repair [9], prepuce mucosal island onlay and Nesbit repair [6], Two-stage Byar repair and ventral dermal graft (3 patients). 13/59 patients were repaired elsewhere, and their records were unavailable. Surgical correction included one stage dorsal replication and skin detethering (32patients), one-stage urethral mobilization and corporal/dermal grafts (12patients), staged corporal/dermal graft and skin coverage followed by urethroplasty (Grafted TIP) 8-12 months later (15patients), and 11/59 had urethral fistula, which were repaired concomitantly.

Result(s): 55 out of 59 patients were followed up for 6-48 months (median 30 months) by periodic office visits, and 2-4 weeks in 4 patients whose subsequent follow up was by email. Of the 55 patients, 53 healed well, and 2patients developed wound breakdown and scarring, which resulted in mild recurrent curvature but to a lesser degree than preoperatively. The other 4 patients who

corresponded by email were pleased with the surgical outcome. Of the total 59 patients, 28 reported satisfactory sexual activity. None of the patients who had corporal/dermal grafts reported erectile abnormalities.

Conclusion(s): Recurrent curvature in adolescents following hypospadias repair, may be caused by peri-urethral and skin fibrosis and/or disproportionate growth of the relatively hypoplastic ventral corporal wall or the reconstructed urethra. Surgical correction of symptomatic patients by dorsal shortening or ventral lengthening procedures depends on the degree of curvature following skin degloving of the penis. We have been recommending to parents of children born with proximal hypospadias who had what appeared to be a good surgical result to follow up after puberty. [Table presented]

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Institution

(Abosena) Tanta University, Tanta, Egypt (Talab) Rutgers New Jersey Medical School Newark, NJ, United States

(Abosena, Hanna) New York Presbyterian Weill-Cornell Hospital, New York, United States

Publisher

Elsevier Ltd

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78.

Time to Complication Detection after Primary Pediatric Hypospadias Repair: A Large, Single-Center, Retrospective Cohort Analysis.

Lucas J., Hightower T., Weiss D.A., Van Batavia J., Coelho S., Srinivasan A.K., Shukla A.R., Zderic S.A., Kolon T.F., Zaontz M.R., Canning D.A., Long C.J.

Embase

The Journal of urology. (pp 101097JU00000000000000762), 2020. Date of Publication: 23 Jan 2020.

[Article]

AN: 630701327

INTRODUCTION AND OBJECTIVE: Controversy remains within the pediatric urology community regarding adequate duration of follow up after hypospadias repair. Some have suggested that minimal long-term follow-up is necessary due to a low incidence of late-term complications. The objective of this study was to delineate time to complication detection for primary hypospadias repairs.

METHOD(S): We queried our prospectively-maintained hypospadias database and identified all patients undergoing primary hypospadias repair from June 2007 to June 2018. Patients were excluded if they had undergone primary repair at an outside institution or did not have a follow up visit. Complications were defined by the need for an additional unplanned surgical procedure.

Kaplan-Meier analysis was performed to assess time to complication by degree of hypospadias.

RESULT(S): A total of 1280 patients met inclusion criteria: 976 distal (dHR; 68.9%), 64 mid-shaft (mHR; 4.9%), and 240 proximal (pHR; 18.8%). Complication rates were 10.7% (n=104), 18.8% (n=12), and 53.8% (n=129) ($p<0.0001$) for dHR, mHR, and pHR, respectively. Only 47% of complications were detected within the first year post-operatively. The median time to complication for all repair types was 69.2 months (IQR 23-131.9 months): 83.1 months (IQR 42.0-131 months) for patients with distal repair, and 29.4 months (IQR 11.9-82.1 months) for patients with proximal repair (figure 2, $p<0.001$).

CONCLUSION(S): In our large single institution series of pediatric patients undergoing hypospadias repair, fewer than half of our complications presented within the first year post-operatively. Long-term follow-up is recommended for patients undergoing hypospadias repair to adequately detect and address complications.

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Institution

(Lucas) Department of Urology, Einstein Healthcare Network, Philadelphia, PA, United States
(Hightower, Weiss, Van Batavia, Coelho, Srinivasan, Shukla, Zderic, Kolon, Zaontz, Canning, Long) Division of Urology, Children's Hospital of Philadelphia, Philadelphia, PA, United States

Publisher

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Year of Publication

2020

79.

Analysis of a 7-year national online audit of the management of open reconstructive urethral surgery in men.

Payne S.R., Fowler S., Mundy A.R.

Embase

BJU International. 125 (2) (pp 304-313), 2020. Date of Publication: 01 Feb 2020.

[Article]

AN: 2003430460

Objective: To conduct an audit of the management of urethral pathology in men presenting for reconstructive urethral surgery in the UK.

Method(s): Between 1 June 2010 and 31 May 2017, data on men presenting with urethral pathologies requiring reconstruction were entered onto a secure online data platform. Surgeon-entered information was collected in 95 fields regarding the stricture aetiology, prior management, mode of presentation, type of surgery and outcomes, with a potential 283 variable responses in the 95 fields. Data were analysed to compare UK practice with that reported in the contemporary literature and with guidelines.

Result(s): Data on 4809 men were entered by 39 centres and 50 surgeons. Field completeness was 70.7%, 74.3% and 53.7% for preoperative, operative and follow-up data, respectively.

Referral for stricture reconstruction frequently followed two prior endoscopic procedures and the stricture was not always assessed anatomically before surgery. Urinary retention was a common symptom in men awaiting reconstruction. Short unifocal strictures of the anterior urethra were the commonest reason for referral, whilst lichen sclerosus and hypospadias generated a significant volume of revisional stricture surgery. Lower numbers of very complex interventions are required for the management of posterior urethral pathology. Although precise criteria for determining success are not clear, management of urethral reconstruction in the UK was found to have a low risk of Clavien-Dindo grade 3 or higher complications, and was associated with outcomes similar to those reported in contemporary series except in the management of posterior urethral fistulae. Conclusion(s): Online databases can provide volume data on the management of reconstructive urethral surgery across a multiplicity of centres in one country. They can also indicate compliance with accepted standards of, and expected outcomes from, this tertiary practice.

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Author NameID

Payne, Stephen R.; ORCID: <http://orcid.org/0000-0001-5359-1877>

Institution

(Payne, Fowler) British Association of Urological Surgeons, United Kingdom (Mundy) University
College London Hospitals NHS Foundation Trust, London, United Kingdom

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Blackwell Publishing Ltd

Year of Publication

2020

80.

Urothelial or oral mucosa cells for tissue-engineered urethroplasty: A critical revision of the clinical outcome.

Barbagli G., Heidenreich A., Zugor V., Karapanos L., Lazzeri M.

Embase

Asian Journal of Urology. 7 (1) (pp 18-23), 2020. Date of Publication: January 2020.

[Review]

AN: 2002193155

Objective: To report the clinical outcome of urethral reconstruction by cultured urothelial or oral mucosa cells for tissue-engineered urethroplasty.

Method(s): We systematically searched for studies reporting the use of tissue-engineered techniques for hypospadias and urethral stricture repair in humans in PubMed and Embase (OvidSP) through January, 1990 to June, 2018. We excluded studies based on titles that clearly were not related to the subject, studies in which tissue-engineered biomaterial were used only in laboratory or experimental animals, and in the absence of autologous cultured epithelial cells. Studies were also excluded if they were not published in English, had no disease background and adequate follow-up. Finally, we search all relevant abstract presented at two of the main urological meetings in the last 10 years: European Association of Urology (EAU) and American Urological Association (AUA).

Result(s): A total of six articles, reporting the clinical use of tissue-engineered techniques in humans, were fully reviewed in our review. The epithelial cells were harvested from the urethra

(10 patients), the bladder (11 patients) and the mouth (104 patients). The tissue-engineered grafts were used in children for primary hypospadias repair in 16 cases, and in adults for posterior and anterior urethral strictures repair in 109 cases. Tissue-engineered grafts were showed working better in children for primary hypospadias repair than in adults for urethral strictures repair.

Conclusion(s): One hundred and twenty-five patients received tissue-engineered urethroplasty using cultured epithelial cells for primary hypospadias or urethral strictures repair. The studies demonstrate a high degree of heterogeneity respect to epithelial cells (from urethra, bladder, and mouth), type of scaffold, etiology, site of urethral stricture, number of patients, follow-up and outcomes.

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Status

Embase

Institution

(Barbagli) Centro Chirurgico Toscano, Arezzo, Italy (Barbagli) Research Institute for Uro-nephrology, Sechenov, First Moscow State Medical University, Moscow, Russian Federation (Heidenreich, Zugor, Karapanos) University Clinic and Policlinic for Urology, Cologne, Germany (Lazzeri) Istituto Clinico Humanitas IRCCS, Clinical and Research Hospital, Rozzano, Italy

Publisher

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Year of Publication

2020

81.

Urinary Continence Outcomes in Classic Bladder Exstrophy: A Long-Term Perspective.

Maruf M., Manyevitch R., Michaud J., Jayman J., Kasprenski M., Zaman M.H., Benz K., Eldridge M., Trock B., Harris K.T., Wu W.J., Di Carlo H.N., Gearhart J.P.

Embase

The Journal of urology. 203 (1) (pp 200-205), 2020. Date of Publication: 01 Jan 2020.

[Article]

AN: 629154258

PURPOSE: We investigated surgical approaches to urinary incontinence and long-term continence outcomes after successful bladder reconstruction in a heterogeneous patient population with classic bladder exstrophy. We hypothesized that while most patients will achieve urinary continence after surgery, only a select group will void volitionally per urethra. **MATERIALS AND METHODS:** An institutional database of 1,323 patients with exstrophy-epispadias complex was reviewed for patients with classic bladder exstrophy who underwent successful bladder closure and a subsequent continence procedure between 1975 and 2017. Procedures included bladder neck reconstruction, bladder neck reconstruction with augmentation cystoplasty or continent catheterizable stoma, and bladder neck closure with continent catheterizable stoma. Cloacal exstrophy, epispadias and variant exstrophy cases were excluded from analysis. Continence at last followup was defined as a dry interval of 3 or more hours without nighttime leakage. Those patients with more than 3 months of followup were assessed.

RESULT(S): Overall 432 patients underwent successful bladder closure (primary 71.5%, repeat 28.5%) and a urinary continence procedure. At last followup 162 (37%) underwent bladder neck reconstruction, 76 (18%) underwent bladder neck reconstruction with augmentation cystoplasty or continent catheterizable stoma, 173 (40%) underwent bladder neck closure with continent catheterizable stoma and 18 underwent other procedures. Median followup from the first continence procedure was 7.2 years (IQR 2.3-13.7). Continence was assessed in 350 patients. After isolated bladder neck reconstruction 91 of 142 patients were continent (64%, 95% CI 56-72). After bladder neck closure with continent catheterizable stoma 124 of 133 patients evaluated were continent (93%, 95% CI 87-97).

CONCLUSION(S): Most patients with classic bladder exstrophy require multiple reconstructive procedures to achieve continence. Only about 25% of patients are expected to void normally per urethra without reliance on catheterization or urinary diversion.

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Institution

(Maruf, Manyevitch, Michaud, Jayman, Kasprinski, Zaman, Benz, Eldridge, Harris, Wu, Di Carlo, Gearhart) Robert D. Jeffs Division of Pediatric Urology, Johns Hopkins Medical Institutions, Baltimore, MD, United States (Trock) Brady Urological Institute, Johns Hopkins Medical Institutions, Baltimore, MD, United States

Publisher

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82.

Disorders of sex development: Genetic characterization of a patient cohort.

Garcia-Acero M., Moreno-Nino O., Suarez-Obando F., Molina M., Manotas M.C., Prieto J.C., Forero C., Cespedes C., Perez J., Fernandez N., Rojas A.

Embase

Molecular Medicine Reports. 21 (1) (pp 97-106), 2020. Date of Publication: 2020.

[Article]

AN: 2004282101

Disorders of sex development (DSDs) are congenital conditions in which the external appearance of the individual does not coincide with the chromosomal constitution or the gonadal sex. In other words, there is an ambiguous or intermediate condition between the male and female phenotypes of the anatomical sex. These atypical conditions are manifested in several ways, ranging from genital ambiguity to phenotypes that are so attenuated that they can go unnoticed or appear normal. Currently, there is a lack of understanding of the factors responsible for these outcomes; however, they are likely to be conditioned by genetic, hormonal and environmental factors during prenatal and postnatal development. The present study determined the genetic etiology of DSDs in Colombian patients by conventional cytogenetic analysis, FISH and MLPA (for SF1, DAX1, SOX9, SRY and WNT4). A cohort of 43 patients with clinical phenotypes of sex development disorder was used in the present study. Using this multistep experimental approach, a diagnostic percentage of 25.58% was obtained: 17 patients (39.53%) were classified as having gonadal development disorders, the majority of which were ovotesticular disorders with numerical and/or structural alterations of the sex chromosomes, 9 patients (20.93%) were classified as having testicular DSD with a 46,XY karyotype, and 3 patients (6.98%) as having ovarian DSD with a 46,XX karyotype. The remaining 14 patients (32.56%) were classified as 'other' since they could not be grouped into a specific class of gonadal development, corresponding to hypospadias and multiple congenital anomalies. These findings highlight the importance of histological and cytogenetic studies in a gonadal biopsy. In 11/43 cases, the multistep experimental protocol presented in the present study yielded etiological or histological findings that could be used to define the medical management of patients with DSDs. In conclusion, for the etiological diagnosis of DSDs, a broad-spectrum approach that includes endocrinological tests, conventional karyotyping, molecular karyotyping by FISH and, molecular tests is required, in addition to gonadal tissue analyses, to identify genetic alterations.

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Status

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Institution

(Garcia-Acero, Moreno-Nino, Suarez-Obando, Molina, Manotas, Prieto, Rojas) Instituto de Genetica Humana, Facultad de Medicina, Pontificia Universidad Javeriana, Bogota 110231, Colombia (Suarez-Obando) Clinical Genetic Service, Hospital Universitario San Ignacio, Bogota 110231, Colombia

(Forero, Cespedes) Pediatric Endocrinology, Bogota 110231, Colombia

(Perez, Fernandez) Division of Urology, Hospital Universitario San Ignacio, Facultad de Medicina, Pontificia Universidad Javeriana, Bogota 110231, Colombia

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83.

Cryptorchidism. [Review]

Leslie SW; Sajjad H; Villanueva CA.

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StatPearls Publishing. 2019 01.

[Review]

UI: 29261861

Cryptorchidism is the absence of at least one testicle from the scrotum. It is the most common birth defect involving the male genitalia. About 3% of full-term and 30% of premature male infants are born with one or both testicles undescended. Approximately 80% of cryptorchid testes descend by the third month of life. This makes the true incidence around 1%. [1] Cryptorchidism may occur on one or both sides, but more commonly affects the right testicle. The testicle may be anywhere along the "path of descent," such as: [2]: Located high in the retroperitoneal abdomen

to the inguinal ring. In the inguinal canal. Ectopic from the path of descent. Hypoplastic. Dysgenetic. Missing or Absent. Unilateral (two-thirds). The undescended testicle can usually be palpated in the inguinal canal. In a minority of patients, the missing testicle may be located in the abdomen or be nonexistent. Undescended testicles are associated with decreased fertility (bilateral cases), increased testicular germ cell tumors (overall risk under 1%), testicular torsion, inguinal hernias, and psychological problems. Without surgical correction, an undescended testicle may descend during the first three months of life. To reduce risks, undescended testes may be brought into the scrotum with an orchiopexy. Cryptorchidism, hypospadias, testicular cancer, and poor semen quality make up testicular dysgenesis syndrome (TDS). This syndrome is thought to be due to harmful environmental factors that disrupt embryonal programming and gonadal development during fetal life.

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Book Title

StatPearls

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Authors Full Name

Leslie, Stephen W.; Sajjad, Hussain; Villanueva, Carlos A..

Institution

Leslie, Stephen W.. Creighton University Medical Center Sajjad, Hussain. RMU and Allied Hospitals

Villanueva, Carlos A.. Children's Hospital and Medical Center

Publisher

StatPearls Publishing

Year of Publication

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84.

Familial GATA6 mutation causing variably expressed diabetes mellitus and cardiac and renal abnormalities.

Du YT; Moore L; Poplawski NK; De Sousa SMC.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Endocrinology, Diabetes & Metabolism Case Reports. 2019, 2019 May 03.

[Journal Article]

UI: 31051468

A 26-year-old man presented with a combination of permanent neonatal diabetes due to pancreatic aplasia, complex congenital heart disease, central hypogonadism and growth hormone deficiency, structural renal abnormalities with proteinuria, umbilical hernia, neurocognitive impairment and dysmorphic features. His older brother had diabetes mellitus due to pancreatic hypoplasia, complex congenital heart disease, hypospadias and umbilical hernia. Their father had an atrial septal defect, umbilical hernia and diabetes mellitus diagnosed incidentally in adulthood on employment screening. The proband's paternal grandmother had a congenital heart defect. Genetic testing of the proband revealed a novel heterozygous missense variant (Chr18:g.19761441T>C, c.1330T>C, p.Cys444Arg) in exon 4 of GATA6, which is class 5 (pathogenic) using American College of Medical Genetics and Genomics guidelines and is likely to account for his multisystem disorder. The same variant was detected in his brother and father, but not his paternal grandmother. This novel variant of GATA6 likely occurred de novo in the father with autosomal dominant inheritance in the proband and his brother. The case is exceptional as very few families with monogenic diabetes due to GATA6 mutations have been reported to date and we describe a new link between GATA6 and renal pathology. Learning points: Monogenic diabetes should be suspected in patients presenting with syndromic features, multisystem congenital disease, neonatal-onset diabetes and/or a suggestive family history. Recognition and identification of genetic diabetes may improve patient understanding and empowerment and allow for better tailored management. Identification of a genetic disorder may have important implications for family planning.

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1

Status

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Authors Full Name

Du, Yang Timothy; Moore, Lynette; Poplawski, Nicola K; De Sousa, Sunita M C.

Institution

Du, Yang Timothy. Endocrine and Metabolic Unit, Royal Adelaide Hospital. Moore, Lynette.

School of Medicine, University of Adelaide.

Moore, Lynette. SA Pathology, Women's and Children's Hospital.

Poplawski, Nicola K. Adult Genetics Unit, Royal Adelaide Hospital.

De Sousa, Sunita M C. Endocrine and Metabolic Unit, Royal Adelaide Hospital.

De Sousa, Sunita M C. School of Medicine, University of Adelaide.
De Sousa, Sunita M C. Adult Genetics Unit, Royal Adelaide Hospital.
De Sousa, Sunita M C. Center for Cancer Biology, SA Pathology and University of South
Australia Alliance, Adelaide, South Australia, Australia.
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Year of Publication
2019

85.

Renal failure, respiratory distress, and an atypical purpuric rash in a full-term infant with
omphalocele and hypospadias: Answers.

Weiss AJ; Kronforst K.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid

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Pediatric Nephrology. 34(11):2339-2342, 2019 11.

[Journal Article]

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1

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In-Process

Authors Full Name

Weiss, Aaron J; Kronforst, Kenny.

Institution

Weiss, Aaron J. McGaw Medical Center of Northwestern University, Chicago, IL, USA.

AaWeiss@luriechildrens.org. Weiss, Aaron J. Division of Neonatology, Ann & Robert H. Lurie
Children's Hospital of Chicago, Box 45, 225 E Chicago Ave., Chicago, IL, 60611, USA.

AaWeiss@luriechildrens.org.

Kronforst, Kenny. McGaw Medical Center of Northwestern University, Chicago, IL, USA.

KKronforst@luriechildrens.org.

Kronforst, Kenny. Division of Neonatology, Ann & Robert H. Lurie Children's Hospital of Chicago, Box 45, 225 E Chicago Ave., Chicago, IL, 60611, USA. KKronforst@luriechildrens.org.

Year of Publication

2019

86.

Perineal hypospadias repair with preservation of a coincidental vagina or perineal utricle in boys with disorders of sex development.

Wolffenbuttel KP; Holmdahl G.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid

MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Journal of pediatric urology. 15(6):626.e1-626.e5, 2019 Dec.

[Journal Article]

UI: 31582336

INTRODUCTION: An enlarged utricle in patients with proximal hypospadias or disorders of sex development (DSD) is common. The utricle orifice is usually in the posterior urethra near the verumontanum, but in exceptional cases located on the perineum. Concurrence of a perineal hypospadias and perineal utricle or vagina is also known as male vagina, pseudovagina, or blind vaginal pouch. The utricle in such cases is usually excised either before or concomitant with hypospadias repair. The authors developed an alternative approach in which the vagina or perineal utricle is retained during hypospadias repair and report on the results in four patients.

OBJECTIVE: To report a novel technique for perineal hypospadias repair while retaining a concurrent vagina or perineal utricle.

PATIENTS AND METHODS: Between 1999 and 2014, four neonates presented with perineal hypospadias. In all patients, a second perineal opening providing access to either an enlarged utricle or a vagina was identified. Karyotype in peripheral blood was in two patients 46,XY and in the other two 45,X/46,XY of which one showed a complex mosaicism in gonadal tissue. No genetic cause was identified on DNA evaluation in the two patients with 46,XY DSD. All patients were raised as boys. Hypospadias repair was performed in two stages at prepubertal age. During the second stage of surgery, performed between the age of 1.5 and 5 years, the vagina or utricle orifice was incorporated into the neo-urethra, resulting in a retained 'built-in' vagina or utricle.

RESULTS: Surgical procedures were uneventful, and patients remained asymptomatic during a mean postoperative follow-up of 8.5 (range 2-13.5) years. One patient was lost to follow-up after the age of 8 years. At their last visit, the remaining patients, at the age of 4, 15, and 17 years, were able to void in standing position without dribbling. Both adolescent patients reported erections without ejaculations and identified themselves as males without signs of gender dysphoria.

CONCLUSION: Hypospadias repair in boys with perineal hypospadias while leaving a male vagina or perineal utricle in situ has not been reported previously, and the study's preliminary results are favorable. One of the benefits of this approach is that inadvertent injury to adjacent anatomic structures such as urethral sphincter, neurovascular bundles, ureters, vas deferens, and rectum is avoided. The main rationale for adopting this conservative approach however is to minimize genital tissue removal in children with a not yet definite gender identity, which will certainly facilitate unforeseen future gender reassignment surgery.

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Version ID

1

Status

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Authors Full Name

Wolffenbuttel, Katja P; Holmdahl, Gundela.

Institution

Wolffenbuttel, Katja P. Erasmus MC-Sophia Children's Hospital, University Medical Center Rotterdam, Department of Urology and Paediatric Urology, Rotterdam, the Netherlands.

Electronic address: k.wolffenbuttel@erasmusmc.nl. Holmdahl, Gundela. Sahlgrenska Academy, Women's and Children's Health, Gothenburg, Sweden; Queen Silvia's Children's Hospital, Department of Paediatric Surgery, Gothenburg, Sweden.

Year of Publication

2019

Tourniquet and adrenaline use in hypospadias surgery: a survey on the current practice in Turkey.

Ates U; Ekberli G; Tastekin NY; Gollu G; Cakmak M.

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MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Turkish Journal of Urology. 45(3):218-222, 2019 05.

[Journal Article]

UI: 30468426

OBJECTIVE: Aim of the study is to determine the hemostatic techniques among pediatric urologists in Turkey.

MATERIAL AND METHODS: Questionnaire forms were sent to 459 pediatric urologist by e-mail.

RESULTS: Ninety eight of 459 participants answered the questionnaire forms. Eighty-one (84.4%) of the participants were using tourniquet. The participants who didn't use tourniquet stated their justifications as follows: lack of need (n=10: 66.7%), development of edema, ischemia, delay of wound-graft healing and fistula risk (n=5: 33.3%). The indications of tourniquet use were stated as follows: penile (91.4%: n=74), distal (72.8%: n=59), penoscrotal (55.6%: n=45) hypospadias; fistula repair (33.3%: n=27), cripple hypospadias (33.3%: n=27), repair with flaps (30.9%: n=25), repair with grafts (27.2%: n=22), and isolated penile curvature (21%: n=17). Most commonly used tourniquet material (49.9%) was latex glove. Erection test was applied by 43.8% of participants. Scalp vein set was the most commonly (54.8%) used injector during erection test. Only 9.4% of participants were using adrenaline. Adrenaline dosages used at 1/100.000 dilution by 55.6%, lidocaine with 1/100.000 adrenaline by 44.4% of participants.

CONCLUSION: Beside a few experimental ones there is a paucity of studies that can serve as a guideline for using these techniques in the literature. There is a necessity of realizing prospective, randomized studies with long-term follow up to evidence that postoperative complications could develop secondary to hemostatic techniques and also to facilitate safe use of these techniques.

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1

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Author Initials

Ates, Ufuk; ORCID: <http://orcid.org/0000-0001-6591-7168> Ekberli, Gunay; ORCID: <http://orcid.org/0000-0002-0021-5998>

Tastekin, Nil Yasam; ORCID: <http://orcid.org/0000-0002-4632-0881>

Gollu, Gulnur; ORCID: <http://orcid.org/0000-0001-8163-2226>

Cakmak, Murat; ORCID: <http://orcid.org/0000-0002-4870-8361>

Authors Full Name

Ates, Ufuk; Ekberli, Gunay; Tastekin, Nil Yasam; Gollu, Gulnur; Cakmak, Murat.

Institution

Ates, Ufuk. Department of Pediatric Surgery, Ankara University School of Medicine, Ankara, Turkey. Ekberli, Gunay. Department of Pediatric Surgery, Ankara University School of Medicine, Ankara, Turkey.

Tastekin, Nil Yasam. Department of Pediatric Surgery, Ankara University School of Medicine, Ankara, Turkey.

Gollu, Gulnur. Department of Pediatric Surgery, Ankara University School of Medicine, Ankara, Turkey.

Cakmak, Murat. Department of Pediatric Surgery, Ankara University School of Medicine, Ankara, Turkey.

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Year of Publication

2019

88.

Identification of three novel SRD5A2 mutations in Chinese patients with 5alpha-reductase 2 deficiency.

Cheng T; Wang H; Han B; Zhu H; Yao HJ; Zhao SX; Zhu WJ; Zhai HL; Chen FG; Song HD; Cheng KX; Liu Y; Qiao J.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid

MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Asian Journal of Andrology. 21(6):577-581, 2019 Nov-Dec.

[Journal Article. Research Support, Non-U.S. Gov't]

UI: 31031332

In this study, we investigated the genetics, clinical features, and therapeutic approach of 14 patients with 5alpha-reductase deficiency in China. Genotyping analysis was performed by direct sequencing of PCR products of the steroid 5alpha-reductase type 2 gene (SRD5A2). The 5alpha-reductase activities of three novel mutations were investigated by mutagenesis and an in vitro transfection assay. Most patients presented with a microphallus, variable degrees of hypospadias, and cryptorchidism. Eight of 14 patients (57.1%) were initially reared as females

and changed their social gender from female to male after puberty. Nine mutations were identified in the 14 patients. p.G203S, p.Q6X, and p.R227Q were the most prevalent mutations. Three mutations (p.K35N, p.H162P, and p.Y136X) have not been reported previously. The nonsense mutation p.Y136X abolished enzymatic activity, whereas p.K35N and p.H162P retained partial enzymatic activity. Topical administration of dihydrotestosterone during infancy or early childhood combined with hypospadias repair surgery had good therapeutic results. In conclusion, we expand the mutation profile of SRD5A2 in the Chinese population. A rational clinical approach to this disorder requires early and accurate diagnosis, especially genetic diagnosis.

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1

Status

In-Process

Authors Full Name

Cheng, Tong; Wang, Hao; Han, Bing; Zhu, Hui; Yao, Hai-Jun; Zhao, Shuang-Xia; Zhu, Wen-Jiao; Zhai, Hua-Ling; Chen, Fu-Guo; Song, Huai-Dong; Cheng, Kai-Xiang; Liu, Yang; Qiao, Jie.

Institution

Cheng, Tong. Department of Endocrinology, Shanghai Ninth People's Hospital, Shanghai Jiao Tong University School of Medicine, Shanghai 200011, China. Wang, Hao. Department of Endocrinology, Shanghai Ninth People's Hospital, Shanghai Jiao Tong University School of Medicine, Shanghai 200011, China.

Han, Bing. Department of Endocrinology, Shanghai Ninth People's Hospital, Shanghai Jiao Tong University School of Medicine, Shanghai 200011, China.

Zhu, Hui. Department of Endocrinology, Shanghai Ninth People's Hospital, Shanghai Jiao Tong University School of Medicine, Shanghai 200011, China.

Yao, Hai-Jun. Department of Urology, Shanghai Ninth People's Hospital, Shanghai Jiao Tong University School of Medicine, Shanghai 200011, China.

Zhao, Shuang-Xia. Central Laboratory, Clinical Research Center, Shanghai Ninth People's Hospital, Shanghai Jiao Tong University School of Medicine, Shanghai 200011, China.

Zhu, Wen-Jiao. Department of Endocrinology, Shanghai Ninth People's Hospital, Shanghai Jiao Tong University School of Medicine, Shanghai 200011, China.

Zhai, Hua-Ling. Department of Endocrinology, Shanghai Ninth People's Hospital, Shanghai Jiao Tong University School of Medicine, Shanghai 200011, China.

Chen, Fu-Guo. Department of Plastic Surgery, Research Center of Tissue Engineering, Shanghai Ninth People's Hospital, Shanghai Jiao Tong University School of Medicine, Shanghai 200011, China.

Song, Huai-Dong. Central Laboratory, Clinical Research Center, Shanghai Ninth People's Hospital, Shanghai Jiao Tong University School of Medicine, Shanghai 200011, China.

Cheng, Kai-Xiang. Department of Plastic Surgery, Research Center of Tissue Engineering, Shanghai Ninth People's Hospital, Shanghai Jiao Tong University School of Medicine, Shanghai 200011, China.

Liu, Yang. Department of Plastic Surgery, Research Center of Tissue Engineering, Shanghai Ninth People's Hospital, Shanghai Jiao Tong University School of Medicine, Shanghai 200011, China.

Qiao, Jie. Department of Endocrinology, Shanghai Ninth People's Hospital, Shanghai Jiao Tong University School of Medicine, Shanghai 200011, China.

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89.

Renal failure, respiratory distress, and an atypical purpuric rash in a full-term infant with omphalocele and hypospadias: Questions.

Weiss AJ; Kronforst K.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Pediatric Nephrology. 34(11):2337-2338, 2019 11.

[Journal Article]

UI: 31236665

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1

Status

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Author Initials

Weiss, Aaron J; ORCID: <https://orcid.org/0000-0002-3869-9887>

Authors Full Name

Weiss, Aaron J; Kronforst, Kenny.

Institution

Weiss, Aaron J. McGaw Medical Center of Northwestern University, Chicago, IL, USA.
AaWeiss@luriechildrens.org. Weiss, Aaron J. Division of Neonatology, Ann & Robert H. Lurie
Children's Hospital of Chicago, Box 45, 225 E Chicago Ave., Chicago, IL, 60611, USA.
AaWeiss@luriechildrens.org.
Kronforst, Kenny. McGaw Medical Center of Northwestern University, Chicago, IL, USA.
Kronforst, Kenny. Division of Neonatology, Ann & Robert H. Lurie Children's Hospital of Chicago,
Box 45, 225 E Chicago Ave., Chicago, IL, 60611, USA.
Year of Publication
2019

90.

Keloid formation after pediatric male genital surgeries: an uncommon and difficult problem to manage.

Alyami F; Fernandez N; Koyle MA; Salle JP.

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MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Journal of pediatric urology. 15(1):48.e1-48.e8, 2019 Feb.

[Journal Article]

UI: 30206024

INTRODUCTION: Penile and genital keloids are uncommon despite frequent surgeries in the genital area. Keloid scar pathogenesis is not well understood, and a uniform effective therapeutic regimen for keloids has not yet been established. In the present study, the clinical features and subsequent management in cases of severe keloid formation after pediatric genital surgery are described.

METHODS: A retrospective review of keloid cases that had developed after genital procedures between 2000 and 2017 was conducted. Pre-operative characteristics, operative procedures, postoperative management, and follow-up were reviewed for each case. All cases were managed by a multidisciplinary team that included plastic surgeons and dermatologists.

RESULT: Six cases developed genital keloids. The mean age at surgery was 5.6 years (+/- standard deviation 4.6 years). Procedures included phalloplasties, penile curvature correction, penoscrotal transposition, redo hypospadias repair, and circumcision. Treatment options included excision of the keloid +/- topical steroid injections and postoperative use of silicone gel. Two

cases of severe keloid lesions developed after using posterior auricular grafts. Ultimately, a successful outcome was achieved in all cases.

CONCLUSION: Genital keloids are rare and difficult to treat. Many therapeutic options are available with varying degrees of proven clinical success. As a result, pediatric urologists must be aware of advances in other fields such as plastic surgery and dermatology to treat and ideally prevent the occurrence of this serious complication.

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Version ID

1

Status

MEDLINE

Authors Full Name

Alyami, F; Ferandez, N; Koyle, M A; Salle, J P.

Institution

Alyami, F. Division of Urology, The Hospital for Sick Children and University of Toronto, Toronto, Ontario, Canada; Urology Division, Department of Surgery, King Saud University, King Saud University Medical City and College of Medicine, Riyadh, Saudi Arabia. Electronic address: faalyami@ksu.edu.sa. Ferandez, N. Division of Urology, The Hospital for Sick Children and University of Toronto, Toronto, Ontario, Canada.

Koyle, M A. Division of Urology, The Hospital for Sick Children and University of Toronto, Toronto, Ontario, Canada.

Salle, J P. Division of Urology, The Hospital for Sick Children and University of Toronto, Toronto, Ontario, Canada; Department of Surgery, Division of Urology, Sidra Medical and Research Center, Doha, Qatar.

Year of Publication

2019

91.

Analysis of the association between paternity and reoperation for urethral obstruction in adult hypospadias patients who underwent two-stage repair in childhood.

Kanematsu A; Tanaka S; Hashimoto T; Nojima M; Yamamoto S.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid
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BMC Urology. 19(1):88, 2019 Oct 04.

[Journal Article]

UI: 31585530

BACKGROUND: The association between surgical outcome of hypospadias repair and long-term male reproductive function has not been documented. The purpose of this study was to clarify association between paternity in adult hypospadias patients and reoperation for urethral obstruction after two-stage repair during childhood.

METHODS: Ninety hypospadias patients who underwent the same kind of two-stage repair in our institute by a single surgeon, were initially treated at < 18 years old, and who were $\geq$ 18 years old during the survey were included in the study. Present physical, social, and life status were evaluated by a mailed self-entry questionnaire, and clinical background and surgical outcome data were evaluated by medical records. National survey data of the general population were used as external control. The paternity rate of the patient groups was evaluated by Kaplan-Meier curve analysis and log-rank tests.

RESULTS: Twenty-six patients (28.9%) underwent 43 reoperations after completion of the initial repair. Twelve patients were reoperated for obstructive complication (Study group) and were compared with 14 patients who were reoperated only for non-obstructive causes and 64 patients who were not reoperated as Study control group (N = 78). The Study group patients showed sexual intercourse rate and marriage rate not statistically different in comparison with the Study control, although marriage rate at 32.5 years old were lower than the general population ($p = 0.048$, z-test). None of the Study group achieved paternity, which showed a significant difference to the Study control ($p = 0.032$, log-rank test). The difference was also statistically significant in the analysis among the 31 married patients ($p = 0.012$, log-rank test). Patients reoperated for obstructive complication documented worsened Quality of Life score in the International Prostate Symptom Score (2.3 ± 2.0 vs. 1.4 ± 1.2 , $p = 0.031$, t-test) and ejaculation problems (66.7% vs. 17.4%, $p = 0.003$, chi-square test).

CONCLUSIONS: History of reoperation for obstructive complication was associated with lower paternity rate in patients with hypospadias, presumably for multifactorial causes associated with marriage age and ejaculation problems. The present results may implicate importance of uncomplicated urethroplasty during childhood for achieving paternity, although it should be further tested in the future for larger groups of hypospadias patients.

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1

Status

MEDLINE

Author Initials

Kanematsu, Akihiro; ORCID: <http://orcid.org/0000-0002-0669-8986>

Authors Full Name

Kanematsu, Akihiro; Tanaka, Shiro; Hashimoto, Takahiko; Nojima, Michio; Yamamoto, Shingo.

Institution

Kanematsu, Akihiro. Department of Urology, Hyogo College of Medicine, Mukogawacho 1-1, Nishinomiya, Hyogo, 663-8501, Japan. aqui@hyo-med.ac.jp. Tanaka, Shiro. Department of Clinical Biostatistics, Graduate School of Medicine Kyoto University, Yoshida Konoecho, Sakyo, Kyoto, 606-8501, Japan.

Hashimoto, Takahiko. Department of Urology, Hyogo College of Medicine, Mukogawacho 1-1, Nishinomiya, Hyogo, 663-8501, Japan.

Nojima, Michio. Department of Urology, Hyogo College of Medicine, Mukogawacho 1-1, Nishinomiya, Hyogo, 663-8501, Japan.

Yamamoto, Shingo. Department of Urology, Hyogo College of Medicine, Mukogawacho 1-1, Nishinomiya, Hyogo, 663-8501, Japan.

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2019

92.

Complex abdominal wall reconstruction combined with bladder closure in OEIS complex.

Davis R; Stewart D; Maruf M; Lau H; Gearhart JP.

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MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Journal of Pediatric Surgery. 54(11):2408-2412, 2019 Nov.

[Journal Article]

UI: 31079865

PURPOSE: Due to the large abdominal defect from the omphalocele and extreme pubic diastasis in cloacal exstrophy (CE), bioprosthetic material may be used to bridge this gap during abdominal closure in CE. This study examined presurgical factors associated with the use of bioprosthetic materials in CE closure and complications in these patients.

METHODS: An institutional database of exstrophy-epispadias complex patients was reviewed for CE. Inclusion criteria included CE and primary closure performed at the host institution from 1998 to 2018. Data collection included demographics, presurgical factors, use of bioprosthetic material, complications, and outcomes.

RESULTS: All 32 patients had a staged closure and pelvic osteotomy prior to bladder closure. Ten of the 32 patients incorporated a bioprosthetic material during abdominal wall closure. There is at least 3months follow up for all patients, all had successful bladder closure without any postoperative hernias. Those who underwent closure without bioprosthetic material were younger at the time of closure (565 vs 693days, $p=0.043$). The differences in complication rates and mean pubic diastasis was not statistically significant, $p=0.079$ and $p=0.457$ respectively.

CONCLUSIONS: The use of bioprosthetic material is associated with older age at abdominal wall and bladder closure. The use of bioprosthetic material is a useful adjunct for secure abdominal wall closure in the reconstruction of CE.

TYPE OF STUDY: Prognostic.

LEVEL OF EVIDENCE: III.

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1

Status

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Authors Full Name

Davis, Rachel; Stewart, Dylan; Maruf, Mahir; Lau, Henry; Gearhart, John P.

Institution

Davis, Rachel. Robert D. Jeffs Division of Pediatric Urology, James Buchanan Brady Urological Institutions, Johns Hopkins Hospital, Johns Hopkins Medical Institutions, Charlotte Bloomberg Children's Hospital, Baltimore, MD, USA. Stewart, Dylan. Department of Surgery, Johns Hopkins School of Medicine, Johns Hopkins Children's Center, Charlotte Bloomberg Children's Hospital, Baltimore, MD, USA.

Maruf, Mahir. Robert D. Jeffs Division of Pediatric Urology, James Buchanan Brady Urological Institutions, Johns Hopkins Hospital, Johns Hopkins Medical Institutions, Charlotte Bloomberg Children's Hospital, Baltimore, MD, USA.

Lau, Henry. Department of Surgery, Johns Hopkins School of Medicine, Johns Hopkins Children's Center, Charlotte Bloomberg Children's Hospital, Baltimore, MD, USA.

Gearhart, John P. Robert D. Jeffs Division of Pediatric Urology, James Buchanan Brady Urological Institutions, Johns Hopkins Hospital, Johns Hopkins Medical Institutions, Charlotte Bloomberg Children's Hospital, Baltimore, MD, USA. Electronic address: jgearha2@jhmi.edu.

Year of Publication

93.

The failed bladder closure in cloacal exstrophy: Management and outcomes.

Davis R; Sood A; Maruf M; Singh P; Kasprenski MC; DiCarlo HN; Gearhart JP.

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MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Journal of Pediatric Surgery. 54(11):2416-2420, 2019 Nov.

[Journal Article]

UI: 30879754

PURPOSE: Cloacal exstrophy (CE) is the most severe presentation of the Exstrophy-Epispadias Complex (EEC) and is associated with an omphalocele, making the bladder and abdominal wall closure difficult. If the bladder closure fails, a secondary closure is necessary. The objective of this study is to identify patient or surgical factors associated with a successful secondary closure.

METHODS: The institution's EEC database was reviewed for CE patients between 1975 and 2015. Inclusion criteria included a failed primary bladder closure with a secondary closure. Patient demographics, surgical factors and outcomes of the secondary bladder closure were reviewed.

RESULTS: Twenty-four patients met inclusion criteria. 8/8 patients had a successful two-staged closure at the author's institution (100%); 2/16 patients had a successful closure at an outside institution (12.5%). Older median age at secondary closure was associated with outcome, $p=0.045$. Pelvic osteotomy was associated with successful secondary closure, $p=0.013$. Using Buck's immobilization with external fixation was associated with a higher proportion of successful secondary closures compared to Spica cast, $p=0.012$.

CONCLUSION: Successful reclosure in CE patients is associated with the use of osteotomy as well as Buck's immobilization with external fixation. While successful reclosure can be achieved, it is often at the cost of multiple procedures and, therefore, all efforts should be expended to achieve a successful primary closure.

TYPE OF STUDY: Prognostic.

LEVEL OF EVIDENCE: III.

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Authors Full Name

Davis, Rachel; Sood, Amol; Maruf, Mahir; Singh, Prachi; Kasprenski, Matthew C; DiCarlo, Heather N; Gearhart, John P.

Institution

Davis, Rachel. Robert D. Jeffs Division of Pediatric Urology, James Buchanan Brady Urological Institutions, Johns Hopkins Hospital, Baltimore, MD, USA; Johns Hopkins Medical Institutions, Charlotte Bloomberg Children's Hospital, Baltimore, MD, USA. Sood, Amol. Robert D. Jeffs Division of Pediatric Urology, James Buchanan Brady Urological Institutions, Johns Hopkins Hospital, Baltimore, MD, USA; Johns Hopkins Medical Institutions, Charlotte Bloomberg Children's Hospital, Baltimore, MD, USA.

Maruf, Mahir. Robert D. Jeffs Division of Pediatric Urology, James Buchanan Brady Urological Institutions, Johns Hopkins Hospital, Baltimore, MD, USA; Johns Hopkins Medical Institutions, Charlotte Bloomberg Children's Hospital, Baltimore, MD, USA.

Singh, Prachi. Robert D. Jeffs Division of Pediatric Urology, James Buchanan Brady Urological Institutions, Johns Hopkins Hospital, Baltimore, MD, USA; Johns Hopkins Medical Institutions, Charlotte Bloomberg Children's Hospital, Baltimore, MD, USA.

Kasprenski, Matthew C. Robert D. Jeffs Division of Pediatric Urology, James Buchanan Brady Urological Institutions, Johns Hopkins Hospital, Baltimore, MD, USA; Johns Hopkins Medical Institutions, Charlotte Bloomberg Children's Hospital, Baltimore, MD, USA.

DiCarlo, Heather N. Robert D. Jeffs Division of Pediatric Urology, James Buchanan Brady Urological Institutions, Johns Hopkins Hospital, Baltimore, MD, USA; Johns Hopkins Medical Institutions, Charlotte Bloomberg Children's Hospital, Baltimore, MD, USA.

Gearhart, John P. Robert D. Jeffs Division of Pediatric Urology, James Buchanan Brady Urological Institutions, Johns Hopkins Hospital, Baltimore, MD, USA; Johns Hopkins Medical Institutions, Charlotte Bloomberg Children's Hospital, Baltimore, MD, USA. Electronic address: jgearha2@jhmi.edu.

Year of Publication

2019

Penile fracture and investigation of early surgical repair effects on erectile dysfunction.

Kati B; Akin Y; Demir M; Boran OF; Gumus K; Ciftci H.

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MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Urologia (Treviso). 86(4):207-210, 2019 Nov.

[Journal Article]

UI: 31010389

OBJECTIVES: Penile fracture is one of the urological emergencies caused by direct trauma to an erect penis during sexual intercourse, which results in a tear in the tunica albuginea within the corpus cavernosum. Serious complications such as penile curvature and erectile dysfunction may develop due to inappropriate and/or late surgical repair. This study aims to evaluate patients with penile fracture and to describe their demographics, surgical repairs, and long-term outcomes.

MATERIALS AND METHODS: A total of 56 patients who were diagnosed with penile fracture between January 2012 and June 2017 were reviewed. Clinical features, pre-operative assessment, time from injury to surgery, tunica defect properties, and presence of urethral injury were assessed. Early surgical management was performed. Outcomes, including International Index of Erectile Function 5 pre-operation and after 6 months, were evaluated.

RESULTS: The mean age was 30.2 (18-57) years. In etiological questionnaires, 32 (57.2%) patients reported direct trauma to an erect penis during intercourse. The mean size of tunica defects was 1.61 +/- 0.42 (0.3-3.6) cm of the nine (16%) patients, and penile fracture was associated with urethral injury. There was no significant difference in International Index of Erectile Function 5 scores before the surgery and 6 months after surgery. Penile skin necrosis developed in one patient 10 days post-operation.

CONCLUSION: Early surgical repair could be an effective method of achieving post-operative erection success in patients with penile fracture due to direct trauma during intercourse.

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1

Status

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Author Initials

Kati, Bulent; ORCID: <https://orcid.org/0000-0002-4024-5147>

Authors Full Name

Kati, Bulent; Akin, Yigit; Demir, Mehmet; Boran, Omer Faruk; Gumus, Kemal; Ciftci, Halil.

Institution

Kati, Bulent. Department of Urology, Faculty of Medicine, Harran University, Sanliurfa, Turkey.

Akin, Yigit. Department of Urology, Faculty of Medicine, Harran University, Sanliurfa, Turkey.

Akin, Yigit. Department of Urology, Faculty of Medicine, Izmir Katip Celebi University, Izmir, Turkey.

Demir, Mehmet. Department of Urology, Faculty of Medicine, Harran University, Sanliurfa, Turkey.

Boran, Omer Faruk. Urology Clinic, Balikligol Government Hospital, Sanliurfa, Turkey.

Boran, Omer Faruk. Department of Anesthesiology and Reanimation, Kahramanmaras Sutcu imam University, Kahramanmaras, Turkey.

Gumus, Kemal. Urology Clinic, Balikligol Government Hospital, Sanliurfa, Turkey.

Ciftci, Halil. Department of Urology, Faculty of Medicine, Harran University, Sanliurfa, Turkey.

Year of Publication

2019

95.

Diagnostic accuracy of human chorionic gonadotropins (HCG) stimulation test in XY-disorders of sex development (XY-DSD) presented in Armed Forces Institute of Pathology.

Sheikh WH; Asif N; Haroon ZH; Ejaz A; Ain QU; Shehzad N.

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JPMA - Journal of the Pakistan Medical Association. 69(8):1090-1093, 2019 Aug.

[Journal Article]

UI: 31431758

OBJECTIVE: To determine diagnostic accuracy of human chorionic gonadotropins stimulation test in differentiating androgen insensitivity syndrome and 5-alpha reductase deficiency, keeping testosterone to dihydrotestosterone ratio as the gold standard.

METHODS: The cross-sectional study was conducted at the Department of Chemical Pathology and Endocrinology, Armed Forces Institute of Pathology, Rawalpindi, Pakistan, from January to December, 2016, and comprised patients aged 01 day to 20 years having XY chromosomes on karyotyping and with a spectrum of phenotypes. Blood samples were collected from each subject for basal serum testosterone, serum luteinizing hormone and serum follicular stimulating hormone level. Human chorionic gonadotropins stimulation test was performed in every subject as per the protocol. Sandwich chemiluminescence immunoassay technique was used to analyse serum

samples. Serum dihydrotestosterone level was also detected to determine testosterone and dihydrotestosterone ratio. Data was analysed using SPSS 24. .

RESULTS: Of the 104 subjects with a mean age of 1.78 ± 0.95 years, 96 (92.3%) were diagnosed as cases of androgen insensitivity syndrome on the basis of human chorionic gonadotropins stimulation response level, which was 2-9 times of basal serum testosterone level. Also, 8 (7.7%) subjects were diagnosed to have 5-alpha reductase deficiency syndrome. In such subjects, post-human chorionic gonadotropins response level of serum testosterone was more than 10 times of the basal level.

CONCLUSIONS: The human chorionic gonadotropins stimulation test was found to be comparable to testosterone-to dihydrotestosterone ratio in differentiating between case of androgen insensitivity syndrome and 5-alpha reductase deficiency.

Version ID

1

Status

MEDLINE

Authors Full Name

Sheikh, Waqas Hanif; Asif, Naveed; Haroon, Zujaja Hina; Ejaz, Aamir; Ain, Qural Ul; Shehzad, Noreen.

Institution

Sheikh, Waqas Hanif. Armed Forced Institute of Pathology, Rawalpindi. Asif, Naveed. Armed Forced Institute of Pathology, Rawalpindi.

Haroon, Zujaja Hina. Combined Military Hospital, Multan.

Ejaz, Aamir. Rehman Institute of Medical Sciences, Peshawar.

Ain, Qural Ul. Armed Forced Institute of Pathology, Rawalpindi.

Shehzad, Noreen. Armed Forced Institute of Pathology, Rawalpindi.

Year of Publication

2019

96.

Specific characteristics of urethral strictures in a developing country (Brazil).

Astolfi RH; Lebani BR; Krebs RK; Dias-Filho AC; Bissoli J; Cavalcanti AG; Ximenes SF; Bertolla RP; Geminiani JJ.

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World Journal of Urology. 37(4):661-666, 2019 Apr.

[Journal Article]

UI: 30810832

PURPOSE: Evaluate the main etiologies and clinical characteristics of male urethral stricture disease (USD) in Brazil.

METHODS: This multicentric study was performed using retrospective data collected from six Brazilian referral centers of urethral reconstruction. The database comprised data from 899 patients with USD who had undergone surgical treatment from 2008 to 2018. Age, stricture site and primary stricture etiology were identified for each patient.

RESULTS: The mean age was 52.13 +/- 16.9 years. The most common etiology was iatrogenic (43.4%), followed by idiopathic (21.7%), trauma (21.5%) and inflammatory (13.7%). Of the iatrogenic causes, 59% were secondary to urethral instrumentation (60% by urethral catheterization and 40% by transurethral procedures), 24.8% by other procedures (prostatectomy, radiotherapy, postectomy) and 16.2% by failed hypospadias repairs. Pelvic fracture urethral distraction injuries were responsible for most of the trauma-related strictures (62.7%). When stratified by age, the most common stricture etiology was trauma in the 0-39 years old group (42.8%), idiopathic in the 40-59 years old group (32.4%) and iatrogenic in patients over 60 years old (68%). In regard to the stricture site, 80% presented with an anterior urethral stricture and 20% with a posterior stenosis. In the anterior stenosis group, the most common stricture site was bulbar (39.5%).

CONCLUSION: In Brazil, as in many developed countries, the most common cause of urethral stricture diseases is iatrogenic, especially urethral catheterization. These findings emphasize the need of a careful urethral manipulation and a better training of healthcare professionals. Trauma is still responsible for a great proportion of strictures and inflammatory etiologies are now less frequently observed.

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1

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Authors Full Name

Astolfi, R H; Lebani, B R; Krebs, R K; Dias-Filho, A C; Bissoli, J; Cavalcanti, A G; Ximenes, S F; Bertolla, R P; Geminiani, Julio J.

Institution

Astolfi, R H. Department of Surgery, Division of Urology, Sao Paulo Federal University (UNIFESP), Sao Paulo, Brazil. Lebani, B R. Department of Surgery, Division of Urology, Sao Paulo Federal University (UNIFESP), Sao Paulo, Brazil.

Krebs, R K. Division of Urology, Parana Federal University (UFPR), Curitiba, Brazil.

Dias-Filho, A C. Division of Urology, Federal District Base Hospital, Brasilia, Brazil.

Bissoli, J. Division of Urology, University of Sao Paulo Medical School (FMUSP), Sao Paulo, Brazil.

Cavalcanti, A G. Urogenital Research Unit from Rio de Janeiro State University (UERJ), Rio de Janeiro, Brazil.

Ximenes, S F. Department of Surgery, Division of Urology, Sao Paulo Federal University (UNIFESP), Sao Paulo, Brazil.

Ximenes, S F. Oswaldo Ramos Foundation, Sao Paulo, Brazil.

Bertolla, R P. Department of Surgery, Division of Urology, Sao Paulo Federal University (UNIFESP), Sao Paulo, Brazil.

Geminiani, Julio J. Department of Surgery, Division of Urology, Sao Paulo Federal University (UNIFESP), Sao Paulo, Brazil. juliogeminiani@gmail.com.

Year of Publication
2019

97.

Comparison of preputioplasty and circumcision in distal hypospadias correction: long-term follow-up.

van den Dungen IAL; Rynja SP; Bosch JLHR; de Jong TPVM; de Kort LMO.

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Journal of pediatric urology. 15(1):47.e1-47.e9, 2019 Feb.

[Comparative Study. Journal Article]

UI: 30270101

OBJECTIVE: A normal penile cosmesis is an important goal in distal hypospadias repair.

Depending on cultural standards, repairs are combined with a preputioplasty or circumcision to attain a 'normal' penile appearance. Although short-term complication rates of preputioplasty are available, data on long-term outcomes are scarce. Therefore, this study assessed long-term

functional and cosmetic outcomes of distal hypospadias repair with either a preputioplasty or a circumcision.

PATIENTS AND METHODS: Eligible for inclusion were patients with distal hypospadias operated in childhood between 1987 and 1993. Complications and reasons for secondary circumcision were extracted from the medical charts. Participants completed a questionnaire including the International Index of Erectile Function (IIEF-15), the International Prostate Symptom score (IPSS), and additional non-validated questions. Penile cosmesis was judged with the Penile Perception Score (PPS), stretched penile length was measured, and uroflowmetry was performed.

RESULTS: Of the 86 eligible and traceable patients, 40 (47%) participated; of them, 27 had a preputioplasty and 13 a circumcision. Six patients underwent a secondary circumcision due to a preputial defect (n = 2), unsatisfactory cosmetic result (n = 2), religious reason (n = 1), or phimosis (n = 1). Complication rates were similar in both the groups. Long-term outcomes in the preputioplasty and circumcision group were comparable regarding cosmetic, sexual, and micturition outcomes.

CONCLUSIONS: Distal hypospadias correction combined with preputioplasty had complication rates similar to those of hypospadias repair with circumcision. In these patients, preputioplasty had a failure rate of 22%. In both the groups, long-term outcomes of urinary function, sexual function, and cosmesis were good.

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1

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MEDLINE

Authors Full Name

van den Dungen, I A L; Rynja, S P; Bosch, J L H R; de Jong, T P V M; de Kort, L M O.

Institution

van den Dungen, I A L. University Medical Center Utrecht (UMC Utrecht), The Netherlands.

Electronic address: ialdungen@gmail.com. Rynja, S P. University Medical Center Utrecht (UMC Utrecht), The Netherlands. Electronic address: sybrenrynja@hotmail.com.

Bosch, J L H R. University Medical Center Utrecht (UMC Utrecht), The Netherlands. Electronic address: J.L.H.R.Bosch@umcutrecht.nl.

de Jong, T P V M. University Children's Hospital UMC Utrecht, The Netherlands. Electronic address: T.P.V.M.dejong@umcutrecht.nl.

de Kort, L M O. University Medical Center Utrecht (UMC Utrecht), The Netherlands. Electronic address: L.M.O.deKort@umcutrecht.nl.

Comments

Comment in (CIN)

Year of Publication

2019

98.

Risks of 23 specific malformations associated with prenatal exposure to 10 antiepileptic drugs. Blotiere PO; Raguideau F; Weill A; Elefant E; Perthuis I; Goulet V; Rouget F; Zureik M; Coste J; Dray-Spira R.

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Neurology. 93(2):e167-e180, 2019 07 09.

[Journal Article. Research Support, Non-U.S. Gov't]

UI: 31189695

OBJECTIVE: To assess the association between exposure to monotherapy with 10 different antiepileptic drugs (AEDs) during the first 2 months of pregnancy and the risk of 23 major congenital malformations (MCMs).

METHODS: This nationwide cohort study, based on the French health care databases, included all pregnancies ≥ 20 weeks and ending between January 2011 and March 2015. Women were considered to be exposed when an AED had been dispensed between 1 month before and 2 months after the beginning of pregnancy. The reference group included pregnant women with no reimbursement for AEDs. MCMs were detected up to 12 months after birth (24 months for microcephaly, hypospadias, and epispadias). Odds ratios (ORs) were adjusted for potential confounders for MCMs with at least 5 cases. Otherwise, we calculated crude ORs with exact confidence intervals (CIs).

RESULTS: The cohort included 1,886,825 pregnancies, 2,997 of which were exposed to lamotrigine, 1,671 to pregabalin, 980 to clonazepam, 913 to valproic acid, 579 to levetiracetam, 517 to topiramate, 512 to carbamazepine, 365 to gabapentin, 139 to oxcarbazepine, and 80 to phenobarbital. Exposure to valproic acid was associated with 8 specific types of MCMs (e.g., spina bifida, OR 19.4, 95% CI 8.6-43.5), and exposure to topiramate was associated with an increased risk of cleft lip (6.8, 95% CI 1.4-20.0). We identified 3 other signals. We found no

significant association for lamotrigine, levetiracetam, carbamazepine, oxcarbazepine, and gabapentin.

CONCLUSIONS: These results confirm the teratogenicity of valproic acid and topiramate.

Because of the small numbers of cases and possible confounding, the other 3 signals should be interpreted with appropriate caution.

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Version ID

1

Status

MEDLINE

Author Initials

Blotiere, Pierre-Olivier; ORCID: <https://orcid.org/0000-0002-6857-6781> Weill, Alain; ORCID: <https://orcid.org/0000-0001-8687-9092>

Authors Full Name

Blotiere, Pierre-Olivier; Raguideau, Fanny; Weill, Alain; Elefant, Elisabeth; Perthus, Isabelle; Goulet, Veronique; Rouget, Florence; Zureik, Mahmoud; Coste, Joel; Dray-Spira, Rosemary.

Institution

Blotiere, Pierre-Olivier. From the Department of Studies in Public Health (P.-O.B., A.W., J.C.), French National Health Insurance (CNAM), Paris; Universite de Lorraine (P.-O.B.), Universite Paris-Descartes, Apemac, Nancy; Department of Epidemiology of Health Products (F. Raguideau, M.Z., R.D.-S.), French National Agency for Medicines and Health Products Safety, Saint-Denis; Reference Center on Teratogenic Agents (E.E.), Hopital Trousseau, Groupe Hospitalo-Universitaire Est Parisien, Assistance Publique Hopitaux de Paris; Auvergne Registry of Congenital Malformations (I.P.), Centre de reference des Anomalies du Developpement et des maladies rares, Service de genetique medicale, CHU Clermont-Ferrand; Department of Chronic Diseases and Injuries (V.G.), French Public Health Agency, Saint Maurice; Brittany Registry of Congenital Malformations (F. Rouget), Universite de Rennes, CHU Rennes, Inserm, EHESP, Irset (Institut de recherche en sante, environnement et travail)-UMR_S 1085; Versailles Saint-Quentin University (M.Z.); and Biostatistics and Epidemiology Unit (J.C.), Hotel-Dieu Hospital, Assistance Publique-Hopitaux de Paris, and Paris Descartes University, France. pierre-olivier.blotiere@assurance-maladie.fr. Raguideau, Fanny. From the Department of Studies in Public Health (P.-O.B., A.W., J.C.), French National Health Insurance (CNAM), Paris; Universite de Lorraine (P.-O.B.), Universite Paris-Descartes, Apemac, Nancy; Department of Epidemiology of Health Products (F. Raguideau, M.Z., R.D.-S.), French National Agency for Medicines and Health Products Safety, Saint-Denis; Reference Center on Teratogenic Agents (E.E.), Hopital Trousseau, Groupe Hospitalo-Universitaire Est Parisien, Assistance Publique Hopitaux de Paris; Auvergne Registry of Congenital Malformations (I.P.), Centre de reference des Anomalies du

Developpement et des maladies rares, Service de genetique medicale, CHU Clermont-Ferrand; Department of Chronic Diseases and Injuries (V.G.), French Public Health Agency, Saint Maurice; Brittany Registry of Congenital Malformations (F. Rouget), Universite de Rennes, CHU Rennes, Inserm, EHESP, Irset (Institut de recherche en sante, environnement et travail)-UMR_S 1085; Versailles Saint-Quentin University (M.Z.); and Biostatistics and Epidemiology Unit (J.C.), Hotel-Dieu Hospital, Assistance Publique-Hopitaux de Paris, and Paris Descartes University, France.

Weill, Alain. From the Department of Studies in Public Health (P.-O.B., A.W., J.C.), French National Health Insurance (CNAM), Paris; Universite de Lorraine (P.-O.B.), Universite Paris-Descartes, Apemac, Nancy; Department of Epidemiology of Health Products (F. Raguideau, M.Z., R.D.-S.), French National Agency for Medicines and Health Products Safety, Saint-Denis; Reference Center on Teratogenic Agents (E.E.), Hopital Trousseau, Groupe Hospitalo-Universitaire Est Parisien, Assistance Publique Hopitaux de Paris; Auvergne Registry of Congenital Malformations (I.P.), Centre de reference des Anomalies du Developpement et des maladies rares, Service de genetique medicale, CHU Clermont-Ferrand; Department of Chronic Diseases and Injuries (V.G.), French Public Health Agency, Saint Maurice; Brittany Registry of Congenital Malformations (F. Rouget), Universite de Rennes, CHU Rennes, Inserm, EHESP, Irset (Institut de recherche en sante, environnement et travail)-UMR_S 1085; Versailles Saint-Quentin University (M.Z.); and Biostatistics and Epidemiology Unit (J.C.), Hotel-Dieu Hospital, Assistance Publique-Hopitaux de Paris, and Paris Descartes University, France.

Elefant, Elisabeth. From the Department of Studies in Public Health (P.-O.B., A.W., J.C.), French National Health Insurance (CNAM), Paris; Universite de Lorraine (P.-O.B.), Universite Paris-Descartes, Apemac, Nancy; Department of Epidemiology of Health Products (F. Raguideau, M.Z., R.D.-S.), French National Agency for Medicines and Health Products Safety, Saint-Denis; Reference Center on Teratogenic Agents (E.E.), Hopital Trousseau, Groupe Hospitalo-Universitaire Est Parisien, Assistance Publique Hopitaux de Paris; Auvergne Registry of Congenital Malformations (I.P.), Centre de reference des Anomalies du Developpement et des maladies rares, Service de genetique medicale, CHU Clermont-Ferrand; Department of Chronic Diseases and Injuries (V.G.), French Public Health Agency, Saint Maurice; Brittany Registry of Congenital Malformations (F. Rouget), Universite de Rennes, CHU Rennes, Inserm, EHESP, Irset (Institut de recherche en sante, environnement et travail)-UMR_S 1085; Versailles Saint-Quentin University (M.Z.); and Biostatistics and Epidemiology Unit (J.C.), Hotel-Dieu Hospital, Assistance Publique-Hopitaux de Paris, and Paris Descartes University, France.

Perthus, Isabelle. From the Department of Studies in Public Health (P.-O.B., A.W., J.C.), French National Health Insurance (CNAM), Paris; Universite de Lorraine (P.-O.B.), Universite Paris-Descartes, Apemac, Nancy; Department of Epidemiology of Health Products (F. Raguideau, M.Z., R.D.-S.), French National Agency for Medicines and Health Products Safety, Saint-Denis;

Reference Center on Teratogenic Agents (E.E.), Hopital Trousseau, Groupe Hospitalo-Universitaire Est Parisien, Assistance Publique Hopitaux de Paris; Auvergne Registry of Congenital Malformations (I.P.), Centre de reference des Anomalies du Developpement et des maladies rares, Service de genetique medicale, CHU Clermont-Ferrand; Department of Chronic Diseases and Injuries (V.G.), French Public Health Agency, Saint Maurice; Brittany Registry of Congenital Malformations (F. Rouget), Universite de Rennes, CHU Rennes, Inserm, EHESP, Irset (Institut de recherche en sante, environnement et travail)-UMR_S 1085; Versailles Saint-Quentin University (M.Z.); and Biostatistics and Epidemiology Unit (J.C.), Hotel-Dieu Hospital, Assistance Publique-Hopitaux de Paris, and Paris Descartes University, France.

Goulet, Veronique. From the Department of Studies in Public Health (P.-O.B., A.W., J.C.), French National Health Insurance (CNAM), Paris; Universite de Lorraine (P.-O.B.), Universite Paris-Descartes, Apemac, Nancy; Department of Epidemiology of Health Products (F. Raguideau, M.Z., R.D.-S.), French National Agency for Medicines and Health Products Safety, Saint-Denis; Reference Center on Teratogenic Agents (E.E.), Hopital Trousseau, Groupe Hospitalo-Universitaire Est Parisien, Assistance Publique Hopitaux de Paris; Auvergne Registry of Congenital Malformations (I.P.), Centre de reference des Anomalies du Developpement et des maladies rares, Service de genetique medicale, CHU Clermont-Ferrand; Department of Chronic Diseases and Injuries (V.G.), French Public Health Agency, Saint Maurice; Brittany Registry of Congenital Malformations (F. Rouget), Universite de Rennes, CHU Rennes, Inserm, EHESP, Irset (Institut de recherche en sante, environnement et travail)-UMR_S 1085; Versailles Saint-Quentin University (M.Z.); and Biostatistics and Epidemiology Unit (J.C.), Hotel-Dieu Hospital, Assistance Publique-Hopitaux de Paris, and Paris Descartes University, France.

Rouget, Florence. From the Department of Studies in Public Health (P.-O.B., A.W., J.C.), French National Health Insurance (CNAM), Paris; Universite de Lorraine (P.-O.B.), Universite Paris-Descartes, Apemac, Nancy; Department of Epidemiology of Health Products (F. Raguideau, M.Z., R.D.-S.), French National Agency for Medicines and Health Products Safety, Saint-Denis; Reference Center on Teratogenic Agents (E.E.), Hopital Trousseau, Groupe Hospitalo-Universitaire Est Parisien, Assistance Publique Hopitaux de Paris; Auvergne Registry of Congenital Malformations (I.P.), Centre de reference des Anomalies du Developpement et des maladies rares, Service de genetique medicale, CHU Clermont-Ferrand; Department of Chronic Diseases and Injuries (V.G.), French Public Health Agency, Saint Maurice; Brittany Registry of Congenital Malformations (F. Rouget), Universite de Rennes, CHU Rennes, Inserm, EHESP, Irset (Institut de recherche en sante, environnement et travail)-UMR_S 1085; Versailles Saint-Quentin University (M.Z.); and Biostatistics and Epidemiology Unit (J.C.), Hotel-Dieu Hospital, Assistance Publique-Hopitaux de Paris, and Paris Descartes University, France.

Zureik, Mahmoud. From the Department of Studies in Public Health (P.-O.B., A.W., J.C.), French National Health Insurance (CNAM), Paris; Universite de Lorraine (P.-O.B.), Universite Paris-

Descartes, Apemac, Nancy; Department of Epidemiology of Health Products (F. Raguideau, M.Z., R.D.-S.), French National Agency for Medicines and Health Products Safety, Saint-Denis; Reference Center on Teratogenic Agents (E.E.), Hopital Trousseau, Groupe Hospitalo-Universitaire Est Parisien, Assistance Publique Hopitaux de Paris; Auvergne Registry of Congenital Malformations (I.P.), Centre de reference des Anomalies du Developpement et des maladies rares, Service de genetique medicale, CHU Clermont-Ferrand; Department of Chronic Diseases and Injuries (V.G.), French Public Health Agency, Saint Maurice; Brittany Registry of Congenital Malformations (F. Rouget), Universite de Rennes, CHU Rennes, Inserm, EHESP, Irset (Institut de recherche en sante, environnement et travail)-UMR_S 1085; Versailles Saint-Quentin University (M.Z.); and Biostatistics and Epidemiology Unit (J.C.), Hotel-Dieu Hospital, Assistance Publique-Hopitaux de Paris, and Paris Descartes University, France.

Coste, Joel. From the Department of Studies in Public Health (P.-O.B., A.W., J.C.), French National Health Insurance (CNAM), Paris; Universite de Lorraine (P.-O.B.), Universite Paris-Descartes, Apemac, Nancy; Department of Epidemiology of Health Products (F. Raguideau, M.Z., R.D.-S.), French National Agency for Medicines and Health Products Safety, Saint-Denis; Reference Center on Teratogenic Agents (E.E.), Hopital Trousseau, Groupe Hospitalo-Universitaire Est Parisien, Assistance Publique Hopitaux de Paris; Auvergne Registry of Congenital Malformations (I.P.), Centre de reference des Anomalies du Developpement et des maladies rares, Service de genetique medicale, CHU Clermont-Ferrand; Department of Chronic Diseases and Injuries (V.G.), French Public Health Agency, Saint Maurice; Brittany Registry of Congenital Malformations (F. Rouget), Universite de Rennes, CHU Rennes, Inserm, EHESP, Irset (Institut de recherche en sante, environnement et travail)-UMR_S 1085; Versailles Saint-Quentin University (M.Z.); and Biostatistics and Epidemiology Unit (J.C.), Hotel-Dieu Hospital, Assistance Publique-Hopitaux de Paris, and Paris Descartes University, France.

Dray-Spira, Rosemary. From the Department of Studies in Public Health (P.-O.B., A.W., J.C.), French National Health Insurance (CNAM), Paris; Universite de Lorraine (P.-O.B.), Universite Paris-Descartes, Apemac, Nancy; Department of Epidemiology of Health Products (F. Raguideau, M.Z., R.D.-S.), French National Agency for Medicines and Health Products Safety, Saint-Denis; Reference Center on Teratogenic Agents (E.E.), Hopital Trousseau, Groupe Hospitalo-Universitaire Est Parisien, Assistance Publique Hopitaux de Paris; Auvergne Registry of Congenital Malformations (I.P.), Centre de reference des Anomalies du Developpement et des maladies rares, Service de genetique medicale, CHU Clermont-Ferrand; Department of Chronic Diseases and Injuries (V.G.), French Public Health Agency, Saint Maurice; Brittany Registry of Congenital Malformations (F. Rouget), Universite de Rennes, CHU Rennes, Inserm, EHESP, Irset (Institut de recherche en sante, environnement et travail)-UMR_S 1085; Versailles Saint-Quentin University (M.Z.); and Biostatistics and Epidemiology Unit (J.C.), Hotel-Dieu Hospital, Assistance Publique-Hopitaux de Paris, and Paris Descartes University, France.

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<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC6656651>

Year of Publication

2019

99.

Hypospadias Risk from Maternal Residential Exposure to Heavy Metal Hazardous Air Pollutants. White JT; Kovar E; Chambers TM; Sheth KR; Peckham-Gregory EC; O'Neill M; Langlois PH; Jorgez CJ; Lupo PJ; Seth A.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid

MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

International Journal of Environmental Research & Public Health [Electronic Resource]. 16(6), 2019 03 15.

[Journal Article. Research Support, N.I.H., Extramural. Research Support, Non-U.S. Gov't]

UI: 30875870

Objective: Investigate whether residential prenatal exposure to heavy metal hazardous air pollutants (HMHAPs) is associated with an increased risk of hypospadias. Methods: Data on non-syndromic hypospadias cases (n = 8981) and control patients delivered in Texas were obtained from the Texas Birth Defects Registry and matched 1:10 by birth year. Average exposure concentrations of HMHAPs were obtained from the 2005 U.S. Environmental Protection Agency National-Scale Air Toxics Assessment and categorized into quintiles. Odds ratios and 95% confidence intervals were estimated. STROBE reporting guidelines were followed. Results: We observed associations between hypospadias and prenatal HMHAP exposure. Manganese demonstrated significant increased risk of hypospadias at the medium, medium-high and high exposure quintiles; lead in the medium-high and high exposure quintiles. Cadmium, mercury and nickel demonstrated a significant inverted "U-shaped" association for exposures with significant associations in the medium and medium-high quintiles but not in the medium-low and high quintiles. Arsenic and chromium demonstrated a significant bivalent association for risk of hypospadias in a lower quintile as well as a higher quintile with non-significant intermediate quintiles. Conclusions: Using data from one of the world's largest active surveillance birth defects registries, we identified significant associations between hypospadias and HMHAP exposures.

These results should be used in counseling for maternal demographic risk factors as well as avoidance of heavy metals and their sources.

Version ID

1

Status

MEDLINE

Author Initials

White, Jeffrey T; ORCID: <https://orcid.org/0000-0002-9015-7719> Peckham-Gregory, Erin C;
ORCID: <https://orcid.org/0000-0002-7593-8381>

Langlois, Peter H; ORCID: <https://orcid.org/0000-0001-6765-4963>

Jorgez, Carolina J; ORCID: <https://orcid.org/0000-0001-9203-6331>

Authors Full Name

White, Jeffrey T; Kovar, Erin; Chambers, Tiffany M; Sheth, Kunj R; Peckham-Gregory, Erin C;
O'Neill, Marisol; Langlois, Peter H; Jorgez, Carolina J; Lupo, Philip J; Seth, Abhishek.

Institution

White, Jeffrey T. Pediatric Urology, Norton Children's Hospital, Louisville, KY 40207, USA.
jeffrey.white@nortonhealthcare.org. White, Jeffrey T. Department of Urology, University of

Louisville School of Medicine, Louisville, KY 40202, USA. jeffrey.white@nortonhealthcare.org.

Kovar, Erin. Section of Hematology-Oncology, Department of Pediatrics, Baylor College of
Medicine, Houston, TX 77030, USA. emkovar@bcm.edu.

Chambers, Tiffany M. Section of Hematology-Oncology, Department of Pediatrics, Baylor College
of Medicine, Houston, TX 77030, USA. Tiffany.Chambers@bcm.edu.

Sheth, Kunj R. Scott Department of Urology, Baylor College of Medicine, Houston, TX 77030,
USA. Kunj.Sheth@bcm.edu.

Sheth, Kunj R. Division of Pediatric Urology, Department of Surgery, Texas Children's Hospital,
Houston, TX 77030, USA. Kunj.Sheth@bcm.edu.

Peckham-Gregory, Erin C. Section of Hematology-Oncology, Department of Pediatrics, Baylor
College of Medicine, Houston, TX 77030, USA. Erin.Peckham@bcm.edu.

Peckham-Gregory, Erin C. Department of Pediatrics, Texas Children's Hospital, Houston, TX
77030, USA. Erin.Peckham@bcm.edu.

O'Neill, Marisol. Department Molecular and Cellular Biology, Baylor College of Medicine,
Houston, TX 77030, USA. Marisol.oneill@ucsf.edu.

Langlois, Peter H. Birth Defects Epidemiology and Surveillance Branch, Texas Department of
State Health Services, Austin, TX 78751, USA. peter.langlois@dshs.texas.gov.

Jorgez, Carolina J. Scott Department of Urology, Baylor College of Medicine, Houston, TX 77030,
USA. cj129804@bcm.edu.

Jorgez, Carolina J. Division of Pediatric Urology, Department of Surgery, Texas Children's Hospital, Houston, TX 77030, USA. cj129804@bcm.edu.

Lupo, Philip J. Section of Hematology-Oncology, Department of Pediatrics, Baylor College of Medicine, Houston, TX 77030, USA. Philip.Lupo@bcm.edu.

Lupo, Philip J. Department of Pediatrics, Texas Children's Hospital, Houston, TX 77030, USA. Philip.Lupo@bcm.edu.

Seth, Abhishek. Scott Department of Urology, Baylor College of Medicine, Houston, TX 77030, USA. axseth@texaschildrens.org.

Seth, Abhishek. Division of Pediatric Urology, Department of Surgery, Texas Children's Hospital, Houston, TX 77030, USA. axseth@texaschildrens.org.

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100.

Impact of Surgical Treatment of Penile Fracture on Sexual Function.
Barros R; Schul A; Ornellas P; Koifman L; Favorito LA.
OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present
Urology. 126:128-133, 2019 04.
[Journal Article]
UI: 30605691
OBJECTIVE: To conduct a comprehensive assessment of sexual function of patients undergoing surgical treatment of penile fracture (PF), covering psychological aspects related to trauma.
METHODS: Patients undergoing surgical treatment of PF from January 2014 to August 2017 were followed-up in our department for at least 6 months. The patients underwent a detailed clinical follow-up, including physical examination and were interviewed about any evidence of erectile dysfunction, penile nodules, or curvature acquired after surgery, besides psychological sexual problems.
RESULTS: A total of 58 patients conducted the follow-up. The mean age was 38.5 years (range: 18-66 years). Eight (13.7%) patients complained of penile curvature after surgery. Postoperative

erectile function was recovered after 6 months in 50 (86.2%) cases. After the last evaluation at 18 months, only 1 patient developed persistent erectile dysfunction (ED) and color duplex Doppler ultrasound excluded a vascular etiology. Psycho-sexual evaluations showed that 45 (77.5%) patients feared a new episode of PF. Changes in sexual habits, such as avoiding vigorous sexual intercourse, was reported by 40 (68.9%) patients. Finally, patients with performance anxiety and those who reported a negative impact on sexual life were more susceptible to the development of ED ($P=.0337$ and $P=.0418$, respectively).

CONCLUSION: Sexual complications after surgical treatment of PF are unusual but may occur in the late postoperative period and should be treated. Psychological sequela is very common, causing fear of recurrence and psychogenic ED, resulting in negative impact on the sexual life of these patients, which should be monitored closely.

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1

Status

MEDLINE

Authors Full Name

Barros, Rodrigo; Schul, Alex; Ornellas, Paulo; Koifman, Leandro; Favorito, Luciano A.

Institution

Barros, Rodrigo. Souza Aguiar Municipal Hospital, State University of Rio de Janeiro, Rio de Janeiro, RJ, Brazil. Schul, Alex. Souza Aguiar Municipal Hospital, State University of Rio de Janeiro, Rio de Janeiro, RJ, Brazil.

Ornellas, Paulo. Souza Aguiar Municipal Hospital, State University of Rio de Janeiro, Rio de Janeiro, RJ, Brazil.

Koifman, Leandro. Souza Aguiar Municipal Hospital, State University of Rio de Janeiro, Rio de Janeiro, RJ, Brazil.

Favorito, Luciano A. Souza Aguiar Municipal Hospital, State University of Rio de Janeiro, Rio de Janeiro, RJ, Brazil. Electronic address: lufavorito@Yahoo.com.br.

Comments

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Year of Publication

2019

Pericardium in Reconstructive Urologic Surgeries: A Systematic Review and Meta-Analysis.

Hosseini J; Hosseini S; Hosseini MA; Rezaei Y.

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Urologia Internationalis. 102(2):131-144, 2019.

[Journal Article. Meta-Analysis. Systematic Review]

UI: 30517943

INTRODUCTION: The use of pericardium has been expanded into different surgical modalities; however, there are scarce data regarding the feasibility of the pericardium in reconstructive urologic surgeries. We systematically reviewed the literature on the effectiveness of the pericardial tissue for reconstructive urologic surgeries.

MATERIALS AND METHODS: PubMed and Scopus were searched online for evidence on the use of the pericardium in urologic surgeries. Through the methodology recommended by the Preferred Reporting Items for Systematic Reviews and Meta-analysis guidelines, 38 of 4,071 studies were identified.

RESULTS: A total of 715 patients and 139 animals underwent reconstructive urologic surgeries using the pericardium. Bladder, urethral, and renal reconstructions were successful in 100% of the human cases. The rates of dissatisfaction, glans hypoesthesia, and penile shortening were comparable between the pericardial graft surgeries and the other operations during penile straightening, but there was a trend among the patients with pericardial grafts toward having a more penile curvature at follow-up (risk ratio [RR] 2.03, 95% CI 0.90-4.61, $p = 0.09$; $I^2 = 0\%$). Among the animal studies, there were 4 reports of penile reconstruction, 7 studies of bladder reconstruction, and 1 study of urethroplasty. Bladder reconstruction and urethroplasty were successful in 83 and 20% of the animals, respectively. The pooled result of the stimulated intracorporeal pressure 5 V significantly favored pericardial grafts during penile reconstruction (RR 2.61, 95% CI 1.26-3.97, $p = 0.0002$; $I^2 = 0\%$).

CONCLUSIONS: Our systematic review demonstrates the feasibility of the pericardium, regardless of its type, in urologic surgeries. It, however, seems that urethral substitution needs further investigation. Given the lower cost, easier handling, and less immunogenicity of the pericardium, further studies are required to examine its pros and cons.

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1

Status

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Authors Full Name

Hosseini, Jalil; Hosseini, Saeid; Hosseini, Mohammad Ali; Rezaei, Yousef.

Institution

Hosseini, Jalil. Infertility and Reproductive Health Research Center, Shahid Beheshti University of Medical Sciences, Tehran, Iran. Hosseini, Saeid. Heart Valve Disease Research Center, Rajaie Cardiovascular Medical and Research Center, Iran University of Medical Sciences, Tehran, Iran. Hosseini, Mohammad Ali. Student Research Committee, Qazvin University of Medical Sciences, Qazvin, Iran, smahoseini@gmail.com.

Rezaei, Yousef. Heart Valve Disease Research Center, Rajaie Cardiovascular Medical and Research Center, Iran University of Medical Sciences, Tehran, Iran.

Year of Publication

2019

102.

Single Nucleotide Polymorphisms of HAAO and IRX6 Genes as Risk Factors for Hypospadias. Kojima Y; Koguchi T; Mizuno K; Sato Y; Hoshi S; Hata J; Nishio H; Hashimoto D; Matsushita S; Suzuki K; Miyagawa S; Hui CC; Tanikawa C; Murakami Y; Yamada G; Hayashi Y; Matsuda K.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid

MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Journal of Urology. 201(2):386-392, 2019 02.

[Journal Article]

UI: 30063927

PURPOSE: We evaluated the association of hypospadias and 17 susceptibility loci previously identified by a European genome-wide association study in a cohort of Japanese patients. We also examined the expression of candidate genes in male mouse embryos to determine the possible underlying mechanisms of this disease.

MATERIALS AND METHODS: We enrolled 169 Japanese patients (mean age at surgery 3.7 years) who underwent repair of hypospadias. Genotyping of 17 single nucleotide polymorphisms was performed using a multiplex polymerase chain reaction invader assay. We also performed in situ hybridization to determine whether candidate genes were expressed in the male genital tubercle during embryonic development of the external genitalia in mice.

RESULTS: Single nucleotide polymorphism rs3816183 of HAAO was significantly associated with susceptibility to hypospadias in general ($p = 0.0019$) and to anterior/middle hypospadias ($p = 0.0283$) and posterior hypospadias ($p = 0.0226$), while single nucleotide polymorphism rs6499755 of IRX6 showed an association with susceptibility to anterior/middle hypospadias ($p = 0.0472$). In mouse embryos there was no significant upregulation of HaaO expression in the developing male external genitalia. *Irx3* and *Irx5*, which are linked to *Irx6* within the *IrxB* cluster, were expressed in the mesenchyme remote from the urethral plate epithelium during the critical embryonic period for masculinization. *Irx6* was expressed in the ectodermal epithelium, demonstrating prominent dorsal ectodermal expression without expression in the ventral ectoderm adjacent to the urethral plate during the same period.

CONCLUSIONS: Genetic variations of HAAO and IRX6 influence susceptibility to hypospadias in the Japanese population. Further research is needed to clarify the mechanism by which variations in these genes contribute to the pathogenesis of hypospadias.

Version ID

1

Status

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Authors Full Name

Kojima, Yoshiyuki; Koguchi, Tomoyuki; Mizuno, Kentaro; Sato, Yuichi; Hoshi, Seiji; Hata, Junya; Nishio, Hidenori; Hashimoto, Daiki; Matsushita, Shoko; Suzuki, Kentaro; Miyagawa, Shinichi; Hui, Chi Chung; Tanikawa, Chizu; Murakami, Yoshimori; Yamada, Gen; Hayashi, Yutaro; Matsuda, Koichi.

Institution

Kojima, Yoshiyuki. Department of Urology, Fukushima Medical University School of Medicine, Fukushima, Japan. Koguchi, Tomoyuki. Department of Urology, Fukushima Medical University School of Medicine, Fukushima, Japan.

Mizuno, Kentaro. Department of Pediatric Urology, Nagoya City University Graduate School of Medical Sciences, Nagoya, Japan.

Sato, Yuichi. Department of Urology, Fukushima Medical University School of Medicine, Fukushima, Japan.

Hoshi, Seiji. Department of Urology, Fukushima Medical University School of Medicine, Fukushima, Japan.

Hata, Junya. Department of Urology, Fukushima Medical University School of Medicine, Fukushima, Japan.

Nishio, Hidenori. Department of Pediatric Urology, Nagoya City University Graduate School of Medical Sciences, Nagoya, Japan.

Hashimoto, Daiki. Department of Developmental Genetics, Institute of Advanced Medicine, Wakayama Medical University, Kimiidera, Wakayama, Japan.

Matsushita, Shoko. Department of Developmental Genetics, Institute of Advanced Medicine, Wakayama Medical University, Kimiidera, Wakayama, Japan.

Suzuki, Kentaro. Department of Developmental Genetics, Institute of Advanced Medicine, Wakayama Medical University, Kimiidera, Wakayama, Japan.

Miyagawa, Shinichi. Department of Developmental Genetics, Institute of Advanced Medicine, Wakayama Medical University, Kimiidera, Wakayama, Japan.

Hui, Chi Chung. Program in Developmental and Stem Cell Biology, Research Institute, Hospital for Sick Children, Toronto, Ontario, Canada.

Tanikawa, Chizu. Laboratory of Molecular Medicine, Human Genome Center, Institute of Medical Science, University of Tokyo, Tokyo, Japan.

Murakami, Yoshimori. Division of Molecular Pathology, Institute of Medical Science, University of Tokyo, Tokyo, Japan.

Yamada, Gen. Department of Developmental Genetics, Institute of Advanced Medicine, Wakayama Medical University, Kimiidera, Wakayama, Japan.

Hayashi, Yutaro. Department of Pediatric Urology, Nagoya City University Graduate School of Medical Sciences, Nagoya, Japan.

Matsuda, Koichi. Laboratory of Molecular Medicine, Human Genome Center, Institute of Medical Science, University of Tokyo, Tokyo, Japan.

Comments

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Year of Publication

2019

103.

Common Practice of Hypospadias Management by Pediatric Urologists in Indonesia: A Multi-center Descriptive Study from Referral Hospitals.

Duarsa GWK; Tirtayasa PMW; Daryanto B; Nurhadi P; Renaldo J; Tarmono T; Utomo T; Yuri P; Siregar S; Wahyudi I; Situmorang GR; Palinrungi MAA; Hutasoit YI; Hutahaeen AYA; Zulfiqar Y; Sigumonrong YH; Mirza H; Rodjani A.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid
MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present
Open Access Macedonian Journal of Medical Sciences. 7(14):2242-2245, 2019 Jul 30.

[Journal Article]

UI: 31592011

BACKGROUND: Hypospadias is the second most common congenital anomalies among human congenital disabilities. There are over 300 surgery techniques being introduced to treat hypospadias. The successful of hypospadias repair is assessed by several outcomes as well as complications following surgery.

AIM: This study aims to show the multicenter hypospadias data in Indonesia descriptively.

METHODS: All the data were compiled based on questionnaires, which were distributed to Indonesian pediatric urologists. The questionnaire includes several questions containing demographic aspect, preferred techniques being used, and complications being found regarding hypospadias repair.

RESULTS: Eighteen Indonesian pediatric urologists from 12 centres involved in this study. The data were collected from June - September 2018 based on the surgeon's experience throughout 2017. From 591 cases based on the returned questionnaire, penile-type hypospadias was the most common type of hypospadias being treated (35.7%) followed by penoscrotal (28.9%) and scrotal-type (12.9%). Moderate severity of chordee was mostly seen among all cases (40.6%). Tubularised incised plate (TIP), + Thiersch Duplay, was the most common technique being used to treat hypospadias (44.3%), followed by onlay island preputial flap (14.9%) and two-stage technique (14%). The incidence of urethrocuteaneous fistulae in this study was 13.9%.

CONCLUSION: This study showed how Indonesian pediatric urologists dealt with hypospadias cases. TIP + Thiersch Duplay procedure being the preferred technique used by most participants and the rate of urethrocuteaneous fistulae as one of the complications was comparable with previous studies.

Copyright: © 2019 Gede Wirya Kusuma Duarsa, Pande Made Wisnu Tirtayasa, Besut Daryanto, Pradana Nurhadi, Johan Renaldo, Tarmono Tarmono, Trisulo Utomo, Prahara Yuri, Safendra Siregar, Irfan Wahyudi, Gerhard Reinaldi Situmorang, Muhammad Asykar A. Palinrungi, Yonas Immanuel Hutasoit, Andre Yudha Alfanius Hutahaeen, Yevri Zulfiqar, Yacobda H. Sigumonrong, Hendy Mirza, Arry Rodjani.

Version ID

1

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Authors Full Name

Duarsa, Gede Wirya Kusuma; Tirtayasa, Pande Made Wisnu; Daryanto, Besut; Nurhadi, Pradana; Renaldo, Johan; Tarmono, Tarmono; Utomo, Trisulo; Yuri, Prahara; Siregar, Safendra; Wahyudi, Irfan; Situmorang, Gerhard Reinaldi; Palinrungi, Muhammad Asykar A; Hutasoit, Yonas Immanuel; Hutahaeen, Andre Yudha Alfanius; Zulfihar, Yevri; Sigumonrong, Yacobda H; Mirza, Hendy; Rodjani, Arry.

Institution

Duarsa, Gede Wirya Kusuma. Division of Urology, Department of Surgery, Faculty of Medicine Universitas Udayana, Sanglah Hospital, Denpasar, Indonesia. Tirtayasa, Pande Made Wisnu. Division of Urology, Department of Surgery, Faculty of Medicine Universitas Udayana, Sanglah Hospital, Denpasar, Indonesia.

Daryanto, Besut. Department of Urology, Faculty of Medicine Universitas Brawijaya, Saiful Anwar Hospital, Malang, Indonesia.

Nurhadi, Pradana. Department of Urology, Faculty of Medicine Universitas Brawijaya, Saiful Anwar Hospital, Malang, Indonesia.

Renaldo, Johan. Department of Urology, Faculty of Medicine Universitas Airlangga, Soetomo Hospital, Surabaya, Indonesia.

Tarmono, Tarmono. Department of Urology, Faculty of Medicine Universitas Airlangga, Soetomo Hospital, Surabaya, Indonesia.

Utomo, Trisulo. Department of Urology, Faculty of Medicine Universitas Gadjah Mada, Sardjito Hospital, Yogyakarta, Indonesia.

Yuri, Prahara. Department of Urology, Faculty of Medicine Universitas Gadjah Mada, Sardjito Hospital, Yogyakarta, Indonesia.

Siregar, Safendra. Department of Urology, Faculty of Medicine Universitas Padjajaran, Hasan Sadikin Hospital, Bandung, Indonesia.

Wahyudi, Irfan. Department of Urology, Faculty of Medicine Universitas Indonesia, Cipto Mangunkusumo Hospital, Jakarta, Indonesia.

Situmorang, Gerhard Reinaldi. Department of Urology, Faculty of Medicine Universitas Indonesia, Cipto Mangunkusumo Hospital, Jakarta, Indonesia.

Palinrungi, Muhammad Asykar A. Division of Urology, Department of Surgery, Faculty of Medicine Universitas Hasannudin, Wahidin Sudirohusodo Hospital, Makassar, Indonesia.

Hutasoit, Yonas Immanuel. Department of Surgery, Fatmawati Hospital, Jakarta, Indonesia.

Hutahaeen, Andre Yudha Alfanius. Department of Surgery, Harapan Kita Hospital, Jakarta, Indonesia.

Zulfihar, Yevri. Division of Urology, Department of Surgery, Faculty of Medicine, Universitas Andalas, M. Djamil Hospital, Padang, Indonesia.

Sigumonrong, Yacobda H. Department of Urology, Faculty of Medicine, Universitas Sumatera Utara, Adam Malik Hospital, Medan, Indonesia.

Mirza, Hendy. Department of Surgery, Persahabatan Hospital, Jakarta, Indonesia.
Rodjani, Arry. Department of Urology, Faculty of Medicine Universitas Indonesia, Cipto
Mangunkusumo Hospital, Jakarta, Indonesia.

PMID

<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC6765069>

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2019

104.

Expression of androgen, estrogen, and progesterone hormone receptors in the penile tissues of children with different types of hypospadias.

Celayir A; Moralioglu S; Cetiner H; Kir G; Celayir S.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid

MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Northern Clinics of Istanbul. 6(2):110-116, 2019.

[Journal Article]

UI: 31297475

OBJECTIVE: Androgen (AR), Estrogen (ER) and Progesterone (PR) hormones play an important role in the prenatal and postnatal development of urogenital tract and especially the penis. The expressions of AR, ER and PR receptors in penile tissues in children with hypospadias had also been shown previously. In this leading study, to demonstrate of the sex hormone receptor expression in cases with different types of hypospadias were aimed.

METHODS: This study was designed in children operated due to hypospadias without DSD. Biopsy samples of 3 mm's were obtained from three different sites as the lateral parametatal tissue and the anterior corner of the prepuce, and inner layer of posterior prepuce. The presence of AR, ER and PR receptors was investigated immunohistochemically.

RESULTS: Mean age was 5.4 years in 18 children with hypospadias; in totally 33 specimens were taken in 5 subcoronal as 5 specimens, and 7 penile as 15 specimens, and 6 penoscrotal as 13 specimens. According to sites of samples; 13 samples were from lateral para-meatal tissues, and 13 were from anterior corners of prepuces, and 7 were from inner layers of posterior prepuces. In regard to receptor expression; ER and AR receptors were positive in 29 (87.8%) and 12 (36.4%) respectively; PR receptors were negative.

CONCLUSION: This study emphasized the dominant expression of estrogen receptors in penile tissues of children with hypospadias. Although there was not a manifest correlation of androgen receptors absence in regard to the severity of hypospadias patients, there was a marked estrogen receptors presence in penile tissues. These findings suggest that the disrupted androgen and estrogen receptor interaction and/or balance could play a role during the development of external genitalia in hypospadias patients. Progesterone receptor was not present and therefore the active role in the postnatal development of hypospadias is still debatable.

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1

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PubMed-not-MEDLINE

Authors Full Name

Celayir, Aysenur; Moralioglu, Serdar; Cetiner, Handan; Kir, Gozde; Celayir, Sinan.

Institution

Celayir, Aysenur. Department of Pediatric Surgery, Health Science University Istanbul Zeynep Kamil Maternity and Children's Diseases Health Training and Research Center, Istanbul, Turkey.

Moralioglu, Serdar. Department of Pediatric Surgery, Health Science University Istanbul Zeynep Kamil Maternity and Children's Diseases Health Training and Research Center, Istanbul, Turkey.

Cetiner, Handan. Department of Pathology, Health Science University Istanbul Zeynep Kamil Maternity and Children's Diseases Health Training and Research Center, Istanbul, Turkey.

Kir, Gozde. Department of Pathology, Istanbul Medeniyet University Goztepe Training and Research Hospital, Istanbul, Turkey.

Celayir, Sinan. Department of Pediatric Surgery, Istanbul University Cerrahpasa Faculty of Medicine, Istanbul, Turkey.

PMID

<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC6593914>

Year of Publication

2019

105.

Anatomical Explanations of the Pathogenesis of Proximal Hypospadias.

Acimi S.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid

MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Journal of Indian Association of Pediatric Surgeons. 24(2):97-99, 2019 Apr-Jun.

[Journal Article]

UI: 31105393

Aims: The aim of this study was to try to explain the pathogenesis of proximal hypospadias based on anatomical and histological findings.

Methods: During 9 years, we performed systematic biopsies (in the lateral areas of the urethral plate, as well as under this plate) in 81 patients treated for proximal hypospadias. The histological study was performed by routine coloring, hematoxylin and eosin, and Masson's trichrome, which colors the collagen fibers in blue, and monoclonal antibody against alpha-smooth muscle actin.

Results: There is a fibrosis tissue abnormally present on the ventral side of the penis. This tissue consists of a mixture of fibrous connective tissue, nerve nets, short vessels, and smooth muscle fibers. The penis' dartos does not contain smooth muscle fibers. These fibers can come from a blood vessel or spongy tissue which existed during the neonatal period in the distal part of the penis before disappearing.

Conclusions: The proximal hypospadias is due presumably to avascular necrosis of the distal part poorly vascularized of the corpus spongiosum.

Version ID

1

Status

PubMed-not-MEDLINE

Authors Full Name

Acimi, Smail.

Institution

Acimi, Smail. Department of Visceral Surgery, Faculty of Medicine, Children's Hospital Canastel, University of Oran, Oran, Algeria.

PMID

<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC6417047>

Year of Publication

2019

Functional characterization of two new variants in the bone morphogenetic protein 7 prodomain in two pairs of monozygotic twins with hypospadias.

Bouty A., Walton K., Listyasari N.A., Robevska G., Van Den Bergen J., Santosa A., Faradz S.M.H., Harrison C., Ayers K.L., Sinclair A.H.

Embase

Journal of the Endocrine Society. 3 (4) (pp 814-824), 2019. Date of Publication: 2019.

[Article]

AN: 628081894

Context: Variants in bone morphogenetic protein 7 (BMP7) have been reported in patients with hypospadias. Here we report and analyze two variants in the BMP7 prodomain in monozygotic twins with hypospadias.

Material(s) and Method(s): Patients with hypospadias were prospectively recruited. After informed consent was obtained, DNA was extracted from blood. The coding regions of 1034 genes [including 64 known diagnostic genes and candidate genes for disorder/difference of sex development (DSD)] were sequenced using a targeted capture approach (HaloPlex, Agilent, Santa Clara, CA), combined with massively parallel sequencing. The resulting variants were filtered for rarity in the general population (<1%) and in our screen. Quality, depth of the reads, and predicted pathogenicity were also considered. The consequences of the identified mutations on BMP7 expression was determined by Western blot analysis on culture media from transfected cells, and activity measured using a SMAD 1/5-responsiveness luciferase assay.

Result(s): We analyzed DNA from 46 patients with hypospadias. Two variants in BMP7 were identified in two pairs of monozygotic concordant twins exhibiting proximal hypospadias. Both variants are heterozygous, nonsynonymous, and affect highly conserved amino acids in the prodomain of BMP7 in regions predicted to be important for BMP7 assembly/folding. Functional analyses demonstrated that both variants disrupt BMP7 synthesis or secretion.

Conclusion(s): Through our targeted DSD panel we have identified two variants in the prodomain of BMP7 in hypospadias. By decreasing BMP7 synthesis, these variants are likely to limit BMP7 bioavailability during closure of the urethral plate. Further analysis of patients with hypospadias may uncover additional variants that cause this DSD.

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Status

Embase

Institution

(Bouty, Robevska, Van Den Bergen, Ayers, Sinclair) Murdoch Children's Research Institute,
Parkville 3052, Australia (Bouty) Urology Department, Royal Children's Hospital, 50 Flemington
Road, Parkville, VIC 3052, Australia

(Walton, Harrison) Department of Physiology, Growth Factor Therapeutics Laboratory, Monash
University, Clayton 3168, Australia

(Listyasari, Faradz) Centre for Biomedical Research, Faculty of Medicine, Diponegoro University,
Semarang 50242, Indonesia

(Santosa) Urology Section, Department of Surgery, Dr Kariadi General Hospital, Semarang
50242, Indonesia

(Ayers, Sinclair) Department of Paediatrics, University of Melbourne, Parkville 3010, Australia
Publisher

Oxford University Press (E-mail: Chris.Payne@oup.com)

Year of Publication

2019

107.

Baseline characteristics of infants with atypical genital development: Phenotypes, diagnoses, and
sex of rearing.

Finlayson C., Rosoklija I., Aston C.E., Austin P., Bakula D., Baskin L., Chan Y.-M., Delozier A.M.,
Diamond D.A., Fried A., Greenfield S., Kolon T., Kropp B., Lakshmanan Y., Meyer S., Meyer T.,
Nokoff N., Mullins L.L., Palmer B., Perez M.N., Poppas D.P., Reddy P., Reyes K.J.S., Schulte M.,
Sharkey C.M., Yerkes E., Wolfe-Christensen C., Wisniewski A.B., Cheng E.Y.

Embase

Journal of the Endocrine Society. 3 (1) (pp 264-272), 2019. Date of Publication: 2019.

[Article]

AN: 628079133

Purpose: Little is known about the phenotypes, diagnoses, and sex of rearing of infants with
atypical genital development in the United States. As part of a multicenter study of these infants,
we have provided a baseline report from US difference/disorder of sex development clinics
describing the diagnoses, anatomic features, and sex of rearing. We also determined whether
consensus guidelines are followed for sex designation in the United States.

Method(s): Eligible participants had moderate-to-severe genital atypia, were aged, 3 years, and had not undergone previous genitoplasty. Karyotype, genetic diagnosis, difference/disorder of sex development etiology, family history, and sex of rearing were collected. Standardized examinations were performed.

Result(s): Of 92 subjects, the karyotypes were 46,XX for 57%, 46,XY for 34%, and sex chromosome abnormality for 9%. The median age at the baseline evaluation was 8.8 months. Most 46,XX subjects (91%) had congenital adrenal hyperplasia (CAH) and most 46,XY subjects (65%) did not have a known diagnosis. Two individuals with CAH underwent a change in sex of rearing from male to female within 2 weeks of birth. The presence of a uterus and shorter phallic length were associated with female sex of rearing. The most common karyotype and diagnosis was 46,XX with CAH, followed by 46,XY with an unknown diagnosis. Phenotypically, atypical genitalia have been most commonly characterized by abnormal labioscrotal tissue, phallic length, and urethral meatus location.

Conclusion(s): An increased phallic length was positively associated with rearing male. Among the US centers studied, sex designation followed the Consensus Statement recommendations. Further study is needed to determine whether this results in patient satisfaction.

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Status

Embase

Institution

(Finlayson, Rosoklija, Meyer, Yerkes, Cheng) Ann and Robert H. Lurie Children's Hospital of Chicago, Northwestern University, Feinberg School of Medicine, 225 East Chicago Avenue, Chicago, IL 60611, United States (Aston) University of Oklahoma, Health Sciences Center, Oklahoma City, OK 73104, United States

(Austin) Texas Children's Hospital, Baylor College of Medicine, Houston, TX 77030, United States

(Bakula, Delozier, Mullins, Perez, Sharkey) Oklahoma State University, Stillwater, OK 74078, United States

(Baskin) University of California San Francisco, San Francisco, CA 94143, United States

(Chan, Diamond) Boston Children's Hospital, Harvard Medical School, Boston, MA 02115, United States

(Fried, Greenfield, Meyer) Women and Children's Hospital of Buffalo, Buffalo, NY 14202, United States

(Kolon) Children's Hospital of Philadelphia, Philadelphia, PA 19104, United States

(Kropp, Palmer, Reyes, Wolfe-Christensen, Wisniewski) Cook Children's Hospital, Fort Worth, TX 76104, United States

(Lakshmanan) Children's Hospital of Michigan, Detroit, MI 48201, United States

(Nokoff) Children's Hospital Colorado, Denver, CO 80045, United States

(Poppas) Comprehensive Center for Congenital Adrenal Hyperplasia, New York-Presbyterian Hospital-Weill Cornell Medicine, New York, NY 10021, United States

(Reddy, Schulte) Cincinnati Children's Hospital Medical Center, Cincinnati, OH 45229, United States

Publisher

Oxford University Press (E-mail: Chris.Payne@oup.com)

Year of Publication

2019

108.

Exploring disease-specific methylated CpGs in human male genital abnormalities by using methylated-site display-amplified fragment length polymorphism (MSD-AFLP).

Aiba T., Saito T., Hayashi A., Sato S., Yunokawa H., Fukami M., Hayashi Y., Mizuno K., Sato Y., Kojima Y., Ohsako S.

Embase

The Journal of reproduction and development. 65 (6) (pp 491-497), 2019. Date of Publication: 18 Dec 2019.

[Article]

AN: 629179580

The incidence of male reproductive system disorders, especially hypospadias, has been increasing in developed countries since the latter half of the 20th century. Endocrine-disrupting chemicals from the environment are considered to be involved in hypospadias onset through epigenetic alterations. This pilot study aimed to explore disease-specific methylated CpGs in human patient samples using the methylated-site display-amplified fragment length polymorphism (MSD-AFLP) technique developed by our research group [1]. We compared clinical samples from hypospadias and phimosis patients. Foreskin and blood samples were collected from one- to two-year-old patients with hypospadias (N = 3) and phimosis (N = 3) during surgical treatment. MSD-AFLP analysis showed significantly decreased CpG-methylation levels of genes such as MYH11 and increased CpG-methylation levels of genes such as PLA2G15 in hypospadias patients. Hierarchical clustering analysis showed that genes with significantly altered CpG levels were more markedly altered in DNA from blood than from foreskin. Because of the small number of

samples, further investigation is necessary to elucidate the association between variations in CpG levels in foreskin and blood DNA and male genital abnormalities. However, our MSD-AFLP method appears to be a useful tool for exploring disease-specific methylated-CpGs in human epidemiological studies.

PMID

31462596 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=31462596>]

Institution

(Aiba, Ohsako) Laboratory of Environmental Health Science, Center for Disease Biology and Integrative Medicine, Graduate School of Medicine, University of Tokyo, Tokyo 113-8655, Japan
(Aiba, Saito, Hayashi) Department of Radiation Effects Research, National Institutes for Quantum and Radiological Science and Technology, Chiba 263-8555, Japan
(Sato, Yunokawa) MazeTokyo 193-0835, Japan

(Fukami) Department of Molecular Endocrinology, National Research Institute for Child Health and Development, Tokyo 157-8535, Japan

(Hayashi, Mizuno) Department of Pediatric Urology, Nagoya City University, Graduate School of Medical Sciences, Nagoya 467-8601, Japan

(Sato, Kojima) Department of Urology, Fukushima Medical University, School of Medicine, Fukushima 960-1295, Japan

Publisher

NLM (Medline)

Year of Publication

2019

109.

Probability of Bladder Augmentation, Diversion and Clean Intermittent Catheterization in Classic Bladder Exstrophy: A 36-Year, Multi-Institutional, Retrospective Cohort Study.

Szymanski K.M., Fuchs M., McLeod D., Rosoklija I., Strine A.C., Vanderbrink B., Whittam B., Yerkes E., Gargollo P.C.

Embase

Journal of Urology. 202 (6) (pp 1256-1262), 2019. Date of Publication: 01 Dec 2019.

[Article]

AN: 631776710

Purpose:We assessed the probability of bladder augmentation/diversion and clean intermittent catheterization in classic bladder exstrophy in a multi-institutional cohort.

Materials and Methods:We included children born from 1980 to 2016 with bladder exstrophy and treated across 5 centers (exclusion criteria less than 1 year followup after birth, isolated epispadias, bladder exstrophy variants etc). Outcomes were probability of bladder augmentation/diversion after bladder closure and proportion of patients performing clean intermittent catheterization at last followup. Survival analysis was used.

Result(s):Of 216 patients 63.4% were male (median followup 14.4 years). Overall 4 patients (1.9%) underwent primary diversion and 212 underwent primary closure (72.6% in first week of life). After primary closure 50.9% underwent augmentation, 4.7% diversion and 44.8% neither. By age 18 years 88.5% underwent a bladder neck procedure (synchronous augmentation 27.3%). On survival analysis the probability of bladder augmentation/diversion was 14.9% by age 5 years, 50.7% by 10 years and 70.1% by 18 years. Probability of bladder augmentation/diversion varied significantly between centers ($p=0.01$). Probability of bladder augmentation/diversion was 60.7% 10 years after bladder neck procedure. At last followup of the entire cohort 67.4% performed clean intermittent catheterization. Among 95 patients with intact native bladders 30.5% performed clean intermittent catheterization (channel 72.4%). Among 76 adults without a diversion 85.5% performed clean intermittent catheterization (augmented bladder 100.0% clean intermittent catheterization, native bladder 31.3%). Fifteen patients underwent diversion (continent 8, ureterosigmoidostomy 5, incontinent 2).

Conclusion(s):On long-term followup probability of bladder augmentation/diversion increased with age, with 1 in 2 patients by age 10 years and the majority in adulthood. Probability of bladder augmentation/diversion differed among institutions. Almost a third of patients, including adults, with a closed native bladder performed clean intermittent catheterization. Considering all adults only 14% did not perform clean intermittent catheterization.

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PMID

31526259 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=31526259>]

Status

Embase

Institution

(Szymanski, Whittam) Division of Pediatric Urology, Riley Hospital for Children, Indiana University Health, Indianapolis, IN, United States (Fuchs, McLeod) Section of Pediatric Urology, Nationwide Children's Hospital, Columbus, OH, United States

(Strine, Vanderbrink) Division of Pediatric Urology, Cincinnati Children's Hospital Medical Center, Cincinnati, OH, United States

(Rosoklija, Yerkes) Division of Pediatric Urology, Ann and Robert H. Lurie Children's Hospital of Chicago, Chicago, IL, United States

(Gargollo) Division of Pediatric Urology, Mayo Clinic, Mayo Medical School, Rochester, MN, United States

Publisher

Lippincott Williams and Wilkins (E-mail: kathiest.clai@apta.org)

Year of Publication

2019

110.

A Systematic Review of Core Outcomes for Hypospadias Surgery.

Leunbach T.L., O'Toole S., Springer A., Williamson P.R., Ahmed S.F.

Embase

Sexual development : genetics, molecular biology, evolution, endocrinology, embryology, and pathology of sex determination and differentiation. 13 (4) (pp 165-170), 2019. Date of Publication: 2019.

[Article]

AN: 630403406

Although the outcome of hypospadias surgery requires careful scrutiny, there is little consensus on what clinical outcome parameters should be considered essential as part of routine clinical practice. To understand the extent of variation in reported outcome measures, Pubmed was searched to identify outcomes in boys with hypospadias published in the English language from 2008 to 2017. Eligibility criteria were met in 268 publications. Outcomes were inconsistently reported and only 9 (13%) were reported in more than 25% of papers. Outcome studies describing younger boys were overrepresented. Urethrocutaneous fistula was reported in about 90% of publications that included boys aged <16 years. Among younger boys meatal stenosis, dehiscence, and urethral strictures were next most frequent. In the older age groups, an increasing frequency of cosmesis, meatal shape/location, and genital skin changes was described. Outcomes reflecting sexual health, erection, and relationship status including paternity were reported in those aged >16 years. The current study identifies the range of parameters that are measured to assess outcome after hypospadias surgery. The results can be used to promote

the development of an age-specific core outcome set that can be applied as a standardized assessment tool in future routine clinical settings.

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PMID

31865321 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=31865321>]

Publisher

NLM (Medline)

Year of Publication

2019

111.

The use of tubularized incised plate urethroplasty to repair distal hypospadias in a peripheral state hospital.

Aydin A., Sonmez M.G., Buyukserbetci M., Salar R., Ozcan S., Goger Y.E., Balasar M.

Embase

Journal of Urological Surgery. 6 (3) (pp 231-237), 2019. Date of Publication: 2019.

[Article]

AN: 2005104843

Objective: Hypospadias is a congenital anomaly of the male urethra that is becoming increasingly prevalent. In this paper, we share the results of our series of 45 distal hypospadias cases treated with tubularized incised plate urethroplasty (TIPU) at a single center in Sanliurfa, Turkiye.

Material(s) and Method(s): Hypospadias repairs were made by two urologists who had completed their urology residencies at the same clinic in a state hospital. The TIPU technique was used for all 45 patients. To prevent bladder contractions and reduce anal itching during hospitalization, the patients were administered low-dose oxybutynin and 2 mg/mL hydroxyzine hydrochloride; they were also followed up postoperatively for a mean duration of 9.3 (6-12) months.

Result(s): The average age of the patients was 9.5+/-0.5 (2-24) years. Ten patients had been circumcised previously, preoperative skin chordee was observed in 35, and none experienced preoperative complications. Postoperative catheterization lasted seven days. Postoperatively, one patient developed a fistula and two developed meatal strictures. No other complications were observed in the other patients.

Conclusion(s): Hypospadias is an anomaly frequently seen in pediatric urology practice and requires considerable attention and experience. TIPU is an ideal technique for correcting hypospadias, especially distal hypospadias, due to its low complication rate and favorable cosmetic results. Although hypospadias surgery requires experience, it can be done quite easily in peripheral hospitals by surgeons who have trained in clinics experienced in this field, provided that they follow the rules pertaining to surgery.

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Status

Embase

Institution

(Aydin, Sonmez, Goger, Balasar) Necmettin Erbakan University, Faculty of Medicine, Department of Urology, Konya, Turkey (Buyukserbetci) Bigadic State Hospital, Clinic of Urology, Balikesir, Turkey

(Salar) Mehmet Akif Inan Training and Research Hospital, Clinic of Urology, Sanliurfa, Turkey

(Ozcan) Izmir Katip Celebi University, Faculty of Medicine, Department of Urology, Izmir, Turkey

Publisher

Galenos Publishing House (E-mail: info@balkanmedicaljournal.org)

Year of Publication

2019

112.

Risk factors affecting post-pubertal high serum follicle-stimulating hormone in patients with hypospadias.

Moriya K., Nakamura M., Kon M., Nishimura Y., Kanno Y., Kitta T., Shinohara N.

Embase

World journal of urology. 37 (12) (pp 2795-2799), 2019. Date of Publication: 01 Dec 2019.

[Article]

AN: 629923193

PURPOSE: The factors affecting spermatogenesis in adulthood in patients with hypospadias (HS) are not clearly understood. In the present study, risk factors affecting post-pubertal high serum follicle-stimulating hormone (FSH) were evaluated in patients with HS. MATERIALS AND

METHODS: Among those with a history of HS surgery, patients in whom endocrinological evaluation regarding pituitary-gonadal axis was performed at 15 years of age or older between March 2004 and April 2018 were enrolled in the present study. High serum FSH was defined as greater than 10 mIU/ml. The severity of HS was divided into mild and severe. Factors affecting the post-pubertal high serum FSH were estimated.

RESULT(S): Seventy-nine patients were included in the present study. The severity of HS was mild in 35 and severe in 44. History of undescended testis (UDT) was confirmed in 12. High serum FSH was detected in nine. On logistic regression model analysis, a history of UDT was the only significant factor for high serum FSH. The incidence of high serum FSH in patients with UDT was significantly higher than that in those without UDT (58.3% vs 7.5%, $p < 0.01$). When stratified by severity of HS and the presence of UDT, high serum FSH was detected in 70% in patients with severe HS and UDT, whereas less than 10% in other groups.

CONCLUSION(S): A history of UDT was a significant factor for post-pubertal high serum FSH in patients with HS. Accordingly, the presence of UDT may be a marker for impaired spermatogenesis in patients with HS, especially in severe cases.

PMID

30820650 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=30820650>]

Author NameID

Moriya, Kimihiko; ORCID: <http://orcid.org/0000-0001-6161-9403>

Institution

(Moriya, Nakamura, Kon, Nishimura, Kanno, Kitta, Shinohara) Department of Renal and Genitourinary Surgery, Hokkaido University Graduate School of Medicine, North-15, West-7, Kita-Ku, Sapporo, 060-8638, Japan

Publisher

NLM (Medline)

Year of Publication

2019

113.

2-octyl cyanoacrylate hypospadias repair dressing: a retrospective, controlled comparison.

Brockway W.J., Bradsher A.J., Hook J.E., Patel A.S., Zamilpa I., Canon S.J.

Embase

The Canadian journal of urology. 26 (1) (pp 9675-9679), 2019. Date of Publication: 01 Feb 2019.
[Article]

AN: 626524662

INTRODUCTION: 2-octyl cyanoacrylate (OC) has been shown to be a viable option for usage following standard circumcision but data on its utilization following hypospadias repair is limited. Both OC and a standard waterproof transparent dressing (WD) are used following hypospadias repair at our children's hospital. Our hypothesis is that patients with distal hypospadias repair using OC for surgical dressing have similar outcomes as compared to patients with WD.

MATERIALS AND METHODS: A retrospective study was performed evaluating all patients with distal hypospadias repair during a 2 year period. OC was primarily used by one of the three physicians in the practice with the other two primarily used WD for surgical dressing. The primary endpoints evaluated include hematoma requiring surgical drainage, infection, meatal stenosis, urethrocutaneous fistula, dehiscence, and diverticulum. Standard follow up after hypospadias repair includes a 1 week follow up for patients requiring urethral stent removal and reevaluation for all patients 3-4 months after surgery. REDCap was used in order to compile the database used in this study.

RESULT(S): A total of 280 patients underwent distal hypospadias repair during this interval. One hundred twenty-two patients had OC used with 3 (2.4%) having complications: 2 fistulas and 1 with both meatal stenosis and fistula. One hundred fifty-eight patients were dressed with WD with 5 (3.2%) complications: 4 fistulas and 1 meatal stenosis. No patients had hematoma, wound dehiscence, diverticulum, or infection.

CONCLUSION(S): A low rate of complication was observed following distal hypospadias repair using both 2-octyl cyanoacrylate and a standard waterproof transparent dressing. 2-octyl cyanoacrylate is a safe option for surgical dressing following distal hypospadias repair but its utilization in this setting is surgeon dependent.

PMID

30797251 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=30797251>]

Institution

(Brockway) Department of Urology, University of Arkansas for Medical Sciences, Little Rock, AR, United States

Publisher

NLM (Medline)

Year of Publication

2019

Langerhans cells in hypospadias: an analysis of Langerin (CD207) and HLA-DR on epidermal sheets and full thickness skin sections.

Haid B., Reider D., Nagele F., Spinoit A.-F., Pechriggl E., Romani N., Fritsch H., Oswald J.

Embase

BMC urology. 19 (1) (pp 114), 2019. Date of Publication: 12 Nov 2019.

[Article]

AN: 629848008

BACKGROUND: Hypospadias are among the most common genital malformations. Langerhans Cells (LCs) play a pivotal role in HIV and HPV infection. The migration of LC precursors to skin coincides with the embryonic period of hypospadias development and genetic alterations leading to the formation of hypospadias impact the development of ectodermally derived tissues. We hypothesized that this might be associated with a difference in frequency or morphology of epidermal and dermal LCs in hypospadias patients.

METHOD(S): A total of 43 patients from two centers were prospectively included into this study after parental consent and ethics approval. Epidermal and dermal sheets were prepared from skin samples of 26 patients with hypospadias, 13 patients without penile malformations and 4 patients with penile malformations other than hypospadias. Immunofluorescence staining of sheets was performed with anti-HLA-DR-FITC and anti-CD207/Langerin-A594 antibodies. Skin sections from 11 patients without penile malformation and 11 patients with hypospadias were stained for Langerin. Frequencies as well as morphology and distribution of epidermal and dermal LCs on sheets and sections were microscopically evaluated. Cell counts were compared by unpaired t-tests.

RESULT(S): There was no difference in frequency of epidermal LCs, Neither on sheets (873 ± 61 vs. 940 ± 84 LCs/mm², $p=0.522$) nor on sections (32 ± 3 vs. 30 ± 2 LCs/mm², $p=0.697$). Likewise, the frequency of dermal LCs (5.9 ± 0.9 vs. 7.5 ± 1.3 LCs/mm², $p=0.329$) was comparable between patients with hypospadias and without penile malformation. No differences became apparent in subgroup analyses, comparing distal to proximal hypospadias ($p=0.949$), younger and older boys ($p=0.818$) or considering topical dihydrotestosterone treatment prior to surgery ($p=0.08$). The morphology of the LCs was not different comparing hypospadias patients with boys without penile malformations.

CONCLUSION(S): LCs are present in similar frequencies and with a comparable morphology and distribution in patients with hypospadias as compared to children without penile malformations.

This suggests that patients with hypospadias are not different from patients with normal penile development considering this particular compartment of their skin immunity.

PMID

31718599 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=31718599>]

Author NameID

Haid, Bernhard; ORCID: <http://orcid.org/0000-0003-1691-7510>

Institution

(Haid, Oswald) Department of Pediatric Urology, Hospital of the Sisters of Charity, Ordensklinikum Linz, Seilerstatte 4, Linz 4020, Austria (Haid) Department of Urology, Ludwig Maximilians University, Marchioninistrasse 15, Munich, Germany

(Reider, Romani) Department for Dermatology and Venereology, Medical University Innsbruck, Anichstrasse 35, Innsbruck 6020, Austria

(Nagele, Fritsch) Section for clinical and functional Anatomy, Medical University Innsbruck, Mullerstrasse 59, Innsbruck 6020, Austria

(Spinoit) Department of Urology, University Clinic Gent, Gent 9000, Belgium

(Pechriggl) Department of Plastic, Reconstructive and Aesthetic Surgery, Medical University Innsbruck, Innsbruck, Austria

Publisher

NLM (Medline)

Year of Publication

2019

115.

Effect of anesthesia for hypospadias repair on perioperative complications.

Splinter W.M., Kim J., Kim A.M., Harrison M.-A.

Embase

Paediatric Anaesthesia. 29 (7) (pp 760-767), 2019. Date of Publication: 01 Jul 2019.

[Article]

AN: 2004376119

Background: Recent publications from the United States, India, and Korea report that children undergoing hypospadias repair with caudal regional anesthesia/analgesia could have increased postoperative surgical complications.

Aim(s): The purpose of this retrospective cohort study was to assess the impact between caudal regional anesthesia, other regional anesthesia, and no regional anesthesia on complications after hypospadias repair at a tertiary care children's hospital in Ottawa, Canada, with an expectation to changing practices if a link was found.

Method(s): We reviewed the health records of 827 children with hypospadias undergoing penile surgery from January 1991-June 2017. The final sample size for the analysis consisted of 764 patients and 825 procedures.

Result(s): The overall complications were almost identical when considering anesthesia effects, and this similarity persisted when we assessed specifically for only surgical complications. We had 716, 94, and 15 subjects who had a caudal block, penile block, and general anesthesia only, respectively, and their complication rates were 28, 31, and 27%, respectively, and their fistula formation rates were 10, 6, and 0%, respectively, and their stricture formation rates were 8, 7, and 20%, respectively. Hypospadias type and surgical repair technique were marked predictors of complications in the postoperative period.

Conclusion(s): Anesthesia technique appears to have minor impact on complications after hypospadias repair, while surgical technique and type of hypospadias impact complications after hypospadias surgery in children. Based upon these results, we will not change our current practice of using a variety of regional anesthesia techniques for children undergoing hypospadias repair.

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PMID

31063627 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=31063627>]

Status

Embase

Author NameID

Splinter, William M.; ORCID: <http://orcid.org/0000-0002-9293-2889>

Institution

(Splinter, Kim, Kim) Department of Anesthesiology and Pain Medicine, University of Ottawa and Children's Hospital of Eastern Ontario, Ottawa, ON, Canada (Harrison) Children's Hospital of Eastern Ontario Research Institute, Ottawa, ON, Canada

Publisher

Blackwell Publishing Ltd

Year of Publication

2019

116.

Modified Hinderer's Technique for Serious Proximal Hypospadias with Ventral Curvature:
Outcomes and Our Experience.

Ciftci I., Gunduz M., Sekmenli T.

Embase

Urology journal. 16 (5) (pp 478-481), 2019. Date of Publication: 21 Oct 2019.

[Article]

AN: 629652384

PURPOSE: Hypospadias is a congenital anomaly that includes deficient ventral structure of the penis. Proximal hypospadias cases make up 20% of all hypospadias cases. The choice of operative technique for hypospadias repair depends on the severity, and it is influenced by the surgeon's experience and perception of where priorities should lie. Several other factors interact to determine the type of repair, such as meatal site, presence of chordee, availability of the prepuce, and quality of the urethral plate and in addition surgeon's experience affects the type of repair. **MATERIALS AND METHODS:** The treatment records of 42 penoscrotal and perineal hypospadias cases that were treated in our clinic from 1998 to 2017 were reviewed retrospectively. Cases with penoscrotal and perineal meatus were included in the study at the beginning of the urethroplasty. All cases had surgical intervention via Hinderer's technique. **RESULT(S):** Acceptable cosmetic results were obtained in 37 (85%) patients with an objective scoring system (HOSE) for evaluating the results of hypospadias surgery score. The mean score after surgery was 14.8. Fistula and wound breakdown occurred in 7 out of the 42 cases. **CONCLUSION(S):** In conclusion, the modified Hinderer's technique is a safe and reliable technique for both proximal and perineal hypospadias. Low complication rates and application in a single surgical session increase the comfort of both the patient and the surgeon.

PMID

30520010 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=30520010>]

Institution

(Ciftci, Gunduz, Sekmenli) Department of Pediatric Surgery, Selcuk University Medical Faculty,
Konya, Turkey

Publisher

NLM (Medline)

Year of Publication

2019

117.

Outcomes of hypospadias surgery performed by different surgeons under the supervision of an experienced pediatric urology surgeon.

Ozman O., Kuru M., Gezer M., Gevher F., Onal B.

Embase

Journal of Urological Surgery. 6 (2) (pp 144-147), 2019. Date of Publication: 2019.

[Article]

AN: 2005104819

Objective: Hypospadias is one of the most common congenital anomalies among males.

Hypospadias repair is still a challenging issue due to its results and frequently seen complications. The most commonly used techniques are the meatal advancement with glansplasty incorporated and tubularized incised plate urethroplasty. The experience of the surgeon is an important factor that influence the success of the surgery. In the literature, there are not enough data about the success of these operations done by different surgeons under the supervision of a pediatric urologist.

Material(s) and Method(s): Records of 98 patients who underwent hypospadias surgery performed under the supervision of a pediatric urologist between 2008 and 2015 were retrospectively investigated. Patients who developed fistula after operation and those who did not were divided into two groups. The two groups were compared according to hyposdias type, history, operation type and age.

Result(s): A total of 98 patient were included in the study. The mean age was 8.58+/-8.3 years. Eighty six (88%) patients had distal and 12 (12%) had proximal hypospadiasis. Twenty two (22%) patients developed fistula. There was no statistically significant difference in history and type of hypospadias and age group (child vs adult) between the two groups. The mean age of the non-fistula group was statistically significantly lower than the fistula group ($p=0.0169$).

Conclusion(s): The rates of success and complications of the hypospadias surgeries performed by different surgeons under the supervision of an experienced pediatric urologist are similar with the series done by experienced pediatric urologists.

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Status

Embase

Author NameID

Ozman, Oktay; ORCID: <http://orcid.org/0000-0003-2499-8947>

Institution

(Ozman) University of Health Sciences, Gaziosmanpasa Taksim Training and Research Hospital, Clinic of Urology, Istanbul, Turkey (Kuru) Yeditepe University Faculty Medical, Department of

Urology, Istanbul, Turkey

(Gezer) Adiyaman University Faculty Medical, Training and Research Hospital, Clinic of Urology, Adiyaman, Turkey

(Gevher) Bezmialem Vakif University, Faculty Medical, Department of Urology, Istanbul, Turkey

(Onal) Istanbul University Cerrahpasa, Faculty Medical, Department of Urology, Istanbul, Turkey

Publisher

Galenos Publishing House (E-mail: info@balkanmedicaljournal.org)

Year of Publication

2019

118.

Ultrasonographic Assessment of Bladder Volumes in Children Undergoing Penile Surgery: Does the Type of Anesthesia Matter?.

Ekstein M., Bar-Yosef Y., Ben-Chaim J., Flaishon R., Weinbroum A.A.

Embase

American Journal of Therapeutics. 26 (3) (pp E314-E320), 2019. Date of Publication: 01 May 2019.

[Article]

AN: 631074620

Background: There are scant data reporting postoperative ultrasonographically measured bladder volumes in children undergoing penile surgery. Study Question: We studied the effect of various anesthesia techniques on return of micturition after penile surgery in children, using ultrasonographically measured bladder volumes.

Data Sources: Ultrasonographically measured postvoiding residual bladder volumes indexed to age-appropriate capacity, and time elapsed between the end of surgery and spontaneous voiding after pediatric circumcision, distal hypospadias repair, or repair of urethrocutaneous fistula, were studied.

Study Design:Children between 4 months and 12 years were randomized to caudal block, intravenous (IV) fentanyl or penile block, in association with inhaled general anesthesia. Bladder volumes were measured before surgery and immediately after voiding for the first time. Time to first postsurgery void was also recorded.

Result(s):Thirty-one children completed all assessments; 12 underwent caudal block, 9 IV fentanyl anesthesia, and 7 were given penile block. The mean first postvoid bladder residual volumes were highest in the caudal and lowest in the penile block children (27.5 vs. 17.3 mL, $P = 0.003$). The time elapsing between the end of surgery and first voiding was the longest in the fentanyl group compared with caudal and penile blocks (232, 178, 150 minutes, respectively, $P = 0.02$).

Conclusion(s):None of the anesthetic techniques provoked postoperative urinary retention after minor penile surgery in children. The penile block appears superior to caudal block or to IV fentanyl-based anesthesia with regard to postoperative recovery of normal micturition.

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PMID

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Status

Embase

Institution

(Ekstein, Flaishon, Weinbroum) Departments of Anesthesiology and Critical Care Medicine and Sackler, Faculty of Medicine, Tel-Aviv University, Tel Aviv, Israel (Bar-Yosef, Ben-Chaim) Departments of Urology-Pediatric Urology Unit, Sackler Faculty of Medicine, Tel-Aviv University, Tel Aviv, Israel

Publisher

Lippincott Williams and Wilkins (E-mail: kathiest.clai@apta.org)

Year of Publication

2019

119.

Cardiac disorders and structural brain abnormalities are commonly associated with hypospadias in children with neurodevelopmental disorders.

Gazdag G.E., Wang C., McGowan R., Tobias E.S., Ahmed S.F.

Embase

Clinical Dysmorphology. 28 (3) (pp 112-117), 2019. Date of Publication: 01 Jul 2019.

[Article]

AN: 631095618

The objective of our study was to use an established cohort of boys to investigate common patterns of malformations in those with hypospadias. We performed a retrospective review of the phenotype of participants in the Deciphering Developmental Disorders Study with neurodevelopmental delay and an 'Abnormality of the genital system'. This group was divided into two subgroups: Those with hypospadias and without hypospadias. Associated phenotypes of the two subgroups were compared and analysed. Of the 166 Deciphering Developmental Disorders participants with hypospadias and neurodevelopmental delay, 47 (28%) had cardiovascular and 40 (24%) had structural brain abnormalities. The rate of cardiovascular abnormalities in those with neurodevelopmental delay and genital abnormalities other than hypospadias (N = 645) was lower at 19% (P = 0.001). In addition, structural brain malformations were higher at 24% in the hypospadias group versus 15% in the group without hypospadias (P = 0.002). The constellation of these features occurred at a higher rate in the hypospadias group versus the no hypospadias group (P = 0.038). In summary, this is the first study to indicate that cardiovascular and brain abnormalities are frequently encountered in association with hypospadias in children with neurodevelopmental disorders. Not only do these associations provide insight into the underlying aetiology but also they highlight the multisystem involvement in conditions with hypospadias.

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Status

Embase

Institution

(Gazdagh, Wang, McGowan, Tobias, Ahmed) Developmental Endocrinology Research Group, School of Medicine, University of Glasgow, United Kingdom (Wang) Academic Unit of Medical Genetics and Clinical Pathology, School of Medicine, University of Glasgow, United Kingdom (McGowan) West of Scotland Regional Genetics Service, Queen Elizabeth University Hospital, Glasgow, Glasgow, United Kingdom

(Tobias) Wellcome Sanger Institute, Wellcome Genome Campus, Hinxton, Cambridge, United Kingdom

Publisher

Lippincott Williams and Wilkins (E-mail: agents@lww.com)

Year of Publication

2019

120.

Phthalate-induced fetal leydig cell dysfunction mediates male reproductive tract anomalies.

Wang Y., Ni C., Li X., Lin Z., Zhu Q., Li L., Ge R.-S.

Embase

Frontiers in Pharmacology. 10 (no pagination), 2019. Article Number: 01309. Date of Publication: 2019.

[Review]

AN: 629884409

Male fetal Leydig cells in the testis secrete androgen and insulin-like 3, determining the sexual differentiation. The abnormal development of fetal Leydig cells could lead to the reduction of androgen and insulin-like 3, thus causing the male reproductive tract anomalies in male neonates, including cryptorchidism and hypospadias. Environmental pollutants, such as phthalic acid esters (phthalates), can perturb the development and differentiated function of Leydig cells, thereby contributing to the reproductive toxicity in the male. Here, we review the epidemiological studies in humans and experimental investigations in rodents of various phthalates. Most of phthalates disturb the expression of various genes encoded for steroidogenesis-related proteins and insulin-like 3 in fetal Leydig cells and the dose-additive effects are exerted after exposure in a mixture.

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Status

In-Process

Institution

(Wang, Ni, Li, Ge) Department of Anesthesiology, Second Affiliated Hospital, Wenzhou Medical University, Wenzhou, China (Lin, Zhu, Li, Ge) Center of Scientific Research, Second Affiliated Hospital, Wenzhou Medical University, Wenzhou, China

Publisher

Frontiers Media S.A. (E-mail: info@frontiersin.org)

Year of Publication

2019

121.

Reliability of thermocautery-assisted circumcision: retrospective analysis of circumcision performed voluntarily in countries of low socioeconomic status.

Cakiroglu B., Gozukucuk A., Arda E., Tas T.

Embase

Therapeutic Advances in Urology. 11 (no pagination), 2019. Date of Publication: 2019.

[Article]

AN: 2003340526

Objective: The objective of this study was to evaluate the reliability of thermocautery-assisted circumcision performed voluntarily in patients of poor countries.

Material(s) and Method(s): Between 2016 and 2019, 32,000 children aged 7 days to 17 years were circumcised in multiple countries. The patients' urological examinations were done before the administration of local anaesthesia. Patients revealed to have undescended testicle, inguinal hernia, hypospadias, varicocele, penile rotation anomaly, epispadias and infection were not circumcised. All procedures were performed under local anaesthesia by using thermocautery. Afterwards, mucosa and skin were sutured using absorbable suture and the circumcised penis was dressed. Patients were instructed to remove the dressing after 3 days.

Result(s): Bleeding, requiring surgical intervention and drug reactions were not observed. The most observed complication was mucosal oedema, which occurred in approximately one-quarter of patients (26%, 8320/32,000) and continued for 3-5 days after the surgery. The most serious complication was a trapped penis, which occurred in 25 patients (0.078%). In six (0.018%) cases, meatal stenosis developed. Wound infection developed in only 10 (0.03%) cases, through the formation of an aseptic environment. Penile adhesion was seen in 35 cases (0.1%) and improved with anti-inflammatory treatment without any surgical intervention.

Conclusion(s): Thermocautery-assisted circumcision can be used as an effective, safe and useful technique with few complications and rapid healing rates.

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Status

In-Process

Author NameID

Cakiroglu, Basri; ORCID: <http://orcid.org/0000-0001-5337-5226>

Institution

(Cakiroglu) Department of Urology, Hisar Intercontinental Hospital, Saray Mah, Siteyolu
Cad.No.7-9, Istanbul 34768, Turkey (Gozukucuk) Department of Pediatric Surgery, Hisar
Intercontinental Hospital, Istanbul, Turkey
(Arda) Medical Faculty, Department of Urology, Trakya University, Edirne, Turkey
(Tas) Istanbul Cerrahi Hospital, Department of Urology, Esenyurt University, Istanbul, Turkey
Publisher
SAGE Publications Inc. (E-mail: claims@sagepub.com)
Year of Publication
2019

122.

Genital tract and reproductive characteristics in daughters of women and men prenatally exposed to diethylstilbestrol (DES).

Wautier A., Tournaire M., Devouche E., Epelboin S., Pouly J.-L., Levadou A.

Embase

Therapie. (no pagination), 2019. Date of Publication: 2019.

[Article]

AN: 2004060927

Objective: Prenatal exposure of women to diethylstilbestrol (DES) has been associated with reproductive tract anomalies, menstrual irregularity, infertility and pregnancy complications. In prenatally exposed men, adverse effects included genital anomalies and possible risk of infertility. In children of prenatally exposed women, i.e the third generation, an increased incidence of genital defects was observed in sons (hypospadias), but not in daughters. In daughters of prenatally exposed men, the incidence of genital anomalies was in the normal range. Experimental studies in mice evidenced an increased incidence of reproductive tract anomalies in the female descendants of females and males prenatally exposed to DES, indicative of transgenerational transmission of DES defects. The aim of this study is to assess genital tract defects, fertility and pregnancy outcome, in daughters of women and men prenatally exposed to DES.

Method(s): In a retrospective observational analysis, 759 daughters of prenatally exposed women and men reported their genital and reproductive characteristics that were compared with those of:

1) general population in France; 2) two cohorts of daughters of exposed women reported in previous publications; 3) women prenatally exposed to DES.

Result(s): An increased incidence of uterine defects was observed, with both doubling of uterus and bicornuate and aplastic uterus which constitutes the Mayer-Rokitansky-Kuster-Hauser syndrome (MRKHS). No specific anomalies described in prenatally exposed women such as T-shape or hypoplastic uterus were reported. Infertility appeared to be in the normal range.

Pregnancy outcomes of our 121 pregnancies of women born to DES exposed mothers and two other published cohorts presented inconsistent results for ectopic pregnancy, miscarriage and preterm delivery. Early and late miscarriages were higher than expected in general population in our cohort but not in the two others.

Conclusion(s): These results must be considered as preliminary, due to the small numbers of patients, limited follow-up duration after birth due to young age of the studied population, and observational methods. An important point is that the high risk of reproductive dysfunction of women prenatally exposed to DES was not observed in their daughters. There is a signal on the high incidence of uterine defects, especially aplastic uterus, and its possible link with DES exposure through epigenetic effects is discussed in our findings. Inconsistent findings regarding pregnancy outcomes in the third generation are worthy of further examination.

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Status

Article-in-Press

Institution

(Wautier, Levadou) Association reseau DES France, 1052, rue de la Ferme du Carboue, Mont-de-Marsan 40000, France (Tournaire) AP-HP, Paris 75004, France

(Tournaire, Devouche) Descartes University, Paris 75006, France

(Devouche) Laboratoire psychopathologie et processus de sante, Research Unit 4057, Boulogne-Billancourt 92100, France

(Epelboin) Hopital Bichat-Claude-Bernard, AP-HP, Paris 75018, France

(Epelboin) Paris 7 Diderot University, Paris 75013, France

(Pouly) Hopital Estaing de Clermont Ferrand, Clermont-Auvergne University, Clermont-Ferrand 63000, France

Publisher

Elsevier Masson SAS (62 rue Camille Desmoulins, Issy les Moulineaux Cedex 92442, France)

Year of Publication

2019

123.

Free Tube Graft Urethroplasty for Repair of Hypospadias.

Obara K., Hoshii T., Hoshino S., Yamana K., Anraku T., Maruyama R., Kuroki H., Ishizaki F., Yamazaki H., Tomita Y.

Embase

Urologia Internationalis. (no pagination), 2019. Date of Publication: 2019.

[Article]

AN: 630113582

Introduction: We aimed to assess the outcome of free tube graft urethroplasty for single-stage repair of hypospadias with chordee in children.

Material(s) and Method(s): We retrospectively evaluated a series of 56 patients (16 months to 9 years old, median 24 months) who underwent free graft urethroplasty for repair of hypospadias with chordee between May 2005 and November 2017. The median follow-up was 7 years (range 1-11).

Result(s): After releasing the chordee, the hypospadiac orifice was retracted to become penile in 32 patients (57%), penoscrotal in 18 patients (32%), and scrotal in 6 patients (11%). Single-stage repair was achieved without complications in 42 patients (75%). Of the remaining 14 patients with postoperative complications requiring surgical intervention, 2 had meatal stenosis, 9 had urethrocutaneous fistula, 1 had urethral diverticulum without meatal stenosis, and 1 had meatal regression. One patient who complained the urine stream went upwards in an arc underwent cutback meatoplasty to correct the stream. In all patients, a neomeatus with a vertically oriented slit-like appearance was eventually achieved at the tip of the glans.

Conclusion(s): A free graft is an appropriate choice for repairing hypospadias with chordee. Our procedure achieved favorable functional and cosmetic outcomes with a low postoperative morbidity rate.

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PMID

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Status

Article-in-Press

Institution

(Obara, Hoshii, Hoshino, Yamana, Anraku, Maruyama, Kuroki, Ishizaki, Yamazaki, Tomita)
Division of Urology, Niigata University Graduate School of Medical and Dental Sciences,
Asahimachi 1-757, Niigata 951-8510, Japan

Publisher

S. Karger AG

Year of Publication

2019

124.

Fetal Safety of Dydrogesterone Exposure in the First Trimester of Pregnancy.

Koren G., Gilboa D., Katz R.

Embase

Clinical Drug Investigation. (no pagination), 2019. Date of Publication: 2019.

[Article]

AN: 2003506744

Background: The progestin dydrogesterone (DYD) is widely used for threatened and recurrent miscarriages, as well as for dysfunctional bleeding, infertility and other obstetric and gynecological indications. While its apparent efficacy has been compared to other progestins, its fetal safety has been only sparsely investigated.

Objective(s): To follow up fetal outcome after gestational exposure to DYD and compare it to a non-exposed comparison group.

Objective(s): To follow up fetal outcome after gestational exposure to DYD and compare it to a non-exposed comparison group.

Patients and Methods: Using the 2.5 million patient database of Maccabi Health Services, we compared rates of congenital malformations among babies exposed in utero during the first trimester of pregnancy to DYD between Jan 1999 and December 2016, to a comparison group not receiving this medication. From the DYD group we excluded all cases with concomitant exposure to in vitro fertilization (IVF) and other forms of assisted reproductive technology (ART).

Result(s): There were 8508 children exposed to DYD during the first trimester of pregnancy (4417 males, 4091 females) out of 777,422 cases in the database. After excluding from the DYD group cases with concomitant exposure to IVF and other ART, DYD exposure was associated with increased risk for hypospadias [OR 1.28 (95% confidence interval 1.06-1.55)], for overall

cardiovascular malformations [OR 1.18 (91.06-1.33)], spina bifida [OR 2.29 (1.32-3.97)] and hydrocephalus [OR 2.04 (1.28-3.25)]. In a sensitivity analysis, including also cases exposed to IVF and ART in addition to DYD, there were also increased risks for cryptorchidism [1.37 (1.19-1.58)] and congenital dislocation of the hip [OR 1.58 (1.42-1.78)].

Conclusion(s): DYD confers teratogenic effects after exposure to the recommended doses in pregnant women. The risks of hypospadias and cryptorchidism have biological plausibility by the known effects on male genitalia, as is the risk for spina bifida, by the proven decrease in folic acid levels. Some of these adverse fetal effects appear to be further augmented by concomitant use of IVF and ART.

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Status

Article-in-Press

Author NameID

Koren, Gideon; ORCID: <http://orcid.org/0000-0002-9234-0875>

Institution

(Koren) Adelson Faculty of Medicine, Ariel University, Ariel 40700, Israel (Koren) Motherisk Israel Program, Zerifin, Israel

(Koren, Gilboa, Katz) Kahn-Sagol-Maccabi Institute for Research and Innovation, Tel Aviv, Israel

Publisher

Springer International Publishing

Year of Publication

2019

125.

Is a vegetarian diet safe to follow during pregnancy? A systematic review and meta-analysis of observational studies.

Tan C., Zhao Y., Wang S.

Embase

Critical reviews in food science and nutrition. 59 (16) (pp 2586-2596), 2019. Date of Publication: 2019.

[Article]

AN: 629272532

Background: Whether a vegetarian diet is appropriate for pregnancy remains unclear.

Objective(s): This study aimed to determine the association between vegetarian diet during pregnancy and various maternal-fetal outcomes.

Method(s): PubMed-Medline, EMBASE, and Cochrane Library databases were searched for relevant articles published by August 30, 2017. Quantitative data were analyzed by a random-effects model with pooled odds ratios or weighted mean difference (WMD) and 95% confidence interval as aggregate estimations.

Result(s): A total of 19 observational studies were identified for each of meta-analysis and narrative review. The overall estimated relation between vegetarian pregnancy and low birth weight (LBW) was marginally significant (1.27 (0.98, 1.65), $P=0.07$, $I^2=0\%$). Asian (India/Nepal) vegetarian mothers exhibited increased risks to deliver a baby with LBW (1.33 (1.01, 1.76), $P=0.04$, $I^2=0\%$). However, the WMD of neonatal birth weight in five studies suggested no difference between vegetarians and omnivores. Given the high heterogeneity of the included studies, lack of high-quality evidence, and limited studies included for each category, we failed to reach conclusive results regarding the risks of hypospadias, intrauterine growth retardation, maternal anemia, and gestational diabetes mellitus.

Conclusion(s): Asian vegetarian mothers presented increased risks to deliver babies with LBW than those of omnivores. Large-scale prospective studies focusing on pre- and/or early gestational nutrition will help clarify the correlation between vegetarian diet and various pregnancy outcomes.

PMID

29621406 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=29621406>]

Institution

(Tan, Wang) Department of Nutrition and Food Hygiene, School of Health Sciences, Wuhan

University, Wuhan, China (Zhao) Department of Computer Science, Volgenau School of

Engineering, George Mason University, Fairfax, VA, United States

(Wang) Hubei Provincial Key Laboratory for Applied Toxicology, Hubei Provincial Academy for Preventive Medicine, Wuhan 430060, China

Publisher

NLM (Medline)

Year of Publication

2019

126.

The structure-activity relationship (SAR) for phthalate-mediated developmental and reproductive toxicity in males.

Li X., Mo J., Zhu Q., Ni C., Wang Y., Li H., Lin Z.-K., Ge R.-S.

Embase

Chemosphere. 223 (pp 504-513), 2019. Date of Publication: May 2019.

[Review]

AN: 2001620406

Testicular dysgenesis syndrome includes the hypospadias, cryptorchidism and abnormal fetal testis in male neonate. This is possibly caused by the environmental phthalates, which down-regulate the expression of androgen synthetic genes and *Ins13* or directly inhibits steroidogenic enzymes. There are distinct structure-activity relationships (SARs) for phthalate-mediated developmental and reproductive toxicity. Here, we review the SAR for phthalate-mediated testicular dysgenesis syndrome. Of phthalates of straight side chains, C5-C6 ones are the most potent, C4 or C7 are moderate, C3 is weakest, and C1-2 or C8-13 are ineffective. The branching and unsaturation of side chains increases the toxicity. The cycling of side chains does not increase the toxicity.

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Status

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Institution

(Li, Mo, Zhu, Ni, Wang, Li, Lin, Ge) Department of Anesthesiology, The Second Affiliated Hospital and Yuying Children's Hospital of Wenzhou Medical University, Wenzhou, Zhejiang, China

Publisher

Elsevier Ltd

Year of Publication

2019

127.

Congenital anomalies of the kidneys, collecting system, bladder, and urethra.

Janjua H.S., Lam S.K., Gupta V., Krishna S.

Embase

Pediatrics in Review. 40 (12) (pp 619-624), 2019. Date of Publication: 01 Dec 2019.

[Article]

AN: 2004518807

* Based on strong research evidence, hydronephrosis is the most common cause of abnormal prenatal ultrasonography and abdominal masses in newborns. The urinary tract dilation classification system and the Society for Fetal Urology classification system are widely used for disease severity classification and management. (20)(22) * Based on strong research evidence, vesicoureteral reflux is the most common pediatric urologic condition, affecting up to 30% of children with a febrile urinary tract infection and 0.4% to 1.8% of the general pediatric population. (24) Management remains a controversial topic and is the subject of ongoing research. * Based on consensus, ultrasonography is the imaging of choice for initial evaluation prenatally and after birth to evaluate congenital anomalies of the kidney and urinary tract and is the image of choice for the initial evaluation of various diagnoses, to classify severity of disease, and to guide future monitoring. * Based on strong research evidence, posterior urethral valves are most commonly diagnosed prenatally with ultrasonography findings of bilateral hydronephrosis, dilated posterior urethra, and/or oligohydramnios. (45) Based on some research evidence as well as consensus, the treatment of choice is transurethral valve ablation. (47) * Based on strong research evidence, hypospadias is the most common malformation of the male genitalia. Newborn circumcision is contraindicated in these patients, and surgical repair is ideally performed at 6 to 12 months of age. (60).

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Status

Embase

Institution

(Janjua) Center for Pediatric Nephrology, Cleveland Clinic Children's, Cleveland, OH, United States (Lam, Krishna) Department of Pediatric Hospital Medicine, Cleveland Clinic Children's, Cleveland, OH, United States

(Gupta) Department of Pediatrics, Cleveland Clinic Children's, Cleveland, OH, United States

Publisher

American Academy of Pediatrics (141 Northwest Point Blvd, P.O. Box 927, Elk Grove Village IL 60007-1098, United States)

Year of Publication

2019

128.

Tunica vaginalis flap repair for urethrocutaneous fistulae.

Bhat S., Nair C., Shetty S., Paul F.

Embase

Journal of Clinical and Diagnostic Research. 13 (12) (pp PC06-PC08), 2019. Date of Publication: 2019.

[Article]

AN: 2004581401

Introduction: Hypospadias is a common condition which needs to be repaired within the first few years of life. However, the surgery is usually associated with several complications of which Urethrocutaneous Fistula (UCF) is the most common. Several surgical techniques have been described to correct these UCF. Use of a vascularized flap to repair this fistula is the current standard.

Aim(s): To assess the feasibility of using tunica vaginalis flap in repair of UCFs.

Material(s) and Method(s): The present study was an observational study where 32 patients with UCF resulting from a previous failed hypospadias repair were subjected to tunica vaginalis flap repair. These patients were followed-up to a mean duration of 18 months to look for recurrence of fistula and other fresh complications.

Result(s): The study included 32 patients of whom 19 had a fistula after primary surgery, 9 had undergone 2 failed surgeries and 4 had undergone 3 previous attempts at hypospadias repair. Majority (29 patients) had a single UCF. No scrotal haematoma or testicular complications were noted. Penile cosmesis was satisfactory as determined by HOSE scoring system. Erection seemed to be unaffected but could be a spurious result as majority of the patients were of pre-pubertal age. After median follow-up of 18 months, two cases had recurrence of fistula; both occurred within 1 month of the surgery.

Conclusion(s): Tunica vaginalis interposition flap repair of urethrocutaneous fistula has good outcome.

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Status

Embase

Institution

(Bhat, Nair, Shetty, Paul) Department of Urology, Government Medical College, Kottayam, Kerala, India

Publisher

Journal of Clinical and Diagnostic Research (No 3, 1/9 Roop Nagar, GT Karnal Road, Delhi 110007, India)

Year of Publication

2019

129.

Analysis of 1478 Cases of Hypospadias Repair: The Incidence of Requiring Repeated Anesthetic Exposure as Well as Exploration of the Involvement of Trainees on Case Duration.

Adler A.C., Chandrakantan A., Sawires Y., Lee A.D., Hart M., Koh C.J., Janzen N.K., Austin P.F., Andropoulos D.B.

Embase

Anesthesia and analgesia. (no pagination), 2019. Date of Publication: 09 Dec 2019.

[Article]

AN: 630582301

BACKGROUND: Recently, there has been significant focus on the effects of anesthesia on the developing brain. Concern is heightened in children <3 years of age requiring lengthy and/or multiple anesthetics. Hypospadias correction is common in otherwise healthy children and may require both lengthy and repeated anesthetics. At academic centers, many of these cases are performed with the assistance of anesthesia and surgical trainees. We sought to identify both the incidence of these children undergoing additional anesthetics before age 3 as well as to understand the effect of trainees on duration of surgery and anesthesia and thus anesthetic exposure (AE), specifically focusing on those cases >3 hours.

METHOD(S): We analyzed all cases of hypospadias repair from December 2011 through December 2018 at Texas Children's Hospital. In all, 1326 patients undergoing isolated

hypospadias repair were analyzed for anesthesia time, surgical time, provider types involved, AE, caudal block, and additional AE related/unrelated to hypospadias.

RESULT(S): For the primary aim, a total of 1573 anesthetics were performed in children <3 years of age, including 1241 hypospadias repairs of which 1104 (89%) were completed with <3 hours of AE. For patients with <3 hours of AE, 86.1% had a single surgical intervention for hypospadias. Of patients <3 years of age, 17.3% required additional nonrelated surgeries. There was no difference in anesthesia time in cases performed solely by anesthesia attendings versus those performed with trainees/assistance (16.8 vs 16.8 minutes; $P = .98$). With regard to surgery, cases performed with surgical trainees were of longer duration than those performed solely by surgical attendings (83.5 vs 98.3 minutes; $P < .001$). Performance of surgery solely by attending surgeon resulted in a reduced total AE in minimal alveolar concentration (MAC) hours when compared to procedures done with trainees (1.92 vs 2.18; $P < .001$). Finally, comparison of patients undergoing initial correction of hypospadias with subsequent revisions revealed a longer time (117.7 vs 132.2 minutes; $P < .001$) and AE during the primary stage.

CONCLUSION(S): The majority of children with hypospadias were repaired within a single AE. In general, most children did not require repeated AE before age 3. While presence of nonattending surgeons was associated with an increase in AE, this might at least partially be due to differences in case complexity. Moreover, the increase is likely not clinically significant. While it is critical to maintain a training environment, attempts to minimize AE are crucial. This information facilitates parental consent, particularly with regard to anesthesia duration and the need for additional anesthetics in hypospadias and nonhypospadias surgeries.

PMID

31913910 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=31913910>]

Institution

(Adler, Chandrakantan, Sawires, Lee, Hart, Andropoulos) From the Department of Anesthesiology, Perioperative and Pain Medicine, Texas Children's Hospital, Baylor College of Medicine, Houston, TX (Koh, Janzen, Austin) Division of Pediatric Urology, Department of Surgery, Texas Children's Hospital, Baylor College of Medicine, Houston, TX

Publisher

NLM (Medline)

Year of Publication

2019

130.

Megameatus intact prepuce treated with urethral plate-preserving surgery: A retrospective study of an unusual hypospadias variant.

Duan S., Jiang X., Zhang X., Ou W., Fu M., Chen K., Li J., Ma S.

Embase

Translational Andrology and Urology. 8 (6) (pp 583-590), 2019. Date of Publication: 2019.

[Article]

AN: 630509785

Background: Megameatus intact prepuce (MIP) is a unique variant of hypospadias and is a clinically rare condition. Due to the anatomical characteristics of the MIP hypospadias variant presenting a unique challenge to surgeons, no single urethroplasty method provides a universal solution for all patients. The purpose of this study was to evaluate the outcomes of hypospadias after MIP repair by urethral plate-preserving urethroplasty.

Method(s): A retrospective study was performed on 25 coronal or distal MIP patients, with a median age of 8, with most deficiencies being discovered during their first hospital visit for phimosis. Correction with urethroplasty was performed for all patients; 5 underwent the Mathieu procedure, 13 underwent the tubularized incised plate (TIP) procedure, and 7 underwent the Duplay procedure. The 25 patients were followed up for 6 to 36 months to evaluate the surgical outcomes.

Result(s): There were no significant differences in intraoperative bleeding, hospital stays, postoperative analgesia rate, and cure rate among the three surgical procedures. The operative time for the Mathieu procedure was longer than that for the TIP and Duplay procedures, which did not differ. Complications occurred in 4 of the 25 patients (16.0%), and the overall complication-free survival rate at 1 year after surgery was 80.5%. The age at the time of surgery, urethral plate width, urethroplasty length, surgical procedures, or meatal location (coronal or distal penis) were not independently predictive of complications.

Conclusion(s): The clinical manifestations of MIP are often concealed and then accidentally discovered during hospital visits for phimosis; thus, the actual incidence of MIP might be higher. The urethral plate should be preserved during MIP-correcting treatment, especially for coronal or distal MIP. The same satisfactory outcomes can be obtained with Mathieu, TIP, or Duplay urethroplasty.

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Status

Embase

Institution

(Duan) Department of Pediatric Surgery, First Affiliated Hospital of Shantou University Medical College, Shantou 515041, China (Duan, Ou, Fu, Chen, Li) Department of Pediatric Surgery, Second Affiliated Hospital of Shantou University Medical College, Shantou 515041, China (Jiang, Zhang) Department of Pediatric Surgery, Woman's and Children's Hospital, Shenzhen University and, Pingshan District, Shenzhen 518122, China (Ma) Department of Radiology, First Affiliated Hospital of Shantou University Medical College, Shantou 515041, China

Publisher

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Year of Publication

2019

131.

Dorsal vertical island flap urethroplasty in children with hypospadias: A single center's experience 75 patients.

Chalapathi G., Ramaiah K.S., Veeraswamy J.

Embase

World Journal of Pediatric Surgery. 2 (1) (no pagination), 2019. Article Number: e000021. Date of Publication: February 2019.

[Article]

AN: 630447715

Objective: To assess the results and complications of dorsal vertical island flap (DVIF) urethroplasty.

Method(s): A total of 175 children were operated on for hypospadias. Out of these, 41 with proximal hypospadias with severe chordee required two-stage urethroplasty. In 18 babies with glanular hypospadias, a meatal advancement and glanuloplasty procedure was done. In 25 babies with mid-penile and distal penile hypospadias, tubularized incised urethral plate (TIP) urethroplasty was the option. 16 babies with unhealthy urethral plate and chordee were chosen for dorsal vertical tube urethroplasty after excision of the urethral plate. The rest of the 75 babies with proximal, mid-penile or distal penile hypospadias with no or minimal chordee after degloving and poor urethral plate were chosen for DVIF urethroplasty. These 75 babies with DVIF were followed up from 3 months to 5 years to assess complications such as urethrocutaneous fistula,

meatal stenosis, glans dehiscence, megalourethra or urethral diverticulum, stricture, and penile torsion/rotation.

Result(s): A total of 75 patients with proximal, mid-penile, or distal penile hypospadias in whom DVIF was used during the study period were included. The mean age was 3.7 years, ranging from 8 months to 14 years. Fourteen patients developed complications (18.6%). The most common complication was urethrocutaneous fistula, which was seen in seven (9.3%) patients. Glans dehiscence was seen in five patients (6.6%), and one had meatal stenosis with diverticulum formation. Skin necrosis was observed in one patient. In 61 patients, stream was good, with no torsion, and good cosmetic appearance was observed.

Conclusion(s): DVIF is a good alternative to TIP in midpenile and distal penile hypospadias. Our early experience with DVIF urethroplasty showed an acceptable rate of complications and good cosmetic results.

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Status

Embase

Institution

(Chalapathi, Ramaiah, Veeraswamy) Kurnool Medical College, Kurnool, India

Publisher

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Year of Publication

2019

132.

Undermining the epidermis around the U-shaped skin incision preserves thick, well-vascularized tissue that effectively prevents posturethroplasty complications.

Ochi T., Seo S., Yazaki Y., Murakami H., Takeda M., Lane G.J., Yamataka A.

Embase

Journal of Pediatric Surgery. 54 (12) (pp 2627-2630), 2019. Date of Publication: December 2019.

[Article]

AN: 2002789795

Purpose: We modified the U-shaped skin incision technique (UIT) during urethroplasty (UP) to prevent post-UP complications (post-UPC).

Patients and Methods: Our modified UIT (mUIT) involves undermining the epidermis for 5-7 mm around the U-shaped incision to create a thicker, better vascularized tissue layer for suturing the neourethra compared with UIT. To prevent injury to underlying connective tissue and avoid compromising blood perfusion, the scalpel should be held at a more slanted angle than is conventional. After the urethral plate is incised in the midline, stay sutures are placed to the middle of the penile shaft as a landmark, and three to five sutures are placed through the thick layer created by undermining. To facilitate mobilizing connective tissue during neourethroplasty, relaxing incisions may be used on the edges of the connective tissue. We compared 207 hypospadias patients who had primary or staged UP using either mUIT (n = 110) or UIT (n = 97) between 2003 and 2017 for incidence of post-UPC.

Result(s): mUIT had significantly less post-UPC than UIT after a mean follow-up of 6.44 +/- 0.26 years after final UP, 1/110 (0.9%) versus 15/97 (15.5%) (p < .0001).

Conclusion(s): Undermining the edges of the U-shaped incision during mUIT would appear to prevent post-UPC.

Level of Evidence: Level III.

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Status

Embase

Institution

(Ochi, Seo, Yazaki, Murakami, Takeda, Lane, Yamataka) Department of Pediatric General and Urogenital Surgery, Juntendo University School of Medicine, Tokyo, Japan

Publisher

W.B. Saunders

Year of Publication

2019

133.

Assessment of the effect of urethral plate width on outcome of hypospadias repair.

Chukwubuike K.E., Obianyo N.E.N., Ekenze S.O., Ezomike U.O.

Embase

Journal of Pediatric Urology. 15 (6) (pp 627.e1-627.e6), 2019. Date of Publication: December 2019.

[Article]

AN: 2003553802

Background: The paradigm for hypospadias repair is a straight penis with a vertical meatus at the tip of the glans that provides satisfactory urination and is cosmetically acceptable to the parents of the patient.

Objective(s): To determine the cosmetic and functional outcomes of hypospadias repair in relation to the width of the urethral plate.

Patients and Methods: This study was a prospective evaluation of patients operated for hypospadias. The urethral plate width (UPW) of the patients were measured preoperatively using vernier calipers. The patients were categorized into groups A and B. Group A patients have a UPW <8 mm, whereas group B patients have a UPW ≥8 mm. The width of the urethral plate was correlated to the cosmetic outcome (using hypospadias objective penile evaluation [HOPE]) and functional outcome (using the urinary stream) of hypospadias repair.

Result(s): Overall, 47 patients had their hypospadias repaired during the study period. But, only 42 patients who had their distal hypospadias repaired using tubularized incised plate urethroplasty were evaluated. There were 20 patients (47.6%) in group A and 22 patients (52.4%) in group B, with a mean urethral plate of 7.3 mm +/- 0.50 SD. The mean UPW in group A was 5.6 mm +/- 1.22 SD and the mean UPW in group B was 8.8 mm +/- 0.88 SD. Overall, mean HOPE score was 40.0 +/- 6.83 SD. Group A patients had a mean HOPE score of 38.7 +/- 7.49 SD, whereas Group B patients had a mean HOPE score of 41.2 +/- 6.08 SD. P-value was 0.725, which is not statistically significant. Relating good urinary stream (15 in group A and 20 in group B) with the width of the urethral plate statistically (using Spearman correlation technique) gave a P-value of 0.03 (P < 0.05), which is statistically significant.

Conclusion(s): Our findings indicate that the cosmetic outcome of hypospadias repair may not be determined by UPW, but the functional outcome may be predicted by the width of the urethral plate. [Table presented]

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Status

Embase

Institution

(Chukwubuike) Paediatric Surgery Unit, Department of Surgery, Enugu State University Teaching Hospital, Park Lane, Enugu, Enugu State, Nigeria (Obianyo, Ekenze, Ezomike) Sub-Department

of Paediatric Surgery, College of Medicine, University of Nigeria, Enugu Campus, Enugu State,
Nigeria

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Year of Publication

2019

134.

Association of Maternal First Trimester Serum Levels of Free-Beta Human Chorionic
Gonadotrophin and Hypospadias: A Population-Based Study.

Peycelon M., Lelong N., Carlier L., Monn F.M., De Chalus A., Bonnard A., Rachid M., Houang M.,
Paye-Jaouen A., Ali L., Lecourbe A., Grapin C., Audry G., Legendre M., Muller F., Dreux S., El
Ghoneimi A., Benachi A., Khoshnood B., Siffroi J.-P.

Embase

The Journal of urology. (pp 101097JU0000000000000708), 2019. Date of Publication: 19 Dec
2019.

[Article]

AN: 630344216

PURPOSE: The Human Chorionic Gonadotrophin (hCG) stimulates fetal testosterone production and contributes to normal development of male genitalia. Using population-based data, we hypothesized that differences in maternal free beta-hCG (hCGb) may be associated with hypospadias. MATERIALS AND METHODS: Data were obtained from the Paris Registry of Congenital Malformations (REMAPAR) (2011-2016). The initial study population included 3,172 pregnant women who gave birth to a singleton live born male infant with a congenital malformation. After exclusion of cases with unknown hCGb and those with chromosomal or genetic abnormalities, the study population included 194 boys with isolated hypospadias and 1,075 controls. For cases with operative note (N=125), we obtained data on type of (proximal/distal) hypospadias. Using quantile regression, we compared median values of multiple of median (MoM) hCGb measured for first trimester Down syndrome screening (10th-13th gestational weeks) for overall, as well as by type of hypospadias versus controls. We also took into account possible effects of placental dysfunction (maternal age, intrauterine growth retardation and preterm births) as potential factors of confusion.

RESULT(S): Overall, the median hCGb MoM was comparable for women who had an infant with hypospadias versus controls (0.99 versus 0.95, $p=0.3$). However, proximal hypospadias was associated with a statistically significant higher median MoM than distal hypospadias or unspecified (1.49 versus 0.92 versus 1.05, $p=0.02$). The estimates were comparable after adjustment for placental dysfunction.

CONCLUSION(S): Our findings support the hypothesis that an alteration in maternal hCGb levels is associated with hypospadias. However, this association appears to be limited to proximal hypospadias.

PMID

31855125 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=31855125>]

Institution

(Peycelon, Paye-Jaouen, Ali, Grapin, El Ghoneimi) Department of Pediatric Surgery and Urology, Robert-Debre University Hospital, APHP, Paris, France (Peycelon, El Ghoneimi) Universite Paris Diderot, Sorbonne Paris Cite, Paris, France

(Peycelon, Paye-Jaouen, Ali, Grapin, El Ghoneimi) Centres de Reference Maladies Rares, Maladies Endocriniennes Rares de la Croissance (CRMERC), Paris, France

(Peycelon, Carlier, De Chalus, Bonnard, Rachid, Legendre, Siffroi) Sorbonne Universite, INSERM, APHP, Hopital d'Enfants Armand Trousseau, Paris, France

(Peycelon, Monn) Department of Pediatric Urology, Riley Children Hospital at Indiana University Health, Indianapolis, IN, United States

(Peycelon, Monn) Indiana University School of Medicine, Department of Urology, Indianapolis, IN, United States

(Lelong, Lecourbe, Khoshnood) Universite de Paris, Perinatal and Pediatric Epidemiology Research Team (EPOPe), INSERM, INRA, Paris, France

(Houang) Department of Pediatric Endocrinology, Armand Trousseau Children Hospital, APHP, Paris, France

(Grapin) Universite Paris 13, Sorbonne Paris Cite, Paris, France

(Audry) Sorbonne Universite, Department of Pediatric Surgery, Armand Trousseau Children Hospital, APHP, Paris, France

(Muller, Dreux) Department of Biochemistry, Robert-Debre University Hospital, APHP, Paris, France

(Benachi) Department of Gynecology-Obstetrics, Antoine Beclere Hospital, APHP, Clamart, France

(Benachi) Universite Paris Sud, Clamart, France

Publisher

NLM (Medline)

Year of Publication

135.

Residential sources of pesticide exposure during pregnancy and the risks of hypospadias and cryptorchidism: The French ELFE birth cohort.

Cognez N., Warembourg C., Zaros C., Metten M.-A., Bouvier G., Garlantezec R., Charles M.-A., Beranger R., Chevrier C.

Embase

Occupational and Environmental Medicine. 76 (9) (pp 672-679), 2019. Date of Publication: 01 Sep 2019.

[Article]

AN: 628983138

Objectives Prenatal occupational exposure to pesticides has been associated with male reproductive tract abnormalities. Little is known about the possible impact of non-occupational pesticide exposure on fetal and child development in the general population. Using data from a nationwide birth cohort, we aimed to assess the association between residential sources of prenatal pesticide exposure and the risks of hypospadias and cryptorchidism. **Methods** Of the 9281 boys in ELFE (French Longitudinal Study of Children), the national French birth cohort, 53 were diagnosed with hypospadias and 137 with cryptorchidism. We assessed residential exposure sources from self-reported domestic use of eight types of pesticide products and French spatial land use data with acreage within a 1000 m radius around each family's home for 21 crop types. We used logistic regression modelling, adjusted for possible confounders that included estimated dietary pesticide intake. Multiple imputations were used to handle missing data. **Results** An increased risk of hypospadias was associated with domestic pesticide use against fleas and ticks (OR=2.28, 95% CI 1.09 to 4.75); no associations were found between cryptorchidism and any domestic pesticide use. Slightly increased risks of cryptorchidism were observed in association with all crop acreages near homes during pregnancy, especially for orchards, and no association was observed for hypospadias. **Conclusions** Our results suggest a possible increased risk of hypospadias associated with prenatal use of some domestic pesticide products, likely to contain insecticides, and of cryptorchidism with nearby orchard acreage (crops repeatedly sprayed with pesticides). This work is limited by its modest number of cases.

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Status

Embase

Institution

(Cognez, Warembourg, Chevrier) UnivRennes, Inserm, EHESP, Irset-UMR-S 1085, Rennes 35042, France (Warembourg) ISGlobal, Barcelona, Spain

(Zaros) Ined, Inserm, EFS, ELFE Joint Unit, Rennes, France

(Metten, Garlantezec, Beranger) UnivRennes, CHU Rennes, Inserm, EHESP, Irset-UMR-S 1085, Rennes, France

(Bouvier) University of Bordeaux, Inserm UMR 1219 EPICENE Team, Bordeaux Population Health Research Center, Bordeaux, France

(Charles) Ined, Inserm, EFS, ELFE Joint Unit, Villejuif, France

Publisher

BMJ Publishing Group (E-mail: subscriptions@bmjgroup.com)

Year of Publication

2019

136.

Effects of pre-gestational diabetes mellitus and gestational diabetes mellitus on macrosomia and birth defects in Upstate New York.

Yang G.-R., Dye T.D., Li D.

Embase

Diabetes Research and Clinical Practice. 155 (no pagination), 2019. Article Number: 107811.

Date of Publication: September 2019.

[Article]

AN: 2002647398

Aims: To evaluate the effects of pre-gestational diabetes mellitus (PGDM) and gestational diabetes mellitus (GDM) on macrosomia and birth defects.

Method(s): Existing birth registry data from the Perinatal Data System in Upstate New York was analysed. 650,914 women with a singleton term pregnancy (≥ 37 weeks) aged 18-55 years from 2004 to 2016 were included.

Result(s): The prevalence of macrosomia in infants born to women with PGDM and GDM were 26.0% and 16.4%, respectively, higher than that in the controls (11.2%). Compared with the controls (0.8%), the PGDM and GDM groups had higher prevalence of any birth defect (1.8% and 1.0%). The PGDM group had the highest prevalence of cyanotic heart disease (0.6%). Moreover, the PGDM group had higher prevalence of cleft lip and palate, cleft palate alone, hypospadias and limb reduction defect compared to the GDM and control groups ($p < 0.05$). However, these birth defects in the GDM group were similar to those in the controls. Both the PGDM and GDM groups had significantly elevated odds of macrosomia, cyanotic heart disease and any birth defect than controls. The PGDM group had higher odds of cleft lip and palate, cleft palate alone, hypospadias and limb reduction defect.

Conclusion(s): Using the Perinatal Data System database, PGDM and GDM, especially PGDM, was associated with higher prevalence of macrosomia, cyanotic heart disease and any birth defect in singleton term pregnancy in Upstate New York. PGDM, not GDM had higher prevalence of cleft lip and palate, cleft palate alone, hypospadias and limb reduction defect.

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Status

Embase

Institution

(Yang) Department of Endocrinology, Beijing Tongren Hospital, Capital Medical University, Beijing 100730, China (Yang, Dye, Li) Clinical and Translational Science Institute, School of Medicine and Dentistry, University of Rochester, Rochester, New York, NY 14620, United States

Publisher

Elsevier Ireland Ltd

Year of Publication

2019

Oral contraceptive use and genital anomalies in sons. A Danish cohort study.

Buur L.E., Laurberg V.R., Ernst A., Arendt L.H., Nybo Andersen A.-M., Olsen J., Ramlau-Hansen C.H.

Embase

Reproductive Toxicology. 89 (pp 67-73), 2019. Date of Publication: October 2019.

[Article]

AN: 2002275832

Exposure to exogenous sex hormones with estrogenic or anti-androgen properties may influence intrauterine development of male genitals. This population-based cohort study based on data from 44,408 live-born singleton sons in the Danish National Birth Cohort (DNBC) aimed to investigate whether maternal use of oral contraceptives prior to or during early pregnancy increase the risk of cryptorchidism or hypospadias. We found no consistent association between use of oral contraceptives and cryptorchidism or hypospadias, neither in those exposed any time four months prior to conception [cryptorchidism: adjusted Odds Ratio (aOR): 1.06 (95% CI: 0.91; 1.23), hypospadias: 0.74 (95% CI: 0.53; 1.03)] nor in those exposed any time during the first trimester of pregnancy [cryptorchidism: aOR: 0.93 (95% CI: 0.53; 1.62), hypospadias: 1.02 (95% CI: 0.32; 3.23)]. Despite relatively strong exposure levels from oral contraceptive use in pregnancy, this study revealed no evidence of an increased risk of either two genital malformations.

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Status

Embase

Institution

(Buur, Laurberg, Ernst, Arendt, Olsen, Ramlau-Hansen) Department of Public Health, Aarhus University, Aarhus, Denmark (Nybo Andersen) Department of Public Health, Section of Epidemiology, University of Copenhagen, Copenhagen, Denmark (Olsen) Department of Clinical Epidemiology, Aarhus University Hospital, Aarhus, Denmark

Publisher

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Year of Publication

2019

138.

Postoperative pain in small-for-gestational age infants after hernia repair, orchidopexy and urethral reconstruction surgery: A pilot study.

Schussler S.C., Kussmann F., Fahlbusch F.B., Munster T., Hirsch K., Carbon R., Albrecht S., Dotsch J., Rascher W.

Embase

Early Human Development. 136 (pp 39-44), 2019. Date of Publication: September 2019.

[Article]

AN: 2002256970

Background: Small-for-gestational-age (SGA) birth bears an enhanced risk of developing hypertension, obesity, insulin resistance and mental health disorders in later life as a consequence of adaptive processes in utero. Only a small number of studies on pain perception in SGA infants exist. These are indicative of a blunted stress response to pain in SGA newborns.

Aim(s): We initiated a pilot study investigating differences in postoperative pain perception between SGA and appropriate-for-gestational-age (AGA) infants.

Method(s): Pain and alertness levels of 10 formerly SGA and 14 AGA infants at the age 0.5-2 years were evaluated by the FLACC scale, Steward and Aldrete Scores following hernia repair, reconstructive surgery of hypospadias and orchidopexy. In addition, the postoperative consumption of non-steroidal anti-inflammatory drugs was compared between SGA and AGA.

Result(s): Postoperative pain and alertness levels were not significantly different in SGA and AGA children. We did not observe significant group differences regarding the consumption of non-steroidal anti-inflammatory drugs.

Conclusion(s): While previous studies were suggestive of a suppressed stress response to pain in SGA newborns, these findings did not fully translate into an altered response to pain beyond the newborn age. Further studies in a larger cohort seem necessary to verify this finding.

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Status

Embase

Institution

(Schussler, Kussmann, Fahlbusch, Rascher) Department of Pediatrics and Adolescent Medicine, University Hospital, Erlangen, Germany (Munster, Albrecht) Department of Anaesthesiology, University Hospital, Erlangen, Germany

(Carbon) Department of Pediatric Surgery, University Hospital, Erlangen, Germany

(Hirsch) Department of Urology, University Hospital, Erlangen, Germany

(Dotsch) Department of Pediatrics, University Hospital, Koln, Germany

Publisher

Elsevier Ireland Ltd

Year of Publication

2019

139.

Long-term consequences of androgen insensitivity syndrome.

Kosti K., Athanasiadis L., Goulis D.G.

Embase

Maturitas. 127 (pp 51-54), 2019. Date of Publication: September 2019.

[Review]

AN: 2002124218

Androgen insensitivity syndrome (AIS) is one of the most common sexual developmental disorders. According to the grade of the remaining androgen receptor (AR) function, AIS is classified as complete (CAIS), partial (PAIS) or mild (MAIS). In CAIS, the prevalence of germ cell tumours is increased compared with the general population. Although patients with CAIS used to undergo gonadectomy before puberty, nowadays a gonadectomy is recommended after spontaneous puberty, and up to 15% of patients retain their gonads. Nevertheless, the risk of germ cell tumour increases gradually after puberty. Annual follow-up with ultrasound or magnetic resonance imaging (MRI) is recommended. Unfortunately, these imaging methods are not sensitive enough for the diagnosis of an in situ germ cell tumour. In PAIS, the risk of germ cell tumour is higher than in CAIS; therefore, an early gonadectomy or an orchidopexy is indicated. Optimal hormone replacement therapy (HRT) is necessary for long-term health. The risks of osteopenia and of regimen osteoporosis are higher, ESPECIALLY in patients with early gonadectomy. Infertility is the rule in CAIS and PAIS. A few mutations do not affect fertility detrimentally, and these are responsible for MAIS. In PAIS leading to a predominantly male phenotype or ambiguous genitalia, multiple surgical procedures for gynaecomastia and/or hypospadias are required. Some small studies have found a higher risk of obesity, hyperlipidaemia and impaired insulin sensitivity. Psychological support is essential, as the prevalence of psychiatric disorders is increased. In conclusion, the diagnosis of AIS has long-

term consequences for which shared decision-making (physicians, patients, parents) is appropriate.

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Status

Embase

Institution

(Kosti, Goulis) Unit of Reproductive Endocrinology, 1st Department of Obstetrics and Gynaecology, Medical School, Aristotle University of Thessaloniki, Greece (Athanasiadis) Third Department of Psychiatry, Medical School, Aristotle University of Thessaloniki, Greece

Publisher

Elsevier Ireland Ltd

Year of Publication

2019

140.

Level of agreement on postoperative complications after one-stage hypospadias correction comparing medical records and parent reports.

Dokter E.M., Goosen E.E., van der Zanden L.F., Kortmann B.B., de Gier R.P., Roeleveld N., Feitz W.F., van Rooij I.A.

Embase

Journal of Pediatric Surgery. 54 (9) (pp 1825-1831), 2019. Date of Publication: September 2019.

[Article]

AN: 2001658241

Purpose: To analyze agreement on postoperative complications after hypospadias surgery according to medical records and parents' reports.

Material(s) and Method(s): In this retrospective cohort study, data were collected from 409 children who received an initial one-stage hypospadias correction in the Radboudumc, The Netherlands. Postoperative complications according to medical records were compared with parent-reported complications in an online questionnaire. Main complications studied were

wound-related complications, urinary tract infections, fistulas, stenosis, and prepuce-related complications. Agreement was determined by Cohen's kappa coefficient.

Result(s): Slightly less complications were mentioned in medical records (37%) compared to parents' reports (42%). Overall agreement was moderate (kappa = 0.50, 95% confidence interval (CI):0.41-0.59), but poor for some specific complications. Agreement was higher for complications that needed reoperation compared to when no reoperation was performed (kappa = 0.53, 95% CI: 0.43-0.62 and kappa = 0.18, 95% CI: 0.06-0.31) and for patients with recent surgery (< 5 years before questionnaire completion) compared to less recent surgeries (kappa = 0.69, 95% CI: 0.55-0.84 and kappa = 0.43, 95% CI: 0.33-0.54).

Conclusion(s): Agreement on complications according to medical records and parents' reports was poor to moderate, but better after reoperation and more recent surgery. Some complications mentioned in medical records were missing from parents' reports and the other way around. Better agreement will give physicians and parents a more reliable view on postoperative outcome after hypospadias surgery.

Type of Study: Diagnostic test.

Level of Evidence: Level III.

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Status

Embase

Institution

(Dokter, Goosen, van der Zanden, Roeleveld, van Rooij) Radboud Institute for Health Sciences, Department for Health Evidence, Radboud University Medical Center, Nijmegen, Netherlands
(Goosen, Kortmann, de Gier, Feitz) Department of Urology, Pediatric Urology, Radboudumc Amalia Children's Hospital, Nijmegen, Netherlands

Publisher

W.B. Saunders

Year of Publication

2019

Male subfertility and the risk of major birth defects in children born after in vitro fertilization and intracytoplasmic sperm injection: A retrospective cohort study.

Jwa S.C., Jwa J., Kuwahara A., Irahara M., Ishihara O., Saito H.

Embase

BMC Pregnancy and Childbirth. 19 (1) (no pagination), 2019. Article Number: 192. Date of Publication: 03 Jun 2019.

[Article]

AN: 627947538

Background: Children born after intracytoplasmic sperm injection (ICSI) are at increased risk of specific major birth defects compared with children born after in vitro fertilization (IVF). However, whether this risk is due to the treatment itself (i.e., IVF or ICSI) or underlying male subfertility is unknown. This study investigated the associations between male subfertility and the risk of major birth defects in children born after IVF and ICSI.

Method(s): We conducted a retrospective cohort study using data from the Japanese assisted reproductive technology registry between 2007 and 2014. Fresh embryo transfer cycles registered from 2007 to 2014 that resulted in singleton live births, still births, or selective terminations were included (n = 59,971). Major birth defects were defined by the US Centers for Disease Control and Prevention guidelines, excluding chromosomal abnormalities. Odds ratios (ORs) and 95% confidence intervals (CIs) were calculated using generalized estimating equations adjusting for potential confounders.

Result(s): Major birth defects were reported in 626/59,971 (1.04%) cases. Among IVF cycles, male subfertility was associated with significantly greater risks of hypospadias (3/3163 [0.09%] vs 4/28,671 [0.01%], adjusted OR = 6.85, 95% CI 2.05-22.9, P = 0.002) and atrial septal defects (4/3163 [0.13%] vs 9/28,671 [0.03%], adjusted OR = 3.98, 95% CI 1.12-14.1, P = 0.03) compared with fertile men. Subgroup analysis using sperm parameters showed that oligozoospermia (i.e., sperm concentrations < 15 million/mL) was significantly associated with a greater risk of ventricular septal defects compared with normal sperm concentrations in IVF pregnancies (5/868 [0.58%] vs 60/28,090 [0.21%], adjusted OR = 2.68, 95% CI 1.15-6.27, P = 0.02), and severe oligozoospermia (i.e., sperm concentrations < 5 million/mL) was significantly associated with an increased risk of hypospadias compared with normal sperm concentrations in ICSI pregnancies (5/3136 [0.16%] vs 5/16,865 [0.03%], adjusted OR = 3.88, 95% CI 1.14-13.2, P = 0.03).

Conclusion(s): The results of this exploratory study suggest that underlying male subfertility may play a role in the risk of major birth defects related to ICSI and IVF. Further research, including systematic reviews adjusting for confounders, is required to confirm the associations between male subfertility and major cardiac and urogenital birth defects.

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PMID

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Embase

Author NameID

Jwa, Seung Chik; ORCID: <http://orcid.org/0000-0002-8815-5714>

Institution

(Jwa, Jwa, Saito) Division of Reproductive Medicine, Center of Maternal-Fetal, Neonatal and Reproductive Medicine, National Center for Child Health and Development, 2-10-1, Okura, Setagaya-ku, Tokyo 157-8535, Japan (Jwa, Ishihara) Department of Obstetrics and Gynecology, Saitama Medical University, 38 Morohongo, Moroyama, Saitama 350-0495, Japan (Kuwahara, Irahara) Department of Obstetrics and Gynecology, Graduate School of Biomedical Sciences, University of Tokushima, 3-18-5, Kuramoto-chou, Tokushima-shi, Tokushima 770-8503, Japan

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BioMed Central Ltd. (E-mail: info@biomedcentral.com)

Year of Publication

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142.

Outcome of staged buccal mucosal graft for repair of long segment anterior urethral stricture.

Selim M., Salem S., Elsherif E., Badawy A., Elshazely M., Gawish M.

Embase

BMC Urology. 19 (1) (no pagination), 2019. Article Number: 38. Date of Publication: 16 May 2019.

[Article]

AN: 627732277

Background: Long anterior urethral stricture due to variable etiological factors constitutes a challenge for reconstruction. We evaluated our centers experience with cases of long anterior urethral stricture due to different etiologies that were managed by 2-stage substitution urethroplasty using buccal mucosal graft procedure.

Method(s): During the period between November 2009 and November 2016. All cases with long anterior urethral stricture that were planned for substitution urethroplasty in our department were

enrolled in this study. The first stage was excision of most fibrotic areas of the urethral plate, the remaining of the urethra is laid open and augmented with buccal mucosal graft for second stage closure after 6-9 months.

Result(s): The study included 123 patients who underwent first stage, 105 patients of them underwent second stage urethroplasty. Eighteen cases were missed after first stage. The mean (range) age was 38.4 (17-60 years). The mean (range) stricture length was 8.3 (4-13 cm). The cause of stricture was idiopathic in 47, inflammatory in 15, lichen sclerosus in 26 and post failed hypospadias repair in 35 patients. First stage was complicated by graft contracture in 11 (8.9%) patients that needed re-grafting, 5(4.1%) patient had bleeding from the buccal mucosa site that needed haemostatic sutures, oral numbness was reported in 7 (5.7%) patients. Second stage was complicated by wound dehiscence in 2(1.9%) patients, resticture in 11 (10.5%), fistula in 6 (5.7%) patients, meatal stenosis in 3 (2.9%). The overall success rate was 79.1% (83 cases out of 105) with a mean (range) follow-up of 34.7 (10-58 months).

Conclusion(s): Staged urethroplasty using buccal mucosal graft procedure is an effective surgical option for patients with long anterior urethral strictures especially for patients with lichen sclerosus and those with failed previous surgical repair.

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Author NameID

Gawish, Maher; ORCID: <http://orcid.org/0000-0002-8586-1488>

Institution

(Selim, Salem, Elsherif, Badawy, Elshazely) Department of Urology, Faculty of Medicine, Menoufia University, Governorate, Menoufia, Egypt (Gawish) Department of Urology, Faculty of Medicine, Alazhar University, Governorate, Assiut, Egypt

Publisher

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Year of Publication

2019

Health-related quality of life among children, adolescents, and adults with bladder exstrophy-epispadias complex: a systematic review of the literature and recommendations for future research.

Dellenmark-Blom M., Sjostrom S., Abrahamsson K., Holmdahl G.

Embase

Quality of Life Research. 28 (6) (pp 1389-1412), 2019. Date of Publication: 15 Jun 2019.

[Review]

AN: 626296310

Purpose: Bladder exstrophy-epispadias complex (BEEC) is a rare spectrum of genitourinary malformations. Children risk long-term urinary and genital dysfunctions. To achieve a comprehensive understanding, this study aimed to review the literature on generic and disease-specific health-related quality of life (HRQOL) in BEEC patients, and methodologies used.

Method(s): A literature search was conducted in Pubmed/CINAHL/Embase/PsycINFO/Cochrane, from inception to May 2018. A meta-analysis of HRQOL in BEEC patients compared to healthy references was performed.

Result(s): Twenty-one articles (published 1994-2018), describing HRQOL of children and adolescents (n = 5) and adults only (n = 5), or integrated age populations (n = 11), were identified (median sample size 24, loss to follow-up 43%, response rate 84%). Overall HRQOL was reduced in BEEC patients compared to healthy references in 4/4 studies. Impaired physical or general health in BEEC patients has been described in 9 articles, diminished mental health in 11, restricted social health in 10, and sexual health/functioning or body perception impairments in 13 articles. Urinary incontinence was the most common factor related to worse HRQOL (12 studies). In six studies, HRQOL was better than healthy norms. In eligible studies (n = 5), the pooled estimate of the effect of BEEC indicated worse HRQOL for children and adults (0 > effect sizes < 0.5). Thirty-six HRQOL assessments were used, none developed and validated for BEEC.

Conclusion(s): HRQOL in BEEC patients may be negatively impacted, particularly considering mental and social HRQOL. Sexual health/functioning or body perception impairments may be present in adolescents and adults. However, HRQOL is heterogeneously assessed and subsequent findings are differently reported. Additional research is warranted and can be improved.

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Status

Embase

Institution

(Dellenmark-Blom, Sjostrom, Abrahamsson, Holmdahl) Department of Pediatric Surgery, The Queen Silvia Children's Hospital, Sahlgrenska University Hospital, Gothenburg 416 85, Sweden

(Dellenmark-Blom, Sjostrom, Abrahamsson, Holmdahl) Department of Pediatrics, The Queen Silvia Children's Hospital, Institute of Clinical Sciences, Sahlgrenska Academy, Gothenburg University, Gothenburg 416 86, Sweden

Publisher

Springer International Publishing

Year of Publication

2019

144.

Psychiatric symptoms in men with hypospadias - preliminary results of a cross-sectional cohort study.

Ortqvist L., Engberg H., Strandqvist A., Nordenstrom A., Holmdahl G., Nordenskjold A., Frisen L.

Embase

Acta Paediatrica, International Journal of Paediatrics. 108 (6) (pp 1156-1162), 2019. Date of Publication: June 2019.

[Article]

AN: 625545030

Aim: Population studies have shown an increased risk of neurodevelopmental disorders in males born with the congenital condition hypospadias, where the opening of the urethra is on the underside of the penis. We investigated overall psychiatric morbidity in cases and matched controls.

Method(s): This study compared 167 men born with hypospadias from 1959 to 1994 in Stockholm or Gothenburg in Sweden using hospital registers. They were compared with controls from the Swedish population registry, who were contacted by regular mail and students who were recruited by local advertisements. The total sample had a mean age of 33.5 years (range: 19-54). They completed self-rating scales for depressive, anxiety and obsessive-compulsive symptoms and symptoms of attention deficit hyperactivity disorder. In addition, 33 cases and 47 controls underwent psychiatric morbidity interviews that covered the 17 most common psychiatric diagnoses.

Result(s): A fifth (21%) of both the cases and controls reported current or previous psychiatric symptoms. There were no significant differences in self-rated depression, anxiety or obsessive-compulsive disorder symptoms between the patients and controls or between the different phenotype groups. The distribution was not significantly affected by the severity of hypospadias.

Conclusion(s): Psychiatric morbidity was no higher in men with hypospadias than population-based controls.

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Status

Embase

Author NameID

Ortqvist, Lisa; ORCID: <http://orcid.org/0000-0002-7767-237X>

Institution

(Ortqvist, Strandqvist, Nordenstrom, Nordenskjold) Department of Women's and Children's Health, Karolinska Institutet, Stockholm, Sweden (Ortqvist, Nordenskjold) Department of Paediatric Surgery, Astrid Lindgren Children's Hospital, Karolinska University Hospital, Stockholm, Sweden

(Engberg, Frisen) Department of Clinical Neuroscience, Karolinska Institutet, Stockholm, Sweden

(Nordenstrom) Department of Pediatric Endocrinology, Astrid Lindgren Children's Hospital, Karolinska, Stockholm, Sweden

(Holmdahl) Women's and Children's Health, University Hospital Sahlgrenska Academy, Gothenburg, Sweden

(Holmdahl) Department of Pediatric Surgery, Queen Silvia's Children's Hospital, Gothenburg, Sweden

Publisher

Blackwell Publishing Ltd

Year of Publication

2019

Conditional deletion of platelet derived growth factor receptor alpha (Pdgfra) in urorectal mesenchyme causes mesenchyme apoptosis and urorectal developmental anomalies in mice. Qian C., Wu Z., Ng R.C.-L., Garcia-Barcelo M.-M., Yuan Z.-W., Wong K.K.Y., Tam P.K.H., Lui V.C.H.

Embase

Cell Death and Differentiation. 26 (8) (pp 1396-1410), 2019. Date of Publication: 01 Aug 2019.

[Article]

AN: 624409252

In mammals, urorectal development starts at early embryonic stage, defective urorectal development results in anorectal malformations, which are common congenital developmental defects of the anus and the urethra in newborns. The etiology and embryology of the defects are still largely unknown. Platelet-derived growth factor receptor alpha (Pdgfra) is a cell surface receptor tyrosine kinase, upon binding to its ligands (Pdgfa-d), mediates intracellular signaling and regulates embryonic development. The expression of Pdgfra is tightly regulated in the developing urorectal mesenchyme, and its dysregulation is associated with urorectal defects in animals with urorectal defects. Knockout of Pdgfra induces early embryo lethality which precludes investigation of Pdgfra in urorectal development. To address the temporal requirement of Pdgfra in urorectal development, we conditionally deleted Pdgfra in Pdgfra-expressing tissues using a tamoxifen inducible Cre-loxP approach in mice, examined the urorectal development in Pdgfra conditional knockout (Pdgfra-cKO) embryos. We showed that conditional deletion of Pdgfra in Pdgfra-expressing tissues at E10-E11 caused cloaca septation defect, anteriorly displaced anus, defective urogenital folds development and abnormal urethra tubularization in both male and female mice. Furthermore, we showed that Pdgfra was required for the survival of urorectal mesenchyme, deletion of Pdgfra caused apoptosis in the peri-cloacal, the peri-urethra and the urorectal septum mesenchyme of Pdgfra-cKO mutants, associated with an induction of p53, Ndr1 and activation of caspase-3 in Pdgfra-cKO embryos. In conclusion, Pdgfra is required for the development and survival of the urorectal mesenchyme in embryo, dysregulated Pdgfra signaling induced urorectal defects in mice resembling human congenital diseases of anorectal malformations and hypospadias. Perturbation of PDGFRA signaling may contribute to anorectal malformations and hypospadias in human.

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Status

Embase

Institution

(Qian, Wu, Ng, Garcia-Barcelo, Wong, Tam, Lui) Department of Surgery, LKS Faculty of Medicine, The University of Hong Kong, Hong Kong (Qian) Department of Obstetrics and Gynecology, Renji Hospital Affiliated to Shanghai Jiaotong University School of Medicine, Shanghai, China

(Ng) Department of Medicine, LKS Faculty of Medicine, The University of Hong Kong, Hong Kong (Garcia-Barcelo, Tam, Lui) Dr Li Dak-Sum Research Centre, The University of Hong Kong-Karolinska Institutet Collaboration in Regenerative Medicine, Hong Kong

(Yuan) Key Laboratory of Health Ministry for Congenital Malformation, Shengjing Hospital, China Medical University, Shengyang, China

Publisher

Nature Publishing Group (Houndmills, Basingstoke, Hampshire RG21 6XS, United Kingdom)

Year of Publication

2019

146.

Anatomy of Classic Bladder Exstrophy: MRI Findings and Surgical Correlation.

Dunn E.A., Kasprinski M., Facciola J., Benz K., Maruf M., Zaman M.H., Gearhart J., Di Carlo H., Tekes A.

Embase

Current Urology Reports. 20 (9) (no pagination), 2019. Article Number: 48. Date of Publication: 01 Sep 2019.

[Review]

AN: 2002262553

Purpose of Review: The exstrophy-epispadias complex (EEC) represents a group of congenitally acquired malformations involving the musculoskeletal, gastrointestinal, and genitourinary systems. Classic bladder exstrophy (CBE) is the most common and best studied entity within the EEC. In this review, imaging features of CBE anatomy will be presented with surgical correlation. Recent Findings: Magnetic resonance imaging (MRI) has emerged as a useful modality for pre- and postnatal assessment of the abdominal wall, pelvic floor, and gastrointestinal and genitourinary systems of children with CBE. The authors' experience supports use of preoperative MRI, in conjunction with navigational software, as a method for identifying complex CBE anatomy.

Summary: Imaging facilitates surgical approach and improves visualization of complex anatomy, potentially helping to avoid complications. Continued investigation of imaging guidance in CBE repair is needed as surgical techniques improve.

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Status

Embase

Institution

(Dunn, Tekes) Division of Pediatric Radiology, Russell H. Morgan Department of Radiology and Radiological Science, Johns Hopkins University School of Medicine, Baltimore, MD, United States (Kasprenski, Benz, Maruf, Zaman, Gearhart, Di Carlo) Robert D. Jeffs Division of Pediatric Urology, James Buchanan Brady Urological Institutions, Charlotte Bloomberg Children's Hospital, Johns Hopkins Hospital, Baltimore, MD, United States

(Facciola) Russell H. Morgan Department of Radiology and Radiological Science, Johns Hopkins University School of Medicine, Baltimore, MD, United States

Publisher

Current Medicine Group LLC 1 (E-mail: info@phl.cursci.com)

Year of Publication

2019

147.

Staged repair of redo and crippled hypospadias: analysis of outcomes and complications.

Badawy H., Soliman A., Moussa A., Youssef M., Fahmy A., Dawood W., Elmesiry M., Assem A., Elsayed S., Abulfotooh Eid A., Orabi S.

Embase

Journal of Pediatric Urology. 15 (2) (pp 151.e1-151.e10), 2019. Date of Publication: April 2019.

[Article]

AN: 2001647395

Introduction: Residual curvature, scarred or absent urethral plate, shortage of skin, and paucity of vascularized tissues and flaps are all obstacles to overcome during repair of redo and cripple

hypospadias after failed reconstruction. Limited articles address the outcome of repair of these cases using different grafts.

Objective(s): An analysis of outcomes and complications after the repair of redo and cripple hypospadias in a cohort of children operated by a single surgeon is presented, and data are retrieved from a prospectively designed database. **Study design:** Thirty-one children with a median age of 96 months (18-216, interquartile range [IQR]: 78), who underwent previous surgeries three to five times, were operated in the period from late 2011 to August 2017 in a single center by a single surgeon (first author); the first-stage repair was performed by using an inner prepuce graft in three children and oral grafts in 28 children. Penile straightening by degloving and removal of ventral scarred tissues are followed by development of glanular wings and grafting of the ventral surface.

Result(s): Eleven distal penile hypospadias and 20 posterior hypospadias were operated. First-stage repair was revised in three children; 25 children, eight distal and 17 posterior hypospadias, underwent second-stage repair with a median age of 84 months (18-216, IQR: 60). The success rate after the second-stage repair was 56% (14 children), and complications were encountered in 11 children in the form of penoscrotal fistulae in four, complete dehiscence in one, and glanular dehiscence in six children. After closure of fistulae, the overall success rate increased to 72%. Although complications were more common among children with posterior hypospadias (nine children) than children with distal hypospadias (two children), no statistical significance was reached ($p = .234$), with no effect of age on complications ($p = .233$), no effect of the position of the meatus on glanular dehiscence ($p = .624$), and no effect of age on glanular dehiscence ($p = .114$). **Conclusion(s):** Repair of redo and crippled hypospadias using staged graft repair in children could be achieved with a satisfactory overall success rate of 72%. Glanular dehiscence is the main complication; however, it is not considered by parents of children in the series, necessitating intervention. The lowest complication rate is expected among those with a position of the meatus more distal, however, not proven statistically in the series.[Figure presented]

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Status

Embase

Author NameID

Abulfotooh Eid A.; ORCID: <http://orcid.org/0000-0002-2791-8028>

Institution

(Badawy, Soliman, Moussa, Youssef, Fahmy, Dawood, Elmesiry, Assem, Abulfotooh Eid, Orabi)

Department of Urology, University of Alexandria, Egypt (Elsayed) Department of Pediatrics,

University of Alexandria, Egypt

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Year of Publication
2019

148.

The use of postoperative prophylactic antibiotics in stented distal hypospadias repair: a systematic review and meta-analysis.

Chua M.E., Kim J.K., Rivera K.C., Ming J.M., Flores F., Farhat W.A.

Embase

Journal of Pediatric Urology. 15 (2) (pp 138-148), 2019. Date of Publication: April 2019.

[Article]

AN: 2001358549

Introduction: The current literature on the use of antibiotics perioperatively for many pediatric procedures, including hypospadias, is inconsistent. There is currently no clear evidence for the use of postoperative antibiotic prophylaxis for stented distal hypospadias repair.

Objective(s): This study aims to synthesize and assess the available literature on the use versus non-use of postoperative antibiotic prophylaxis for stented distal hypospadias repair.

Methodology: Systematic literature search was performed on March 2018 for evaluation of trials that assessed the use and non-use of postoperative prophylactic antibiotics among stented distal hypospadias repair in children. Methodological quality of the studies was assessed according to the study design as recommended by the Cochrane Collaboration. The outcome assessed includes composite overall posthypospadias repair complications of infection and wound healing complications. The event rate for each treatment group was extracted to extrapolate intervention relative risk (RR) and corresponding 95% confidence interval (CI). Mantel-Haenszel method with random effect model was used in pooling of effect estimates from the included studies.

Heterogeneity was assessed with subgroup analysis performed according to the study design.

Publication bias was likewise determined. The protocol of this review was registered in PROSPERO (CRD42018087301) and reported in accordance with preferred reporting items for systematic reviews and meta-analyses (PRISMA) guidelines.

Result(s): A total of seven studies (four cohorts, three randomized controlled trials) with 986 stented distal hypospadias repairs (408 with no post-operative prophylactic antibiotics and 578

given postoperative prophylactic antibiotics) were included for the meta-analysis. Moderate to serious risk of bias was noted among the cohort studies, while the included randomized controlled trials (RCT) were of high risk of bias. Inconsistencies of effect estimates between subgroups and publication bias with small study effect were likely present. The overall pooled effect estimates comparing treatment groups showed no significant difference for outcomes of overall composite postoperative complication (RR 0.93, 95% CI 0.45, 1.93). Assessment of composite infection related complications and wound healing complications likewise did not show any significant between-group differences (RR 1.28, 95% CI 0.49, 3.35 and RR 1.01, 95% CI 0.48, 2.12; respectively) (Table). Asymptomatic bacteriuria was noted to be significantly higher among the intervention group with no postoperative prophylactic antibiotics (RR 4.01, 95% CI 1.11, 14.54).

Conclusion(s): The available evidence to date was assessed to be of high risk. The low level of evidence generated suggests that there is limited utility in the use of postoperative prophylactic antibiotics to prevent clinically significant posthypospadias repair [Table presented]

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Status

Embase

Institution

(Chua, Rivera) Institute of Urology, St. Luke's Medical Center-Quezon City, Philippines (Chua, Kim, Farhat) Division of Urology, The Hospital for Sick Children, Toronto, Canada

(Kim) Faculty of Medicine, University of Toronto, Toronto, Canada

(Ming) Department of Surgery, Section of Urology, University of New Mexico, United States

(Flores) Department of Surgery, Section of Urology, Philippines Children's Medical Center, Philippines

Publisher

Elsevier Ltd

Year of Publication

2019

Comparative study of urethroplasties to reduce urethral strictures in patients with severe hypospadias.

Huang L.-Q., Wang J., Ge Z., Ma G., Lu R.-G., Deng Y.-J., Huang S.-M., Guo Y.-F.

Embase

Journal of International Medical Research. 47 (4) (pp 1620-1627), 2019. Date of Publication: 01 Apr 2019.

[Article]

AN: 627136015

Objective: Transverse island pedicle flap (TIPF) plus transected urethral plate-preserving urethroplasty is increasingly used for treatment of severe hypospadias. We aimed to reduce the occurrence of urethral strictures in patients undergoing such procedures.

Method(s): Sixty-five patients with severe hypospadias were enrolled. Thirty-two patients underwent onlay-tube-onlay urethroplasty (Group A), and 33 patients underwent modified Duplay urethroplasty (Group B). Postoperative complications were recorded, including fistulas, urethral strictures, and diverticula.

Result(s): Three patients (9.4%) in Group A and 10 patients (30.3%) in group B had urethrocuteaneous fistulas. Three patients (9.4%) in Group A and 0 patients (0%) in Group B had urethral strictures. No patient in the two groups had symptoms of diverticulum or penile chordee. The results of uroflowmetry were better in Group B than Group A, when comparing uroflow patterns.

Conclusion(s): TIPF plus transected urethral plate-preserving urethroplasty can lower the occurrence of stricture, which is a challenging complication. The occurrence of stricture was lower in patients who underwent modified Duplay urethroplasty, and neourethral function and quality were better in these patient. Thus, this modified procedure can be used for treatment of severe hypospadias.

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Status

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Author NameID

Huang, Li-Qu; ORCID: <http://orcid.org/0000-0001-7718-6446>

Institution

(Huang, Wang, Ge, Ma, Lu, Deng, Guo) Department of Urology, Children's Hospital of Nanjing Medical University, Nanjing, China (Huang) Department of Nephrology, Children's Hospital of Nanjing Medical University, Nanjing, China

Publisher

SAGE Publications Ltd (E-mail: info@sagepub.co.uk)

Year of Publication

2019

150.

Prostatic utricles without external genital anomalies in children: Our experience, literature review, and pooling analysis.

Liu B., He D., Zhang D., Liu X., Lin T., Wei G.

Embase

BMC Urology. 19 (1) (no pagination), 2019. Article Number: 21. Date of Publication: 03 Apr 2019.

[Article]

AN: 627053074

Background: It has been recognized that the incidence of prostatic utricle in boys is increasing and is closely associated with diseases such as hypospadias. However, the clinical features of prostatic utricle with normal external genitalia have received little attention.

Method(s): Based on this, a series of 22 male children with prostatic utricles has been compiled by adding our 3 patients to 19 cases reported. All children enrolled had normal external genitalia. Clinical data of the case was reviewed.

Result(s): Urinary tract infection, purulent urethral discharge and pyuria were the most common presenting chief complaint (41%), irritative lower urinary tract symptoms were present in 17% of cases, obstructive lower urinary tract symptoms were noted in 14%. Urinary retention has been reported in 18% and epididymitis has been reported in 14%. Relatively rare clinical symptoms are abdominal pain, hematuria, and hematospermia. A case of calculus formation and a case of neoplasia within the prostatic utricle has been reported. A cystic mass found by digital rectal examination is the most common presenting sign. A utricular lesion posterior to the bladder was revealed by imaging examination. Unilateral renal agenesis was associated in 32% of reports. Non-surgical approach was chosen in 3 cases, transrectal ultrasonography guided aspiration has been reported in 1 case. Endoscopic techniques were used in 3 cases. Open excision was used in 11 cases. The laparoscopic excision was chosen in 3 cases and Robot-assisted laparoscopy was reported in 1 case. Symptoms and signs disappeared after treatment in all children, and no recurrence occurred during follow-up.

Conclusion(s): Prostate utricles without external genital anomalies are rarely reported in children, and are easily missed and misdiagnosed, often accompanied by recurrent urinary tract infections, lower urinary tract symptoms, epididymitis, dysuria and other symptoms. Imaging studies can confirm the diagnosis. Symptomatic and large utricles should be actively treated. The treatment program should be based on the age, clinical symptoms, and size and location of the utricle.

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Status

Embase

Institution

(Liu, He, Zhang, Liu, Lin, Wei) Department of Urology, Children's Hospital of Chongqing, Medical University, No. 136, Zhongshan 2 RD, Yuzhong District, Chongqing 400014, China (Liu, He, Lin, Wei) Ministry of Education Key Laboratory of Child Development and Disorders, Chongqing Key Laboratory of Pediatrics, China Intl. Science and Technology Cooperation Base of Child Development and Critical Disorders, Chongqing Key Laboratory of Children Urogenital Development and Tissue Engineering, Chongqing 400014, China

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BioMed Central Ltd. (E-mail: info@biomedcentral.com)

Year of Publication

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151.

Complete penile disassembly in epispadias repair.

Acimi S., Acimi M.A.

Embase

International Urology and Nephrology. 51 (4) (pp 579-583), 2019. Date of Publication: 01 Apr 2019.

[Article]

AN: 626496974

Purpose: To report current results of complete penile disassembly technique in epispadias repair.

Method(s): In ten years, we have preformed 31 complete penile disassembly for proximal epispadias repair. Twenty-four patients had epispadias after primary repair of bladder exstrophy and 7 isolated penopubic epispadias. The age of the patients ranged from 10 months to 6 years (median 3 years).

Result(s): The shortening of urethral plate was found in 30 patients (97% of cases), and this shortening varied between 6 and 16 mm. However, in one patient we found a lengthening of the urethral plate of 8 mm. The narrowing of urethral plate was found in all patients, and this narrowing varied between 30 and 50% of the width of the plate. Postoperative complications encountered in our patients were dominated by fistulas and dehiscence, particularly in patients who had bladder exstrophy-epispadias complex. After dehiscence and fistulas repair, the cosmetic results were satisfying in 25 patients (80.5% of cases) with conical glans and meatus in orthotopic position without any necrosis of the glans. However, the urinary continence ≥ 1 h was observed in 6 patients (19% of cases) and only 3 patients (9.7% of cases) had a urinary continence ≥ 3 h. The mean follow-up was 61 months.

Conclusion(s): The complete penile disassembly remains one of the best techniques for epispadias repair. However, we noticed a reappearance of the dorsal curvature of the penis in a large number of patients treated for isolated epispadias and the impact of this technique on urinary incontinence remains uncertain.

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Status

Embase

Author NameID

Acimi, Smail; ORCID: <http://orcid.org/0000-0002-2764-0302> Acimi, Mohammed Ali; ORCID: <http://orcid.org/0000-0003-3038-1230>

Institution

(Acimi) Department of Visceral Surgery, Children's Hospital Canastel, Faculty of Medicine, University Of Oran, Oran, Algeria (Acimi) Department of Surgery, EHU Oran, Faculty of Medicine, University Of Oran, Oran, Algeria

Publisher

Springer Netherlands (E-mail: rbk@louisiana.edu)

Year of Publication

2019

152.

Surgical management of hypospadias in cases with concomitant disorders of sex development.
Ochi T., Ishiyama A., Yazaki Y., Murakami H., Takeda M., Seo S., Sueyoshi R., Lane G.J.,
Haruna H., Shimizu T., Yamataka A.

Embase

Pediatric Surgery International. 35 (5) (pp 611-617), 2019. Date of Publication: 01 May 2019.

[Article]

AN: 626412327

Introduction: To review the surgical treatment of hypospadias (HP) associated with disorders of sex development (DSD).

Patients and Methods: HP cases were assessed for DSD by gross examination for atypical external genitalia, and assessment of hormone levels and karyotype. There were 58 HP cases with concomitant DSD treated between 1999 and 2017. DSD classification, type of HP, sex assignment, hormonal abnormality, surgical strategy, and post-urethroplasty complications (post-UPC) were reviewed.

Result(s): DSD were sex chromosome abnormalities (n = 4), 46,XY (n = 51), 46,XX (n = 1), and 47,XY + 21 (n = 2). HP was perineal: (n = 26), scrotal: (n = 16), penoscrotal: (n = 15), and midshaft: (n = 1); repair was primary (n = 6) or staged (n = 52). Mean age at final urethroplasty (UP) was 4.12 +/- 0.21 years; all cases had soft tissue interposition at UP. At mean follow-up 5.16 +/- 0.56 years after final UP, observed post-UPC (n = 8; 13.8%) were urethral stenosis (n = 3), urethral diverticulum (n = 2), urethrocutaneous fistula (n = 2), and curvature (n = 1). Mean onset of post-UPC was 1.24 +/- 0.77 years (range 0.1-6.3). The second half of our cases (n = 29; treated 2015 ~) had significantly less post-UPC (0/29; 0%) than the first half (8/29; 27.6%) (p = 0.0075).

Conclusion(s): Although UP for HP + DSD was formidably challenging, we achieved a significant decrease in post-UPC through a combination of surgical techniques and experience.

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Embase

Institution

(Ochi, Ishiyama, Yazaki, Murakami, Takeda, Seo, Sueyoshi, Lane, Yamataka) Department of Pediatric General and Urogenital Surgery, Juntendo University School of Medicine, 2-1-1 Hongo,

Bunkyo-ku, Tokyo 113-8421, Japan (Haruna, Shimizu) Department of Pediatrics and Adolescent Medicine, Juntendo University School of Medicine, Tokyo, Japan

Publisher

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Year of Publication

2019

153.

Application of quality indicators to data from the National Network of Congenital Anomalies of Argentina.

Groisman B., Barbero P., Mastroiacovo P., Botto L.D., Bidondo M.P., Liascovich R.

Embase

Birth Defects Research. 111 (6) (pp 333-340), 2019. Date of Publication: 01 Apr 2019.

[Article]

AN: 626373176

Background: In Argentina, birth defects are a leading cause of infant deaths. In 2009, the National Network for Congenital Anomalies of Argentina (RENAC) was established as a hospital-based surveillance system of major structural birth defects. To assess and enhance the system's data quality, we previously developed data quality indicators (DQI). Our aim was to evaluate quality indicators in RENAC.

Method(s): We applied the DQI presented in a related publication to the 2016 RENAC data.

Result(s): Among the DQI of description, spina bifida coverage and talipes had the lowest results. Regarding prevalence of hypospadias, it was lower than the defined threshold. RENAC did not achieve the ascertainment threshold of a prevalence of 21.5 per 10,000 for critical congenital heart defects. There was a high ratio of spina bifida to anencephaly.

Conclusion(s): DQI provide a focus for improving quality, making decisions, and advocating for interventions. Examples include advocating for newborn screening of critical congenital heart defects with pulse oximetry; developing training programs for clinicians to improve the detection of isolated hypospadias; and developing visual tools and checklists to improve the completeness and accuracy of case description for spina bifida, talipes, and other major malformations. After the interventions, it is important to track the impact by measuring again the DQI.

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Status

Embase

Author NameID

Groisman, Boris; ORCID: <http://orcid.org/0000-0001-6263-2562> Barbero, Pablo; ORCID: <http://orcid.org/0000-0002-6026-744X>

Bidondo, Maria Paz; ORCID: <http://orcid.org/0000-0002-4792-4775>

Liascovich, Rosa; ORCID: <http://orcid.org/0000-0002-7953-3197>

Mastroiacovo, Pierpaolo; ORCID: <http://orcid.org/0000-0002-8318-8664>

Botto, Lorenzo D.; ORCID: <http://orcid.org/0000-0002-5322-7116>

Institution

(Groisman, Barbero, Bidondo, Liascovich) National Network of Congenital Anomalies of Argentina (RENAC), National Center of Medical Genetics, National Administration of Health Laboratories and Institutes, National Ministry of Health, Buenos Aires, Argentina (Groisman, Liascovich) National Scientific and Technical Research Council (CONICET), Argentina (Mastroiacovo, Botto) International Centre on Birth Defects (ICBD), Rome, Italy (Botto) Division of Medical Genetics, University of Utah, Salt Lake City, United States

Publisher

John Wiley and Sons Inc. (P.O.Box 18667, Newark NJ 07191-8667, United States)

Year of Publication

2019

154.

Autologous platelet-rich plasma covering urethroplasty versus dartos flap in distal hypospadias repair: A prospective randomized study.

Mahmoud A.Y., Gouda S., Gamaan I., Baky Fahmy M.A.

Embase

International Journal of Urology. 26 (4) (pp 475-480), 2019. Date of Publication: April 2019.

[Article]

AN: 626254696

Objective: To compare the outcome and complication rate of the platelet-rich plasma applied as a coverage layer and dartos flap layer during primary repair of distal hypospadias.

Method(s): A prospective randomized study was carried out comprising 180 boys (age range 12-65 months) from October 2011 to December 2016 at Al-Azhar University Hospitals, Cairo, Egypt. A single surgeon carried out all urethroplasty. Patients were randomly divided into two groups: group A (tubularized incised plate urethroplasty with platelet-rich plasma coverage layer) and group B (ventral dartos flap). Complication rates were compared between two groups.

Result(s): There was a significant difference in the occurrence of complications between the two groups. A total of 36 (20%) complications were recorded in 26 patients, just 12 (13.3%) reported in group A, but 24 (26.7%) complications were reported in group B. Urethrocutaneous fistula was observed in nine patients (10%) in group A, and 12 (13.3%) in group B. Partial glans dehiscence occurred in one patient in group A, and four patients in group B. No patient in group A had a superficial wound infection, compared with six patients in group B. One case of meatal stenosis and urethral stricture was recorded in each group, all of which were managed conservatively. The resultant urinary stream was single and good in 154 patients of both groups.

Conclusion(s): Platelet-rich plasma sheet might be considered as an alternative coverage layer for distal hypospadias repair, especially in the absence of a healthy layer.

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Embase

Institution

(Mahmoud, Gouda, Gamaan, Baky Fahmy) Pediatric Surgery Department, Al-Azhar University Hospitals, Cairo, Egypt

Publisher

Blackwell Publishing

Year of Publication

2019

Maternal residential exposure to specific agricultural pesticide active ingredients and birth defects in a 2003-2005 North Carolina birth cohort.

Rappazzo K.M., Warren J.L., Davalos A.D., Meyer R.E., Sanders A.P., Brownstein N.C., Luben T.J.

Embase

Birth Defects Research. 111 (6) (pp 312-323), 2019. Date of Publication: 01 Apr 2019.

[Article]

AN: 625728589

Background: Previously we observed elevated odds ratios (ORs) for total pesticide exposure and 10 birth defects: three congenital heart defects and structural defects affecting the gastrointestinal, genitourinary and musculoskeletal systems. This analysis examines association of those defects with exposure to seven commonly applied pesticide active ingredients.

Method(s): Cases were live-born singleton infants from the North Carolina Birth Defects Monitoring Program linked to birth records for 2003-2005; noncases served as controls (total n = 304,906). Pesticide active ingredient exposure was assigned using a previously constructed metric based on crops within 500 m of residence, dates of pregnancy, and likely chemical application dates for each pesticide-crop combination. ORs (95% CI) were estimated with logistic regression for categories of exposure compared to unexposed. Models were adjusted for maternal race/ethnicity, age at delivery, education, marital status, and smoking status.

Result(s): Associations varied by birth defect and pesticide combinations. For example, hypospadias was positively associated with exposures to 2,4-D (OR50th to <90th percentile: 1.39 [1.18, 1.64]), mepiquat (OR50th to <90th percentile: 1.10 [0.90, 1.34]), paraquat (OR50th to <90th: 1.14 [0.93, 1.39]), and pendimethalin (OR50th to <90th: 1.21 [1.01, 1.44]), but not S-metolachlor (OR50th to <90th: 1.00 [0.81, 1.22]). Whereas atrial septal defects were positively associated with higher levels of exposure to glyphosate, cyhalothrin, S-metolachlor, mepiquat, and pendimethalin (ORs ranged from 1.22 to 1.35 for 50th to <90th exposures, and 1.72 to 2.09 for >90th exposures); associations with paraquat were null or inconsistent (OR 50th to <90th: 1.05 (0.87, 1.27).

Conclusion(s): Our results suggest differing patterns of association for birth defects with residential exposure to seven pesticide active ingredients in North Carolina.

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Status

Embase

Author NameID

Rappazzo, Kristen M.; ORCID: <http://orcid.org/0000-0002-9939-0883>

Institution

(Rappazzo, Luben) Office of Research and Development, U.S. Environmental Research Triangle Park, Research Triangle Park, NC, United States (Rappazzo) Oak Ridge Institute for Science and Education at the U.S. Environmental Protection Agency, National Center for Environmental Assessment, U.S. Environmental Protection Agency, Research Triangle Park, NC, United States (Warren) Department of Biostatistics, Yale School of Public Health, New Haven, CT, United States

(Davalos) Department of Biostatistics, Gillings School of Global Public Health University of North Carolina at Chapel Hill, Chapel Hill, NC, United States

(Meyer) Division of Public Health, North Carolina Department of Health and Human Services, Raleigh, NC, United States

(Meyer) Department of Maternal and Child Health, Gillings School of Global Public Health University of North Carolina at Chapel Hill, Chapel Hill, NC, United States

(Sanders) Department of Pediatrics, Icahn School of Medicine at Mount Sinai, New York, NY, United States

(Sanders) Department of Environmental Medicine and Public Health, Icahn School of Medicine at Mount Sinai, New York, NY, United States

(Brownstein) Department of Behavioral Sciences and Social Medicine, College of Medicine, Florida State University, Tallahassee, FL, United States

(Brownstein) Department of Statistics, Florida State University, Tallahassee, FL, United States
Publisher

John Wiley and Sons Inc. (P.O.Box 18667, Newark NJ 07191-8667, United States)

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156.

Risk Factors for hypospadias in Northwest Russia: A Murmansk county birth registry study.

Kovalenko A.A., Brenn T., Odland J.O., Nieboer E., Krettek A., Anda E.E.

Embase

PLoS ONE. 14 (4) (no pagination), 2019. Article Number: e0214213. Date of Publication: April 2019.

[Article]

AN: 2001789651

Background Hypospadias is the most common congenital anomaly of the penis, but its causes are mainly unknown. Of the risk factors identified, the most plausible are hormonal and genetic. The aim of this study was to identify risk factors for hypospadias in Northwest Russia based on registry data. **Methods** The study population included male infants registered in the Murmansk County Birth Registry between 1 January 2006 and 31 December 2011 (n = 25 475). These infants were followed-up for 2 years using the Murmansk Regional Congenital Defects Registry to identify cases of hypospadias not diagnosed at birth. We used logistic regression analysis to examine the contributions of hypospadias risk factors. **Results** Out of 25 475 male infants born during the study period, 148 had isolated hypospadias. The overall prevalence rate was 54.2 (95% CI 53.6-54.8) per 10 000 male infants. Those born to mothers with preeclampsia (OR = 1.65; 95% CI 1.03-2.66) or infant birthweight < 2500 g (OR = 2.06; 95% CI 1.18-3.60) exhibited increased risk for hypospadias. Maternal age, smoking during pregnancy, folic acid intake during pregnancy or hepatitis B surface antigen positivity did not associate with increased risk of hypospadias. **Conclusions** Combining data from a birth registry with those from a congenital defects registry provided optimal information about the prevalence of hypospadias and its association with low infant birthweight and preeclampsia. These factors have in common changes in hormone levels during pregnancy, which in turn may have contributed to hypospadias development.

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Status

Embase

Institution

(Kovalenko, Brenn, Odland, Krettek, Anda) Department of Community Medicine, UiT -The Arctic University of Norway, Tromsø, Norway (Kovalenko) International School of Public Health, Northern State Medical University, Arkhangelsk, Russian Federation

(Nieboer) Department of Biochemistry and Biomedical Sciences, McMaster University, Hamilton, ON, Canada

(Krettek) Department of Biomedicine and Public Health, School of Health and Education, University of Skövde, Skövde, Sweden

(Krettek) Department of Internal Medicine and Clinical Nutrition, Institute of Medicine, Sahlgrenska Academy at University of Gothenburg, Gothenburg, Sweden

Publisher

157.

No difference in cognitive performance or gender role behavior between men with and without hypospadias.

Strandqvist A., Ortqvist L., Frisen L., Nordenskjold A., Herlitz A., Nordenstrom A.

Embase

Hormones and Behavior. 109 (pp 64-70), 2019. Date of Publication: March 2019.

[Article]

AN: 2001595826

Background: Hypospadias is a common malformation of the male external genitalia that results in urethral displacement with different levels of severity. Male genital development during the fetal period is dependent on androgen function, while the etiology of hypospadias differs and can be multifactorial. The psychosocial outcome is sometimes affected, but according to several studies acceptable. The question of whether hypospadias is associated with differences in psychosexual development has been investigated previously, with mixed results. There are no previous investigations of cognitive abilities in men with hypospadias.

Objective(s): The aim of this study was to investigate whether hypospadias is associated with differences in performance on cognitive tests and/or gender role behavior.

Participant(s): Eighty-six men with hypospadias were compared to male and female controls from the general population. Procedure: Cognitive tasks, previously shown to yield group level sex differences and questions regarding self-reported childhood gender role behavior, were administered either at an outpatient clinic visit or via online participation.

Result(s): The cognitive performance of men and women in the control groups differed significantly in the expected directions. Men and women also differed on self-reported childhood gender role behavior questions. There were no significant differences between men with and without hypospadias on any of the measures. Men with proximal hypospadias performed slightly lower on many of the cognitive tasks in comparison to men with distal hypospadias and controls.

Conclusion(s): In general, hypospadias is not associated with differences in performance on cognitive tests that typically yield sex differences or with altered gender role behavior in

childhood. Further studies on cognitive abilities in boys and men with proximal hypospadias are warranted.

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Status

Embase

Author NameID

Strandqvist, Anna; ORCID: <http://orcid.org/0000-0003-1138-3927>

Institution

(Strandqvist, Ortqvist, Nordenskjold, Nordenstrom) Department of Women's and Children's Health, Karolinska Institutet, Stockholm, Sweden (Strandqvist, Herlitz) Division of Psychology, Department of Clinical Neuroscience, Karolinska Institutet, Stockholm, Sweden

(Frisen) Department of Clinical Neuroscience, Karolinska Institutet, Stockholm, Sweden

(Frisen) Child and Adolescent Psychiatry Research Center, Stockholm, Sweden

(Nordenskjold) Center for Molecular Medicine, Karolinska Institutet, Stockholm, Sweden

(Ortqvist, Nordenskjold) Department of Pediatric Surgery, Astrid Lindgren Children's Hospital, Karolinska University Hospital, Stockholm, Sweden

(Nordenstrom) Department of Pediatric Endocrinology, Astrid Lindgren Children's Hospital, Karolinska University Hospital, Stockholm, Sweden

Publisher

Academic Press Inc. (E-mail: apjcs@harcourt.com)

Year of Publication

2019

158.

Disparities in Pediatric Operative Experience during Urology Residency Training.

Silvestre J., Hernandez J.M., Lee D.I.

Embase

Urology. 127 (pp 24-29), 2019. Date of Publication: May 2019.

[Article]

AN: 2001591954

Objective: To understand trends in pediatric cases performed during urology residency including intraresident variability and cases performed relative to required minimums.

Material(s) and Method(s): Case logs of urology residents graduating from 2010 to 2018 were analyzed. Temporal trends in reported pediatric case volume were assessed via ANOVA tests and calculation of compound annual growth rates (CAGRs). Percent differences between the 90th and 10th percentiles of residents were calculated to assess intraresident variability.

Reported case volumes were compared with minimum requirements with t tests.

Result(s): 1072 residents from 306 urology residency programs were represented in this study. Minor pediatric cases increased from 2010 to 2018 (105.4 +/- 54 vs 124.6 +/- 65, $P = .004$, CAGR = 2.1%) while major pediatric cases decreased (83.9 +/- 40 vs 60.8 +/- 30, $P < .001$, CAGR = -3.9%). Orchiopexy (range, 23%-27%), hypospadias (range, 19%-21%), and hydrocele / hernia (range, 15%-19%) were the highest volume case categories. Mean intraresident variability in reported case volumes was 338% for minor pediatric (CAGR = 0%) and 382% for major pediatric (CAGR = 1.8%). Mean reported case volumes exceeded the minimum requirement for each case category by several fold ($P < .001$, range, percent difference 232-675%). All urology residents reported minimum pediatric case requirements in 2018.

Conclusion(s): Urology residents report more cases than minimum requirements for pediatric urology by several folds. Future research is needed to understand the implications of increasing intraresident case volume variabilities on residency training in pediatric urology.

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Status

Embase

Author NameID

Silvestre, Jason; ORCID: <http://orcid.org/0000-0002-5831-0760>

Institution

(Silvestre, Hernandez, Lee) The Perelman School of Medicine at the University of Pennsylvania, Philadelphia, PA, United States

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Year of Publication

2019

Is Degloving the Best Method to Approach the Penile Corporoplasty With Yachia's Technique?.

Dell'Atti L., Polito M., Galosi A.B.

Embase

Urology. 126 (pp 204-208), 2019. Date of Publication: April 2019.

[Article]

AN: 2001541165

Objective: To assess functional outcomes after surgical correction of congenital and acquired penile curvatures using Yachia's technique (YT) with degloving (DG) and without degloving (WDG) of the penis.

Material(s) and Method(s): A penile deformity with angle $\geq 30^\circ$ difficulty in vaginal penetration, and severity of erectile dysfunction secondary to penile curvatures were the indications for a surgical treatment with YT. The preoperative characteristics of the patients, postoperative outcomes (change in angulation, palpation of sutures, penile shortening, and patient satisfaction), operative time, and hospital stay were recorded. A total of 64 patients were included in this review and divided into 2 groups: 34 in group I (YT with DG) and 30 in group II (YT-WDG).

Result(s): The mean operative time was 65.87 $\pm$ 21.32 minutes for group I and 48.17 $\pm$ 23.82 minutes for group II ($P < .02$). The mean hospital stay was 3.09 $\pm$ 0.96 and 2.87 $\pm$ 0.93 days in DG and WDG, respectively ($P = .324$). There were no significant differences in recurrence rates and complications (palpation of sutures: group I: 14.7% vs. group II: 13.3%; penile shortening: group I: 8.9% vs. group II: 10%). At follow-up of 20.8 months, all treated patients were able to insert the penis in the partner's vagina, were satisfied overall with sexual intercourse.

Conclusion(s): The outcomes of the DG and WDG techniques were similar, even if the YT-WDG presents better results in terms of less healing and operative time.

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Status

Embase

Author NameID

Dell'Atti, Lucio; ORCID: <http://orcid.org/0000-0002-8471-9965>

Institution

(Dell'Atti) Department of Urology, University Hospital "Ospedali Riuniti", Ancona, Italy (Polito)

Andrology Unit, University Hospital "Ospedali Riuniti", Ancona, Italy

(Galosi) Department of Urology, University Hospital "Ospedali Riuniti", Marche Polytechnic University, Ancona, Italy

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Year of Publication

2019

160.

The biomechanical properties of the urethra in boys with hypospadias: a preliminary study.

Faurschou I., Ernst A., Zhao J., Liao D., Olsen L.H.

Embase

Journal of Pediatric Urology. 15 (1) (pp 62.e1-62.e7), 2019. Date of Publication: February 2019.

[Article]

AN: 2001319065

Purpose: The ventral aspect of the penis in boys with hypospadias is composed of dysplastic tissue of the skin and the urethra. The aim of this study was to assess the pre-operative size and biomechanical properties of urethrae in boys with and without hypospadias using a more objective catheter-based system.

Material(s) and Method(s): In this non-blinded clinical observation study, the study population consisted of 19 boys with hypospadias-the case group (median age 13.9 months [range: 12.2-21.3])-and seven boys without hypospadias-the control group (median age 8.5 months [range: 3.8-18.1]). Modified measurements of impedance were used to assess the size, compliance and viscoelasticity of the urethrae under stepwise increased pressures (between 0, 40 and 60 cmH₂O) using a customised Endolumenal Functional Lumen Imaging probe (EndoFLIP).

Result(s): The sizes of the urethrae in boys with hypospadias are variable but tend towards being narrower and less compliant than those of the control subjects i.e. median diameter for meatus urethra was 3.2 mm (range: 2.98-3.92) in the hypospadias group compared with 3.64 (range: 3.22-4.44) in the control group at 40 cmH₂O, and the median change in diameter at meatus urethra was 0.08 mm (range: -0.02 to 0.52) in the hypospadias group compared with 0.23 mm (range: -0.02 to 0.34) when the pressure was increased from 40 to 60 cmH₂O. This biomechanical analysis found that there was no significant viscoelasticity of the urethral meatus in both the groups, whereas the remainder of the urethral structure generally had viscoelastic

properties in the control group, seen as a creep on the time/diameter curves (Figure). In the group of boys with hypospadias, evaluations of the urethrae revealed varying viscoelastic abilities, ranging from abilities that were comparable with those of the control subjects to no sign of viscoelasticity at all.

Conclusion(s): This study is the first to measure the biomechanical properties of the urethra in children, which might help to provide an understanding as to the structural and functional changes associated with hypospadias. The urethrae in the subjects with hypospadias were variable in diameter but tended to be narrower overall, especially in the distal portion of the urethra. Furthermore, the urethrae in boys with hypospadias were frequently less viscoelastic than those of controls. Clinical relevance: The EndoFLIP system may be a future way of objectively estimating the severity of a urethral obstruction and could potentially be included in the postoperative assessment of patients with signs of hampered voiding.[Figure presented]

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Institution

(Faurschou, Olsen) Department of Clinical Medicine, Aarhus University, Palle Juul-Jensens Blvd. 82, Aarhus N DK-8200, Denmark (Faurschou, Ernst, Olsen) Department of Urology, Section of Paediatric Urology, Aarhus University Hospital, Palle Juul-Jensens Blvd. 99, Aarhus N DK-8200, Denmark

(Ernst) Department of Public Health, Section for Epidemiology, Aarhus University, Bartholins Alle 2 - Building 1260, Aarhus C DK-8000, Denmark

(Zhao, Liao) Giome Academia, Department of Clinical Medicine, Aarhus University, Palle Juul-Jensens Blvd. 82, Aarhus N DK-8200, Denmark

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A retrospective population-based cohort study to evaluate the impact of an older sibling with undescended testis and hypospadias on the known maternal and fetal risk factors for undescended testis and hypospadias in Ontario, Canada, 1997-2007.

Dave S., Liu K., Clark R., Garg A.X., Shariff S.Z.

Embase

Journal of Pediatric Urology. 15 (1) (pp 41.e1-41.e9), 2019. Date of Publication: February 2019.

[Article]

AN: 2001290928

Introduction: There are several reported risk factors for undescended testis (UDT) and hypospadias (HYP). Also, a family history of UDT or HYP has not been accounted for in prior studies, and doing so may influence these independent risk estimates. Study design: A population-based retrospective cohort study was conducted using linked administrative databases in Ontario, Canada, to identify all live male newborns born between 1997 and 2007, and it was determined whether they underwent an orchidopexy or HYP repair within 5 years of birth. Baseline maternal and fetal risk factors were obtained using appropriate ICD codes. A statistical analysis using a generalized estimating equation with a logit link was performed, adjusting for clustering in mothers with a previous child born in the 5 years before the proband with UDT or HYP, to evaluate the adjusted risk factors of UDT and HYP.

Result(s): A total of 709,968 male infants were followed up from birth for 5 years, of which 5830 underwent an orchidopexy and 2722 had an HYP repair. On multivariable analysis, factors associated with a higher risk of UDT included prematurity, small for gestational age (SGA), associated HYP, gestational hypertension, use of assisted fertility techniques, increased maternal age, Cesarean section, previous sibling with UDT, and disorders of sexual differentiation (DSDs). After adjusting for clustering in mothers with a previous baby with UDT, DSD, associated HYP (odds ratio [OR], 2.0; 95% confidence interval [CI], 1.0-4.1), and a previous sibling with UDT (OR, 3.6; 95% CI, 2.5-5.2) remained significant risk factors. The risk factors on multivariable analysis predicting the risk of HYP included SGA, prematurity, higher income families, and associated anomalies such as UDT. After adjusting for clustering in mothers with a previous sibling with HYP, SGA (OR, 1.8; 95% CI, 1.0-3.1), higher income families (OR, 1.5-1.6), associated UDT (OR, 7.1; 95% CI, 4.9-10.0), and a previous sibling with HYP (OR, 12.8; 95% CI, 9.1-18.1) remained significant risk factors.

Discussion(s): Studies estimating risk factors for UDT and HYP have used variable methodologies to identify index cases and perform statistical analysis. This study suggests that having an older sibling with UDT or HYP is a significant independent risk factor. Performing an analysis adjusting for clustering in mothers with a previous child with UDT or HYP leads to loss of statistical significance for other described risk factors.

Conclusion(s): Underlying genetic or similar environmental exposures may be a key risk factor for UDT and HYP, which confounds known maternal and fetal risk factors for these anomalies.

[Table presented]

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Status

Embase

Author NameID

Dave S.; ORCID: <http://orcid.org/0000-0003-1462-647X>

Institution

(Dave, Clark) Department of Surgery and Paediatrics, Division of Urology, Western University, London, Ontario, Canada (Dave, Liu, Clark, Garg, Shariff) Institute for Clinical Evaluative Sciences, London, Ontario, Canada

(Garg, Shariff) Arthur Labatt Family School of Nursing, Western University, London, Ontario, Canada

(Garg, Shariff) Lawson Health Research Institute, London, Ontario, Canada

(Garg, Shariff) Department of Medicine, Division of Nephrology, Western University, London, Ontario, Canada

(Garg, Shariff) Department of Epidemiology and Biostatistics, Western University, London, Ontario, Canada

Publisher

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Year of Publication

2019

162.

Delayed access to care and unmet burden of pediatric surgical disease in resource-constrained African countries.

Yousef Y., Lee A., Ayele F., Poenaru D.

Embase

Journal of Pediatric Surgery. 54 (4) (pp 845-853), 2019. Date of Publication: April 2019.

[Review]

AN: 2000948163

Background: The purpose of this study was to estimate the unmet burden of surgically correctable congenital anomalies in African low- and middle-income countries (LMICs).

Method(s): We conducted a chart review of children operated for cryptorchidism, isolated cleft lip, hypospadias, bladder exstrophy and anorectal malformation at an Ethiopian referral hospital between January 2012 and July 2016 and a scoping review of the literature describing the management of congenital anomalies in African LMICs. Procedure numbers and age at surgery were collected to estimate mean surgical delays by country and extrapolate surgical backlog. The unmet surgical need was derived from incidence-based disease estimates, established disability weights, and actual surgical volumes.

Result(s): The chart review yielded 210 procedures in 207 patients from Ethiopia. The scoping review generated 42 data sets, extracted from 36 publications, encompassing: Benin, Egypt, Ghana, Ivory Coast, Kenya, Nigeria, Madagascar, Malawi, Togo, Uganda, Zambia, and Zimbabwe. The largest national surgical backlog was noted in Nigeria for cryptorchidism (209,260 cases) and cleft lip (4154 cases), and Ethiopia for hypospadias (20,188 cases), bladder exstrophy (575 cases) and anorectal malformation (1349 cases).

Conclusion(s): These data support the need for upscaling pediatric surgical capacity in LMICs to address the significant surgical delay, surgical backlog, and unmet prevalent need.

Type of Study: Retrospective study and review article Level of evidence: III

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Embase

Institution

(Yousef, Lee, Poenaru) Division of Pediatric General and Thoracic Surgery, The Montreal Children's Hospital, McGill University Health Centre, 1001 Decarie Blvd, Room B04.2028, Montreal, H4A3J1, Quebec, Canada (Ayele) MyungSung Christian Medical Center, MyungSung Medical College, Box 15478, Addis Ababa, PO, Ethiopia

Publisher

W.B. Saunders

Year of Publication

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163.

Urethral pull-up operation for distal and mid-penile hypospadias: a new safe method.

Jawale S.A., Jesudian G., Nehete R.

Embase

Tropical Doctor. 49 (1) (pp 65-67), 2019. Date of Publication: 01 Jan 2019.

[Article]

AN: 625822309

This case series of 51 patients (age range = 11-17 years; 30-month follow-up) describes a new modified approach to treatment of distal and mid-penile hypospadias that has fewer complications. The meatal locations were sub-coronal (51%), coronal (45%) and mid-penile (4%). Forty patients had chordee. Urethra with the corpus spongiosum was dissected off the corpus cavernosum up to the peno-scrotal junction. The distal 5 mm stenotic and avascular part of the urethral tube was excised. The elastic urethra was then stretched and sutured to form a neo-meatus. The urethra was anchored to the Buck's fascia at the glandular, sub-coronal and mid-penile levels. Meatal regression in one patient (2%), subcutaneous hematomas in two patients (4%) and wound dehiscence in two patients (4%) were the complications in the immediate postoperative period. None developed fistula, meatal stenosis or residual chordee in the 30-month follow-up.

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PMID

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Status

Embase

Institution

(Jawale) CEO, Jawale Institute of Pediatric Surgery, Jalgaon, Maharashtra, India (Jesudian)

Surgical Innovation and Consultant Urologist & Laparoscopic Surgeon, Karunya Rural Community Hospital, Coimbatore, Tamil Nadu, India

(Nehete) Head of Department, Vedant Plastic, Cosmetic, Hand & Orthopedic Hospital, Nashik, India

Publisher

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Year of Publication

2019

164.

Variant Presentations of the Exstrophy-Epispadias Complex: A 40-Year Experience.

Maruf M., Benz K., Jayman J., Kasprenski M., Michaud J., Di Carlo H.N., Gearhart J.P.

Embase

Urology. 125 (pp 184-190), 2019. Date of Publication: March 2019.

[Article]

AN: 2001429881

Objective: To investigate the diagnosis, surgical management, and outcomes in patients with variant EEC. Variant presentations of the exstrophy-epispadias complex (EEC) span a wide range of abnormalities. The rarity and diversity of EEC variants can lead to challenges in the diagnosis and subsequent management of this population.

Method(s): The authors reviewed an institutional database of 1336 EEC patients from 1975 to 2018 for variant presentations of EEC. Variant presentations included those with skin covered bladder exstrophy (BE), duplicate bladders, superior vesical fistula, and epispadias with major bladder prolapse. Surgical management and outcomes were assessed.

Result(s): In total, 44 EEC variants were identified. Nineteen (43%) presented with a skin-covered BE variant. Five patients presented with duplicate BE, while 6 presented with superior vesical fistula. Fourteen patients (32%) presented with epispadias with major bladder prolapse. Overall, 36 (82%) EEC variants underwent primary bladder closure, at a median of 135 days after birth (range 1-2010), with 21 (58%) undergoing pelvic osteotomy. Primary closures were successful in 89% of cases. Continence procedures were performed in 17 patients. This includes 5 patients who underwent bladder augmentation. However even without a continence procedure, continence with volitional voiding was found in 8 patients.

Conclusion(s): The most common EEC variant is the skin-covered form of BE. In order to expedite appropriate management, accurate diagnosis upon initial presentation is crucial. Still, successful surgical reconstruction often results in continence that is similar to, or better than, nonvariant EEC presentations.

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Embase

Institution

(Maruf, Benz, Jayman, Kasprenski, Michaud, Di Carlo, Gearhart) Robert D. Jeffs Division of Pediatric Urology, Charlotte Bloomberg Children's Center, James Buchanan Brady Urological Institute, The Johns Hopkins Medical Institutions, Baltimore, MD, United States

Publisher

Elsevier Inc. (E-mail: usjcs@elsevier.com)

Year of Publication

2019

165.

Isoflavone Intake in Early Pregnancy and Hypospadias in the Japan Environment and Children's Study.

Michikawa T., Yamazaki S., Ono M., Kuroda T., Nakayama S.F., Suda E., Isobe T., Iwai-Shimada M., Kobayashi Y., Yonemoto J., Tamura K., Kawamoto T., Nitta H.

Embase

Urology. 124 (pp 229-236), 2019. Date of Publication: February 2019.

[Article]

AN: 2001351439

Objective: To explore the association between isoflavone intake in early pregnancy (the critical window of masculinisation) and hypospadias. Since oestrogen is likely to contribute to the differentiation of male external genitalia, dietary intake of isoflavone, which has a similar structure to human oestrogen, may be associated with the occurrence of hypospadias. However, there has been little evidence of this association.

Material(s) and Method(s): We used data of a nationwide birth cohort study, which recruited women as early in pregnancy as possible throughout Japan between 2011 and 2014. From the response to a self-administered food-frequency questionnaire, the daily intake of genistein (as a representative for isoflavone) was estimated. Information on hypospadias cases that were diagnosed until the first month after birth was obtained from medical records. Odds ratios (ORs) of hypospadias were estimated using a logistic regression model.

Result(s): Among 41,578 mothers, who delivered singleton live male births, the median genistein intake was 15.3 mg/day, and a total of 51 cases of hypospadias were identified. Compared with mothers in the reference group (genistein intake 11th-89th percentiles), those in the low intake

group (≤ 10 th percentile) had an elevated risk of their sons having hypospadias (multivariable-adjusted OR = 2.8, 95% confidence interval = 1.4-5.8). Adverse or beneficial effects of genistein on hypospadias were not observed in the high intake group (≥ 90 th percentile) (OR = 0.9, 95% confidence interval = 0.4-2.4).

Conclusion(s): Low maternal intake of isoflavone in early pregnancy was associated with an elevated risk of hypospadias.

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Status

Embase

Author NameID

Michikawa, Takehiro; ORCID: <http://orcid.org/0000-0002-2298-5133> Isobe, Tomohiko; ORCID: <http://orcid.org/0000-0001-9235-1227>

Institution

(Michikawa, Yamazaki, Ono, Nakayama, Suda, Isobe, Iwai-Shimada, Kobayashi, Yonemoto, Tamura, Kawamoto, Nitta) Centre for Health and Environmental Risk Research, National Institute for Environmental Studies, Tsukuba, Japan (Kuroda) Department of Paediatric Surgery, Keio University School of Medicine, Tokyo, Japan

(Kawamoto) Department of Environmental Health, University of Occupational and Environmental Health, Kitakyushu, Japan

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Year of Publication

2019

166.

Distal Urethroplasty and Glanuloplasty Procedure Can be Suitable for All Types of Glanular/Subcoronal Hypospadias.

Kawai S., Hyuga T., Nakamura S., Nakai H.

Embase

Urology. 124 (pp 248-253), 2019. Date of Publication: February 2019.

[Article]

AN: 2001288153

Objective: To correct all types of glanular/subcoronal hypospadias, we performed surgery named the distal urethroplasty and glanuloplasty procedure (DUG procedure). We analyzed cases that we have experienced.

Method(s): A vertical incision with the meatal margin was made in the 12 o'clock direction, with the margin of the external urethral meatus as the center. By this meatoplasty according to Heineke-Mikulicz principle, we changed all type of glanular/subcoronal hypospadias to the hypospadias with wide meatus and wide glans. And then Thiersh-Duplay procedure was performed.

Result(s): Consecutive 24 underwent modified DUG procedure. The mean age at the time of surgery was 19.0 +/- 11.9 months and the mean preoperative glans width was 13 +/- 1.5 mm. The external urethral meatus was located glanular in 13 and subcoronal in 11. Hypoplastic urethra (HU) was observed in 7. Skin chordee in 10, penile torsion in 6 and meatal stenosis in 10 were observed. The mean surgical duration was 106 +/- 25.4 minutes and the mean postoperative observation period was 40.5 +/- 26.2 months. All patients with preoperative skin chordee, penile torsion, and meatal stenosis were improved postoperatively, and in all cases, apart from the patient with meatal regression with longest HU from glanular to distal penile, the slit-like shape of the external urethral meatus was achieved.

Conclusion(s): DUG procedure can be used for any type of glanular/subcoronal hypospadias but care should be taken not to indicate too aggressively for glanular/subcoronal hypospadias accompanying long HU to distal penile shaft.

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Status

Embase

Institution

(Kawai, Hyuga, Nakamura, Nakai) Department of Pediatric Urology, Jichi Children's Medical Center Tochigi, Tochigi, Shimotsuke, Japan

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Elsevier Inc. (E-mail: usjcs@elsevier.com)

Year of Publication

2019

167.

Ultrasound-Guided versus Conventional Caudal Block in Children: A Prospective Randomized Study.

Karaca O., Pinar H.U., Gokmen Z., Dogan R.

Embase

European Journal of Pediatric Surgery. 29 (6) (pp 533-538), 2019. Date of Publication: 2019.

[Article]

AN: 629942154

Background Injection to the accurate area without any complications is the main factor for the efficiencies of caudal block. The aim of this study was to compare success and the complications of conventional and ultrasound method for caudal block in children. Materials and Methods Two-hundred sixty-six American Society of Anesthesiologists (ASA) category 1 children aged between 6 months and 6 years undergoing hypospadias, circumcision, or both surgeries were randomly allocated two groups (Group C or Group H, n = 133). About 0.25% bupivacaine with 1/200000 adrenaline (total volume: 0.5 mL/kg) was injected after the needle was inserted into the sacral canal in Group C, or right after the needle pierced the sacrococcygeal ligament under longitudinal ultrasound view in Group H. Success rate of block, block performing time, number of needle puncture, success at first puncture, complication rate, age and weight of the patients encountering these complications were recorded. Results The success rate of block was similar between two groups (94.7% in Group C vs 96.2% in Group U, $p > 0.05$). Success at first puncture was higher in Group U than in Group C (90.2 vs 66.2%, respectively; $p < 0.001$). Number of needle puncture, blood aspiration, subcutaneous bulging, and bone contact was higher in Group C but none in Group U ($p < 0.001$) and these complications were occurred in children weighing < 16 kg and less younger than 6 years old. Conclusion We observed that the complications were not encountered, number of needle puncture was lesser, and the success rate of first puncture was higher under ultrasound with longitudinal view.

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Status

Embase

Institution

(Karaca, Pinar, Dogan) Department of Anaesthesiology and Reanimation, Baskent Universitesi Konya Uygulama Ve Arastirma Merkezi, Hocacihan Neighborhood, Saray Avenue, No:1 Selcuklu,

Konya 42080, Turkey (Gokmen) Department of Pediatrics, Baskent Universitesi Konya
Uygulama Ve Arastirma Merkezi, Konya, Turkey

Publisher

Georg Thieme Verlag (E-mail: iaorl@iaorl.org)

Year of Publication

2019

168.

A hospital-based birth defects surveillance system in Kampala, Uganda.

Mumpe-Mwanja D., Barlow-Mosha L., Williamson D., Valencia D., Serunjogi R., Kakande A.,
Namale-Matovu J., Nankunda J., Birabwa-Male D., Okwero M.A., Nsungwa-Sabiiti J., Musoke P.

Embase

BMC Pregnancy and Childbirth. 19 (1) (no pagination), 2019. Article Number: 372. Date of
Publication: 22 Oct 2019.

[Article]

AN: 629850828

Background: In 2010, the World Health Assembly passed a resolution calling upon countries to prevent birth defects where possible. Though birth defects surveillance programs are an important source of information to guide implementation and evaluation of preventive interventions, many countries that shoulder the largest burden of birth defects do not have surveillance programs. This paper shares the results of a hospital-based birth defects surveillance program in Uganda which, can be adopted by similar resource-limited countries.

Method(s): All informative births, including live births, stillbirths and spontaneous abortions; regardless of gestational age, delivered at four selected hospitals in Kampala from August 2015 to December 2017 were examined for birth defects. Demographic data were obtained by midwives through maternal interviews and review of hospital patient notes and entered in an electronic data collection tool. Identified birth defects were confirmed through bedside examination by a physician and review of photographs and a narrative description by a birth defects expert. Informative births (live, still and spontaneous abortions) with a confirmed birth defect were included in the numerator, while the total informative births (live, still and spontaneous abortions) were included in the denominator to estimate the prevalence of birth defects per 10,000 births.

Result(s): The overall prevalence of birth defects was 66.2/10,000 births (95% CI 60.5-72.5). The most prevalent birth defects (per 10,000 births) were: Hypospadias, 23.4/10,000 (95% CI 18.9-28.9); Talipes equinovarus, 14.0/10,000 (95% CI 11.5-17.1) and Neural tube defects, 10.3/10,000 (95% CI 8.2-13.0). The least prevalent were: Microcephaly, 1.6/10,000 (95% CI 0.9-2.8); Microtia and Anotia, 1.6/10,000 (95% CI 0.9-2.8) and Imperforate anus, 2.0/10,000 (95% CI 1.2-3.4).

Conclusion(s): A hospital-based surveillance project with active case ascertainment can generate reliable epidemiologic data about birth defects prevalence and can inform prevention policies and service provision needs in low and middle-income countries.

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Status

Embase

Author NameID

Mumpe-Mwanja, Daniel; ORCID: <http://orcid.org/0000-0002-5670-5371>

Institution

(Mumpe-Mwanja, Barlow-Mosha, Serunjogi, Namale-Matovu, Musoke) Makerere University,
Johns Hopkins University Research Collaboration, Kampala, Uganda (Williamson, Valencia) US
Centers for Disease Control and Prevention (CDC), Atlanta, United States

(Kakande) Medical Research Council, Uganda Virus Research Institute, London School of
Hygiene and Tropical Medicine Uganda Research Unit, Entebbe, Uganda

(Nankunda, Birabwa-Male, Musoke) Makerere University College of Health Sciences, Kampala,
Uganda

(Nankunda, Birabwa-Male) Mulago National Referral Hospital, Kampala, Uganda

(Okwero) US Centers for Disease Control and Prevention (CDC), Kampala, Uganda

(Nsungwa-Sabiiti) Community Health Department, Ministry of Health, Kampala, Uganda

Publisher

BioMed Central Ltd. (E-mail: info@biomedcentral.com)

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2019

Identification of three novel SRD5A2 mutations in Chinese patients with 5alpha-reductase 2 deficiency.

Cheng T., Wang H., Han B., Zhu H., Yao H.-J., Zhao S.-X., Zhu W.-J., Zhai H.-L., Chen F.-G., Song H.-D., Cheng K.-X., Liu Y., Qiao J.

Embase

Asian Journal of Andrology. 21 (6) (pp 577-581), 2019. Date of Publication: 01 Nov 2019.

[Article]

AN: 629754425

In this study, we investigated the genetics, clinical features, and therapeutic approach of 14 patients with 5alpha-reductase deficiency in China. Genotyping analysis was performed by direct sequencing of PCR products of the steroid 5alpha-reductase type 2 gene (SRD5A2). The 5alpha-reductase activities of three novel mutations were investigated by mutagenesis and an in vitro transfection assay. Most patients presented with a micropallus, variable degrees of hypospadias, and cryptorchidism. Eight of 14 patients (57.1%) were initially reared as females and changed their social gender from female to male after puberty. Nine mutations were identified in the 14 patients. p.G203S, p.Q6X, and p.R227Q were the most prevalent mutations. Three mutations (p.K35N, p.H162P, and p.Y136X) have not been reported previously. The nonsense mutation p.Y136X abolished enzymatic activity, whereas p.K35N and p.H162P retained partial enzymatic activity. Topical administration of dihydrotestosterone during infancy or early childhood combined with hypospadias repair surgery had good therapeutic results. In conclusion, we expand the mutation profile of SRD5A2 in the Chinese population. A rational clinical approach to this disorder requires early and accurate diagnosis, especially genetic diagnosis.

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Institution

(Cheng, Wang, Han, Zhu, Zhu, Qiao) Department of Plastic Surgery, Research Center of Tissue Engineering, Shanghai Ninth People's Hospital, Shanghai Jiao Tong University, School of Medicine, Shanghai 200011, China (Yao) Department of Urology, Shanghai Ninth People's Hospital, Shanghai Jiao Tong University, School of Medicine, Shanghai 200011, China

(Zhao, Chen) Central Laboratory, Clinical Research Center, Shanghai Ninth People's Hospital, Shanghai Jiao Tong University, School of Medicine, Shanghai 200011, China

(Zhai, Song, Cheng, Liu) Department of Plastic Surgery, Research Center of Tissue Engineering, Shanghai Ninth People's Hospital, Shanghai Jiao Tong University, School of Medicine, Shanghai 200011, China

Publisher

Wolters Kluwer Medknow Publications (B9, Kanara Business Centre, off Link Road, Ghatkopar (E), Mumbai 400 075, India)

Year of Publication

2019

170.

Timing of pubertal development in boys born with cryptorchidism and hypospadias: A nationwide cohort study.

Arendt L., Ernst A., Braskhoj Lauridsen L., Brix N., Olsen Jo., Ramlau-Hansen C.

Embase

Asian Journal of Andrology. 21 (6) (pp 551-556), 2019. Date of Publication: 01 Nov 2019.

[Article]

AN: 629754411

Pubertal development may be altered in boys with cryptorchidism and hypospadias, but existing knowledge is inconsistent. Therefore, we investigated the association between cryptorchidism and hypospadias and pubertal development in a large cohort study. Boys in the Puberty Cohort, a cohort nested within the Danish National Birth Cohort, were included in this study. Information on cryptorchidism and hypospadias was retrieved from the Danish National Patient Register. From 11 years until 18 years or full pubertal development, information on physical markers of pubertal development was provided biannually, including Tanner stages, axillary hair, acne, voice break, and first ejaculation. In multivariate regression models for interval censored data, the mean (95% confidence intervals [CIs]) differences in months in obtaining the pubertal markers between boys with and without the anomalies were estimated. Among 7698 boys, 196 (2.5%) had cryptorchidism and 60 (0.8%) had hypospadias. Boys with hypospadias experienced first ejaculation and voice break 7.7 (95% CI: 2.5-13.0) months and 4.5 (95% CI: 0.3-8.7) months later than boys without hypospadias. The age at attaining the Tanner stages for gonadal and pubic hair growth was also higher, though not statistically significant. Pubertal development seemed unaffected in boys with mild as well as severe cryptorchidism. In conclusion, hypospadias may be associated with delayed pubertal development, but pubertal development seems unaffected by cryptorchidism. The relation between hypospadias and later pubertal development may be due to the underlying shared in utero risk or genetic factors.

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Status

Embase

Institution

(Arendt, Ernst, Braskhoj Lauridsen, Brix, Ramlau-Hansen) Department of Public Health, Section for Epidemiology, Aarhus University, Aarhus 8000, Denmark (Arendt) Perinatal Epidemiology Research Unit, Department of Paediatric and Adolescent Medicine, Aarhus University Hospital, Aarhus 8000, Denmark

(Olsen) Department of Clinical Epidemiology, Aarhus University Hospital, Aarhus 8000, Denmark

Publisher

Wolters Kluwer Medknow Publications (B9, Kanara Business Centre, off Link Road, Ghatkopar (E), Mumbai 400 075, India)

Year of Publication

2019

171.

Repair of the penile suspensory ligament for congenital and acquired pathology.

Ralph O., Shroff N., Anfosso M., Blecher G., Ralph D.

Embase

BJU International. 124 (4) (pp 687-692), 2019. Date of Publication: 01 Oct 2019.

[Article]

AN: 627060939

Objective: To review the mechanisms and patient outcomes for men presenting with abnormalities of the penile suspensory ligament (PSL) and their correction.

Patients and Methods: We conducted a retrospective review of a total of 118 patients who presented with a variety of PSL abnormalities that necessitated surgical repair from 1993 to 2018.

The patients mean (range) age at presentation was 29 (12-60) years with a mean (range) follow up of 8 months (3 months-12 years). The diagnosis was made clinically, often with a history of penile instability, pain or curvature/torsion, which was confirmed on artificial erection testing. Nocturnal tumescent testing and magnetic resonance imaging were used where necessary. The

surgical repair was performed by placing non-absorbable sutures between the tunica albuginea of the penis and the symphysis pubis. Postoperative outcomes were reported in the clinic by direct questioning and a repeat of the investigations above when patients were unsatisfied with the result.

Result(s): The aetiologies and surgical indications in the 118 patients included: abnormality subsequent to sexual trauma (n = 66); congenital curvature of penis and/or congenital absence/laxity of the ligament (n = 37); Peyronie's disease (n = 8); and venogenic erectile dysfunction (ED; n = 7). A good surgical result, as defined by stabilization and straightening of the penis and a return to normal sexual function, was achieved in 85% of the patients and 82% were very satisfied with the outcome. Complications included long-term ED in four patients.

Conclusion(s): Abnormalities of the PSL are uncommon and often subtle, but with the appropriate diagnosis, a good cosmetic and functional result is usually achievable.

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Status

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Author NameID

Ralph, Oliver; ORCID: <http://orcid.org/0000-0003-1967-5983>

Institution

(Ralph, Shroff, Anfosso, Blecher, Ralph) University College London Hospitals, London, United Kingdom

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Year of Publication

2019

172.

Dermal regeneration sheet Integra in management of recurrent Urethrocutaneous fistula after hypospadias surgery.

Casal-Beloy I., Somoza Argibay I., Garcia Gonzalez M., Garcia-Novoa M.A., Miguez Fortes L.M., Dargallo Carbonell T.

Embase

Journal of Pediatric Urology. 15 (6) (pp 634.e1-634.e6), 2019. Date of Publication: December 2019.

[Article]

AN: 2003618159

Background: Urethrocutaneous fistula UCF is the most common complication following surgical repair of hypospadias. Currently, the surgical technique mostly used to prevent recurrence employs preputial dartos or testicular tunica vaginalis flaps as a urethral covering. However, autologous tissues are limited in patients with multiple surgeries, and the use of biomaterials as a urethral coverage may represent a good alternative.

Objective(s): The goal of the present study is to assess the results and complications of recurrent UCF correction using a dermal bovine regeneration sheet as a urethral covering.

Material(s) and Method(s): From May 2016 to January 2019, all patients with recurrent UCF of the authors center were repaired using this technique. The inclusion criteria were patients who had undergone one or more unsuccessful UCF repair surgeries and the absence of preputial tissue. The informed consent has been signed by all the patients. Patients were examined in outpatient consultations where their urinary stream was evaluated and a physical examination of the penis was conducted.

Result(s): A total of 12 patients and 13 UCFs were included in the study. The median follow-up was 18 months, (range: 4-26), and only two patients (15%) developed a recurrence of UCF. No complications were observed in the remaining patients (85%) during their evolution. No patient developed a fibrosis increase or loss of elasticity of the tissues in contact with the dermal matrix.

Conclusion(s): The use of an Integra sheet as a urethral covering during urethral fistula surgery appears to be a safe, effective, and easily reproducible option. However, prospective studies with larger numbers of patients should be performed to corroborate these results. [Table presented]

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Status

Embase

Institution

(Casal-Beloy, Somoza Argibay, Garcia Gonzalez, Garcia-Novoa, Miguez Fortes, Dargallo Carbonell) Pediatric Urology Division, Pediatric Surgery Department, University Children's Hospital of A Coruna, A Coruna CP: 15006, Spain

Publisher

Elsevier Ltd
Year of Publication
2019

173.

Goniometer not better than unaided visual inspection at estimating ventral penile curvature on plastic models.

Villanueva C.A.

Embase

Journal of Pediatric Urology. 15 (6) (pp 628-633), 2019. Date of Publication: December 2019.

[Article]

AN: 2003584701

Introduction and objective: The assessment of penile curvature is a key component of hypospadias surgery, as it often determines if a one- or two-stage procedure should be performed. The objective of this study was to compare unaided visual inspection (UVI) against goniometry estimations of ventral penile curvature (VC) among pediatric urologists.

Method(s): A total of nine different penile models (1.5 cm wide and 5-6 cm long) representing each decile of curvature from 10degree to 90degree were created. The nine models were divided in two groups: one with five models (group 1: 10degree, 30degree, 50degree, 70degree, and 90degree) and the other with four models (group 2: 20degree, 40degree, 60degree, and 80degree). Each subject measured the VC of each model in group 1 using one method (i.e. UVI) and the curvature of each model in group 2 using the other method (i.e. goniometry). The next subject then used the opposite method for group 1 (goniometry) and group 2 (UVI), and so on, alternating the methods used to measure each group in between the subjects (Figure). The mean error (difference in between the true curvature and the subject estimation) was compared in between the two measurement methods (UVI and goniometry). A statistician calculated that 20-30 subjects would be needed to detect more than 10degree difference in between the methods.

Result(s): A total of 25 subjects were recruited for the study (24 pediatric urologists and 1 adult urologist). Mean errors for all degrees of penile curvature and methods ranged from 3.5degree (90degree model) to 13.6degree (30degree model). The mean error was not statistically different in between UVI and goniometry methods for any degree of curvature. A subgroup analysis of only goniometry estimations comparing subjects with and without prior experience with goniometry

showed a statistically significant difference only for the 60degree model. If choosing the correct surgery depended on determining if the curvature was ≤ 30 degree or > 30 degree, all subjects would have chosen the right surgery for all except the 20degree, 30degree, and 40degree models, where wrong surgery was chosen in 6/25, 15/25, and 7/25, respectively.

Conclusion(s): In this preliminary study, goniometry was not superior to UVI at estimating VC. There is pressing need in the field of hypospadias surgery to develop a tool that can measure VC in a reproducible and reliable fashion.[Formula presented]

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Status

Embase

Author NameID

Villanueva, Carlos A.; ORCID: <http://orcid.org/0000-0002-1733-6390>

Institution

(Villanueva) Phoenix Children's Hospital, 1920 E. Cambridge Ave., Ste. 302, Phoenix, AZ 85006, United States

Publisher

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Year of Publication

2019

174.

Prenatal imaging of genital defects: clinical spectrum and predictive factors for severe forms.

Fuchs F., Borrego P., Amouroux C., Antoine B., Ollivier M., Faure J.-M., Lopez C., Forgues D., Faure A., Merrot T., Boulot P., Jeandel C., Philibert P., Gaspari L., Sultan C., Paris F., Kalfa N.

Embase

BJU International. 124 (5) (pp 876-882), 2019. Date of Publication: 01 Nov 2019.

[Article]

AN: 2003484562

Objectives: To report the clinical spectrum of genital defects diagnosed before birth, identify predictive factors for severe phenotypes at birth, and determine the rate of associated malformations.

Patients and Methods: A retrospective study (2008-2017) of 4580 fetuses, identified prenatally with abnormalities evaluated by our Reference Center for Fetal Medicine, included cases with fetal sonographic findings of abnormal genitalia or uncertainty of fetal sex determination. Familial, prenatal and postnatal data were collected via a standardised questionnaire.

Result(s): In all, 61 fetuses were included. The positive predictive value (PPV) of the prenatal diagnosis of genital defects was 90.1%. Most cases were 46,XY-undervirilized boys, 42 cases (68.8%), which included 29 with mid-penile or posterior hypospadias, nine with anterior hypospadias, and epispadias, micropenis, scrotal transposition, and buried penis (one each). In all, 46,XX-virilized girls were identified in seven cases (11.5%), which included four with congenital adrenal hyperplasia, two with isolated clitoromegaly, and one with ovotestis. Other defects included prune belly syndrome and persistent cloaca (six cases). Early detection during the second trimester (58.1% vs 18.8%, $P = 0.03$), intra-uterine growth restriction (IUGR) (45.2% vs 9.1%, $P = 0.06$), and curvature of the penis (38.7% vs 0%, $P = 0.02$), were more frequently related to severe defects in male newborns. Associated malformations (14 cases, 22.9%) and genetic defects (six) were frequent in undervirilized boys.

Conclusion(s): Prenatal imaging of genital defects leads to a wide range of phenotypes at birth. Its PPV is high and extra-urinary malformations are frequent. Early diagnosis during the second trimester, associated IUGR, and curvature of the genital tubercle, should raise suspicion of a severe phenotype and may justify delivery near a multidisciplinary disorders/differences of sex development team.

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Status

Embase

Author NameID

Faure, Alice; ORCID: <http://orcid.org/0000-0002-3327-6391>

Institution

(Fuchs, Antoine, Faure, Boulot) Service de Gynecologie-Obstetrique, Hopital Arnaud de Villeneuve - CHU Montpellier, France (Borrego, Ollivier, Lopez, Forgues, Kalfa) Unite de Chirurgie Viscerale et Urologique Pediatrique, Hopital Lapeyronie - CHU Montpellier, Montpellier, France

(Borrego, Amouroux, Ollivier, Lopez, Forgues, Jeandel, Philibert, Gaspari, Sultan, Paris, Kalfa)
Universite de Montpellier, Montpellier, France

(Amouroux, Jeandel, Philibert, Gaspari, Sultan, Paris) Unite d'Endocrinologie-Gynecologie
Pediatrique, Departement de Pediatrie, Hopital Arnaud de Villeneuve - CHU Montpellier,
Montpellier, France

(Amouroux, Ollivier, Lopez, Jeandel, Philibert, Gaspari, Sultan, Paris, Kalfa) National Reference
Network DSD DevGen, Centre Constitutif Sud. - CHU Montpellier, Montpellier, France

(Faure, Merrot) Service de Chirurgie Pediatrique, Hopital La Timone, APHM, Marseille, France
Publisher

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175.

Yield of modern genetic evaluation for patients with proximal hypospadias and descended
gonads.

Rowe C.K., Adam M.P., Ahn J.J., Merguerian P.A., Shnorhavorian M.

Embase

Journal of Pediatric Urology. 15 (5) (pp 527.e1-527.e6), 2019. Date of Publication: October 2019.

[Article]

AN: 2002906197

Introduction and background: Although the pediatric urologic community has embraced a multidisciplinary genetic and endocrine evaluation for newborns with ambiguous genitalia, this approach has been reserved for the most severe cases of undervirilized 46,XY individuals despite growing evidence that genetic differences are found even in patients whose only genitourinary anomaly appears to be proximal hypospadias. Identifying these genetic differences is vital for counseling patients as they move through puberty to parenthood as well as parents on future pregnancies.

Objective(s): The primary objective was to evaluate genetic diagnosis in patients with proximal hypospadias. The authors hypothesized the more sensitive genetic evaluation available in the modern era will reveal a high rate of patients with proximal hypospadias and descended testicles who are found to have a genetic difference, supporting a thorough genetic evaluation in these

patients. Study design: A retrospective review was performed of all patients who underwent surgical correction for proximal hypospadias at a single institution from January 1, 2010, to December 31, 2016. Those with midshaft hypospadias were excluded as were patients whose primary surgery was performed at an outside institution. Patient characteristics, including demographics, clinical presentation, genetic evaluation, and referral to a multidisciplinary difference of sex development (DSD) clinic, were collected. The chi-squared test and t-test were used for analysis.

Result(s): There were 112 patients with proximal hypospadias who met the inclusion criteria. Of these, 91 had bilaterally descended testicles, whereas 21 had one or more undescended testicles. Thirty-three percent of patients with isolated proximal hypospadias received genetic testing of some kind, with 24% seen in the multidisciplinary DSD clinic. Four patients had an associated genetic syndrome identified, and 5 had a genetic difference of unknown clinical significance. Overall, 10% of patients with proximal hypospadias and descended testicles had an identifiable genetic difference vs 33% with associated cryptorchidism. Of these, one patient with proximal hypospadias and descended testicles had a genetic difference of known clinical significance that was likely to have been missed in the absence of an evaluation by a geneticist.

Discussion and conclusion: There was a high rate of identifiable genetic differences in patients whose only genitourinary abnormality was proximal hypospadias, especially with the 1% risk of a likely missed diagnosis. These findings support the discussion of a genetic evaluation for all patients with proximal hypospadias, regardless of the testicular location.[Formula presented]

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Status

Embase

Institution

(Rowe) Divison of Pediatric Urology, Connecticut Children's Hospital, Hartford, CT 06106, United States (Adam, Ahn, Merguerian, Shnorhavorian) Division of Pediatric Urology, Seattle Children's Hospital, NE, Seattle, WA 98105, United States

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176.

Chronological changes in uroflowmetry after hypospadias repair: An observational study.

Al Adl A.M., Omar R.G., Mohey A., El Mogy A.A.E.N., El Karamany T.M.

Embase

Research and Reports in Urology. 11 (pp 269-276), 2019. Date of Publication: 2019.

[Article]

AN: 2002864238

Aim: Urinary flow after urethroplasty is of paramount importance. The aim of this study is to evaluate the progression of uroflowmetry (UF) parameters after different distal and proximal hypospadias repair techniques.

Method(s): In this cohort study, cases that underwent primary hypospadias repair at our institution between March 2010 and December 2018 were included when uncomplicated, asymptomatic and toilet-trained. UF findings and post void residual were described after each specific technique.

Result(s): In all, 88 patients were eligible. Time to last UF ranged from 35 to 138 months postoperatively. Significant increase started 36 months after distal tubularized incised plate urethroplasty (TIP) and afterwards than Mathieu technique. While was noticed 24 and 36 months after Onlay technique and proximal TIP, respectively; however, TIP showed steady significant increase at all time intervals. Duckett repair exhibited insignificant change in maximum flow rate (Qmax) values, buccal mucosal graft (BMG) and inner preputial graft (IPG), significant increase in the Qmax values after 6 and 24 months, respectively, then remained steady high. Transposed preputial flap (TPF) showed significant increase at 6-12 months only, then remained steady lower than the other two techniques. Obstructed flow was 37% after distal TIP, 30% after Mathieu, 25% after proximal TIP, 66.7% after Duckett repair, and 33.3% after TPF. There were no obstructed cases after BMG and IPG.

Discussion(s): Improvement by time varied between techniques. After repair most cases are below the 50th percentile, implying that the reconstructed urethra is not functioning as a normal urethra. Staged repair for proximal hypospadias is preferable to a heroic one-stage procedure.

Conclusion(s): Choice of the surgical technique for hypospadias repair had impact on the improvement of Qmax values. TIP improved 36 months postoperatively. However, for proximal cases staged graft repair had earlier improvement and higher Qmax values than proximal TIP and Onlay techniques.

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Status

Embase

Institution

(Al Adl, Omar, Mohey, El Mogy, El Karamany) Urology Department, Faculty of Medicine, Benha University, Benha, Egypt

Publisher

Dove Medical Press Ltd. (PO Box 300-008, Albany, Auckland, New Zealand)

Year of Publication

2019

177.

Coping styles in patients with hypospadias.

Rynja S.P., Bosch J.L.H.R., de Jong T.P.V.M., van der Werf-Kok E.T., de Kort L.M.O.

Embase

Journal of Pediatric Urology. 15 (6) (pp 625.e1-625.e8), 2019. Date of Publication: December 2019.

[Article]

AN: 2002862345

Objective: The objective of this study is to investigate the coping styles used by patients with hypospadias. Long-term hypospadias studies generally show satisfying outcomes, but some report a lower quality of life (QoL) or poorer psychosocial adjustment, particularly in patients with proximal hypospadias. A lower QoL or psychosocial adjustment was found to be associated with passive coping styles in other pediatric patient populations. Hypothetically, patients with hypospadias also develop different coping styles due to medical experiences in childhood, leading to the lower QoL outcomes on the long term. However, coping styles of patients with hypospadias have never been analyzed.

Patients and Methods: Adult men with hypospadias repair in childhood were recruited (n = 55; aged 19.9 [IQR 19.2-22.1]). Coping styles were determined with the Utrecht Coping List (UCL) and results compared with a reference group of male students (n = 55, age 20-30 years, no medical history). Sub analysis of coping styles of the hypospadias groups was done based on three items: severity of hypospadias, time of last hypospadias surgery and occurrence of postoperative complications.

Result(s): Compared to the reference groups, patients with hypospadias had higher scores on Avoidance ($P < 0.05$), particularly patients who had ≥ 1 postoperative complication or proximal hypospadias. Patients with proximal hypospadias also had lower scores on Seeking Social

Support compared to the reference group ($P < 0.05$). Within the hypospadias group, coping style scores did not differ based on severity of hypospadias, timing of surgery or postoperative complications.

Conclusion(s): Patients with hypospadias, particularly those with proximal hypospadias or a postoperative complication, more often use an avoiding coping style compared to a reference population. Attention to coping styles during the follow-up of patients with hypospadias might help to improve the QoL in these patients.

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Status

Embase

Institution

(Rynja, Bosch, de Kort) Department of Urology, University Medical Center Utrecht, Utrecht, Netherlands (de Jong) Department of Pediatric Urology, Wilhelmina Children's Hospital, University Medical Center Utrecht and Emma Children's Hospital, Amsterdam UMC, Netherlands (van der Werf-Kok) Taylor's University, School of Medicine, 1, Jalan Taylor's, Selangor D.E., Malaysia

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2019

178.

Outcome of snodgrass urethroplasty in Distal and mid-penile hypospadias.

Bhutta M.R.

Embase

Pakistan Paediatric Journal. 43 (3) (pp 189-192), 2019. Date of Publication: September 2019.

[Article]

AN: 2002837095

Objective: To know the outcome of Snodgrass urethroplasty in distal and mid penile

hypospadias Study design: Prospective cohort study. Place and duration of study: Department of

pediatric and neonatal surgery Quaid-e-Azam medical college Bahawalpur, September 2015 to February 2018.

Material(s) and Method(s): Eighty-nine patients with distal and mid-penile hypospadias were managed. Patients operated previously and with severe chordae were excluded from the study. Age range was 2 4 yrs to 7 yrs. These patients were managed in pediatric surgery department On an average 9th post-operative day (range 8 to 10 day) per urethra urinary drainage tube was removed. Assessment for complications was made at discharge, and then on 15 days after discharge to 3, 6 and 12 months postoperatively.

Result(s): Mean operative time was 55 +/- 13.2 minutes Age of patients was 2.4 years to 7 years. Urinary drainage tube was removed on day 9 +/- 1.4 days. A total of 7 patients (7.86%) out of 89 developed complications; urethrocutaneous fistula 5 patients, meatal stenosis 1 patient and total disruption 1 patient. Cosmetic results were also very good with slit like urethral meatus.

Conclusion(s): Outcome of Snodgrass urethroplasty as regards to complications and cosmetic results is excellent and is recommended in mid and distal penile hypospadias.

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Status

Embase

Institution

(Bhutta) Department of Pediatric Surgery, Quaid e-Azam Medical College, Bahawai Victoria hospital, Bahawalpur, Pakistan

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Pakistan Pediatric Journal

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2019

179.

The hypospadias phenotype with a distal meatus in the presence of distal penile penoscrotal angle fixation.

Wong Y.S., Pang K.K.Y., Tam Y.H.

Embase

Research and Reports in Urology. 11 (pp 255-260), 2019. Date of Publication: 2019.

[Article]

AN: 2002754164

Objective: Hypospadias patients may present with the phenotype that features the migration of scrotum to distal penile shaft below a coronal/subcoronal meatus. Patients with this phenotype differ widely in the severity of the hypospadias and the complexity of the surgical repair. We aimed to investigate the operative findings and the outcomes of consecutive patients who presented with this phenotype.

Method(s): We retrospectively reviewed the medical charts of 31 consecutive patients who underwent hypospadias repairs from January 2014 to May 2017, and the hypospadias was characterized by i) the external urethral meatus at coronal/subcoronal region, ii) scrotal skin encroaching distally resulting in fixation of penoscrotal angle at distal penile shaft, and iii) urethral plate below the glans on penile shaft invisible or barely visible.

Result(s): The median age at the time of surgery was 15.5 months (10-63). The division of corpus spongiosum was noted at distal penile shaft (n=2; 6.5%), mid-shaft (n=5; 16.1%) and proximal location (n=24; 77.4%). The median ventral curvature before degloving was 45 degrees (10-90). Twenty-eight and 3 patients underwent tubularized incised plate and 2-stage preputial flap repairs, respectively. Twenty-nine of 31 patients required cutback of the hypoplastic urethra. At the time of urethroplasty, 2 (6.5%), 7 (22.6%) and 22(70.9%) patients underwent distal, midshaft and proximal repairs, respectively. At a median follow-up of 30 months (14-50), 6 (19.4%) patients developed one or more complications (fistula=3; meatal stenosis=5).

Conclusion(s): Patients affected by this particular phenotype likely require non-distal hypospadias repair with possibly higher complication rate and should be treated by surgeons with expertise in complex hypospadias repair.

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Institution

(Wong, Pang, Tam) Division of Paediatric Surgery and Paediatric Urology, Department of Surgery, Prince of Wales Hospital, The Chinese University of Hong Kong, Shatin, NT, Hong Kong
Publisher

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180.

Ultrasound-guided dorsal penile nerve block vs neurostimulator-guided pudendal nerve block in children undergoing hypospadias surgery: A prospective, randomized, double-blinded trial.

Aksu C., Akay M.A., Sen M.C., Gurkan Y.

Embase

Paediatric Anaesthesia. 29 (10) (pp 1046-1052), 2019. Date of Publication: 01 Oct 2019.

[Article]

AN: 2002661856

Background and Aims: Hypospadias is a common congenital malformation in pediatric patients. Surgical repair of this malformation is a painful procedure and has long-term effects. Pudendal and penile nerve blocks are commonly preferred techniques for maintaining postoperative analgesia. However, the conventional landmark-based penile block technique involves numerous potential complications and provides a shorter analgesic period compared to the pudendal block. A promising ultrasound-guided dorsal penile nerve block was recently described. We aimed to compare the analgesic effectiveness of ultrasound-guided penile nerve block with that of neurostimulator-guided pudendal nerve block.

Method(s): Thirty-three patients aged 1-7 years were included in this prospective, double-blinded, randomized controlled trial. Patients were divided into two groups and received either ultrasound-guided dorsal penile nerve block or neurostimulator-guided pudendal nerve block. All blocks were performed by the same two anesthesiologists, and the same surgeons performed the surgical procedures. The Face, Legs, Activity, Cry, and Consolability (FLACC) scale was used for postoperative pain management. The primary outcome of the study was time to first analgesic requirement. Secondary outcomes were FLACC scores at different time points, and types and cumulative doses of analgesic drugs.

Result(s): Dorsal penile nerve block provided longer analgesia than pudendal nerve block (32.29 +/- 5.47 hours and 21.13 +/- 3.53 hours, respectively; differences in mean: 11.16, 95% CI: 7.873-14.465) (P <.001). FLACC scores at the time of first analgesic requirement were significantly lower in dorsal penile nerve block group than pudendal nerve block group (median [IQR]: 2 [2-2.5] and 3 [3-5], respectively; differences in median: -1, 95% CI: -1.851 to -0.149) (P <.001).

Conclusion(s): Ultrasound-guided dorsal penile nerve block provided a longer analgesic period and reduced opioid consumption compared to neurostimulator-guided pudendal nerve block.

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Status

Embase

Author NameID

Aksu, Can; ORCID: <http://orcid.org/0000-0002-4389-4257>

Institution

(Aksu, Gurkan) Department of Anesthesiology and Reanimation, Kocaeli University, Faculty of Medicine, Kocaeli, Turkey (Akay, Sen) Department of Pediatric Surgery, Kocaeli University, Faculty of Medicine, Kocaeli, Turkey

Publisher

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2019

181.

The use of high-resolution melting techniques for mutation screening of diseases caused by trinucleotide repeats expansion, with emphasis on the AR gene.

Juniarto A.Z., Ariani M.D., Harumsari S., Listyasari N.A., Hardian, Utari A., Faradz S.M.H.

Embase

Medical Journal of Indonesia. 28 (2) (pp 116-122), 2019. Date of Publication: June 2019.

[Article]

AN: 2002640119

BACKGROUND Trinucleotide repeat expansion (TRE) diseases are genetic diseases caused by an increase in the number of CAG, CGG, and CTG codons. CAG repeat expansion in exon 1 of the androgen receptor (AR) gene is known to be associated with disorders of sex development (DSD) and spinal and bulbar muscular atrophy (SBMA). Because the traditional Southern blot for CAG repeat expansion is laborious and time-consuming, this study was aimed to use high-resolution melting (HRM) analysis to screen the CAG repeat length of the AR gene in Indonesian patients with DSD. **METHODS** In total, 30 male patients with DSD (46, XY), one male patient with SBMA, and 30 healthy males (control) were included in the study. The CAG repeat length was determined using HRM analysis, and Sanger sequencing was used to confirm the CAG repeat length. **RESULTS** For the DSD cases and controls, the melting temperature (T_m) was within the normal range of 89-91.05degreeC; however, T_m was 92.65degreeC for the SBMA case. Sanger sequencing confirmed that DSD cases had 13-27 CAG repeats, and the SBMA case had 54 CAG repeats. **CONCLUSIONS** HRM analysis using polymerase chain reaction is a sensitive, effective,

and rapid technique for screening CAG repeat expansion in exon 1 of the AR gene. This is the first technique for AR gene screening that may be applicable to other TRE diseases.

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Embase

Institution

(Juniarto, Ariani, Harumsari, Listyasari, Utari, Faradz) Center for Biomedical Research (CEBIOR), Faculty of Medicine, Universitas Diponegoro, Semarang, Indonesia (Hardian) Department of

Physiology, Faculty of Medicine, Universitas Diponegoro, Semarang, Indonesia

(Utari) Department of Pediatrics, Faculty of Medicine, Universitas Diponegoro, Semarang, Indonesia

Publisher

Faculty of Medicine, Universitas Indonesia (E-mail: mji@ui.ac.id)

Year of Publication

2019

182.

Occupational exposure to endocrine-disrupting chemicals and other parental risk factors in hypospadias and cryptorchidism development: a case-control study.

Estors Sastre B., Campillo Artero C., Gonzalez Ruiz Y., Fernandez Atuan R.L., Bragagnini Rodriguez P., Frontera Juan G., Gracia Romero J.

Embase

Journal of Pediatric Urology. 15 (5) (pp 520.e1-520.e8), 2019. Date of Publication: October 2019.

[Article]

AN: 2002527146

Aim of the study: Endocrine-disrupting chemicals (EDCs) are exogenous agents that are capable of altering the endocrine system functions, including the regulation of developmental processes. The aim of this study was to investigate the association between EDC exposure and other parental factors in the etiology of hypospadias and cryptorchidism.

Method(s): A case-control study was conducted. Cases (n = 210) were infants aged between 6 months and 14 years diagnosed with hypospadias or cryptorchidism who attended the authors' hospital over a period of 18 months, and controls (n = 210) were infants within the same range of

age and without any urological disorders who attended the outpatient clinic of the same hospital during the same time period. Their selection was independent of exposures. Data on parental occupational exposure to EDCs and other sociodemographic variables were collected through face-to-face interviews and systematically for both cases and controls. Crude and adjusted odds ratios (ORs) were estimated to control for confounding with their 95% confidence interval (CI) by means of logistic regressions. Specifically, three final models of a dichotomous outcome were constructed: one for cryptorchidism, one for hypospadias, and the third considering both malformations together. The Hosmer-Lemeshow test was used to assess the goodness of fit of the models. Their discriminatory accuracy (DA) was ascertained by estimating their areas under the receiver operating characteristic curves area under the curve (AUC) along with their 95% CI. Result(s): Associations were found between advanced maternal age (OR adjusted = 1.82; 95% CI: 1.14-2.92), mother's consumption of anti-abortives (OR = 5.40; 95% CI: 1.40-38.5) and other drugs (OR = 2.02; 95% CI: 1.31-3.16) during pregnancy, maternal and paternal occupational exposure to EDCs (OR = 4.08; 95% CI: 2.03-8.96 and OR = 3.90; 95% CI: 2.41-6.48, respectively), fathers smoking (OR = 2.0; 95% CI: 1.33-2.99), and fathers with urological disorders (OR = 2.31; 95% CI: 1.15-4.90). Maternal and paternal high educational level could be protective of cryptorchidism (OR = 0.47; 95% CI: 0.28-0.76 and OR = 0.63; 95% CI: 0.42-0.93, respectively). The DA of the models for the whole sample (AUC = 0.75; 95% CI: 0.70-0.79) for cryptorchidism (AUC = 0.76; 95% CI: 0.71-0.82) and for hypospadias (AUC = 0.75; 95% CI: 0.69-0.81) was moderately high.

Conclusion(s): Advanced age, some parental occupational exposure to EDCs, some drug consumption, smoking, and the father's history of urological disorders may increase risk and predict the developments of these malformations. Studies with higher samples sizes are needed to assess associations between individual EDC occupational exposures and drugs and these malformations. [Table presented]

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Status

Embase

Author NameID

Estors Sastre B.; ORCID: <http://orcid.org/0000-0001-9064-3496> Gonzalez Ruiz Y.; ORCID: <http://orcid.org/0000-0002-2536-3559>

Bragagnini Rodriguez P.; ORCID: <http://orcid.org/0000-0002-3053-3436>

Institution

(Estors Sastre) Department of Pediatric Surgery, Hospital Universitari Son Espases, Palma de Mallorca, Spain (Campillo Artero) Balearic Health Service, Palma de Mallorca, Center for

Research in Health and Economics, Barcelona School of Management, Universitat Pompeu Fabra, Barcelona, Spain

(Gonzalez Ruiz, Fernandez Atuan, Bragagnini Rodriguez, Gracia Romero) Department of Pediatric Surgery, Hospital Universitario Miguel Servet, Zaragoza, Spain

(Frontera Juan) Clinical Trials and Methodology Support Platform-IDISBA, Clinical Pharmacology Department, Hospital Universitari Son Espases, Palma de Mallorca, Spain

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Year of Publication

2019

183.

The role of anatomic pelvic dissection in the successful closure of bladder exstrophy: an aid to success.

Davis R., Maruf M., Dunn E., DiCarlo H., Gearhart J.P.

Embase

Journal of Pediatric Urology. 15 (5) (pp 559.e1-559.e7), 2019. Date of Publication: October 2019.

[Article]

AN: 2002465975

Introduction: Classic bladder exstrophy is one of the rarest congenital anomalies compatible with life. Surgical treatment of bladder exstrophy has progressed, but the goal of surgery remains a successful primary bladder closure. Several factors have been identified to decrease the risk of failed closure, including appropriate use of osteotomy and adequate postoperative immobilization and analgesia. However, the role of the radical anatomic pelvic dissection, including dissection of the urogenital diaphragm fibers, in a successful closure has not yet been extensively explored.

Objective(s): The objective of this study was to examine the role of radical anatomic pelvic dissection, including dissection of the urogenital diaphragm fibers, in patients with classic bladder exstrophy. Study design: This was a retrospective study based on an institutional database.

Method(s): A retrospective review from an institutional approved database of more than 1,300 patients with epispadias-exstrophy complex was performed. The inclusion criteria included patients with classic bladder exstrophy with at least one failed bladder closure and a reclosure at the authors' institution with a single senior surgeon. Data collection included demographics,

clinical variables, and status of urogenital diaphragm fibers. Magnetic resonance imaging (MRI) scans, if available, were reviewed with a pediatric radiologist to identify urogenital diaphragm fibers.

Result(s): From the database, 93 patients met inclusion criteria. Of these patients, 74 had urogenital diaphragm fibers completely intact at the time of repeat closure, whereas 19 patients did not. There was no association with age or gender and status of urogenital diaphragm fibers. There was no association with osteotomy, the type of primary bladder closure, surgeon subspecialty, and the status of the urogenital fibers. Fourteen patients had at least two prior closures; surprisingly, 11 of these repeat closure patients still had intact urogenital fibers even after two prior closures.

Discussion(s): The recent development and application of 3D MRI-guided pelvic dissection in a large group of patients led the authors to investigate whether adequate pelvic floor dissection had been accomplished at primary or secondary closure. Several patients had MRI scans performed before repeat closure in which the urogenital diaphragm fibers were identified to be intact on imaging; this was corroborated with surgical findings. Approximately 80% of patients had their urogenital diaphragm fibers completely intact and, therefore, did not have an adequate pelvic dissection during their primary or secondary bladder closure, putting the success of their previous closures at risk.

Conclusion(s): Inadequate pelvic diaphragm dissection, defined as intact urogenital diaphragm fibers, demonstrated in a large group of patients with failed exstrophy closure, may be a decisive factor in bladder closure failure. The use of 3D intra-operative image guidance may aid in a safer and more successful pelvic dissection.[Formula presented]

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Status

Embase

Author NameID

DiCarlo H.; ORCID: <http://orcid.org/0000-0003-2396-4796>

Institution

(Davis, Maruf, DiCarlo, Gearhart) Robert D. Jeffs Division of Pediatric Urology, James Buchanan Brady Urological Institutions, Johns Hopkins Hospital, Johns Hopkins Medical Institutions, Charlotte Bloomberg Children's Hospital, Baltimore, MD, United States (Dunn) Department of Radiology, Pediatrics, Johns Hopkins School of Medicine, Johns Hopkins Children's Center, Charlotte Bloomberg Children's Hospital, Baltimore, MD, United States

Publisher

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184.

Parental perspectives on decision-making about hypospadias surgery.

Chan K.H., Panoch J., Carroll A., Wiehe S., Downs S., Cain M.P., Frankel R.

Embase

Journal of Pediatric Urology. 15 (5) (pp 449.e1-449.e8), 2019. Date of Publication: October 2019.

[Article]

AN: 2002456345

Introduction: Many parents who choose hypospadias repair for their son experience decisional conflict and regret. The utilization of a shared decision-making process may address the issue of decisional conflict and regret in hypospadias repair by engaging both parents and physicians in decision-making.

Objective(s): The objective of this study was to develop a theoretical framework of the parental decision-making process about hypospadias surgery to inform the development of a decision aid.

Study design: We conducted semistructured interviews were conducted with parents of children with hypospadias to explore their role as proxy decision-makers, inquiring about their emotions/concerns, informational needs, and external/internal influences. Interviews were conducted until no new themes were identified, analyzing them iteratively using open, axial, and selective coding. The iterative approach entails a cyclical process of conducting interviews and analyzing transcripts while the data collection process is ongoing. This allows the researcher to make adjustments to the interview guide as necessary based on preliminary data analysis in order to explore themes that emerge from early interviews with parents. Grounded theory methods were used to develop an explanation of the surgical decision-making process.

Result(s): Sixteen mothers and one father of seven preoperative and nine postoperative patients (n = 16) with distal (8) and proximal (8) meatal locations were interviewed. Four stages of the surgical decision-making process were identified: (1) processing the diagnosis, (2) synthesizing information, (3) processing emotions and concerns, and (4) finalizing the decision (Extended Summary Figure). Core concepts in each stage of the decision-making process were identified. Primary concerns included anxiety/fear about the child not waking up from anesthesia and their inability to be present in the operating room. Parents incorporated information from the Internet,

medical providers, and their social network as they sought to relieve confusion and anxiety while building trust/confidence in their child's surgeon.

Discussion(s): The findings of this study contribute to our understanding of decision-making about hypospadias surgery as a complex and multifaceted process. The overall small sample size is typical and expected for qualitative research studies. The primary limitation of the study, however, is the underrepresentation of fathers, minorities, and same-sex couples.

Conclusion(s): This study provides an initial framework of the parental decision-making process for hypospadias surgery that will inform the development of a decision aid. Future stages of decision aid development will focus on recruitment of fathers, minorities, and same-sex couples in order to enrich the perspectives of our work.[Formula presented]

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Status

Embase

Author NameID

Downs S.; ORCID: <http://orcid.org/0000-0003-1843-108X>

Institution

(Chan, Panoch, Cain) Department of Urology, Indiana University School of Medicine, Department of Pediatrics, United States (Chan, Carroll) Center for Pediatric and Adolescent Comparative Effectiveness Research, United States

(Wiehe, Downs) Children's Health Services Research Center, United States

(Frankel) Department of Medicine, Indiana University School of Medicine, Indianapolis, Indiana and Cleveland Clinic Learner Institute, Cleveland, OH, United States

Publisher

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185.

A less invasive technique for delayed bladder exstrophy closure without fascia closure and immobilisation: can the need for prolonged anaesthesia be avoided?.

Nikolaev V.V.

Embase

Pediatric Surgery International. 35 (11) (pp 1317-1325), 2019. Date of Publication: 01 Nov 2019.

[Article]

AN: 2002410405

Introduction: It is believed that the main factors enhancing security of the bladder exstrophy closure are use of osteotomy, pubic bones approximation or transferred flaps for rectus fascia closure. However, these methods increase operating time, surgical trauma and carry risks for the patient.

Objective(s): To demonstrate that the goal of secure bladder exstrophy closure can be achieved easier technically and safer for the child than previously thought. The paper examines the hypothesis that less invasive bladder exstrophy closure achieved without fascia closure can reduce pain and avoid the need for immobilization and prolonged analgesia. Study design: Patients aged 34 days to 15 years (n = 36) from 37 who consecutively referred to the institution with classical bladder exstrophy between 2004 and 2016 underwent modified delayed primary (25) or redo (11) closure. One boy with low weight was excluded. Patient and treatment features were analysed to determine needs for immobilisation and anaesthesia in the postoperative period, and outcomes. Procedure: Bladder exstrophy closure with proximal urethroplasty was performed with the detachment of crura from the ishiopubic rami and levators-from obturator internus muscle. Abdominal wall closure was accomplished with skin and subcutaneous fat mobilisation without rectus fascia closure. No method of immobilization was applied. Results and limitations: Bladder closures have been successful in all 36 children in this report after 37 months (22-138) follow up. The surgeries took time between 126 and 215 min (mean - 148). After 1 day in the ICU the majority of the patients (34/36) were returned to the ward. No bladder spasms or signs of acute pain were noted in the ward; therefore, no local anesthesia or opioids were needed. Intravenous analgesia with non-narcotic analgesics was used for all patients in the ward for an average period 2.2 days (95% CI 2-4 days). Complications: Minor complications: two fistulas, which closed spontaneously; three bladder outlet obstructions, each required one endoscopic incision. No major complications of exstrophy closure such as dehiscence or bladder prolapse were occurred.

Conclusion(s): The proposed less invasive technique with relieved postoperative program is the way to obtain successful bladder exstrophy closure as well as to reduce some risks for the patients. Absence of major complications, and avoiding the need for immobilisation and prolonged analgesia, contribute to the benefits of this approach.

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Status

Embase

Author NameID

Nikolaev, Vasily V.; ORCID: <http://orcid.org/0000-0001-7815-4825>

Institution

(Nikolaev) Department of Paediatric Surgery, Pirogov Russian National Research Medical University (RNRMU), Ostrovitianov Str. 1, Moscow 117997, Russian Federation (Nikolaev) Departments of Paediatric Surgery and Uroandrology, Russian Children's Clinical Hospital, Leninsky Prosp. 117, Moscow 119571, Russian Federation

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186.

Postoperative outcomes in distal hypospadias: a meta-analysis of the Mathieu and tubularized incised plate repair methods for development of urethrocutaneous fistula and urethral stricture. Winberg H., Arnbjornsson E., Anderberg M., Stenstrom P.

Embase

Pediatric Surgery International. 35 (11) (pp 1301-1308), 2019. Date of Publication: 01 Nov 2019.

[Article]

AN: 2002365774

Purpose: To compare the two major complications, namely postoperative urethrocutaneous fistula and urethral stricture, between the Mathieu and tubularized incised plate (TIP) repair methods for distal hypospadias.

Method(s): In this meta-analysis, electronic databases were searched for comparative studies on the two techniques. The Oxford Centre for Evidence-based Medicine Levels of Evidence was used to evaluate the included studies. The main outcome measure was the frequency of postoperative fistula and urethral stricture. RevMan 5.3 was used for statistical analyses, with $P < 0.05$ indicating statistical significance.

Result(s): A total of 17 studies, which included 1572 patients, met the inclusion criteria. The frequency of urethrocutaneous fistula did not differ between the Mathieu [115 (13%)] and TIP [90

(13%)] methods [odds ratio (OR) 1.1, 95% confidence intervals (CI) 0.6-1.9; P = 0.73]. Urethral stricture was less frequent after the Mathieu [15 (2%)] method than after the TIP [37 (5%)] method (OR 0.5, 95% CI 0.3-0.8; P < 0.01), even after the subgroup analysis of eight randomized controlled trials was included. Overall, the quality of the included studies was determined to be satisfactory. The levels of evidence on which this review was based ranged from 1b to 2b using the CEBM Levels of Evidence.

Conclusion(s): Compared with TIP repair, Mathieu repair for hypospadias had a significantly lower risk for urethral stricture; however, the risk for urethrocutaneous fistula was similar.

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Embase

Institution

(Winberg, Arnbjornsson, Anderberg, Stenstrom) Department of Pediatric Surgery, Skane University Hospital, Lund 221 85, Sweden (Winberg, Arnbjornsson, Anderberg, Stenstrom) Department of Clinical Sciences, Pediatrics, Lund University, Lund, Sweden

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187.

Risk of re-operation after outpatient distal hypospadias repair in a large, multistate cohort.

Sebastiao Y.V., Brown C.T., Cooper J.N., McLeod D.J., DaJusta D.G.

Embase

Journal of Pediatric Urology. 15 (5) (pp 471.e1-471.e6), 2019. Date of Publication: October 2019. [Article]

AN: 2002324205

Background: Repair of distal hypospadias is one of the most common pediatric urology procedures in the US. However, the risk of postsurgical complications has been reported primarily from single-institution and tertiary center studies, with short duration of patient follow-up.

Objective(s): The aim of the study was to examine the incidence of re-operation and risk factors for re-operation following outpatient distal hypospadias repair in a large, representative sample of US children.

Method(s): A retrospective cohort study of patients aged 0-18 years undergoing single-stage distal hypospadias repair was conducted. Data were obtained from the State Ambulatory Surgery and Services Databases of 9 participating states. Patients who underwent outpatient surgery in 2008-2013 were identified using Current Procedural Terminology (CPT) codes. Patients with records suggesting prior surgery for hypospadias (CPT) were excluded, as were patients who underwent the initial repair <2 years before the end of state data availability. Return outpatient surgery visits across institutions within each of the 9 states were tracked to identify re-operations after the single-stage repair, using CPT codes for surgical treatment of hypospadias complications in 2008-2015. Time-to-event analyses were used to estimate the probability (risk) of re-operation over time and to examine whether patient and institutional characteristics were predictive of re-operation (age, race/ethnicity, health insurance, facility ownership, and institutional volume of hypospadias repair).

Result(s): A total of 4673 children treated across 148 institutions were included. The median follow-up time was 4.1 years (range: 2-7.9). Most patients were <1 year of age at the time of initial repair (53%). The risk of re-operation was 2.6% (95% confidence interval [CI]: 2.1-3.0%) at 1 year and 6.7% (95% CI: 6.0-7.5%) at 5 years after initial repair (Figure). Approximately 13% of re-operation patients had the re-operation at a different institution. None of the patient or institutional factors examined was a significant predictor of the risk of re-operation.

Discussion(s): In this population-based cohort, the estimated 5-year risk of re-operation following single-stage distal hypospadias repair was 6.7% (95% CI: 6.0-7.5). Most re-operations occurred after the first year, informing long-term expectations about postoperative complications. This study was limited by a lack of data on severity of hypospadias and surgeon characteristics and the inability to track re-operations outside of the state in which the original repair was performed.

Conclusion(s): Approximately 7% of children undergoing distal hypospadias repair undergo a re-operation within 5 years. None of the factors studied were predictive of re-operations.[Formula presented]

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Status

Embase

Author NameID

Sebastiao Y.V.; ORCID: <http://orcid.org/0000-0002-4268-3095>

Institution

(Sebastiao, Cooper, McLeod) Center for Surgical Outcomes Research, The Abigail Wexner Research Institute at Nationwide Children's Hospital, Columbus, OH, United States (Brown, McLeod, DaJusta) Division of Urology, Nationwide Children's Hospital, Columbus, OH, United States

(Cooper) Department of Pediatrics, The Ohio State University College of Medicine, Columbus, OH, United States

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188.

Outcomes of staged lingual mucosal graft urethroplasty for redo hypospadias repair.

Aldaquadossi H.A., Shaker H., Youssef H., Kotb Y., Eladawy M.

Embase

Journal of Pediatric Urology. 15 (5) (pp 519.e1-519.e7), 2019. Date of Publication: October 2019.

[Article]

AN: 2002255982

Background: The objective of this study was to present the outcomes for redo hypospadias repair using lingual mucosal graft (LMG).

Patients and Methods: Between June 2012 and February 2017, 47 patients underwent staged LMG urethroplasty for redo hypospadias repair. The inclusion criteria were previous failed hypospadias repair with a paucity of local skin that interferes with correction using skin flaps and demands graft urethroplasty. During the first stage, a well-vascularized bed on the tunica albuginea was created. Then, the harvested LMG was secured to the prepared bed. The second-stage urethroplasty was carried out after six months. In this stage, tubularization of the previously implanted LMG was performed. In four cases, tubularization was difficult owing to graft contracture. This difficulty was managed by using the dorsally degloved penile skin as the onlay island flap in three cases and the buccal mucosa onlay graft in the fourth case. In all cases, a second protective layer from the dartos or tunica vaginalis was developed to cover the neourethra.

Result(s): The median (interquartile range [IQR]) age of patients at the first stage was 5 (4-6) years, and the median (IQR) duration between both stages was 7 (6-8) months. The median (IQR) follow-up after the second stage was 15 (13-16) months. The median (IQR) number of previous operations was 2 (2-3). The median (IQR) length of the LMG was 3 (2.5-4) cm, and the median (IQR) width was 1 (1-2) cm. No donor-site major complications, but mild oral discomfort in the first week after graft harvesting, were reported in 39 (83%) patients. After the second stage, complications were reported in nine (19.2%) patients, meatal stenosis in five and fistula in four. The reported success rate was 80.9%.

Discussion(s): Reconstruction of previously failed hypospadias is a challenge owing to local tissue scarring and a paucity of adjacent healthy tissue. In this study, the LMG was used in two-stage redo hypospadias repair after previous repair failure. In the present study, a success rate of 80.9% was reported after the second stage. According to this study and the published series, harvesting the LMG is associated with minimal immediate donor-site complications and no long-term morbidity. Another advantage of the LMG is easy harvesting with effortlessly reachable tongue in comparison with the buccal mucosa that is deep and requires application of a mouth retractor.

Conclusion(s): Two-stage LMG urethroplasty is a reliable procedure for salvage urethroplasty. Lingual mucosal graft harvesting is easy, with minor oral complications.[Formula presented]

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Status

Embase

Institution

(Aldaquadossi, Shaker, Youssef, Eladawy) Fayoum University, Fayoum, Egypt (Kotb) Ain Shams University, Ain Shams, Egypt

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Experience of pediatric urogenital tract inserted objects: 10-year single-center study.

He Y., Zhang W., Sun N., Feng G., Ni X., Song H.

Embase

Journal of Pediatric Urology. 15 (5) (pp 554.e1-554.e8), 2019. Date of Publication: October 2019.

[Article]

AN: 2002255883

Introduction: Urogenital tract foreign bodies (FBs) have been rarely reported in children, and the management is still challenging.

Objective(s): The aim of this study is to review a 10-year experience with urogenital tract FBs in a single center.

Patients and Methods: The authors reviewed the records of children suspected with urogenital tract FBs and first admitted to the hospital, including demographic characteristics, presenting symptoms, methods of diagnosis, and management. The authors compared the surgery strategies in different locations of FBs and age, and the locations of FBs in different age groups.

Result(s): Two hundred and thirty-nine cases were reviewed, and 188 were confirmed to retain urogenital tract FBs (150 girls and 38 boys). The number of the patients increased progressively in the last 10 years and mainly concentrated in spring and summer in the last 4 years. The peak ages were 3-5 years old and 9-13 years old. General anesthesia surgeries were performed on 20 patients (Fig. 1). Vagina FBs were more likely to require day surgery, whereas bladder FBs required surgery in hospital. Patients younger than 6 years were more likely to be girls with vagina FBs, and patients older than 11 years were more likely to be boys with bladder FBs.

Discussion(s): Urogenital tract FBs in children is a great challenge. As the vagina is shorter and wider than the urethra, girls with vagina FBs are usually treated by day surgery and adolescent boys of urethra FBs are treated by hospital surgery. Misdiagnosis may occur when patients conceal FBs insert history, have severe urinary tract infections, or have previous surgery history. Ultrasonography helps to reduce misdiagnosis. FBs should be taken into consideration when patients have new symptoms after hypospadias repair, and postoperative changes of hypospadias repair, such as urinary calculi, have been excluded. Appropriate surgery techniques, based on the size, nature, and location of FBs, should be performed for complete removal of FBs with minimal complications to reduce secondary injury. Sharp FBs could be migrated among the digestive system, urogenital system, and deep pelvic. If the procedure is difficult, patients with a stable needle can be conservatively managed with close follow-up. Nevertheless, symptomatic patients should be treated actively.

Conclusion(s): The awareness of potential severity of pediatric urogenital tract FBs should be raised. Appropriate toys and timely sex education help prevent children from urogenital tract FBs insertion. Selecting appropriate techniques for particular situations is the best way to reduce

secondary injury, especially for cases with migrated FBs (needles), magnetic FBs, and postoperative FBs.

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Status

Embase

Author NameID

Song H.; ORCID: <http://orcid.org/0000-0001-6686-8505>

Institution

(He, Zhang, Sun, Song) Department of Urology, Beijing Children's Hospital, Capital Medical University, National Center for Children's Health, Beijing 100045, China (Ni) Department of Surgery, Beijing Children's Hospital, Capital Medical University, National Center for Children's Health, Beijing 100045, China

(Feng) Center for Clinical Epidemiology & Evidence-Based Medicine, Beijing Children's Hospital, Capital Medical University, National Center for Children's Health, Beijing 100045, China

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190.

Community engagement of adolescents in the development of a patient-centered outcome tool for adolescents with a history of hypospadias repair.

Chan K.H., Panoch J., Carroll A., Downs S., Cain M.P., Frankel R., Cockrum B., Moore C., Wiehe S.

Embase

Journal of Pediatric Urology. 15 (5) (pp 448.e1-448.e8), 2019. Date of Publication: October 2019.

[Article]

AN: 2002105788

Introduction: Hypospadias may lead to long-term issues with urination, sexual function and psychosocial well-being. Limited evidence exists regarding the healthcare communication preferences of male adolescents regarding sensitive topics.

Objective(s): The purpose of this qualitative study was to explore the healthcare communication preferences of male adolescents regarding sensitive topics (e.g. urinary and sexual issues) and engage them in the initial stages of development of a patient-centered outcome tool for adolescents with a history of hypospadias repair. Study design: A multidisciplinary team with communication design expertise, pediatric urology experts, and health services researchers developed a self-reported toolkit for adolescent patients who had hypospadias repair as children. The toolkit featured short writing/diagramming exercises and scales to facilitate participant reflections about genital appearance, urination, sexual function, and psychosocial well-being. We recruited students from two local high schools for two focus groups to obtain feedback about the usability/acceptability of the toolkit's appearance/content. We inquired about language preferences and preferred format and/or setting for sharing sensitive information with researchers. The focus groups were audio recorded, professionally transcribed, checked for accuracy, and analyzed by two coders using qualitative content analysis. Major themes and subthemes were identified, and representative quotes were selected.

Result(s): We conducted two focus groups in January 2018 with 33 participants, aged 14-18 years. Participants preferred language that would make patients feel comfortable and serious, clinical language rather than slang terms/sexual humor (Extended Summary Table). They recommended avoidance of statements implying that something is wrong with a patient or statements that would pressure the patient into providing answers. They suggested fill-in-the-blank and open-ended responses to encourage freedom of expression and colorful graphics to de-emphasize the test-like appearance of the toolkit. Most participants preferred a toolkit format to a one-on-one interview to discuss sensitive topics such as urinary or sexual issues. Participants would prefer either a male interviewer or would like to have a choice of interviewer gender for individual qualitative interviews, and they recommended a focus group leader with a history of hypospadias repair.

Discussion(s): This study provides a rich description of a group of male high school students' experiences with healthcare providers and researchers. Its qualitative design limits generalizability, and our findings may not be similar to those of adolescents with a history of hypospadias repair.

Conclusion(s): We used focus group feedback on the toolkit prototype to refine the tool for use in a future study of adolescents with a history of hypospadias repair. [Table presented]

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Author NameID

Downs S.; ORCID: <http://orcid.org/0000-0003-1843-108X> Cockrum B.; ORCID:

<http://orcid.org/0000-0002-3560-5566>

Institution

(Chan, Panoch, Cain) Department of Urology, Indiana University School of Medicine, Cleveland, OH, United States (Chan, Carroll) Department of Pediatrics: Center for Pediatric and Adolescent Comparative Effectiveness Research, Cleveland, OH, United States

(Downs, Wiehe) Children's Health Services Research Center, Cleveland, OH, United States

(Frankel) Department of Medicine, Indiana University School of Medicine, Indianapolis, Indiana and Cleveland Clinic Learner Institute, Cleveland, OH, United States

(Cockrum, Moore) Children's Health Services Research, Department of Pediatrics, IU School of Medicine, 410 West 10th Street, HITS, Suite 2000, Indianapolis, IN 46202, United States

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191.

Combined spinal/caudal catheter anesthesia: extending the boundaries of regional anesthesia for complex pediatric urological surgery.

Jayanthi V.R., Spisak K., Smith A.E., Martin D.P., Ching C.B., Bhalla T., Tobias J.D., Whitaker E.

Embase

Journal of Pediatric Urology. 15 (5) (pp 442-447), 2019. Date of Publication: October 2019.

[Article]

AN: 2001943206

Background: Spinal anesthesia (SA) is an established anesthetic technique for short outpatient pediatric urological cases. To avoid general anesthesia (GA) and expand regional anesthetics to longer and more complex pediatric surgeries, the authors began a program using a combined spinal/caudal catheter (SCC) technique. Study design: The authors retrospectively reviewed the charts of all patients scheduled for surgery under SCC between December 2016 and April 2018

and recorded age, gender, diagnosis, procedure, conversion to GA/airway intervention, operative time, neuraxial and intravenous medications administered, complications, and outcomes. The SCC technique typically involved an initial intrathecal injection of 0.5% isobaric bupivacaine followed by placement of a caudal epidural catheter. At the discretion of the anesthesiologist, patients received 0.5 mg per kilogram of oral midazolam approximately 30 min prior to entering the operating room. One hour after the intrathecal injection, 3% chloroprocaine was administered via the caudal catheter to prolong the duration of surgical block. Intra-operative management included either continuous infusion or bolus dosing of dexmedetomidine, as needed, for patient comfort and to optimize surgical conditions. Prior to removal of caudal catheter in the post-anesthesia care unit, a supplemental bolus dose of local anesthesia was given through the catheter to provide prolonged post-operative analgesia.

Result(s): Overall, 23 children underwent attempted SCC. SA was unsuccessful in three patients, and surgery was performed under GA. The remaining 20 children all had successful SCC placement. There were 11 girls and nine boys, with a mean age of 16.5 months (3.3-43.8). Surgeries performed under SCC included seven ureteral reimplantations, two ureterocele excisions/reimplantations, two megaureter repairs, four first-stage hypospadias repairs, one distal hypospadias repair, one second-stage hypospadias repair, two feminizing genitoplasties, and one open pyeloplasty. Average length of surgery was 109 min (range 63-172 min). Pre-operative midazolam was given in 13/20 (65%). All SCC patients were spontaneously breathing room air during the operation, and there were no airway interventions. Only one SCC patient received opioids intra-operatively. There were no intra-operative or perioperative complications.

Discussion(s): This pilot study shows that the technique of SCC allows one to do more complex urologic surgery under regional anesthesia than what would be possible under pure SA alone. The main limitations of the study include the relatively small number of patients and the small median length of the operative procedures. As a proof of concept, however, this does show that complex genital surgery bladder level procedures such as ureteral reimplantation can be performed under regional anesthesia.

Conclusion(s): SCC allows for more complex surgeries to be performed exclusively under regional anesthesia, thus obviating the need for airway intervention, minimizing or eliminating the use of opioids, and thus avoiding known and potential risks associated with GA. The latter is of particular importance given current concerns regarding hypothetical neurocognitive effects of GA on children aged below 3 years.[Formula presented]

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Author NameID

Jayanthi V.R.; ORCID: <http://orcid.org/0000-0002-6865-9026> Whitaker E.; ORCID:
<http://orcid.org/0000-0002-1362-1653>

Institution

(Jayanthi, Ching) Section of Urology, Nationwide Children's Hospital, United States (Smith, Martin, Bhalla, Tobias, Whitaker) Department of Anesthesiology and Pain Medicine, Nationwide Children's Hospital, United States

(Spisak) Department of Anesthesiology, Dayton Children's Hospital, United States

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192.

Urethro-cutaneous Fistula after Hypospadias Repair: A single institution study.

Jumbi T., Shahbal S., Mugo R., Osawa F., Mwika P., Lessan J.

Embase

Annals of African Surgery. 16 (2) (pp 59-63), 2019. Date of Publication: 2019.

[Article]

AN: 629112811

Urethro-cutaneous fistula (UCF) is one of the most frequently seen complications of hypospadias surgery requiring re-operation; it occurs with an incidence of between 4% and 28%. Risk factors associated with the development of UCF can be classified as preoperative, intraoperative or postoperative. The aim of this study was to determine the association of peri-operative risk factors and the development of urethrocutaneous fistula after hypospadias repair. A retrospective review of patients who had undergone hypospadias repair at Kenyatta National Hospital between 2013 and 2017 was conducted. 114 patient records were retrieved. The incidence of UCF was 47%. Risk factors that were significantly associated with UCF are hypospadias type ($p=0.028$), lack of a protective intermediate layer ($p=0.002$), and presence of postoperative complications ($p=0.001$). Age at surgery, suture material, type of repair and use of catheter/stents were not significant factors. Multivariate analysis showed wound infection and meatal stenosis as the most significant factors associated with UCF development.

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Status

Embase

Institution

(Jumbi, Shahbal, Mugo, Osawa, Mwika) University of Nairobi, Pediatric Surgery, School of Medicine, University of Nairobi, PO Box 1059-00618, Ruaraka, Nairobi, Kenya (Lessan)
Kenyatta National Hospital, Kenya

Publisher

Surgical Society of Kenya (E-mail: hsaid2ke@yahoo.com)

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193.

Intralesional Collagenase Clostridium histolyticum Causes Meaningful Improvement in Men with Peyronie's Disease: Results of a Multi-Institutional Analysis.

Hellstrom W.J.G., Tue Nguyen H.M., Alzweri L., Chung A., Virasoro R., Tapscott A., Ziegelmann M., Trost L., Gelbard M.

Embase

Journal of Urology. 201 (4) (pp 777-782), 2019. Date of Publication: 01 Apr 2019.

[Article]

AN: 628784155

Purpose:In a multi-institutional setting we studied the efficacy and safety outcomes at multiple high volume centers where collagenase Clostridium histolyticum is used to treat Peyronie's disease.**Materials and Methods:**We collected retrospective data on consecutive patients with Peyronie's disease who underwent treatment with collagenase C. histolyticum between April 2014 and March 2018 at a total of 5 institutions. Included in the study were 918 patients. Main outcomes of interest included the change in curvature after receiving collagenase C. histolyticum therapy and the frequency of serious treatment related adverse events. The 2-Tailed paired Student t-Test was used to compare continuous variables. Univariate and multivariate regression analyses were performed to assess predictors of the success of collagenase C. histolyticum therapy to improve curvature.

Result(s):In the cohort of 918 patients curvature improved from a mean of 48.2 degrees before treatment to 32.9 degrees after treatment, a 30.1% improvement from baseline ($p < 0.0001$). Of the men 68.7% had a 20% or greater improvement in curvature. In the 502 patients who completed 4 or more cycles curvature improved from a mean of 49.7 degrees before to 32.7 degrees after treatment, a 33% improvement from baseline ($p < 0.0001$). Of these men 74.4% experienced a 20% or greater improvement in curvature. A complication of treatment developed in 9% of patients. The number of cycles of collagenase C. histolyticum received was predictive of curvature improvement ($p < 0.0001$).

Conclusion(s):This large multi-institutional analysis confirms the safety and efficacy of collagenase C. histolyticum therapy in men with Peyronie's disease. Intralesional collagenase C. histolyticum for Peyronie's disease according to the IMPRESS (Investigation of Maximal Peyronie's Reduction Efficacy and Safety Studies) trial protocol produced an improvement in penile curvature in men with Peyronie's disease with a low rate of complications.

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Status

Embase

Institution

(Hellstrom, Tue Nguyen, Alzweri) Department of Urology, Tulane University School of Medicine, 1430 Tulane Ave., New Orleans, LA 70112, United States (Chung, Virasoro) Department of Urology, Eastern Virginia Medical School, Norfolk, VA, United States

(Ziegelmann, Trost) Department of Urology, Mayo Clinic, Rochester, MI, United States

(Gelbard) Department of Urology, University of California-Los Angeles, Los Angeles, CA, United States

(Tapscott) Carolina Urology Partners, Huntersville, NC, United States

Publisher

Lippincott Williams and Wilkins (E-mail: kathiest.clai@apta.org)

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2019

The role of genetic variation in DGKK on moderate and severe hypospadias.

Richard M.A., Sok P., Canon S., Brown A.L., Peckham-Gregory E.C., Nembhard W.N., Carmichael S.L., Ehli E.A., Kallsen N.A., Peyton S.A., Davies G.E., Patel A., Zamilpa I., Wyatt R.A., Hobbs C.A., Scheurer M.E., Lupo P.J.

Embase

Birth Defects Research. 111 (13) (pp 932-937), 2019. Date of Publication: 2019.

[Article]

AN: 627811989

Background: Recent genome-wide association studies of hypospadias have implicated the role of genetic variants in or near the diacylglycerol kinase kappa (DGKK) gene. However, these variants are largely identified among samples of mild and moderate hypospadias cases. Therefore, we evaluated previously identified DGKK variants among second- and third-degree hypospadias cases and controls recruited in Arkansas, a state characterized by a high birth prevalence of hypospadias.

Method(s): Second- and third-degree hypospadias non-Hispanic white cases (n = 36 and n = 9, respectively) and controls (n = 45) were recruited at Arkansas Children's Hospital. Preputial tissue was collected on cases and controls between 2013 and 2017. Cases and controls were genotyped using the Illumina Infinium Global Screening Array. We used logistic regression models to assess the association of genotyped and imputed genetic variants mapped to the DGKK region with second- and third-degree hypospadias.

Result(s): All families self-reported as non-Hispanic white and genetic principal component analyses did not demonstrate evidence of population stratification. Five DGKK variants previously reported as associated with hypospadias were identified in the genotype data. None of the variants were associated with second- or third-degree hypospadias (range of odds ratios = 0.7-0.9, all p >.05).

Conclusion(s): In our analyses, genetic variation in DGKK does not play a role in the development of moderate and severe hypospadias. Our findings provide support to the etiologic heterogeneity of hypospadias by all classifications of severity.

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Status

Embase

Author NameID

Richard, Melissa A.; ORCID: <http://orcid.org/0000-0003-0129-9860> Lupo, Philip J.; ORCID: <http://orcid.org/0000-0003-0978-5863>

Nembhard, Wendy N.; ORCID: <http://orcid.org/0000-0002-8318-4049>

Institution

(Richard, Sok, Brown, Peckham-Gregory, Scheurer, Lupo) Department of Pediatrics, Baylor College of Medicine, Houston, TX, United States (Canon, Patel, Zamilpa) Arkansas Children's Hospital, Little Rock, AR, United States

(Canon, Patel, Zamilpa) Department of Urology, University of Arkansas for Medical Sciences, Little Rock, AR, United States

(Nembhard) Department of Epidemiology, Fay W. Boozman College of Public Health, University of Arkansas for Medical Sciences and Arkansas Children's Research Institute, Little Rock, AR, United States

(Carmichael) Department of Pediatrics, Stanford University School of Medicine, CA, United States

(Ehli, Kallsen, Peyton, Davies) Avera Institute for Human Genetics, Sioux Falls, SD, United States

(Wyatt) School of Medicine, University of Arkansas for Medical Sciences, Little Rock, AR, United States

(Hobbs) Division of Birth Defects Research, Department of Pediatrics, College of Medicine, University of Arkansas for Medical Sciences, Arkansas Children's Research Institute, Little Rock, AR, United States

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John Wiley and Sons Inc. (P.O.Box 18667, Newark NJ 07191-8667, United States)

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2019

195.

Schilbach-Rott syndrome associated with 9q22.32q22.33 duplication, involving the PTCH1 gene. Prontera P., Rogaia D., Sallicandro E., Mencarelli A., Imperatore V., Squeo G.M., Merla G., Elisei S., Moretti-Ferreira D., Esposito S., Stangoni G.

Embase

European Journal of Human Genetics. 27 (8) (pp 1260-1266), 2019. Date of Publication: 01 Aug 2019.

[Article]

AN: 627009185

Schilbach-Rott syndrome (SRS, OMIM%164220) is a disorder of unknown aetiology that is characterised by hypotelorism, epichantal folds, cleft palate, dysmorphic face, hypospadias in males and mild mental retardation in some patients. To date, 5 families and 17 patients have exhibited this phenotype, and recurrence in two of these families suggests an autosomal dominant inheritance. SRS overlaps with a mild form of holoprosencephaly (HPE), but array-CGH analysis and sequencing of some HPE-related genes (SEPT9, SHH and TWIST) did not reveal any variants in at least one family. Herein, we investigated by array-CGH analysis a 11-year-old female patient and her father, both exhibiting the typical SRS phenotype, disclosing in the daughter-father couple the same microduplication of chromosome 9q22.32q22.33 [arr[hg19]9q22.32(98,049,611_98,049,636)x3,9q22.33 (99,301,483_99,301,508)x3], involving eight genes, including PTCH1. The duplication segregated with the disease, since it was not found in the healthy paternal grandparents of the proband. The gain-of-function variants of the PTCH1 gene are responsible for a mild form of HPE. This is the first genetic variant found in SRS. This finding reinforces the hypothesis that SRS belongs to the HPE clinical spectrum and suggests to perform array-CGH in patients with SRS phenotype and, if negative, to consider a potential benefit from sequencing of HPE-related genes.

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Status

Embase

Author NameID

Merla, Giuseppe; ORCID: <http://orcid.org/0000-0001-5078-928X>

Institution

(Prontera, Rogaia, Sallicandro, Mencarelli, Imperatore, Stangoni) Medical Genetics Unit, University-Hospital "Santa Maria della Misericordia", Perugia, Italy (Sallicandro, Imperatore) "Mauro Baschirotto" Institute for Rare Diseases (B.I.R.D.), Via B. Bizio, 1, Costozza di Longare, Costozza di Longare VI 36023, Italy

(Squeo, Merla) Division of Medical Genetics, IRCCS Casa Sollievo della Sofferenza, San Giovanni Rotondo, Foggia, Italy

(Elisei) Istituto Serafico, Assisi, Perugia, Italy

(Moretti-Ferreira) Department of Genetics, Institute of Biosciences of Botucatu, Sao Paulo State University - UNESP, Sao Paulo, Brazil

(Esposito) Pediatric Clinic, Department of Surgical and Biomedical Sciences, University of Perugia, Perugia, Italy

Publisher

Nature Publishing Group (Houndmills, Basingstoke, Hampshire RG21 6XS, United Kingdom)

Year of Publication

2019

196.

Postoperative behavioral changes in Chinese children undergoing hypospadias repair surgery: A prospective cohort study.

Luo R., Zuo Y., Liu H.B., Pan Y.

Embase

Paediatric Anaesthesia. 29 (2) (pp 144-152), 2019. Date of Publication: February 2019.

[Article]

AN: 625655508

Background: The perioperative period can be psychologically challenging, and children may exhibit behavioral changes following surgical anesthesia. It is unknown whether children in China have additional risk factors associated with negative behavioral changes.

Objective(s): The aim of this study was to investigate the incidence of behavioral changes in children after hypospadias repair surgery and to identify potential risk factors associated with negative behavioral changes.

Method(s): A prospective cohort of 177 children aged 2-12 years scheduled for hypospadias repair surgery from 2016 to 2017 was studied. The primary outcome was the incidence of behavioral changes on postoperative days 14 and 30 evaluated with the Post-Hospitalization Behavioral Questionnaire. Data collected included demographic data, anesthesia details, procedure details, admission details, child anxiety, child temperament, pain, and emergence delirium. Multivariable logistic regression was used to identify risk factors associated with postoperative negative behavioral changes.

Result(s): A total of 60.5% (107/177) of children exhibited negative postoperative behavioral changes on day 14 and 46.5% (79/170) exhibited changes on day 30 after the surgery.

Approximately 2.3% (4/177) and 2.4% (4/170) of children showed improved behavior on postoperative days 14 and 30, respectively. The frequency of temper tantrums changed the most. The logistic regression results suggested that a younger age (odds ratio: 0.86; 95% confidence interval 0.76-0.96), emotional temperament (odds ratio: 1.1; 95% confidence interval 1.0-1.2) and maternal education (odds ratio: 2.2; 95% confidence interval 1.1-4.5) were associated with negative postoperative behavioral changes on day 14. On day 30, a younger age (odds ratio:

0.87; 95% confidence interval 0.77 to 0.98) was the only factor associated with negative postoperative behavioral changes.

Conclusion(s): For children undergoing hypospadias repair surgery in our institution, approximately three in five showed postoperative behavioral changes. In addition to a younger age and a higher maternal education, an emotional temperament is associated with a high incidence of negative postoperative behavioral changes.

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Status

Embase

Author NameID

Luo, Rong; ORCID: <http://orcid.org/0000-0002-8684-3652> Zuo, YunXia; ORCID:

<http://orcid.org/0000-0003-1743-078X>

Institution

(Luo, Zuo, Liu, Pan) Department of Anesthesiology, West China Hospital of Sichuan University, Chengdu, Sichuan, China

Publisher

Blackwell Publishing Ltd

Year of Publication

2019

197.

Outcome of circumcision for newborns with penoscrotal web: oblique skin incision followed by penis shaft skin physical therapy shows success.

Maizels M., Meade P., Rosoklija I., Mitchell M., Liu D.

Embase

Journal of Pediatric Urology. 15 (4) (pp 404.e1-404.e8), 2019. Date of Publication: August 2019.

[Article]

AN: 2002343546

Short Introduction/Background: and objectives: Although it is widely agreed that newborn circumcision complications are low when the penile anatomy is normal, outcomes are uncertain

when a web of skin attaches the penis to the scrotum. This anomaly, called a penoscrotal web or webbed penis, often leads to surgical reconstruction instead of newborn circumcision.

Objective(s): With this study, the authors compare the circumcision success rate for webbed penis circumcisions using a new, alternate method vs that using the traditional method.

Study Design: Data from circumcision patients presenting to the Division of Urology's circumcision clinic from January 2014 to April 2018 were reviewed. All patients who met the checklist criteria for suitability to circumcise were enrolled in the study. They were grouped into the 'normal' group if they had no penile anomalies or the 'web' group if they had a web with a straight penis. Cases with penile anomalies were excluded. The new circumcision method includes altering the circumcision site planned to be oblique, slant up, to compensate for the web, retaining slightly more ventral than dorsal shaft skin, and including home care skin physical therapy as 'push down' the shaft skin. Postcircumcision evaluation was completed within 2 weeks after circumcision, and families were followed up as needed over the study period, six months after circumcision. Circumcision success was defined as the penis shaft no longer attached to the scrotum and circumcision line below the glans corona.

Result(s): Of 828 boys who presented for circumcision, 652 (79%) were enrolled as they were suitable for circumcision: 355 (43%) in the normal group and 297 (36%) in the web group. The remaining 176 (21%) were excluded because they presented with a penile anomaly: buried penis (125), chordee (40), and hypospadias (11). Follow-up was carried out for 6 months. In the web group, follow-up data were obtained for 263 of 297 (89%) cases, with 261 of 263 (99%) showing success, and in the normal group, follow-up data were obtained for 327 of 355 (92%) cases, with all 327 (100%) showing success. The two web group cases (0.7%) who failed had surgical reconstruction.

Discussion(s): It is believed the high success rate for penoscrotal web circumcisions with oblique incision followed by penis shaft skin physical therapy establishes that surgical reconstruction is not required in most of these cases.

Conclusion(s): It was found that newborns with a penoscrotal web and straight shaft show 99% success for circumcision, which is not different from boys without any penile anomalies ($p =$ not significant). The authors believe the adoption of these new, alternate circumcision methods will enable boys with a web to avoid surgical reconstruction.

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Embase

Institution

(Maizels, Meade, Rosoklija, Mitchell, Liu) Ann and Robert H. Lurie Children's Hospital of Chicago, Division of Urology, Chicago, IL 60611, United States

Publisher

Elsevier Ltd

Year of Publication

2019

198.

Functional and cosmetic urethroplasty outcome, emotional stress after genital examination, post traumatic stress disorder, and ages at the time of urethroplasty as potential risk factor causing psychosocial disorder of hypospadias children.

Duarsa G.W.K., Pratiwi D.A., Tirtayasa P.W., Yudiana W., Santosa K.B., Oka A.A.G., Wahyuni S., Mahadewa T.G.B.

Embase

Open Access Macedonian Journal of Medical Sciences. 7 (9) (pp 1452-1455), 2019. Date of Publication: 15 May 2019.

[Article]

AN: 2002326726

BACKGROUND: Children with hypospadias, being born with congenital abnormalities, having repeated genital examination, hospitalization, and undewent genital surgery, experienced psychological stress that may negatively affect their psychosocial life. Choosing the proper time of surgery as recommended is important, since it may have a positive impact on the psychosocial adaptation. AIM: This study aims to find the risk factors causing psychosocial disorders in post-repair surgery on hypospadias children.

METHOD(S): This is a case control study, from 203 hypospadias patients underwent urethroplasty from 2009 to 2018. Subjets were screened for psychosocial disorders by Pediatric Symptoms Questionnaire 17 (PSC-17) questionnaire to find those with psychosocial disorders, score 15 or more (case group) and those without psychosocial disorders (control group), score 0-14. We traced back the data retrospectively from both group (history of utrethrocuteaneous fistula and meatal stenosis, age upon urethroplasty) and collecting new ones (cosmetic outcome, emotional stress after genital examination, and the existence of PTSD). Fisher's exact test was performed to see the Odds ratio (OR) for each variable.

RESULT(S): Some children with hypospadias show impaired on psychocosomal. Functional and cosmetic outcome not significantly different as potensial risk factor psychosocial disorders, genital examination doesn't trigger psychological stress and also none children show PTSD symtomp after surgery. Comparison time of age urethroplasty did not differ significantly between two group

CONCLUSION(S): Twenty-nine children post urthroplasty show psychosocial disorders. Functional and cosmetic urethroplasty outcomes, emotional stress after genital examination, post-traumatic stress disorder were not risk factors of pshycosocial disorder of hypospadias patients. Ages at time of surgery did not differ significantly between two group and this is contradict to the previous recommendations.

Copyright © 2019 Gede Wirya Kusuma Duarsa, Dinar Ayu Pratiwi, Pande Wisnu Tirtayasa, Wayan Yudiana, Kadek Budi Santosa, Anak Agung Gde Oka, Sri Wahyuni, Tjokorda Gde Bagus Mahadewa.

Status

Embase

Institution

(Duarsa, Tirtayasa, Yudiana, Santosa, Oka) Department of Urology, Faculty of Medicine, Udayana University, Sanglah General Hospital, Bali, Indonesia (Pratiwi, Mahadewa) Department of Surgery, Faculty of Medicine, Udayana University, Sanglah General Hospital, Bali, Indonesia (Wahyuni) Department of Psychiatry, Faculty of Medicine, Udayana University, Sanglah General Hospital, Bali, Indonesia

Publisher

Open Access Macedonian Journal of Medical Sciences (E-mail: mspiroski@id-press.eu)

Year of Publication

2019

199.

Etiology and characteristics of pediatric urethral strictures in a developing country in the 21st century.

Ansari M.S., Yadav P., Srivastava A., Kapoor R., Ashwin Shekar P.

Embase

Journal of Pediatric Urology. 15 (4) (pp 403.e1-403.e8), 2019. Date of Publication: August 2019.

[Article]

AN: 2002255887

Background: Urethral stricture disease in children is not uncommon as assumed; however, most of the information about the etiology, features, and natural history of pediatric strictures is extrapolations from adult series as the literature on this common entity is sparse, and most of the studies are small series.

Objective(s): The current etiology and clinical features of urethral stricture disease in the pediatric population in the developing world were determined.

Material(s) and Method(s): The data of children with urethral stricture disease, who had undergone treatment in the tertiary center from 2001 to 2017, were retrospectively analyzed. After excluding girls, the database was analyzed for clinical presentation, possible causes of stricture, site and number of strictures, and length of stricture and for previous interventions. Subanalysis was performed for stricture etiology by patient age, stricture length, site, previous treatments, and presentation with paraurethral abscess.

Result(s): A total of 195 boys with strictures were identified. The common causes of pediatric urethral stricture were traumatic (36.9%), iatrogenic (31.8%), and idiopathic (28.7%). The anterior urethra was the location of the stricture in 141 patients (72.3%). Iatrogenic causes (due to catheterization, hypospadias repair, and valve fulguration) accounted for the majority of anterior urethral strictures (61/141 or 43.2%). Younger children had a tendency to have an iatrogenic and idiopathic cause for strictures, whereas older children had a traumatic etiology; 18.6% of strictures in children younger than 10 years were secondary to trauma, whereas 44.9% of the strictures in patients older than 10 years were traumatic in origin. Trauma was the major cause of posterior urethral strictures (53/54 or 98.1%) and was always associated with pelvic fracture. Strictures due to lichen sclerosus or infectious cause were rare (5 patients or 2.6%). The length of strictures was longer in pan anterior urethral strictures (mean: 82.0 mm) than that of those due to lichen sclerosus (mean: 42.5 mm) and in patients who had undergone previous treatment (mean: 28.7 mm).

Conclusion(s): Iatrogenic causes such as catheterization and hypospadias repair account for the majority of anterior urethral stricture disease in the pediatric population, especially the younger age-group. However, as the child grows, there is a gradual preponderance of traumatic urethral strictures, including posterior urethral strictures. [Table presented]

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Embase

Institution

(Ansari, Yadav, Srivastava, Kapoor, Ashwin Shekar) Pediatric Urology, Department of Urology and Renal Transplantation, Sanjay Gandhi Postgraduate Institute of Medical Sciences, Lucknow, India

Publisher

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2019

200.

Megaprepuce: presentation of a modified surgical technique with excellent cosmetic and functional results.

Hirsch K., Schwaiger B., Kraske S., Wullich B.

Embase

Journal of Pediatric Urology. 15 (4) (pp 401.e1-401.e6), 2019. Date of Publication: August 2019.

[Article]

AN: 2002252831

Objective: Congenital megaprepuce is a malformation consisting of a great redundancy of the inner preputial skin over a penis with normal shaft and glans and is combined with a severe phimosis. Patients suffer from difficulties in voiding because the urine is trapped in the large dome-shaped megaprepuce. We describe a modification of the surgical technique of reconstructing a megaprepuce initially presented by Leao et al.

Patients and Methods: We retrospectively reviewed 7 patients aged 6-53 months (mean age 17 months, 6 were younger than 18 months) who underwent congenital megaprepuce repair between 02/2014 and 05/2018 in our institution. All these otherwise healthy children suffering from difficulties in voiding and reporting genital ballooning during micturition and urinary retention were referred to our hospital. In all cases, parents needed to express the trapped urine. Four of these patients additionally showed a glanular hypospadias, another one a distal penile hypospadias. In addition to the repair of the megaprepuce, six patients needed correction of a penile curvature, five of whom needed correction of the chordee and one a corporoplasty (Schroder-Essed). The patient showing the distal penile hypospadias additionally underwent hypospadias repair. During the follow-up, we evaluated the cosmetic result and complications

such as secondary concealed penis, difficulties in voiding, urinary retention, and urinary infections.

Result(s): Mean follow-up was 18 months. All patients following surgery showed normal voiding without urinary retention or urinary infections and good cosmetic results resembling a circumcised penis in appearance without reconcealment. No intraoperative complications occurred. One patient had a scrotal hematoma postoperatively. Mild transient edema of the penis was seen in all patients, which disappeared spontaneously within one week after surgery.

Conclusion(s): Our surgical approach is a safe and relatively simple procedure with a low rate of complications, good cosmetic results, and functional outcome. Whether the hypospadias associated with ventral curvature was a coincidence or part of the disease pattern remains unclear but will probably be the object of further investigations.[Figure presented]

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Embase

Institution

(Hirsch, Schwaiger, Kraske, Wullich) Department of Urology and Pediatric Urology, University Hospital Erlangen, Friedrich-Alexander-University Erlangen-Nuremberg, Erlangen 91054, Germany

Publisher

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Year of Publication

2019

201.

Hypospadias Prevalence and Trends in International Birth Defect Surveillance Systems, 1980-2010.

Yu X., Nassar N., Mastroiacovo P., Canfield M., Groisman B., Bermejo-Sanchez E., Ritvanen A., Kiuru-Kuhlefelt S., Benavides A., Sipek A., Pierini A., Bianchi F., Kallen K., Gatt M., Morgan M., Tucker D., Canessa M.A., Gajardo R., Mutchinick O.M., Szabova E., Csaky-Szunyogh M., Tagliabue G., Cragan J.D., Nembhard W.N., Rissmann A., Goetz D., Bower C., Baynam G.,

Lowry R.B., Leon J.A., Luo W., Rouleau J., Zarante I., Fernandez N., Amar E., Dastgiri S., Contiero P., Martinez-de-Villarreal L.E., Borman B., Bergman J.E.H., de Walle H.E.K., Hobbs C.A., Nance A.E., Agopian A.J.

Embase

European Urology. 76 (4) (pp 482-490), 2019. Date of Publication: October 2019.

[Article]

AN: 2002244979

Background: Hypospadias is a common male birth defect that has shown widespread variation in reported prevalence estimates. Many countries have reported increasing trends over recent decades.

Objective(s): To analyze the prevalence and trends of hypospadias for 27 international programs over a 31-yr period. Design, setting, and participants: The study population included live births, stillbirths, and elective terminations of pregnancy diagnosed with hypospadias during 1980-2010 from 27 surveillance programs around the world. Outcome measurements and statistical analysis: We used joinpoint regression to analyze changes over time in international total prevalence of hypospadias across programs, prevalence for each specific program, and prevalence across different degrees of severity of hypospadias. Results and limitations: The international total prevalence of hypospadias for all years was 20.9 (95% confidence interval: 19.2-22.6) per 10 000 births. The prevalence for each program ranged from 2.1 to 39.1 per 10 000 births. The international total prevalence increased 1.6 times during the study period, by 0.25 cases per 10 000 births per year ($p < 0.05$). When analyzed separately, there were increasing trends for first-, second-, and third-degree hypospadias during the early 1990s to mid-2000s. The majority of programs (61.9%) had a significantly increasing trend during many of the years evaluated.

Limitations include known differences in data collection methods across programs.

Conclusion(s): Although there have been changes in clinical practice and registry ascertainment over time in some countries, the consistency in the observed increasing trends across many programs and by degrees of severity suggests that the total prevalence of hypospadias may be increasing in many countries. This observation is contrary to some previous reports that suggested that the total prevalence of hypospadias was no longer increasing in recent decades.

Patient Summary: We report on the prevalence and trends of hypospadias among 27 birth defect surveillance systems, which indicate that the prevalence of hypospadias continues to increase internationally. We report on prevalence and trends of hypospadias among 27 birth defect surveillance systems (1980-2010), which indicate that the prevalence of hypospadias continues to increase internationally.

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Status

Embase

Institution

(Yu, Agopian) School of Public Health, University of Texas Health Science Center at Houston, Houston, TX, United States (Nassar, Baynam) Child Population and Translational Health Research, Children's Hospital at Westmead Clinical School, Faculty of Medicine and Health, University of Sydney, Sydney, Australia

(Mastroiacovo) Centre of the International Clearinghouse for Birth Defects Surveillance and Research, Rome, Italy

(Canfield) Birth Defects Epidemiology and Surveillance Branch, Texas Department of State Health Services, Austin, TX, United States

(Groisman) National Network of Congenital Anomalies of Argentina (RENAC), National Center of Medical Genetics, National Administration of Laboratories and Health Institutes (ANLIS), National Ministry of Health, Buenos Aires, Argentina

(Bermejo-Sanchez) ECEMC, Centro de Investigacion sobre Anomalias Congenitas (CIAC), Instituto de Investigacion de Enfermedades Raras (IIER), Instituto de Salud Carlos III, Madrid, Spain

(Ritvanen, Kiuru-Kuhlefelt) Finnish Register of Congenital Malformations, National Institute for Health and Welfare, Helsinki, Finland

(Benavides) Centro de Registro de Enfermedades Congenitas (CREC), Unidad de Enfermedades Congenitas, Instituto Costarricense de Investigacion y Ensenanza en Nutricion y Salud (INCIENSA), Cartago, Costa Rica

(Sipek) Department of Medical Genetics, Thomayer Hospital, Prague, Czechia

(Pierini, Bianchi) Institute of Clinical Physiology, National Research Council (IFC-CNR), Pisa, Italy

(Pierini, Bianchi) Fondazione Toscana Gabriele Monasterio, Pisa, Italy

(Kallen) National Board of Health and Social Welfare, Stockholm, Sweden

(Gatt) Malta Congenital Anomalies Registry, Directorate for Health Information and Research, G'mangia Hill, G'mangia, Malta

(Morgan, Tucker) Congenital Anomaly Register & Information Service for Wales (CARIS), Public Health Wales, Swansea, Wales, United Kingdom

(Canessa, Gajardo) Regional Register Congenital Malformation Maule Health Service (RRMC-SSM), Maule, Chile

(Mutchinick) Department of Genetics, Registry and Epidemiological Surveillance of Congenital Malformations (RYVEMCE), Instituto Nacional de Ciencias Medicas y Nutricion "Salvador Zubiran", Mexico City, Mexico

(Szabova) Slovak Medical University, FPH, Limbova, Bratislava, Slovakia

(Csaky-Szunyogh) Hungarian Congenital Abnormalities Registry and Rare Disease Centre, National Public Health Center, Budapest, Hungary

(Tagliabue) Lombardy Cancer Registry Fondazione IRCCS Istituto Nazionale Tumori, Milan, Italy

(Cragan) National Center on Birth Defects and Developmental Disabilities, Centers for Disease Control and Prevention, Atlanta, GA, United States

(Nembhard) Fay W. Boozman College of Public Health, University of Arkansas for Medical Sciences and Arkansas Reproductive Health Monitoring System, Little Rock, AR, United States

(Rissmann, Goetz) Malformation Monitoring Centre Saxony-Anhalt, Medical Faculty Otto-von-Guericke University Magdeburg, Magdeburg, Germany

(Bower) Telethon Kids Institute, Subiaco, Western Australia, Australia

(Baynam) Western Australian Register of Developmental Anomalies, Western Australian Department of Health, Perth, Australia

(Lowry) Alberta Congenital Anomalies Surveillance System, Department Clinical Genetics, Alberta Children's Hospital, Calgary, AB, Canada

(Leon, Luo, Rouleau) Public Health Agency of Canada, Ottawa, Ontario, Canada

(Zarante) Instituto de Genetica Humana, Pontificia Universidad Javeriana, Bogota, Colombia

(Fernandez) Division of Pediatric Urology, Hospital Universitario San Ignacio, Pontificia Universidad Javeriana, Bogota, Colombia

(Amar) Registre Des Malformations En Rhone-Alpes, REMERA (ex Central-East France Register of Congenital Malformations, IEG), Lyon, France

(Dastgiri) Tabriz Health Services Management Research Centre, Tabriz University of Medical Sciences, Tabriz, Iran, Islamic Republic of

(Contiero) Lombardy Birth Defect Registry, Fondazione IRCCS Istituto Nazionale dei Tumori, Milan, Italy

(Martinez-de-Villarreal) Genetics Department, Medical School, Universidad Autonoma de Nuevo Leon, Nuevo Leon, Mexico

(Borman) New Zealand Birth Defects Registry, Massey University, Wellington, New Zealand

(Bergman, de Walle) Department of Genetics, University of Groningen, University Medical Center Groningen, Groningen, Netherlands

(Hobbs) Rady Children's Institute for Genomic Medicine, San Diego, CA, United States

(Nance) Utah Birth Defect Network, Division of Family Health and Preparedness, Utah Department of Health, Salt Lake City, UT, United States

Publisher
Elsevier B.V.

Year of Publication
2019

202.

Exploration of Practice Patterns in Exstrophy Closures: A Comparison Between Surgical Specialties Using a National and Institutional Database.

Zaman M.H., Davis R., Maruf M., DiCarlo H., Gearhart J.P.

Embase

Urology. 131 (pp 211-216), 2019. Date of Publication: September 2019.

[Article]

AN: 2002244796

Objective: To compare the surgical subspecialties performing bladder exstrophy closures and characterize their practice patterns using both a national and institutional database.

Method(s): The National Surgical Quality Improvement Program Pediatric (NSQIPP) database was reviewed for all bladder exstrophy closures performed from 2012 to 2017. A single institutional exstrophy-epispadias complex database of 1337 patients was reviewed for patients with a bladder closure at a referring institution from 1975 to 2018. Patients with cloacal exstrophy were excluded. The subspecialties of the surgeons performing the closures were identified. Practice patterns such as the use of a pelvic osteotomy and postoperative immobilization, and perioperative outcomes were compared for each subspecialty group.

Result(s): A total of 84 bladder exstrophy patients from NSQIPP and 263 from the author's institutional database met the inclusion criteria. From NSQIPP, 88% of closures were performed by pediatric urologists while 12% were done by other subspecialists. From the institutional database, 75% of referred bladder exstrophy closures were done by a pediatric urologist, and 25% by other services. Gender, race, operation time, length of stay, and postoperative complications were not significantly different between the groups. In one database, pediatric surgeons performed closures earlier, and in another database, pediatric urologists had greater utilization of osteotomy with different immobilization techniques. Pediatric urologists had a higher success rate.

Conclusion(s): Pediatric urologists performed the most bladder exstrophy closures in both databases; they operated on more delayed closures with a greater use of adjunctive procedures and a higher success rate. Differences in surgical training may contribute to the differences in practice patterns.

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Status

Embase

Institution

(Zaman, Davis, Maruf, DiCarlo, Gearhart) Robert D. Jeffs Division of Pediatric Urology, James Buchanan Brady Urological Institution, The Johns Hopkins Hospital, and Charlotte Bloomberg Children's Hospital, Baltimore, MD, United States

Publisher

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Year of Publication

2019

203.

Assessment of the outcome of the tubularized incised plate (T.I.P) technique in the management of distal hypospadias; prospective single-centre study.

Alwan A.A., Obaid A.A., Ajeel H.T.

Embase

International Journal of Research in Pharmaceutical Sciences. 10 (2) (pp 1547-1550), 2019. Date of Publication: 2019.

[Article]

AN: 2002194802

To assess the consequence of tubularized incised plate urethroplasty on primary hypospadias repair. Total of 42 male patients underwent hypospadias repair in AL-Diwaniyah Teaching Hospital/Iraq. from April 2016 to April 2018. The levels of the hypospadias defect, age at operation, type of sutures and dressing, type of catheter and time of removal and complications were verified. Tubularized incised plate urethroplasty done for all patients and mean patients age at operation was 4.4 years (range 1 year to 8year). Postoperative follow up was 1 to 3 months. Generally, meatal stenosis, dehiscence due to infection and an urethrocutaneous fistula occurred in 3, 2 and 6 patients, respectively. T.I.P. urethroplasty has come to be the favourite surgical procedure of distal hypospadias cases at our hospital. The technique has a small number of complications in addition to prove success and adaptability that continue to increase its application.

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International Journal of Research in Pharmaceutical Sciences.

Status

Embase

Institution

(Alwan, Obaid) Department of Surgery, College of Medicine, University of Al-Qadisiyah, Iraq

(Ajeel) Department of Urology, AL-Diwaniyah Teaching Hospital, Iraq

Publisher

J. K. Welfare and Pharmascope Foundation (E-mail: info@pharmascope.org)

Year of Publication

2019

204.

Outcome of surgical management of urethral stricture following hypospadias repair.

Talab S.S., Cambareri G.M., Hanna M.K.

Embase

Journal of Pediatric Urology. 15 (4) (pp 354.e1-354.e6), 2019. Date of Publication: August 2019.

[Article]

AN: 2002186729

Introduction: Reconstruction of urethral strictures in patients with a history of hypospadias repair is controversial. The authors policy has been that if a residual healthy urethral plate is present, single-stage urethroplasty is recommended. However, if the urethral plate is fibrotic or absent or if lichen sclerosus is present, two-stage repair is utilized.

Objective(s): In this study, the authors report their experience in management of patients with urethral stricture and prior hypospadias surgery. Study design: Between 1993 and 2015, 62 patients with urethral stricture and a prior history of hypospadias repair underwent urethroplasty. Patients were divided into two groups; patients in group 1 (n = 39) found to have a healthy residual urethral plate and underwent single-stage urethral stricture repair using either an island skin flap or a buccal mucosa graft. Patients in group 2 (n = 23) had either a scarred urethra or evidence of lichen sclerosus and underwent staged repair using a buccal mucosa graft. Post-operatively, patients were evaluated at 3 months, 6 months, 1 year, and then annually.

Result(s): The median age of the patients was 10.5 years (2.5-33 years). The mean stricture length was 6.3 cm in group 1 and 7.1 cm in group 2. Overall success rate was 87.1% in group 1; a urethral fistula occurred in one patient (7.1%) who underwent skin flap onlay repair and one patient (4.5%) with a buccal mucosa graft. Recurrent urethral stricture was also diagnosed in one patient (7.1%) after repair using an island skin flap and in two patients (9%) following buccal mucosa graft. In group 2, three patients (13%) developed graft contracture and were revised before the second stage. Two patients (8.6%) had glans dehiscence following second stage urethroplasty. The final success rate in group 2 was 90.4%.

Discussion(s): Both single-stage and 2-stage repair showed successful outcome in management of urethral stricture following hypospadias repair. However, the authors continue to believe that the status of the urethral plate dictates the type of surgery to be utilized. In accordance to the previously published data, the study results also further support promising outcomes of application of buccal mucosa in surgical management of these patients.

Conclusion(s): In patients with urethral stricture after hypospadias surgery who have a healthy residual urethral plate, single-stage repair using buccal mucosa graft is a viable option with high success rate. In patients with scarred urethral plate, a 2-stage repair is recommended. [Table presented]

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Status

Embase

Institution

(Talab) Department of Surgery, Division of Urology, Rutgers-New Jersey Medical School, Newark, NJ 07103, United States (Cambareri) Children's Hospital of New Jersey, RWJBarnabas Health, Newark, NJ 07112, United States

(Hanna) Saint Barnabas Medical Center, RWJBarnabas Health, Livingston, NJ 07039, United States

(Hanna) New York Presbyterian-Weill Cornell Medical Center, New York, NY 10065, United States

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2019

205.

Fetal exposure to polybrominated diphenyl ethers and the risk of hypospadias: focus on the congeners involved.

Koren G., Carnevale A., Ling J., Ozsarfati J., Kapur B., Bagli D.

Embase

Journal of Pediatric Urology. 15 (4) (pp 405.e1-405.e6), 2019. Date of Publication: August 2019.

[Article]

AN: 2002166320

Background: Polybrominated diphenyl ethers (PBDEs) are widely used flame retardants, and their endocrine-disrupting properties have focused growing attention regarding their teratogenic potential. We have recently documented that mothers of children born with hypospadias had been exposed to statistically higher levels of PBDE during pregnancy than mothers of healthy controls. However, it is not known which congeners of PBDE are associated with this putative teratogenic effect.

Objective(s): To identify PBDE congeners associated with increased risk for hypospadias. Study methods: Hair samples from mothers were analyzed and compared between hypospadias cases and healthy controls for eight PBDE congeners using gas chromatography mass spectrometry (GC/MS). Polybrominated diphenyl ether levels were measured in the 0- to 3-cm segment closest to the skull of maternal hair as a proxy for in utero exposure of mothers who lived in the same environment for the duration of their pregnancy.

Result(s): Median maternal hair levels of five PBDE congeners (28, 47, 99, 153, and 154) and of total PBDE (PBDE) were significantly higher among mothers of infants with hypospadias (n = 152) than among controls (n = 64). Apparent greater differences in the lower brominated congeners, especially in BDE-47 and BDE-99, may be due to the fact that they had been used in larger amounts, and their persistence properties confer longer exposure.

Conclusion(s): The majority of the lower brominated PBDE congeners measured in maternal hair exhibited higher PBDE body burden during pregnancy in mothers of infants who were born with hypospadias.

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Status

Embase

Author NameID

Koren G.; ORCID: <http://orcid.org/0000-0002-9234-0875>

Institution

(Koren, Ozsarfati, Kapur) Hospital for Sick Children, Department of Pediatrics, Toronto, ON, Canada (Koren, Carnevale, Ling, Kapur) University of Toronto, Department of Pharmacology and Toxicology, Toronto, ON, Canada

(Koren) Western University, Toronto, ON, Canada

(Koren) University of Toronto, Leslie Dan School of Pharmacy, Toronto, ON, Canada

(Koren) Maccabi-Kahn Research Institute, Tel Aviv, Israel

(Bagli) Hospital for Sick Children, Department of Surgery, Toronto, ON, Canada

Publisher

Elsevier Ltd

Year of Publication

2019

206.

Topical estradiol increases epidermal thickness and dermal collagen of foreskin prior to hypospadias surgery - Randomized double blinded controlled trial.

Casali T.G., Christina de Castro Paiva K., Rodrigues M.N., Santana Silva C.E., Avarese de Figueiredo A., de Bessa J., Bastos A.N., Marques Nogueira Castanon M.C., Bastos Netto J.M.

Embase

Journal of Pediatric Urology. 15 (4) (pp 346-352), 2019. Date of Publication: August 2019.

[Article]

AN: 2002164635

Introduction: The use of preoperative topical testosterone stimulation prior to hypospadias correction aims to increase penile size and achieve better surgical results. Topical estradiol has been shown to improve the quality of skin in other sites, but its use in boys with hypospadias has not yet been elucidated.

Objective(s): This study aims to evaluate the primary effects in epidermal thickness and collagen distribution of estradiol compared to testosterone and placebo in skin of prepuce before hypospadias surgery. Materials and methods: Patients were randomized into three groups according to the topical hormone used: TG: Testosterone ointment; EG: Estradiol ointment; CG: Neutral base ointment. Fragments of foreskin were excised, fixed and then sectioned for

histology. For each sample, epidermal thickness and dermal collagen expression was measured by specific computer analysis, P-values of <0.05.

Result(s): Thirty-three patients with a mean age of 4.01 +/- 2.92 years were included. Hypospadias classification was similar in all three groups. Mean epidermal thickness and collagen type I expression in EG were greater than those of the other groups. Collagen type III expression was similar in all groups.

Discussion(s): Foreskin has a fundamental role in many techniques of hypospadias surgery and can be used either as a graft or a flap in the correction of the penile defect. Increase of epidermal thickness and dermal collagen observed in the present study has already been related to use of estradiol in other skin sites, but not yet in foreskin. Further studies are needed to evaluate the real significance of these findings in boys with hypospadias.

Conclusion(s): Use of topical estradiol before hypospadias surgery lead to greater epidermal thickness and increases dermal collagen expression in foreskin.[Figure presented]

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Embase

Institution

(Casali, Christina de Castro Paiva, Santana Silva, Avarese de Figueiredo, Bastos Netto)
Department of Surgery - Federal University of Juiz de Fora (UFJF), Minas Gerais, Brazil (Casali, Christina de Castro Paiva, Bastos Netto) Department of Surgery - Hospital e Maternidade Therezinha de Jesus of the School of Medical Science and Health of Juiz de Fora (HMTJ-SUPREMA), Minas Gerais, Brazil

(Rodrigues, Bastos, Marques Nogueira Castanon) Departament of Morphology - Federal University of Juiz de Fora (UFJF), Minas Gerais, Brazil

(de Bessa) Department of Surgery - State University of Feira de Santana (UEFS), Bahia, Brazil

Publisher

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2019

Use of small intestinal submucosa for corporal body grafting in cases of epispadias and epispadias/exstrophy complex.

Ballesteros N., Moscardi P.R.M., Blachman-Braun R., Salvitti M., Alam A., Castellan M., Kozakowski K., Gosalbez R., Labbie A.

Embase

Journal of Pediatric Urology. 15 (4) (pp 406.e1-406.e6), 2019. Date of Publication: August 2019.

[Article]

AN: 2002125111

Background: Male epispadias is a rare congenital urogenital anomaly in which the meatus is ectopically located along the dorsal midline of the penile shaft. In cases associated with severe curvature, functional and cosmetic outcomes could be accomplished by lengthening the shorter dorsal surface with the use of corporal grafting. Various graft materials have been used in the past for hypospadias repair including tunica vaginalis, dermis, and small intestinal submucosa (SIS). The use of SIS grafting for corporoplasty during epispadias repair has rarely been described in the literature.

Objective(s): To report the experience in the management of dorsal corporal body grafting using SIS in children with severe penile curvature due to epispadias. Study design: The authors retrospectively reviewed the charts of all patients with epispadias or bladder exstrophy/epispadias complex and severe dorsal chordee (>40degree) who underwent epispadias repair with single-layer SIS for corporal body grafting. Clinical variables, surgical technique, and outcomes were analyzed.

Result(s): A total of nine consecutive patients underwent staged epispadias repair with dorsal corporal single-layer SIS grafting (summary figure). Of these, four (44.4%) had primary penopubic epispadias, one (11.1%) had mid-shaft epispadias, and four (44.4%) had bladder exstrophy/epispadias complex. The mean age at surgery was 13.4 +/- 6 months. After phalloplasty with SIS grafting, there were no reported complications related to the graft during the post-operative period or follow-up visits.

Discussion(s): Although traditional techniques for epispadias repair allow some degree of corporal lengthening, they also result in abrupt medial rotation of the corporal bodies leading to torqueing and potential unsatisfactory cosmetic results. In contrast, the authors use single-layer SIS for corporal body grafting, and this study technique results in a more gradual inward rotation thus allowing more anatomical accuracy. Furthermore, an advantage of the use of SIS over other grafting materials is that there is no need to harvest an autologous graft such as tunica vaginalis or dermis.

Conclusion(s): Epispadias repair using single-layer SIS corporal body grafting is an effective, safe, and feasible method, which provides satisfactory cosmesis and correction of dorsal

curvature in congenital epispadias in children. Furthermore, a more normal penis appearance, without a decrease in the corporal length or diameter, is achieved with this technique.

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Status

Embase

Author NameID

Alam A.; ORCID: <http://orcid.org/0000-0002-0590-109X>

Institution

(Ballesteros, Moscardi, Blachman-Braun, Salvitti, Alam, Castellan, Kozakowski, Gosalbez, Labbie) Division of Pediatric Urology, Nicklaus Children's Hospital, Miami, FL, United States
(Salvitti, Alam, Castellan, Kozakowski, Gosalbez, Labbie) Division of Pediatric Urology, Joe DiMaggio Children's Hospital, Hollywood, FL, United States

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2019

208.

Comprehensive evaluation of grafting the preservable narrow plates with consideration of native plate width at primary hypospadias surgery.

Seleim H.M., ElSheemy M.S., Abdalazeem Y., Abdullateef K.S., Arafa M.A., Shouman A.M., Elsaket H., Kaddah S.N., Elbarbary M.M.

Embase

Journal of Pediatric Urology. 15 (4) (pp 345.e1-345.e7), 2019. Date of Publication: August 2019.

[Article]

AN: 2002036413

Introduction: Dorsal inlay graft urethroplasty using inner-face preputial graft was described as an adjunct method to the classic tubularized incised plate (TIP) urethroplasty, aiming at reducing the risk of neourethral stenosis.

Objective(s): To evaluate the impact of dorsal inlay grafting of preservable narrow plates, in relation to native plate width.

Method(s): Consecutive children with penile hypospadias with narrow plate (width < 8 mm) were evaluated prospectively between Jan 2014 and Jun 2018. Included cases were stratified into two groups: group A (plate width: 4 to <8 mm) and group B (plate width: < 4 mm). All cases were approached by TIP urethroplasty, with dorsal inlay grafting (inner-face prepuce). Cases with significant chordee (non-preservable plates), circumcised cases, and redo cases were excluded.

Result(s): A total of 104 hypospadias cases with narrow plates were included in this study. Among group A (n = 81), the need for postrepair urethral dilations was reported in two cases (2.5%) vs seven cases (30%) in group B (n = 23) (P-value < 0.001). Another two cases (2.5%) in group A developed urethrocutaneous fistulae vs three cases (13%) in group B (P-value = 0.0624). Surgical repair of the reported five cases with fistulae, revealed an ample neourethral wall; disclosing well-taken grafts in both groups.

Discussion(s): Few published studies evaluated grafting the incised plate in penile hypospadias. To the authors knowledge, this is the first study that selectively evaluated the impact of grafting narrow plates in relation to its native pre-incision width.

Conclusion(s): According to the presented authors' experience, 4 mm width is the border line of clinical relevance that defines poor urethral plate. Grafting that plate failed to compensate for its native poor characteristics; however, it offered a valuable neourethral wall that proved indispensable when redo surgery deemed necessary, without adding grafting-related problems. Nevertheless, further extended comparative studies came across as a necessity to verify the long-term outcomes of grafting the incised poor plates.[Figure presented] [Table presented]

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Status

Embase

Institution

(Seleim, Arafa) Pediatric Surgery Unit, Tanta University, Egypt (ElSheemy, Shouman) Pediatric Urology Unit, Cairo University Specialized Pediatric Hospital, Cairo, Egypt

(Abdalazeem, Abdullateef, Elsaket, Kaddah, Elbarbary) Pediatric Surgery Unit, Cairo University Specialized Pediatric Hospital, Cairo, Egypt

Publisher

Elsevier Ltd

Year of Publication

2019

209.

Impact of pelvic immobilization techniques on the outcomes of primary and secondary closures of classic bladder exstrophy.

Zaman M., Kasprenski M., Maruf M., Benz K., Jayman J., Friedlander D., Di Carlo H., Sponseller P., Gearhart J.

Embase

Journal of Pediatric Urology. 15 (4) (pp 382.e1-382.e8), 2019. Date of Publication: August 2019.

[Article]

AN: 2001969227

Introduction: A potential determinant of successful bladder closures in patients with classic bladder exstrophy (CBE) is the postoperative pelvic immobilization technique. This study investigates the success rates of primary and secondary bladder closures based on various immobilization techniques from a high-volume exstrophy center.

Method(s): A prospectively maintained institutional exstrophy-epispadias complex database of 1336 patients was reviewed for patients with CBE who have undergone primary or secondary closures between 1975 and 2018 and subsequently had a known method of pelvic immobilization. Patients were divided into two groups: primary and secondary closures.

Associations between closure outcomes and immobilization techniques were determined.

Result(s): A total of 476 patients with primary closures and 101 patients with secondary closures met the inclusion criteria. In total, 343 (72.1%) primary closures were successful. As shown in the table, the success rates of primary closures were highest in patients immobilized with modified Buck's and Bryant's traction (95.0% and 79.3%, respectively) and lowest in those with spica cast (49.6%). A propensity score-adjusted logistic regression (adjusting for osteotomy status, period of closure, location of closure, and closure type) revealed that modified Buck's traction had a 5.60 (95% confidence interval 1.74-23.1, $p = 0.008$) greater odds of success compared to spica casting during the primary closure. For the secondary closure group, there were 92 (92.1%) successful secondary closures. Success rates were highest in modified Buck's traction (97.3%) and lowest with spica casting (66.7%).

Discussion(s): This study confirms previous findings of better outcomes when patients are immobilized with external fixation and Buck's traction after adjusting for potential confounding factors. Immobilization with modified Buck's or Bryant's traction yielded significantly higher primary closure success rates when compared to spica casting. It is the authors' belief that

despite a longer hospital length of stay, external fixation with Buck's traction provides the best chance of a successful closure and, thus, a financially responsible method to care for these children in the postoperative period. [Table presented]

Conclusion(s): Success rates for primary closures were highest when using modified Buck's traction with external fixation and lowest for spica casts. Similarly, for secondary closures, the best outcomes were achieved using modified Buck's traction with external fixation and the lowest success rates were associated with spica casts.

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Status

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Institution

(Zaman, Kasprenski, Maruf, Benz, Jayman, Friedlander, Di Carlo, Gearhart) Robert D. Jeffs
Division of Pediatric Urology, James Buchanan Brady Urological Institutions, Johns Hopkins
Hospital, Johns Hopkins Medical Institutions, Charlotte Bloomberg Children's Hospital, Baltimore,
MD, United States (Sponseller) Department of Orthopedic Surgery, The Johns Hopkins
University, Baltimore, MD, United States

Publisher

Elsevier Ltd

Year of Publication

2019

210.

Persistent or recurrent ventral curvature after failed proximal hypospadias repair.

Snodgrass W., Bush N.C.

Embase

Journal of Pediatric Urology. 15 (4) (pp 344.e1-344.e6), 2019. Date of Publication: August 2019.

[Article]

AN: 2001918784

Purpose: Persistent or recurrent ventral curvature (VC) in patients with complications after proximal hypospadias repair is reported.

Method(s): Records of patients undergoing re-operation for complications after proximal repair performed elsewhere were reviewed, including earlier operative reports when available. Original extent of VC, means used for straightening, and presenting complaints and findings at re-operation were tabulated. Ventral curvature at re-operation was objectively measured by goniometry and classified as due to short ventral skin and/or scarring of skin/dartos, short neourethra, or short ventral corpora (corporal disproportion). The finding of corporal disproportion at re-operation was considered to be failure of initial straightening.

Result(s): There were 73 patients with an average of 2.7 [1-5] prior operations for proximal shaft to perineal hypospadias; of which, 83% had VC at re-operation averaging 50degree (30-90). This was due to short skin/scarring in 7% patients, a short neourethra in 23%, and corporal disproportion in 70%. Initial straightening was performed by chordee excision in 18 patients, dorsal plication in 23, and ventral lengthening in 15. Persistent or recurrent corporal disproportion was significantly more likely after chordee excision or dorsal plication than after ventral lengthening ($p = 0.005$). Of patients with VC, 93% also had urethroplasty complications, including recurrent fistulas and wound dehiscences that appeared related to the curvature.

Discussion(s): The VC that was encountered during proximal hypospadias re-operations was important for several reasons. First, all patients with VC who had completed urethroplasty had complications that included recurrent fistulas and wound dehiscences (Figure). Even if they had healed without complications, this VC exceeded 30degree in all cases, which is associated with sexual dysfunction in adults. This VC was not reported by 37% of caregivers and sometimes not apparent on pre-operative physical examination. It is possible there is selection bias in this series, although nearly all patients were self-referred for complications, and nearly 40% of them were not aware their son had VC. Furthermore, the finding that most initially had dorsal plication agrees with an earlier survey of pediatric urologists' preferences for straightening penile curvature.

Conclusion(s): The most common complication in this series was persistent or recurrent VC, and nearly all these patients also had urethroplasty complications. This VC was more likely when the urethral plate was preserved during straightening and when chordee excision or dorsal plication rather than ventral lengthening was performed. These data suggest that surgeons should objectively measure VC and consider ventral lengthening rather than chordee excision or dorsal plication when it is ≥ 30 degree. Re-operations for urethroplasty complications should include artificial erection.[Figure presented]

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Embase

Institution

(Snodgrass, Bush) PARC Urology, 5680 Frisco Square Blvd, Suite 2300, Frisco, TX 75034,
United States

Publisher

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Year of Publication

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211.

Combination of clonidine-bupivacaine in caudal epidural anesthesia for hypospadias surgery in children: prospective, randomized, blind study. Associacao clonidina-bupivacaina na anestesia peridural caudal para cirurgia de hipospadia em criancas: estudo prospectivo, randomizado, encoberto <Associacao clonidina-bupivacaina na anestesia peridural caudal para cirurgia de hipospadia em criancas: estudo prospectivo, randomizado, encoberto.>

Bonisson A.C.M., Fernandes M.L., Araujo G.F., Vieira F.E., Noronha L.M., Gomez R.S.

Embase

Brazilian Journal of Anesthesiology. 69 (1) (pp 27-34), 2019. Date of Publication: January - February 2019.

[Article]

AN: 2001216644

Background and objectives: The combination of clonidine with local anesthetic administered for epidural anesthesia via caudal route seems to improve the quality of postoperative analgesia, but with conflicting results. This study compared the postoperative analgesia of three different doses of clonidine combined with bupivacaine in caudal epidural anesthesia in children undergoing hypospadias repair.

Method(s): Eighty children aged 1 to 10 years, candidates for surgical repair of hypospadias, were randomly divided into four groups of 20 patients to receive general anesthesia combined with caudal epidural anesthesia with bupivacaine 0.165% alone or in combination with 1, 2 or 3 mug.kg- 1 of clonidine. The primary outcome was morphine consumption in the first 24 hours postoperatively. Mean arterial pressure, heart rate, end-tidal concentration of sevoflurane, time to awakening, pain severity (FLACC scale), level of sedation (RAMSAY), duration of analgesia, and occurrence of adverse effects were also compared.

Result(s): Intraoperatively, there was no difference between groups regarding mean arterial pressure, heart rate, end-tidal concentration of sevoflurane, and time to awakening. Postoperative morphine consumption and pain severity were similar between groups, but the group receiving clonidine (3 mug.kg-1) had lower heart rate and higher sedation level than the group receiving bupivacaine alone.

Conclusion(s): The combination of clonidine at doses of 1, 2 or 3 mug.kg-1 with bupivacaine 0.16% via caudal epidural route did not alter the consumption of morphine in the early postoperative period of children undergoing hypospadias repair.

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Embase

Institution

(Bonisson, Araujo, Vieira, Noronha) Fundacao Benjamin Guimaraes, Hospital da Baleia, Belo Horizonte, MG, Brazil (Bonisson, Fernandes) Universidade Federal de Minas Gerais (UFMG), Hospital das Clinicas, Belo Horizonte, MG, Brazil

(Gomez) Universidade Federal de Minas Gerais (UFMG), Faculdade de Medicina, Belo Horizonte, MG, Brazil

(Gomez) Faculdade de Minas (Faminas), Belo Horizonte, MG, Brazil

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Elsevier Editora Ltda

Year of Publication

2019

212.

Current indications and techniques for the use of bowel segments in pediatric urinary tract reconstruction.

Stein R., Zahn K., Huck N.

Embase

Frontiers in Pediatrics. 7 (JUN) (no pagination), 2019. Article Number: 236. Date of Publication: 2019.

[Review]

AN: 628453311

Medical Center Mannheim, Heidelberg University, Mannheim, Germany Today, there are few indications for the use of bowel in pediatric urology. This is in large extent due to the successful conservative therapy in patients with neurogenic bladder and the improved success of primary reconstruction in patients with the bladder exstrophy-epispadias complex. Only after the failure of the maximum of conservative therapy or after failure of primary reconstruction, bladder augmentation, or urinary diversion should be considered. Malignant tumors of the lower urinary tract (e.g., rhabdomyosarcomas of the bladder/prostate) are other rare indications for urinary diversion. Replacement or reconstruction of the ureter with a bowel segment is also a quite rarely performed procedure. In this review, the advantages and disadvantages of the different options for the use of bowel segments for bladder augmentation, bladder substitution, urinary diversion, or ureter replacement during childhood and adolescence are discussed.

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Institution

(Stein, Zahn, Huck) Department of Pediatric, Adolescent and Reconstructive Urology, Medical Faculty Mannheim, Germany

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Year of Publication

2019

213.

Analgesic efficacy and impact of caudal block on surgical complications of hypospadias repair: A systematic review and meta-analysis.

Zhu C., Wei R., Tong Y., Liu J., Song Z., Zhang S.

Embase

Regional Anesthesia and Pain Medicine. 44 (2) (pp 259-267), 2019. Date of Publication: 01 Feb 2019.

[Article]

AN: 628313314

Background and objectives is commonly used for children undergoing hypospadias repair. However, the safety of caudal block for hypospadias repair in children is controversial in terms of surgical complications such as urethrocutaneous fistula and glans dehiscence. We sought to perform a meta-analysis to estimate the analgesic efficacy and relative complications of caudal block for hypospadias repair in children. Methods We identified comparative studies of caudal block versus peripheral nerve block or no caudal block; studies were published or presented through 1 January 2018, and reports of analgesic efficacy or surgical complications of hypospadias repair in children were identified. Peripheral nerve block includes dorsal nerve penile block and pudendal nerve block. Data were abstracted from studies comparing caudal block with peripheral nerve block or no caudal block; original source data were used when available. We prespecified separate assessments of randomized controlled trials (RCTs) and observational studies given the inherent differences between types of study designs. Data from 298 patients in four RCTs and from 1726 patients in seven observational studies were included. RCT and observational data were analyzed separately. Results In RCTs, caudal blocks (compared with peripheral nerve blocks) showed no detectable differences in terms of need for additional analgesia within 24 hours after the surgery (OR 10.49; 95% CI 0.32 to 343.24; $p=0.19$), but limited data showed lower pain scores 24 hours after the surgery (standardized mean difference (SMD) 1.57; 95% CI 0.29 to 2.84; $p=0.02$), a significantly shorter duration of analgesia (SMD - 3.33; 95% CI -4.18 to -2.48; $p<0.0001$) and analgesics consumption. No significant differences were observed in terms of postoperative nausea and vomiting (OR 3.08; 95% CI 0.12 to 77.80; $p=0.50$) or motor weakness (OR 0.01; 95% CI -0.03 to 0.05; $p=0.56$). Only one randomized study showed that caudal blocks (compared with peripheral nerve blocks) were associated with detectable differences in urethrocutaneous fistula rate (OR 25.27; 95% CI 1.37 to 465.01; $p=0.03$) and parental satisfaction rate (OR 0.07; 95% CI 0.02 to 0.21; $p<0.00001$). In observational studies, caudal block was not associated with surgical complications in all types of primary hypospadias repair (OR 1.83; 95% CI 0.80 to 4.16; $p=0.15$). To adjust for confounding factors and to eliminate potential selection bias involving caudal block indication, we performed subgroup analysis including only patients with distal hypospadias. This analysis revealed similar complication rates in children who received a caudal block and in children not receiving caudal block (OR 1.02; 95% CI, 0.39 to 2.65; $p=0.96$). This result further confirmed that caudal block was not a risk factor for surgical complications in hypospadias repair. The direction of outcomes in all the other subgroup analyses did not change, suggesting stability of our results. Conclusions In RCTs, only limited data showed peripheral nerve blocks providing better analgesic quality compared with caudal blocks. In real-world non-randomized observational studies with greater number of patients (but with admitted the potential for a presence of selection bias and residual

confounders), caudal blocks were not associated with postoperative complications including urethrocutaneous fistula and glans dehiscence.

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Embase

Institution

(Zhu, Wei, Tong, Liu, Song, Zhang) Department of Anesthesiology, Shanghai Children's Hospital, Shanghai Jiao Tong University, Shanghai 200060, China

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BMJ Publishing Group (E-mail: subscriptions@bmjgroup.com)

Year of Publication

2019

214.

Incidence of disorders of sexual development in neonates in Ghana: Prospective study.

Ameyaw E., Asafo-Agyei S.B., Hughes I.A., Zacharin M., Chanoine J.-P.

Embase

Archives of Disease in Childhood. 104 (7) (pp 636-638), 2019. Date of Publication: 01 Jul 2019.

[Article]

AN: 627780196

Objective The incidence of disorders of sexual development (DSD) is unknown in sub-Saharan Africa. We describe the characteristics and incidence of DSD in a cohort of infants born in Ghana. **Design** Trained research assistants performed systematic genital examination at birth. All infants with suspected abnormal genitalia were further examined by a paediatric endocrinologist. **Setting** Komfo Anokye Teaching Hospital, Kumasi, Ghana. **Patients** Consecutive infants born in a single centre over a 1-year period (May 2014 to April 2015). **Main outcome measures** Incidence of DSD. **Results** We examined 9255 infants (93% of all live births) within 72 hours of birth. Twenty-six neonates had a DSD. Nineteen infants had DSD without genital ambiguity: isolated micropenis

(n=2), hypospadias (n=7), cryptorchidism (n=4) and clitoromegaly (n=6). Seven infants had DSD with ambiguity: clitoromegaly with a uterus on ultrasound and elevated 17-hydroxyprogesterone, suggesting XX DSD due to congenital adrenal hyperplasia (CAH)(n=4) and micropenis, hypospadias and gonads in a bifid scrotum or in the inguinal region, consistent with XY DSD (n=3). Conclusion The incidence of atypical genitalia was 28/10,000 (95% CI 17/10 000 to 39/10 000) live births. The incidence of CAH was 4.3/10 000 (95% CI 1.2/10 000 to 11.1/10 000) and was strongly associated with consanguinity.

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Institution

(Ameyaw, Asafo-Agyei) Department of Child Health, Komfo Anokye Teaching Hospital, Kumasi, Ghana (Hughes) Department of Paediatrics, University of Cambridge, Cambridge, United Kingdom

(Zacharin) Pediatrics, Royal Children's Hospital Melbourne, Parkville, VIC, Australia

(Chanoine) Department of Pediatrics, University of British Columbia, Vancouver, BC, Canada

Publisher

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Year of Publication

2019

215.

Review of the phenotypic spectrum associated with haploinsufficiency of MYRF.

Rossetti L.Z., Ginton K., Yuan B., Liu P., Pillai N., Mizerik E., Magoulas P., Rosenfeld J.A., Karaviti L., Sutton V.R., Lalani S.R., Scott D.A.

Embase

American Journal of Medical Genetics, Part A. 179 (7) (pp 1376-1382), 2019. Date of Publication: July 2019.

[Article]

AN: 627705707

The myelin regulatory factor gene (MYRF) encodes a transcription factor that is widely expressed. There is increasing evidence that heterozygous loss-of-function variants in MYRF can lead to abnormal development of the heart, genitourinary tract, diaphragm, and lungs. Here, we searched a clinical database containing the results of 12,000 exome sequencing studies. We identified three previously unreported males with putatively deleterious variants in MYRF: one with a point mutation predicted to affect splicing and two with frameshift variants. In all cases where parental DNA was available, these variants were found to have arisen *de novo*. The phenotypes identified in these subjects included a variety of congenital heart defects (CHD) (hypoplastic left heart syndrome, scimitar syndrome, septal defects, and valvular anomalies), genitourinary anomalies (ambiguous genitalia, hypospadias, and cryptorchidism), congenital diaphragmatic hernia, and pulmonary hypoplasia. The phenotypes seen in our subjects overlap those described in individuals diagnosed with PAGOD syndrome [MIM# 202660], a clinically defined syndrome characterized by pulmonary artery and lung hypoplasia, agonadism, omphalocele, and diaphragmatic defects that can also be associated with hypoplastic left heart and scimitar syndrome. These cases provide additional evidence that haploinsufficiency of MYRF causes a genetic syndrome whose cardinal features include CHD, urogenital anomalies, congenital diaphragmatic hernia, and pulmonary hypoplasia. We also conclude that consideration should be given to screening individuals with PAGOD for pathogenic variants in MYRF, and that individuals with MYRF deficiency who survive the neonatal period should be monitored closely for developmental delay and intellectual disability.

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Author NameID

Rossetti, Linda Z.; ORCID: <http://orcid.org/0000-0002-8953-3246> Pillai, Nishitha; ORCID:

<http://orcid.org/0000-0002-2943-2136>

Rosenfeld, Jill A.; ORCID: <http://orcid.org/0000-0001-5664-7987>

Scott, Daryl A.; ORCID: <http://orcid.org/0000-0003-1460-5169>

Institution

(Rossetti, Ginton, Yuan, Liu, Pillai, Mizerik, Magoulas, Rosenfeld, Sutton, Lalani, Scott)

Department of Molecular and Human Genetics, Baylor College of Medicine, Houston, TX, United States (Yuan, Liu) Baylor Genetics, Houston, TX, United States

(Karaviti) Division of Pediatric Endocrinology, Baylor College of Medicine, Houston, TX, United States

(Scott) Department of Molecular Physiology and Biophysics, Baylor College of Medicine,
Houston, TX, United States

Publisher

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Year of Publication

2019

216.

Staged transverse preputial island flap urethroplasty for proximal hypospadias: a single-center experience.

Wang C., Song H., Zhang W.

Embase

Pediatric Surgery International. 35 (7) (pp 823-827), 2019. Date of Publication: 08 Jul 2019.

[Article]

AN: 627586815

Purpose: To evaluate the intermediate outcomes of our institution's experience with staged TPIF urethroplasty for proximal hypospadias repair.

Method(s): We retrospectively evaluated the medical records of patients who underwent repair of proximal hypospadias using staged TPIF urethroplasty at our hospital from 2011 to 2017.

Result(s): One hundred and two patients were included in the present study. The mean follow-up was 52.4 months (range 13-74 months). The mean age at the time of the first surgery was 13.5 months (range 11-65 months). There were two main types of initial complications including meatal stenosis in four (3.9%) and urethrocutaneous fistula in three (2.9%) patients after the first stage. Surgical complications were seen in 15 patients after second stage, including urethrocutaneous fistulas in 8 (7.8%), urethral strictures in 5 (4.9%), urethral diverticula in 2 (1.9%). Overall complication rates after second stage were 14.7%. The incidence of fistulas was lower in patients who underwent repair with a tunica vaginalis flap (1/29, 3.4%) than with the dartos fascia (7/73, 9.6%; $p = 0.435$).

Conclusion(s): Our results show that staged TPIF urethroplasty is a viable and durable technique for primary severe proximal hypospadias. This procedure was associated with a 14.7% complication rate in the present study. Staged TPIF urethroplasty can reduce the incidence of urethral strictures and diverticula associated with the second stage.

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Institution

(Wang, Song, Zhang) Department of Urology, Beijing Children's Hospital Affiliated to Capital Medical University, Beijing 100045, China

Publisher

Springer Verlag (E-mail: service@springer.de)

Year of Publication

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217.

Epidemiology of sexual disorders in general medical practice: An Italian survey.

De Rose A.F., Gallo F., Bini P.M., Gattuccio I., Chiriaco V., Terrone C.

Embase

Urologia Journal. 86 (2) (pp 79-85), 2019. Date of Publication: 01 May 2019.

[Article]

AN: 627322769

Objective: To report the results of a survey supported by the Italian Andrological Association (ASS.A.I.), aimed at documenting sexual disorders in a large population of patients who visited general practitioners for general health problems.

Method(s): Between April and October 2016, 15,000 questionnaires were distributed to general practitioners throughout Italy and made available to all the people who visited the doctor's offices.

The data were collated separately for age ranges, 15-30, 31-50 and 51-65 years, respectively.

Result(s): A final sample of 5441 questionnaires was evaluable (4000 men and 1441 women).

Sexual disorders were reported by 1795 out of 5441 (33.0%) patients. Among the male population, phimosis, varicocele, prostate and testicular disorders were the most common conditions interfering with sexuality, being reported by 42.0%, 37.0%, 39.0% and 31.0% of the sample, respectively. Furthermore, erectile dysfunction, sexually transmitted diseases, infertility, premature ejaculation and penile curvature were reported by 27.7%, 27.0%, 17.0%, 14.4% and

7.8% of the male sample, respectively. Among the female population, low sexual satisfaction was the more common complaint, reported by 65.0% of the sample. Sexually transmitted diseases, low libido, dyspareunia, infertility and arousal disorder were reported by 32.0%, 29.0%, 24.0%, 24.0% and 19.7% of the female sample, respectively.

Conclusion(s): Our data show sexual disorders among 33.0% of people visiting doctors' offices. The majority of these sexual disorders increased with age among both male and female subjects. Despite the limitations of our study, we consider that these data confirm the importance of sexual function evaluation during a physician's daily clinical practice.

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Status

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Author NameID

Gallo, Fabrizio; ORCID: <http://orcid.org/0000-0003-4239-9200>

Institution

(De Rose, Terrone) Department of Urology, Ospedale Policlinico San Martino, Istituto di Ricerca e Cura a Carattere Scientifico per l'Oncologia, University of Genoa, Genoa, Italy (Gallo)

Department of Urology, San Paolo Hospital, Savona, Italy

(Bini) General Medical Practice, Genoa, Italy

(Gattuccio) Centro Andros Palermo, Palermo, Italy

(Chiriaco) Psychology and Psychotherapy, Rome, Italy

Publisher

SAGE Publications Ltd (E-mail: info@sagepub.co.uk)

Year of Publication

2019

218.

Prevalence of selected birth defects by maternal nativity status, United States, 1999-2007.

Kirby R.S., Mai C.T., Wingate M.S., Janevic T., Copeland G.E., Flood T.J., Isenburg J., Canfield M.A.

Embase

Birth Defects Research. 111 (11) (pp 630-639), 2019. Date of Publication: 01 Jul 2019.

[Article]

AN: 626963233

Objectives: We investigated differences in prevalence of major birth defects by maternal nativity within racial/ethnic groups for 27 major birth defects.

Method(s): Data from 11 population-based birth defects surveillance systems in the United States including almost 13 million live births (approximately a third of U.S. births) during 1999-2007 were pooled. We calculated prevalence estimates for each birth defect for five racial/ethnic groups. Using Poisson regression, crude and adjusted prevalence ratios (aPRs) were also calculated using births to US-born mothers as the referent group in each racial/ethnic group.

Result(s): Approximately 20% of case mothers and 26% of all mothers were foreign-born. Elevated aPRs for infants with foreign-born mothers were found for spina bifida and trisomy 13, 18, and 21, while lower prevalence patterns were found for pyloric stenosis, gastroschisis, and hypospadias.

Conclusion(s): This study demonstrates that birth defects prevalence varies by nativity within race/ethnic groups, with elevated prevalence ratios for some specific conditions and lower prevalence for others. More detailed analyses focusing on a broader range of maternal behaviors and characteristics are required to fully understand the implications of our findings.

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Author NameID

Kirby, Russell S.; ORCID: <http://orcid.org/0000-0002-3489-401X> Mai, Cara T.; ORCID: <http://orcid.org/0000-0002-7300-0672>

Institution

(Kirby) Department of Community and Family Health, College of Public Health, University of South Florida, Tampa, FL, United States (Mai, Isenburg) National Center on Birth Defects and Developmental Disabilities, Centers for Disease Control and Prevention, Atlanta, GA, United States

(Wingate) Department of Health Care Organization and Policy, School of Public Health, University of Alabama at Birmingham, Birmingham, AL, United States

(Janevic) Department of Obstetrics, Gynecology and Reproductive Science, Blavatnik Family Women's Health Research Institute, Icahn School of Medicine at Mount Sinai, New York, NY, United States

(Janevic) Department of Population Health Science and Policy, Blavatnik Family Women's Health Research Institute, Icahn School of Medicine at Mount Sinai, New York, NY, United States

(Copeland) Michigan Birth Defects Surveillance Program, Michigan Department of Health and Human Services, Lansing, MI, United States

(Flood) Arizona Birth Defects Monitoring Program, Arizona Department of Health Services, Phoenix, AZ, United States

(Canfield) Birth Defects Epidemiology and Surveillance Branch, Texas Department of State Health Services, Austin, TX, United States

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John Wiley and Sons Inc. (P.O.Box 18667, Newark NJ 07191-8667, United States)

Year of Publication

2019

219.

Foreskin reconstruction at the time of single-stage hypospadias repair: is it a safe procedure?.

Manuele R., Senni C., Patil K., Taghizadeh A., Garriboli M.

Embase

International Urology and Nephrology. 51 (2) (pp 187-191), 2019. Date of Publication: 03 Feb 2019.

[Review]

AN: 625325404

Introduction: Foreskin reconstruction (FR) is a recognised, yet debated, option for patients undergoing single-stage hypospadias repair (HR).

Method(s): We evaluated the incidence of complications after single-stage HR in our institution.

This is a retrospective review of all single-stage HR. Patients were classified into group 1 (circumcision) and group 2 foreskin reconstruction (FR). Urethroplasty and foreskin complications were recorded. Statistics used are as follows: Mann-Whitney test to compare age at operation and length of follow-up (FU); Chi-Square test to analyse the incidence of urethral complications and need for reoperation; Log rank test to compare the survival curves; p statistically significant < 0.05. Data are presented as median (range).

Result(s): 304 patients were identified, operated between January 2010 and December 2016, and 20 were excluded: 6 already circumcised at the time of the surgery, 3 with megameatus intact

prepuce, 11 lost at FU. 284 patients were included: 161 circumcised and 123 FR. Median age at the operation was 17 months (8-179) (group 1) and 17 months (8-148) (group 2) ($p = 0.71$). Length of FU was 19 months (8-91) (group 1) and 17 months (4-87) (group 2) ($p = 0.45$). The survival curve was homogeneous ($p = 0.28$). Urethroplasty complications occurred in 32/161 (20%) (group 1) and in 21/123 (17%) (group 2) ($p = 0.55$). Foreskin complications occurred in 18/123 (15%). A second operation was required in 33 boys in each group, (20% group 1 and 27% group 2) ($p = 0.21$).

Conclusion(s): FR does not increase the complication rate or the need for a reoperation after single-stage HR. Parents should be offered the option between the two procedures according to their personal preference.

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Author NameID

Manuele, Riccardo; ORCID: <http://orcid.org/0000-0002-2757-8481>

Institution

(Manuele, Senni, Patil, Taghizadeh, Garriboli) Paediatric Urology, Evelina London Children's Hospital, London, United Kingdom

Publisher

Springer Netherlands (E-mail: rbk@louisiana.edu)

Year of Publication

2019

220.

Single-stage repair of obliterated anterior urethral strictures using buccal mucosa graft and dorsal penile skin flap.

Kojovic V., Djordjevic M.L., Vuksanovic A.

Embase

International Journal of Urology. 26 (1) (pp 90-95), 2019. Date of Publication: January 2019.

[Article]

AN: 624272472

Objective: To present a single-stage repair of obliterative urethral strictures by simultaneous use of a buccal mucosa graft and longitudinal dorsal penile skin flap.

Method(s): Between February 2007 and October 2016, 51 patients with obliterative anterior urethral stricture underwent single-stage substitution urethroplasty. A buccal mucosa graft was harvested and fixed to the corpora cavernosa as the dorsal part of the neourethra, and a vascularized dorsal penile skin flap was created, transposed ventrally and sutured to the buccal mucosa graft to form ventral part of the neourethra.

Result(s): The follow-up period was 12-129 months (mean 49 months). The mean age of the patients was 48 years (range 15-71 years). The mean length of the obliterated urethral segment, measured during the operative procedure, was 5.2 cm. The etiology of strictures was: unknown, hypospadias and trauma in 19, 27 and five patients, respectively. Five patients were lost to follow up, and 46 patients were analyzed for the outcome. At the end of the follow-up period, recurrence of the stricture occurred in seven (15.2%) patients, whereas 39 (84.8%) patients did not develop stricture. An additional three (6.5%) patients developed fistula, resulting in overall successful voiding in 36 (78.3%) patients.

Conclusion(s): A combined buccal mucosa graft and longitudinal dorsal penile skin flap could be a good choice for one-stage substitution urethroplasty in complex obliterative urethral strictures, with an acceptable complication rate.

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Institution

(Kojovic, Djordjevic) Medical School, University of Belgrade, Belgrade, Serbia (Vuksanovic)

Clinic of Urology, Clinical Center of Serbia, Belgrade, Serbia

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2019

Comparison of Patient's Satisfaction and Long-term Results of 2 Penile Plication Techniques:
Lessons Learned From 387 Patients With Penile Curvature.

Cayan S., Asci R., Efesoy O., Kocamanoglu F., Akbay E., Yaman O.

Embase

Urology. 129 (pp 106-112), 2019. Date of Publication: July 2019.

[Article]

AN: 2001934272

Objective: To compare the patient's satisfaction and long-term results of 2 penile plication procedures in patients with penile curvature.

Method(s): This retrospective study included 387 patients with congenital penile curvature (n = 260) and Peyronie's disease (n = 127) who underwent surgical correction of penile curvature with penile plication procedures. Of the patients, 202 underwent plication of tunica albuginea with the Lue's 16-dot technique, while 185 underwent highly superficial excision of tunica albuginea with the modified Nesbit corporoplasty. Surgical outcomes and patient's satisfaction were compared between the 2 techniques in all patients.

Result(s): The mean duration of surgery was significantly shorter in the 16-dot plication technique (48.1 +/- 7.5 minutes), compared with the modified Nesbit corporoplasty (63 +/- 16.9 minutes) (P = .001). Complete penile straightening was achieved in 87.6% of the patients who underwent 16-dot plication technique and in 89.7% of the patients who underwent modified Nesbit plication, revealing no difference (P = .514). The rates of penile sensory loss (P = .001) and de-novo erectile dysfunction (P = .016) were significantly higher in the modified Nesbit corporoplasty than in the 16-dot plication technique, but rate of suture related complications was significantly higher in the 16-dot plication technique than in the modified Nesbit corporoplasty (P = .001). The patients with congenital penile curvature had significantly less ratio of postoperative penile length loss and de-novo erectile dysfunction than Peyronie's disease patients.

Conclusion(s): Overall, both surgical techniques have very high success and satisfaction rates with very low complication rates. However, the types of complications are significantly different between the 2 surgical procedures. Therefore, patients with penile curvature should be informed about outcomes of penile plication procedures, and surgical method should be preferred based on patient's preference and surgeon's experience.

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Status

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Institution

(Cayan, Akbay) University of Mersin School of Medicine, Department of Urology, Mersin, Turkey
(Asci, Kocamanoglu) Ondokuz Mayıs University School of Medicine, Department of Urology,
Samsun, Turkey

(Efesoy) Mersin City Training and Research Hospital, Department of Urology, Mersin, Turkey

(Yaman) Ankara University School of Medicine, Department of Urology, Ankara, Turkey

Publisher

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Year of Publication

2019

222.

A single-institution experience of complete primary repair of bladder exstrophy in girls: risk factors for urinary retention.

Sack B.S., Borer J.G.

Embase

Journal of Pediatric Urology. 15 (3) (pp 262.e1-262.e6), 2019. Date of Publication: May 2019.

[Article]

AN: 2001850144

Introduction: Historically after complete primary repair of exstrophy (CPRE) in girls, it had been more likely to observe urinary incontinence than retention. Following recent technical modifications of elongating the urethra and narrowing the bladder neck, the authors have observed a high rate of urinary retention in girls after CPRE.

Objective(s): The aim was to identify factors that may be responsible for this observation by reviewing historical and current outcomes. The authors hypothesized that differences in anatomic dimensions at the time of CPRE may contribute to urinary retention. Study design: A retrospective review of girls who underwent CPRE from December 1998 through September 2016 from a single institution was performed. Patients were deemed in retention if their clinical course was consistent with such, required a procedure to relieve urinary retention, and/or required clean intermittent catheterization.

Result(s): Nineteen girls underwent CPRE during this period. In 2012, a change to delaying CPRE to approximately 2 months of age was made, and this led the authors to divide their experience into CPRE performed as a newborn (<72 h of age, 8 patients) versus delayed (>72 h,

11 patients) subgroups. There were no girls with retention in the newborn group and three (38%) girls with retention in the delayed group. In the delayed group, girls had a longer urethral plate and narrower bladder neck compared with the newborn group. Long-term outcomes greater than 9 years are available for six girls in the newborn group and two (33%) required bladder neck procedures for incontinence. None in the delayed group have required incontinence procedures; however, follow-up is limited at 25 months.

Discussion(s): The absence of retention in the newborn group is concerning for the delayed group incurring a higher risk of retention after CPRE. This may be secondary to excessive compression of the urethra at the time of pubic symphysis approximation potentially leading to urethral ischemia. Different from the newborn CPRE girls, additional technical revision of CPRE, namely, elongation of the urethra and the dissection it involves and narrowing of the bladder neck, may increase the risk for retention.

Conclusion(s): The multiple factors that were identified as potential contributors to post-CPRE urinary retention should result in a cautious reevaluation of female bladder exstrophy management at the time of CPRE. The authors now create a gradual tapered transition at the bladder neck and, similar to their previous experience, a more generous (wider) bladder neck and a shorter length for urethral plate. [Table presented]

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Status

Embase

Institution

(Sack, Borer) The Department of Urology, Boston Children's Hospital, 300 Longwood Avenue, Boston, MA 02115, United States

Publisher

Elsevier Ltd

Year of Publication

2019

Parental home removal of urethral catheters after urological surgery-a prospective benchmarking study.

Braungart S., Goyal A.

Embase

Journal of Pediatric Urology. 15 (3) (pp 252.e1-252.e4), 2019. Date of Publication: May 2019.

[Article]

AN: 2001827587

Introduction: Many urological operations require placement of a urethral Foley catheter. The catheter often needs to remain in situ for a period of time after discharge; and patients subsequently require either a further hospital admission or community nurse review for catheter removal. Parents can easily remove the catheter at home by cutting the balloon port. This disrupts the valve and hence deflates the retaining balloon, thereby facilitating spontaneous passage of the catheter. The authors introduced this practice to their institution.

Aim(s): The aim was to assess safety and success of parental home catheter removal.

Method(s): A prospective data study was performed in a large pediatric urology center over a 12-month time period. Patients <16 years after single-stage hypospadias repair or other penile surgery were included on a voluntary basis. Parents of eligible patients were instructed verbally and with an information leaflet, including date for removal. Telephone follow-up after removal was undertaken to assess the outcome.

Result(s): Thirty-eight patients were included over a 12-month time period. Patient age ranged from 9 months to 12 years (median age 2.5 years). The majority (82%) of patients had required a catheter after hypospadias repair. Home catheter removal was successful in 92% cases. Three children required professional support for catheter removal. Median time until catheter passage was 3 h (range 0-24 h). Considering that cost for day case admission for catheter removal averages at 130 per patient, home catheter removal saved the NHS 4550 in the time period.

Discussion(s): This is the first study to report the safety and feasibility of parental home catheter removal by cutting the balloon port valve in the pediatric population. It offers a number of distinct advantages compared with traditional methods for removal. These include, namely, (i) positive patient experience: catheter removal in a familiar environment by a relative minimizes stressful experiences for the family; (ii) minimal trauma to healing tissues through spontaneous catheter passage; and (iii) health care-related cost savings. This was an initial benchmarking study, so patient numbers were relatively small. Nevertheless, it shows that the method is safe and received positive parental feedback.

Conclusion(s): Parental home removal of a urethral catheter is a feasible and safe alternative to catheter removal by a health-care professional. It minimizes parental anxiety and inconvenience related to the catheter removal appointment and allows for significant cost savings.[Figure presented]

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Institution

(Braungart, Goyal) Department of Paediatric Surgery and Urology, Royal Manchester Children's Hospital, Manchester University NHS Foundation Trust, Manchester, United Kingdom

Publisher

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2019

224.

Long-term follow-up of urethral reconstruction for blunt urethral injury at a young age: urinary and sexual quality of life outcomes.

Baradaran N., McAninch J.W., Copp H.L., Quanstrom K., Breyer B.N., Hampson L.A.

Embase

Journal of Pediatric Urology. 15 (3) (pp 224.e1-224.e6), 2019. Date of Publication: May 2019.

[Article]

AN: 2001783888

Introduction: Little is known about long-term patient-reported outcomes following surgical repair for pediatric blunt urethral trauma.

Objective(s): The purpose was to evaluate long-term urinary outcomes, sexual function, and quality of life (QOL) of patients who undergo urethroplasty for blunt urethral trauma in childhood.

Study Design: After IRB approval, we retrospectively reviewed the records of patients who sustained blunt urethral injury at ≤ 18 years and underwent urethroplasty at our institution between 1978 and 2013. We then used a web-based survey to assess urinary/sexual/ejaculatory function and overall QOL using validated questionnaires.

Result(s): Of 68 eligible patients, 15 were able to be contacted (table). Median age of injury, age at urethroplasty, and age at follow-up were 17 (4-18), 17 (5-20), and 19 (13.5-21.5) years, respectively. The stricture was membranoprostatic in eight and bulbar in seven patients, with

median length of 2 (1.6-2.6) cm. Excision/primary anastomosis was performed in all but three patients who required a buccal graft. Overall, 80% were 'very satisfied' and 20% were 'satisfied' with surgery. One patient reported a subsequent urethral intervention. On urethral stricture surgery patient-reported outcome measurement, the median bother (0 least, 24 worst) was 10 (8-12.5). The force of urine stream (1 strongest, 4 weakest) was 2 (1.5-2), with no report of urinary incontinence. The median Sexual Health Inventory for Men score (0 worst, 25 perfect) was 24 (22.5-24). The median ejaculatory function score (0 worst, 15 normal) was 14 (13-14.75). Six patients had fathered children and none reported infertility. Three patients reported <30degree penile curvature not interfering with sex. Median QOL (0 worse, 10 best) was 8 (7.5-8).

Conclusion(s): Urethroplasty after blunt urethral injury in young adult population is associated with a high long-term success rate with a low rate of long-term urinary and sexual consequences in adulthood. [Table presented]

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Author NameID

Baradaran N.; ORCID: <http://orcid.org/0000-0003-4332-8017>

Publisher

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Year of Publication

2019

225.

Lifelong Congenital Urology: The Challenges for Patients and Surgeons.

Wood D., Baird A., Carmignani L., De Win G., Hoebeke P., Holmdahl G., Manzoni G., Nijman R.J.M., Taylor C., Tekgul S.

Embase

European Urology. 75 (6) (pp 1001-1007), 2019. Date of Publication: June 2019.

[Review]

AN: 2001744733

Context: Patients born with complex congenital genitourinary anomalies (including bladder exstrophy, cloacal exstrophy, epispadias, neurogenic bladder, hypospadias and posterior urethral valves) often require major reconstructive surgery in childhood. These conditions, their treatment and sequelae require lifelong follow-up. This has created the need for adult urologists to provide care as these patients grow into adults.

Objective(s): To evaluate current strategies for transition and provide a current position statement with examples of the challenges faced by patients and their health care teams as a result of these conditions and their treatment.

Evidence Acquisition: Each of the authors was asked to provide a 500-word synthesis, based on current literature; to highlight the challenges faced in an area of their expertise.

Evidence Synthesis: The authors assembled in March 2018 to form a consensus based on the data gathered. The aforementioned sections were reviewed and following the consensus discussion the paper was formulated and reviewed.

Conclusion(s): Lifelong care of congenital problems is challenging and essential for many but not all. Expertise is needed to provide the best care for patients and make the best use of resources. Specialist centres appear to be the most effective and safe model. In the long term it would be ideal to establish an evidence base focused on the common long-term problems with these conditions to ensure excellent care with appropriate expertise.

Patient Summary: Patients born with complex congenital anomalies of the genitourinary system require specialist care in childhood. Many will need lifelong care to manage their condition and the treatment of it. There is growing interest in this area of medicine and this consensus statement addresses the need for lifelong care in this group. The aim is to ensure that all patients that need care at any age are able to find what they need. Congenital urology is a fascinating field involving the lifelong care of patients with congenital anomalies. We provide a current position statement from which a network of interested practitioners will be able to build further.

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PMID

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Status

Embase

Institution

(Wood, De Win) University College London Hospitals, London, United Kingdom (Baird)

University Hospital Aintree and Alder Hey NHS Foundation Trusts, Liverpool, United Kingdom

(Carmignani) Università degli Studi di Milano, Milan, Italy

(De Win) University of Antwerp, Antwerp, Belgium

(Hoebeke) Ghent University Hospital, Gent, Belgium

(Holmdahl) Queen Silvia's Children's Hospital, Gothenburg, Sweden

(Manzoni) Fondazione IRCCS Ca Granda, Ospedale Maggiore Policlinico, Milan, Italy
(Nijman) University Groningen, University Medical Centre Groningen, Groningen, Netherlands
(Taylor) Guys and St Thomas's Hospital, London, United Kingdom
(Tekgul) Hacettepe University, Ankara, Turkey
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Year of Publication
2019

226.

Spinal anesthesia in children: most pediatric urologists are not on board.

Rehfuss A., Bogaert G., Kogan B.A.

Embase

Journal of Pediatric Urology. 15 (3) (pp 263.e1-263.e5), 2019. Date of Publication: May 2019.

[Article]

AN: 2001741500

Objective: In 2016, the Food and Drug Administration issued a warning on general anesthetic medications used for lengthy procedures (>3 h) in children younger than 3 years. Spinal anesthesia can be a safe alternative to general anesthesia for many pediatric urology procedures. It can shorten total operating room (OR) time, provide excellent pain control, and allow parents to reunite with their child immediately after surgery. However, use of spinal anesthesia can also directly affect the operating surgeon (awake patient, time constraints of spinal, and prolonged preoperative time). Members of the Societies for Pediatric Urology (SPU) and European Society of Pediatric Urology (ESPU) were surveyed to get their opinions on the use of spinal anesthesia for routine pediatric urology procedures. It was hypothesized that half of pediatric urologists would favor spinal anesthesia and that SPU members would be more likely to favor spinal anesthesia than their European colleagues.

Material(s) and Method(s): A short survey with five clinical scenarios was created. Scenarios assessed physicians' recommendations regarding timing and the type of anesthesia (general or spinal) for common pediatric urology procedures: undescended testicle, inguinal hernia, hypospadias, phimosis, and phimosis with penoscrotal webbing. Surveys were emailed to

members of the SPU and ESPU. Responses and demographic information were collected and analyzed.

Result(s): The survey was completed by 113 SPU members (46% response rate for members who opened the invitation) and 109 ESPU members. For all clinical scenarios, < 20% of pediatric urologists from the SPU and <25% from the ESPU favor doing any procedure with spinal anesthesia. The majority of respondents practice in children's hospitals with pediatric anesthesiologists, but roughly half of the responders (54% SPU and 43% ESPU) do not think their anesthesia colleagues would be comfortable performing spinal anesthesia. Furthermore, only 51% of SPU and 36% of ESPU members discuss the possible neurodevelopmental side-effects of anesthesia with parents; similarly, less than half of all respondents think their anesthesia colleagues address these potential side-effects when obtaining consent. The only significant difference between SPU and ESPU responses was that ESPU members tended to delay penile surgery more than SPU respondents.

Conclusion(s): Whether general anesthesia has any effect on the developing brain of children undergoing routine pediatric urology procedures is unclear. Yet, few pediatric urologists, independent of their region of practice, prefer spinal to general anesthesia. Collaboration in the OR is the key to success, and it is important that pediatric urologists and pediatric anesthesiologists work together to balance the benefits and risks of general and spinal anesthesia. [Table presented]

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Institution

(Rehfuß, Kogan) Albany Medical College, United States (Bogaert) University Hospitals Leuven, United States

Publisher

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Year of Publication

2019

Association Between Exstrophy-epispadias Complex And Congenital Anomalies: A German Multicenter Study.

Ebert A.-K., Zwink N., Jenetzky E., Stein R., Boemers T.M., Lacher M., Fortmann C., Obermayr F., Fisch M., Mortazawi K., Schmiedeke E., Eisenschmidt V., Schafer M., Hirsch K., Rosch W.H., Reutter H.

Embase

Urology. 123 (pp 210-220), 2019. Date of Publication: January 2019.

[Article]

AN: 2001274277

Objective: To further investigate associated anomalies in exstrophy-epispadias complex (EEC) patients congenital uro-rectal malformations network (CURE-Net) database was systematically screened. In literature the EEC comprises a spectrum of anomalies, mainly occurring "isolated" without additional congenital defects. Nevertheless, previous epidemiological studies indicated a higher association with renal, anorectal, and lower neurotubular anomalies, which may originate from the same developmental morphogenetic fields.

Material(s) and Method(s): Seventy-three prospectively (born since 2009) and 162 cross-sectional recruited EEC patients (born 1948-2008) were analyzed. Associated anomalies were derived from patient's medical data as well as from a physical examination during a physician's interview, classified according to the international statistical classification of diseases and related health problems and grouped with the London Dysmorphology Database. Descriptive statistical analyses were performed.

Result(s): Majority of participants were male (68%) and expressed the classical bladder exstrophy phenotype (71%). Exstrophy variants occurred significantly more often in newborns (21%, $P < .0001$). Anomalies such as inguinal hernias, skeleton, and joint anomalies were equally present in both groups ($P = .65$ and $P = .67$). Heart defects were seen more often in newborns (6%) than in the cross-sectional group (1%; $P = .033$) and the general German population (1%). In total, 59% of the prospective and 48% of the cross-sectional patients had associated anomalies outside the spectrum ($P = .16$).

Conclusion(s): Phenomenological multicenter data confirmed the dimension of associated anomalies inside and outside the EEC spectrum. The detected anomalies are either important in preparing for the primary reconstruction or later in long-term follow-up. Associated anomalies of EEC should be spotlighted during routine check-up in all EEC patients.

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Status

Embase

Institution

(Ebert) Department of Pediatric Urology, University Hospital for Urology and Pediatric Urology

Ulm, Ulm, Germany (Zwink, Jenetzky) Department of Child and Adolescent Psychiatry,

University Medical Center of the Johannes Gutenberg University Mainz, Mainz, Germany

(Jenetzky) Child Center Maulbronn GmbH, Hospital for Pediatric Neurology and Social Pediatrics,
Maulbronn, Germany

(Stein) Department of Pediatric and Adolescent Urology, University of Mannheim, Mannheim,
Germany

(Boemers) Department of Pediatric Surgery and Urology, University Hospital Cologne, Cologne,
Germany

(Lacher) Department of Pediatric Surgery, University of Leipzig, Leipzig, Germany

(Fortmann) Center of Pediatric Surgery Hannover, Hannover Medical School and Bult Children's
Hospital, Hannover, Germany

(Obermayr) Department of Pediatric Surgery and Urology, University Hospital for Child and
Adolescent Medicine Tübingen, Tübingen, Germany

(Fisch) Department of Pediatric Urology, University Hospital Hamburg-Eppendorf, Hamburg,
Germany

(Mortazawi) Department of Pediatric Surgery, Städtisches Klinikum Karlsruhe, Karlsruhe,
Germany

(Schmiedeke) Department of Pediatric Surgery and Urology, Center for Child and Youth Health,
Klinikum Bremen-Mitte, Bremen, Germany

(Eisenschmidt, Rosch) Department of Pediatric Urology, University Medical Center Regensburg,
Regensburg, Germany

(Schafer) Department of Pediatric Surgery and Urology, Cnopf'sche Kinderklinik, Nürnberg,
Germany

(Hirsch) Department of Pediatric Urology, University Hospital Erlangen, Erlangen, Germany

(Reutter) Institute of Human Genetics, University of Bonn, Bonn, Germany

(Reutter) Department of Neonatology and Pediatric Intensive Care, Children's Hospital, University
of Bonn, Bonn, Germany

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Year of Publication

2019

228.

Complications of proximal hypospadias repair with transverse preputial island flap urethroplasty:
A 15-year experience with long-term follow-up.

Wang C.-X., Zhang W.-P., Song H.-C.

Embase

Asian Journal of Andrology. 21 (3) (pp 300-303), 2019. Date of Publication: 01 May 2019.

[Article]

AN: 627396539

There is still debate regarding the optimal surgical approach for proximal hypospadias. This retrospective study aims to evaluate the long-term outcomes using transverse preputial island flap urethroplasty. A total of 320 patients were included, with a mean follow-up of 40.2 months (range: 1-156 months). Complications were encountered in 125 patients (39.1%), including fistulas in 53 (16.6%), urethral strictures in 31 (9.7%), and diverticula in 41 (12.8%). The mean timing of presentation with a complication was 15.8 months (median: 1.7, range: 1-145), of which 79.2% were early complications and 20.8% were late complications. In all, 20.8% of the patients with complications presented after ≥ 1 year, and 12.8% presented after ≥ 5 years. Univariate analysis revealed that age at the time of surgery, flap length, and location of the urethral meatus were not correlated with complications. A stricture was present in 31.7% (13/41) of those with diverticula ($P < 0.001$), while late urethral diverticula were accompanied by urethral strictures in 11.1% (1/9) of cases ($P = 0.213$). These results indicate that transverse preputial island flap urethroplasty still has a high incidence of complications, even when performed by highly experienced physicians. Most complications of hypospadias are diagnosed within 1 year postoperatively, while fistulas and urinary strictures generally occur within 2 months and diverticula tend to be present by 1 year.

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Status

Embase

Institution

(Wang, Zhang, Song) Department of Urology, Beijing Children's Hospital Affiliated to Capital Medical University, Beijing 100045, China

Publisher

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Year of Publication

2019

229.

Preoperative illnesses in children do not increase the risk of complications after hypospadias repair.

Dokter E.M.J., Slikboer K.M.A., Van Der Zanden L.F.M., Rahamat-Langendoen J.C., Henriët S.S.V., Feitz W.F.J., Kortmann B.B.M., Roeleveld N., Van Rooij I.A.L.M.

Embase

Pediatric Infectious Disease Journal. 38 (2) (pp 104-109), 2019. Date of Publication: 01 Feb 2019.

[Article]

AN: 627371384

Background: Preoperative illnesses might induce immunosuppression and subsequently increase morbidity after surgery. Several studies have tried to identify risk factors for complications after hypospadias correction, but effects of illnesses in the weeks just before surgery are unknown. We aimed to determine the associations between preoperative illnesses not severe enough to postpone surgery and short-term complications after hypospadias repair in children.

Method(s): In this retrospective cohort study, data were collected from 681 children with anterior or middle type hypospadias that had initial 1-stage repair in the period 1983-2012 in the Radboudumc, The Netherlands. The associations between common illnesses, such as common cold, fever and ear infection, within 2 weeks before repair, and postoperative complications, such as urethrocuteaneous fistula, wound dehiscence and stenosis, within 2 months and 1 year after surgery, were analyzed using multivariable logistic regression analyses.

Result(s): Of the 681 boys, 22% had preoperative illnesses, most often common cold, and 14% had postoperative complications. Children with preoperative illnesses had fewer postoperative complications within 2 months ($n = 13$, 9%) than children without preoperative illnesses ($n = 79$, 16%), resulting in a 50% risk reduction (odds ratio: 0.49; 95% confidence interval: 0.26-0.93).

Preoperative infections (common cold, fever and ear infection), in particular, reduced the risk of

postoperative infections (wound and urinary tract infections; odds ratio: 0.37; 95% confidence interval: 0.14-0.98). Results were similar for complications within 1 year.

Conclusion(s): Common preoperative illnesses not severe enough to postpone surgery did not increase the postoperative complication risk and even seemed to have a protective effect, especially for postoperative infections. Consequently, there is no reason to alter preoperative screening.

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Institution

(Dokter, Van Der Zanden, Roeleveld, Van Rooij) Radboud Institute for Health Sciences, Department for Health Evidence, Radboud University Medical Center, 133, P.O. Box 9101, Nijmegen, HB 6500, Netherlands (Slikboer, Feitz, Kortmann) Department of Urology, Pediatric Urology, Radboudumc Amalia Children's Hospital, Nijmegen, Netherlands (Rahamat-Langendoen) Radboud Institute for Molecular Life Sciences, Department of Medical Microbiology, Radboud University Medical Center, Nijmegen, Netherlands (Henriet) Department of Pediatric Infectious Diseases and Immunology, Radboudumc Amalia Children's Hospital, Nijmegen, Netherlands

Publisher

Lippincott Williams and Wilkins (E-mail: kathiest.clai@apta.org)

Year of Publication

2019

230.

Klinefelter syndrome: From pediatrics to geriatrics.

Shiraishi K., Matsuyama H.

Embase

Reproductive Medicine and Biology. 18 (2) (pp 140-150), 2019. Date of Publication: April 2019.

[Review]

AN: 627063421

Background: Klinefelter syndrome (KS) is one of the major causes of nonobstructive azoospermia (NOA). Microdissection testicular sperm extraction (micro-TESE) is often performed to retrieve sperm. Infertility specialists have to care for KS patients on a lifelong basis.

Method(s): Based on a literature review and our own experience, male infertility treatment and KS pathophysiology were considered on a lifelong basis. Main findings: Patients diagnosed early often have an increased number of aberrant X chromosomes. Cryptorchidism and hypospadias are often found, and surgical correction is required. Cryopreservation of testicular sperm during adolescence is an issue of debate because the sperm retrieval rate (SRR) in KS patients decreases with age. The SRR in adult KS patients is higher than that in other patients with NOA; however, low testosterone levels after micro-TESE will lower the general health and quality of life. KS men face a number of comorbidities, such as malignancies, metabolic syndrome, diabetes, cardiovascular disease, bone disease, and immune diseases, which ultimately results in increased mortality rates.

Conclusion(s): A deeper understanding of the pathophysiology of KS and the histories of KS patients before they seek infertility treatment, during which discussions with multidisciplinary teams are sometimes needed, will help to properly treat these patients.

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Embase

Author NameID

Shiraishi, Koji; ORCID: <http://orcid.org/0000-0002-3278-7747>

Institution

(Shiraishi, Matsuyama) Department of Urology, Yamaguchi University School of Medicine, Ube, Japan

Publisher

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E-mail: vgorayska@wiley.com)

Year of Publication

2019

Use of a Stent in Distal Hypospadias Repaired by Tubularized Incised Plate Urethroplasty: A Comparative Study.

Karakaya A.E., Dogan A.B., Guler A.G.

Embase

Urologia Internationalis. 102 (3) (pp 336-340), 2019. Date of Publication: 01 Apr 2019.

[Article]

AN: 626329770

Purpose: We present our experience of stented and unstented distal hypospadias repaired by tubularized incised plate urethroplasty (TIPU).

Patients and Methods: Data of 84 patients who were operated by TIPU method in 2 hospitals were retrospectively analyzed, and they were invited to be included in the study. Sixty-six patients agreed to participate in the study and were divided into 2 groups. Group 1 consisted of 38 boys operated in Sutcu Imam University Hospital on using a stent between 2015 and 2017. Group 2 consisted of 28 boys operated in Private Hospital of Megapark on without a stent between 2016 and 2017. The hypospadias objective scoring evaluation (HOSE) scale was used to compare the groups.

Result(s): The median age was 2.81 years in group 1 and 1.95 years in group 2 (*p* = 0.243). The mean follow-up duration was 25.74 +/- 4.62 and 24.5 +/- 4.19 months in groups 1 and 2 respectively (*p* = 0.268). The HOSE scores were similar in both groups with comparable results (*p* = 0.622).

Conclusion(s): No difference was observed between the groups in this study, regarding functional and cosmetic outcomes according to the HOSE score. However, the results should be supported by prospective studies with a sufficient number of patients.

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Embase

Institution

(Karakaya) Department of Pediatric Surgery, Private Hospital of Megapark, Kahramanmaras, Turkey (Dogan, Guler) Department of Pediatric Surgery, School of Medicine, Sutcu Imam University, Kahramanmaras, Onikisubat TR-46100, Turkey

Publisher

S. Karger AG

Year of Publication

2019

232.

MiR-145-modulated SOX9-mediated hypospadias through acting on mitogen-activated protein kinase signaling pathway.

Shang Y., Kang Y., Sun J., Wei P., Yang J., Zhang H.

Embase

Journal of Cellular Physiology. 234 (7) (pp 10397-10410), 2019. Date of Publication: July 2019.

[Article]

AN: 625588955

This study primarily explored how miR-145, mitogen-activated protein kinase (MAPK) signaling and a downstream transcription factor (i.e., SOX9) mediated development of hypospadias. The hypospadias tissues and preputial tissues were isolated from pediatric inpatients postoperatively. Simultaneously, the rat models of hypospadias were established, and spermatogonial stem cells were separated. The expressions of proteins that symbolized cell apoptosis and oxidative stress were quantified via western blot analysis. Furthermore, the apoptosis, proliferation, and viability of cells were evaluated by means of flow cytometry, 3-(4,5-dimethylthiazol-2-yl)-2,5-diphenyltetrazolium bromide (MTT) and colony formation assays. The results of microarray indicated miR-145 as a differentially expressed biomarker between hypospadias tissues and normal tissues ($p < 0.05$). Moreover, rat models of hypospadias were observed with markedly lower vitamins A and E levels, reduced expressions of proteins relevant to oxidative stress (i.e., Nrf2, HO-1, Gpx, and SOD-1), as well as enhanced Bax and cleaved caspase-3 expressions ($p < 0.05$). Furthermore, SOX9 was found to be targeted by miR-145, and it was also modified by phosphorylated extracellular-regulated kinase (p-ERK), a portion of MAPK signaling ($p < 0.05$). The p-ERK was significantly regulated after altering the expression of miR-145 ($p < 0.05$). Moreover, activation of p-ERK and transfection of pcDNA-SOX9 could cause higher expression of apoptins and larger apoptotic proportion of cells ($p < 0.05$), yet transfection of miR-145 mimic led to improved cell apoptosis and depressed cell viability ($p < 0.05$). In conclusion, SOX9, which was regulated by both miR-145 and miR-145/MAPK signaling, could be involved in the pathogenesis of hypospadias.

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Author NameID

Shang, Yafeng; ORCID: <http://orcid.org/0000-0002-8238-8787>

Institution

(Shang, Kang, Sun, Wei, Yang, Zhang) Department of Urology, Luoyang Central Hospital
Affiliated to Zhengzhou University, Luoyang, China

Publisher

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Year of Publication

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233.

Penile urethral stricture disease.

Spilotros M., Venn S., Anderson P., Greenwell T.

Embase

Journal of Clinical Urology. 12 (2) (pp 145-157), 2019. Date of Publication: 01 Mar 2019.

[Review]

AN: 622684001

Patients affected by a urethral stricture account for a considerable cost to all healthcare systems. The estimated prevalence of all urethral stricture in the UK is 10/100,000 men during youth, increasing to about 40/100,000 by age 65 years and to more than 100/100,000 thereafter. A penile urethral stricture is a narrowing of the lumen of the urethra due to ischaemic fibrosis of the urethral epithelium and/or spongiofibrosis of the corpus spongiosum occurring within the penile urethra. Its aetiology is largely idiopathic but other important causes are failed hypospadias repair and lichen sclerosus, which account for 60% of all cases. Strictures of the anterior urethra account for 92% of cases: bulbar strictures are more frequent (46.9%), followed by penile (30.5%) and combined bulbar/penile (9.9%), that is, 40.4% of all men presenting with stricture will have a penile urethral stricture alone or in combination with a bulbar urethral stricture. There are several options for the treatment of penile urethral strictures ranging from less invasive treatments, including urethral dilatation and direct vision internal urethrotomy, to more complex augmentation graft and flap urethroplasty. The aim of the present review is to describe the aetiology and

epidemiology of anterior urethral strictures and the available options reported in literature for their treatment.

Level of Evidence: 1a.

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Status

Embase

Author NameID

Greenwell, Tamsin; ORCID: <http://orcid.org/0000-0002-9074-7604>

Institution

(Spilotros, Greenwell) Department of Urology, University College London Hospital, United Kingdom
(Spilotros, Venn) Department of Urology, Western Sussex Hospitals Foundation Trust and Portsmouth Hospital, United Kingdom

(Spilotros, Anderson) Department of Urology, West Midlands Hospital, United Kingdom

(Spilotros) Department of Emergency and Organ Transplantation, University of Bari, Italy

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Year of Publication

2019

234.

Foreskin and penile problems in childhood.

Abbas T., McCarthy L.

Embase

Surgery (United Kingdom). 37 (4) (pp 231-236), 2019. Date of Publication: April 2019.

[Review]

AN: 2001661161

This article outlines the embryology, natural history and management of different conditions of the foreskin and penis in children. Although the classification of hypospadias is included, the management of this condition is not. Epispadias is not covered at all.

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Institution

(Abbas, McCarthy) Hamad General Hospital, Doha, Qatar; Birmingham Children's Hospital,
Birmingham, UK

Publisher

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235.

Meeting the challenges of reconstructive urology - Where are we now?.

Cuckow P.M., Cao K.X.

Embase

Journal of Pediatric Surgery. 54 (2) (pp 223-228), 2019. Date of Publication: February 2019.

[Conference Paper]

AN: 2001357220

This is based on the 2018 Storz Urology Lecture at the BAPS Conference and is a personal review of three reconstructive paediatric urological conditions: hypospadias, congenital adrenal hyperplasia, and bladder exstrophy from the perspective of changing expectations and outcomes.

Level of Evidence: V (Expert Opinion)

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Status

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Institution

(Cuckow, Cao) Department of Paediatric Urology, Great Ormond Street Hospital for Children,
Great Ormond Street, London, United Kingdom

Publisher

W.B. Saunders

Year of Publication

2019

236.

An evaluation of the readability, quality, and accuracy of online health information regarding the treatment of hypospadias.

Cisu T.I., Mingin G.C., Baskin L.S.

Embase

Journal of Pediatric Urology. 15 (1) (pp 40.e1-40.e6), 2019. Date of Publication: February 2019.
[Article]

AN: 2001287147

Background: Hypospadias is one of the most common genital anomalies. Treatment of hypospadias requires surgical repair, usually in childhood. Patients are increasingly using the internet to learn more about their health or that of their children, which can often empower patients to make well-informed healthcare decisions.

Objective(s): The objective of this study was to evaluate not only the readability but also the quality and accuracy of available online health information for the treatment of hypospadias.

Study design: Search terms for hypospadias treatment were queried on major search engines. Each website was classified into one of four categories: institutional, commercial, charitable organization, or personal website. Content on each website discussing treatment options was analyzed for readability using three readability formulas. A validated tool, the DISCERN instrument, was used to measure the quality of online health information regarding hypospadias treatment. Accuracy was independently assessed by two pediatric urologists on a 1-5 scale, in which 1 and 5 correspond to 0% and 100% of the information in the text being accurate, respectively.

Result(s): A total of 150 search engine results were acquired, of which 46 were analyzed for readability, quality, and accuracy. The mean readability scores across all websites were 14.89 (Gunning-Fog), 11.01 Simple Measure of Goddledygook (SMOG), and 8.44 (Dale-Chall), which correspond to an 11th- to 12th-grade reading level. Most websites (65.2%) were considered of 'good' quality. Readability and quality scores were not statistically different between website categories. Institutional and charitable websites had the highest mean accuracy scores (3.91 and 3.50, respectively), with institutional websites proving to have significantly more accurate information regarding hypospadias treatment than commercial websites (3.91 and 3.42, respectively; $P = 0.001$).

Discussion(s): Pediatric urologists should know what information about hypospadias and its treatment exists on the Internet and understand if it is accurate and of good quality and, more importantly, if the material is written at a reading level comprehensible by the majority of parents. Limitations included analysis of only English-written websites regarding hypospadias treatment specifically, using search engines alone rather than other online resources, not evaluating online videos or illustrations, and not using more than two pediatric urologists for determining content accuracy.

Conclusion(s): This study demonstrates that online health materials regarding hypospadias and its treatment are written at a level far greater than the reading level of most adults. Most websites were considered of adequate quality, and websites from institutions or references had significantly more accurate information than those from commercial websites.[Figure presented]

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Status

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Author NameID

Cisu T.I.; ORCID: <http://orcid.org/0000-0002-3579-8476>

Institution

(Cisu, Mingin) Division of Urology, University of Vermont College of Medicine, 89 Beaumont Ave, Burlington, VT 05405, United States (Baskin) Department of Urology, UCSF Benioff Children's Hospitals, 1825 Fourth St, 5th Floor, San Francisco, CA 94143, United States

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237.

Modified staged repair of bladder exstrophy: a strategy to prevent penile ischemia while maintaining advantage of the complete primary repair of bladder exstrophy.

Chua M.E., Ming J.M., Fernandez N., Varghese A., Farhat W.A., Bagli D.J., Lorenzo A.J., Pippi Salle J.L.

Embase

Journal of Pediatric Urology. 15 (1) (pp 63.e1-63.e7), 2019. Date of Publication: February 2019.

[Article]

AN: 2001276991

Introduction: Penile ischemic injury is a reported catastrophic complication after complete primary repair of exstrophy (CPRE). Aiming to improve the bladder exstrophy-epispadias repair outcomes, the study institution adopted a modified staged exstrophy repair to incorporate the advantages of CPRE by avoiding concurrent epispadias repair and adding bilateral ureteral re-implantation and bladder neck tailoring (staged repair of bladder exstrophy with bilateral ureteral re-implantation [SRBE-BUR]) at the initial repair. It was hypothesized that such modifications minimize penile complications and prevent upper tract deterioration while enhancing bladder resistance and consequent capacity. Here, a comparative series of outcomes between CPRE and SRBE-BUR is reported.

Method(s): A retrospective cohort study including all exstrophy-epispadias male neonates managed in the study institution from January 2000 to December 2014 was performed. Patients were divided into those who underwent CPRE-BUR (group 1) and SRBE-BUR (group 2) (Figure). Baseline characteristics, peri-operative data, and long-term surgical outcomes were collected and analyzed for between-group comparison. Fisher exact and Mann-Whitney U tests were performed for statistical analysis.

Result(s): A total of 21 eligible patients were included: 10 in group 1 and 11 in group 2. Baseline characteristics were comparable. Two patients in group 1 had intra-operative penile ischemic injury (one with subsequent penile tissue loss), whereas none of the group 2 patients had intra-operative complications. No significant difference between the groups was noted for operative time; however, significantly lesser blood loss was noted in group 2. Comparable long-term surgical outcomes such as additional surgical intervention, urinary continence, bladder capacity, vesicoureteral reflux, hydronephrosis and recurrent urinary tract infections (UTIs) were noted. In addition, although subjective, better penile length and cosmesis were achieved by staging the repair (Figure).

Conclusion(s): The SRBE with bilateral ureteral re-implantation is a safe alternative for the repair of the exstrophy-epispadias repair as it prevents the catastrophic complication of penile tissue loss, while having comparable long-term outcomes with the CPRE. Delaying epispadias repair avoids penile injury besides possible improvement of its overall cosmesis.[Figure presented]

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Author NameID

Pippi Salle J.L.; ORCID: <http://orcid.org/0000-0002-0200-3884>

Institution

(Chua, Ming, Fernandez, Varghese, Farhat, Bagli, Lorenzo) Division of Urology, The Hospital for Sick Children, Toronto, Canada (Farhat, Bagli, Lorenzo) Department of Urology, University of Toronto, Toronto, Canada

(Pippi Salle) Division of Urology, Department of Surgery, Sidra Medical and Research Center, Doha, Qatar

Publisher

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Year of Publication

2019

238.

Predictors of a successful primary bladder closure in cloacal exstrophy: A multivariable analysis. Jayman J., Tourchi A., Feng Z., Trock B.J., Maruf M., Benz K., Kasprenski M., Baumgartner T., Friedlander D., Sponseller P., Gearhart J.

Embase

Journal of Pediatric Surgery. 54 (3) (pp 491-494), 2019. Date of Publication: March 2019.

[Article]

AN: 2000954334

Purpose: To investigate the factors affecting primary bladder closure in cloacal exstrophy (CE). A successful primary closure is important for optimizing reconstructive outcomes, and it is a critical first-step in the reconstruction of CE. The authors' hypothesize that a smaller diastasis and use of an osteotomy are independent predictors of a successful closure.

Method(s): A prospectively maintained database of 1332 exstrophy-epispadias complex (EEC) patients was reviewed for CE patients closed between 1975 and 2015. Univariate and multivariable analyses were performed to identify significant factors associated with CE primary bladder closure.

Result(s): Of 143 CE patients identified, 99 patients met inclusion criteria. Median follow-up time was 8.82 [IQR 5.43-14.26] years. In the multivariable model, the odds of having a successful closure are about 4 times greater for the staged cloacal approach compared to the 1-stage

approach (OR, 3.7; 95% CI 1.2-11.5; p-value = 0.023). Also, having an osteotomy increases the chance of a successful closure by almost six-fold (OR, 5.8; 95% CI 1.7-19.6; p-value = 0.004).

Conclusion(s): Using the staged approach with a pelvic osteotomy is paramount to a successful primary closure in CE. The authors strongly recommend using the staged approach and osteotomy as these factors independently increase the chance for a successful primary bladder closure. Study Type: Therapeutic study.

Level of Evidence: Level III, Retrospective comparative study.

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PMID

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Status

Embase

Institution

(Jayman, Tourchi, Maruf, Benz, Kasprenski, Baumgartner, Friedlander, Gearhart) Robert D. Jeffs Division of Pediatric Urology, James Buchanan Brady Urological Institutions, Johns Hopkins Hospital, Johns Hopkins Medical Institutions, Charlotte Bloomberg Children's Hospital, Baltimore, MD, United States (Feng, Trock) The James Buchanan Brady Urological Institute and Department of Urology, Johns Hopkins Hospital, Baltimore, MD, United States (Sponseller) Division of Pediatric Orthopedic Surgery, Johns Hopkins Hospital, Johns Hopkins Medical Institutions, Charlotte Bloomberg Children's Hospital, Baltimore, MD 21287, United States

Publisher

W.B. Saunders

Year of Publication

2019

239.

Clinical and molecular findings in nine new cases of tetrasomy 18p syndrome: FISH and array CGH characterization.

Slimani W., Ben Khelifa H., Dimassi S., Chioukh F.-Z., Jelloul A., Kammoun M., Hannachi H., Bouslah S., Jammali N., Sanlaville D., Saad A., Mougou-Zerelli S.

Embase

Molecular Cytogenetics. 12 (1) (no pagination), 2019. Article Number: 5. Date of Publication: 08 Feb 2019.

[Article]

AN: 626282772

Background: Small Supernumerary Marker Chromosomes (sSMC) are rare chromosomal abnormalities, which have abnormal banding arrangement and take many shapes. Several disorders have been correlated with sSMC presence. The aim of this study is to characterize the sSMC derived from chromosome 18 by Fluorescence in situ hybridization (FISH) and Array Comparative Genomic Hybridization (aCGH).

Result(s): Nine children with dysmorphic features have been investigated. They have these features in common: a triangular face, low-set ears, a large mouth with a thin upper lip, and a horizontal palpebral fissure. Epicanthus and strabismus were present in two patients. In addition, we have noticed microcephaly and mental and/or developmental delay with low birth weight. However, two patients had standard birth weight; one patient had hypospadias; two had skin problems; and three showed different congenital heart defects. One patient had corpus callosum hypoplasia. Systematic karyotype analysis revealed a de novo supernumerary chromosome. Array CGH showed a gain in copy number on the short arm of chromosome 18 in the nine cases. In one case, the sSMC seemed to be in mosaic. The breakpoints of the marker were identified using aCGH and FISH. Thus, the sSMC led to 18p tetrasomy with approximately 14 Mb lengths, between 364344 and 14763575 based on the human genome version 18.

Conclusion(s): These results have been completed by FISH in order to ascertain the shape of the sSMC. Our results confirm the uniqueness and particularity of the iso18p syndrome on the phenotypic as well as on the genetic level.

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Status

Embase

Institution

(Slimani, Ben Khelifa, Dimassi, Jelloul, Kammoun, Hannachi, Saad, Mougou-Zerelli) Laboratory of Human Cytogenetics, Molecular Genetics and Biology of Reproduction, Farhat Hached University Hospital, Sousse, Tunisia (Chioukh) Neonatology Department, Hospital of Fattouma Bourguiba, Monastir, Tunisia

(Bouslah) Pediatric Department, Farhat Hached University Hospital, Sousse, Tunisia

(Jammali) Department of Child Psychiatry, Fattouma Bourguiba Hospital, Monastir, Tunisia

(Sanlaville) Centre de Biologie et Pathologie Est, CHU de Lyon HCL, Lyon, France

(Saad, Mougou-Zerelli) Universite de Sousse, Faculte de Medecine de Sousse, Hopital Farhat Hached, UR035P2, Sousse 4000, Tunisia

Publisher

BioMed Central Ltd. (E-mail: info@biomedcentral.com)

Year of Publication

2019

240.

22q11.2 duplications in a UK cohort with bladder exstrophy-epispadias complex.

Beaman G.M., Woolf A.S., Cervellione R.M., Keene D., Mushtaq I., Urquhart J.E., Stuart H.M., Newman W.G.

Embase

American Journal of Medical Genetics, Part A. 179 (3) (pp 404-409), 2019. Date of Publication: March 2019.

[Article]

AN: 625899990

The bladder exstrophy-epispadias complex (BEEC) comprises of a spectrum of anterior midline defects, all affecting the lower urinary tract, the external genitalia, and the bony pelvis. In extreme cases, the gastrointestinal tract is also affected. The pathogenesis of BEEC is unclear but chromosomal aberrations have been reported. In particular, duplications of 22q11.2 have been identified in eight unrelated individuals with BEEC. The current study aimed to identify chromosomal copy number variants in BEEC. Analyses was performed using the Affymetrix Genome-wide SNP6.0 assay in 92 unrelated patients cared for by two UK pediatric urology centers. Three individuals had a 22q11.2 duplication, a significantly higher number than that found in a control group of 12,500 individuals with developmental delay who had undergone microarray testing ($p < .0001$). Sequencing of CRKL, implicated in renal tract malformations in DiGeorge syndrome critical region at 22q11, in 89 individuals with BEEC lacking 22q11 duplications revealed no pathogenic variants. To date, 22q11.2 duplication is the genetic variant most commonly associated with BEEC. This is consistent with the hypothesis that altered expression of a single, yet to be defined, gene therein is critical to the pathogenesis of this potentially devastating congenital disorder.

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Status

Embase

Author NameID

Beaman, Glenda M.; ORCID: <http://orcid.org/0000-0002-6098-9995> Woolf, Adrian S.; ORCID: <http://orcid.org/0000-0001-5541-1358>

Institution

(Beaman, Stuart, Newman) Evolution and Genomic Sciences, School of Biological Sciences, Faculty of Biology, Medicine and Health, University of Manchester, Manchester, United Kingdom

(Beaman, Urquhart, Stuart, Newman) Manchester Centre for Genomic Medicine, Manchester University NHS Foundation Trust, Manchester, United Kingdom

(Woolf) Division of Cell Matrix Biology & Regenerative Medicine, School of Biological Sciences, Faculty of Biology, Medicine and Health, University of Manchester, Manchester, United Kingdom

(Woolf, Cervellione, Keene) Royal Manchester Children's Hospital, Manchester University NHS Foundation Trust, Manchester, United Kingdom

(Mushtaq) Department of Paediatric Urology, Great Ormond Street Hospital for Children NHS Foundation Trust, London, United Kingdom

(Newman) Peking University Health Sciences Center, Beijing, China

Publisher

Wiley-Liss Inc. (E-mail: info@wiley.com)

Year of Publication

2019

241.

Anomalies of the genitourinary tract in children with 22q11.2 deletion syndrome.

Van Batavia J.P., Crowley T.B., Burrows E., Zackai E.H., Sanna-Cherchi S., McDonald-McGinn D.M., Kolon T.F.

Embase

American Journal of Medical Genetics, Part A. 179 (3) (pp 381-385), 2019. Date of Publication: March 2019.

[Article]

AN: 625654466

The 22q11.2 deletion syndrome (22q11.2DS) involves multiple organ systems with variable phenotypic expression. Genitourinary tract abnormalities have been noted to be present in up to

30-40% of patients. At our institution, an internationally recognized, comprehensive, and multidisciplinary 22q11.2DS care center has been providing care to these children. We sought to report on the incidence of genitourinary tract anomalies in this large cohort and, therefore, retrospectively reviewed all patients who underwent a complete evaluation from 1992 to March 2017. We identified all children with any genital or urinary tract anomaly. For all children with a diagnosis of hydronephrosis, the underlying etiology was determined, when possible. Overall, 1,073 of 1,267 children with 22q11.2DS underwent renal evaluations at our institution. Hundered Sixty-Two (15.1%) children had structural abnormalities of their kidneys/urinary tracts. The majority of children with hydronephrosis (63%) had isolated upper tract dilation without any additional diagnoses. Boys were significantly more likely to be diagnosed with a genital abnormality than girls (7.7 vs. 0.5%, $p < 0.001$). Of the 649 boys in the entire cohort, 24 (3.7%) had cryptorchidism and 24 (3.7%) had hypospadias, which was noted to be mild in all except one boy. Overall, findings of hydronephrosis, unilateral renal agenesis, and multicystic dysplastic kidney occur at higher rates than expected in the general population. Given these findings, in addition to routine physical examination, we believe that all patients with 22q11.2DS warrant screening RBUS at time of diagnosis.

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Author NameID

Van Batavia, Jason P.; ORCID: <http://orcid.org/0000-0002-1592-7824> Crowley, Terrence B.;

ORCID: <http://orcid.org/0000-0002-3272-5458>

Institution

(Van Batavia, Kolon) Division of Urology, Children's Hospital of Philadelphia, Philadelphia, PA,

United States (Crowley, Burrows, Zackai, McDonald-McGinn) Division of Human Genetics,

Children's Hospital of Philadelphia, Philadelphia, PA, United States

(Sanna-Cherchi) Division of Nephrology, New York-Presbyterian Hospital Columbia University

Medical Center, New York, NY, United States

Publisher

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Year of Publication

2019

242.

Family and neighborhood socioeconomic inequality in cryptorchidism and hypospadias: A nationwide study from Sweden.

Li X., Sundquist J., Hamano T., Sundquist K.

Embase

Birth Defects Research. 111 (2) (pp 78-87), 2019. Date of Publication: 15 Jan 2019.

[Article]

AN: 625545678

Objectives: To examine whether there is an association between neighborhood deprivation and incidence of cryptorchidism and hypospadias, after accounting for family-level and individual-level sociodemographic characteristics.

Method(s): All boys born in Sweden between January 1, 2001 and December 31, 2010 were followed. Data were analyzed by multilevel logistic regression, with family-level and individual-level characteristics at the first level and level of neighborhood deprivation at the second level.

Result(s): During the study period, among a total of 497,584 boys, 8,584 (1.7%) and 3,704 (0.7%) were diagnosed with cryptorchidism and hypospadias, respectively. Cumulative rates for cryptorchidism and hypospadias increased with increasing levels of neighborhood deprivation. In the study population, 1.5 per 100 and 2.0 per 100 boys, in the least and most deprived neighborhoods were diagnosed with cryptorchidism and 0.7 per 100 and 0.9 per 100 boys were diagnosed with hypospadias. Incidence of hospitalization for cryptorchidism and hypospadias increased with increasing neighborhood-level deprivation across all family-level and individual-level sociodemographic categories. The odds ratio (OR) for cryptorchidism and hypospadias for those living in high-deprivation neighborhoods versus those living in low-deprivation neighborhoods was 1.13 (95% confidence interval [CI] = 1.05-1.21) and 1.24 (95% CI = 1.12-1.37). High neighborhood deprivation remained significantly associated with higher odds of hypospadias after adjustment for family-level and individual-level sociodemographic characteristics (OR = 1.20, 95% CI = 1.08-1.35).

Conclusion(s): This study is the largest so far on neighborhood influences on cryptorchidism and hypospadias. Our results suggest that neighborhood deprivation is associated with a moderate incidence of hypospadias independent of family-level and individual-level sociodemographic characteristics.

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Author NameID

Li, Xinjun; ORCID: <http://orcid.org/0000-0002-5559-4657>

Institution

(Li, Sundquist, Sundquist) Center for Primary Health Care Research, Lund University, Malmo, Sweden (Sundquist, Sundquist) Department of Family Medicine and Community Health, Icahn School of Medicine at Mount Sinai, New York, NY, United States

(Sundquist, Hamano, Sundquist) Department of Functional Pathology, Center for Community-Based Healthcare Research and Education (CoHRE), School of Medicine, Shimane University, Izumo, Japan

(Hamano) Department of Sports Sociology and Health Sciences, Kyoto Sangyo University, Kyoto, Japan

Publisher

John Wiley and Sons Inc. (P.O.Box 18667, Newark NJ 07191-8667, United States)

Year of Publication

2019

243.

Molecular genetics and phenotype/genotype correlation of 5-alpha reductase deficiency in a highly consanguineous population.

Alswailem M.M., Alzahrani O.S., Alghofaili L., Qasem E., Almohanaa M., Alsagheir A., Bin Abbas B., Attia N.A., Al Shaikh A., Alzahrani A.S.

Embase

Endocrine. 63 (2) (pp 361-368), 2019. Date of Publication: 15 Feb 2019.

[Article]

AN: 624185140

Context and objectives: 5-alpha reductase deficiency is a rare 46,XY disorder of sex development. We present detailed phenotypic and genotypic features of a cohort of 24 subjects from a highly consanguineous population of Saudi Arabia Subjects and Methods: We studied the clinical presentation and hormonal profiles of 24 subjects diagnosed with 5-alpha reductase

deficiency and performed genetic testing on DNA isolated from their peripheral blood using polymerase chain reaction and direct sequencing of the SRD5A2.

Result(s): All subjects had 46,XY karyotype and presented with atypical appearance of external genitalia ranging from clitoromegaly, microphallus with hypospadias, undescended testes to completely normally looking female genitalia. Thirteen (54%) of them had severe under virilization and were assigned female sex at birth. The other 11 subjects were raised as males. Stimulated Testosterone:Dihydrotestosterone ratio was high in all 16 subjects in whom it was measured. The genetic testing revealed 2 nonsense mutations (p.R103X and p.R227X) in 2 unrelated subjects, 3 missense mutations (p.P181L, p.A228T, p.R246Q) in 11 subjects and a splice site mutation (IVS1-2A > G) in 11 other subjects. There was significant phenotypic variability even in subjects with the same mutation and also within the same family.

Conclusion(s): This is the first and largest report of the clinical and molecular genetics of 5-alpha reductase deficiency from the Middle East. It shows weak genotype/phenotype correlation and significant phenotypic heterogeneity. IVS1-2A > G mutation is the most common mutation and is likely to be a founder mutation in this part of the world.

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Institution

(Alswailem, Alghofaili, Qasem, Almohanaa, Alzahrani) Department of Molecular Oncology, King Faisal Specialist Hospital & Research Centre, Riyadh, Saudi Arabia (Alzahrani, Alsagheir, Bin Abbas) Department of Pediatrics, King Faisal Specialist Hospital & Research Centre, Riyadh, Saudi Arabia

(Attia, Al Shaikh) Department of Pediatrics, King Abdulaziz Medical City, Jeddah, Saudi Arabia

(Alzahrani) Department of Medicine, King Faisal Specialist Hospital & Research Centre, Riyadh, Saudi Arabia

Publisher

Humana Press Inc. (E-mail: humana@humanapr.com)

Year of Publication

2019

244.

The genetic factors contributing to hypospadias and their clinical utility in its diagnosis.

Joodi M., Amerizadeh F., Hassanian S.M., Erfani M., Ghayour-Mobarhan M., Ferns G.A., Khazaei M., Avan A.

Embase

Journal of Cellular Physiology. 234 (5) (pp 5519-5523), 2019. Date of Publication: May 2019.

[Review]

AN: 624005340

Hypospadias is among the most common congenital malformations in male neonates. It results from abnormal penile and urethral development, but is a multifactorial disorder that is highly heterogeneous, with several genetic and environmental determinants. Monogenic and chromosomal abnormalities are present in approximately 30% of cases, although the genetic factors contributing to hypospadias remain unknown in 70% of cases. While defects in androgen synthesis can lead to this malformation, mutational analyses have shown several genes, such as sonic hedgehog, fibroblast growth factors, bone morphogenetic proteins, homeobox genes, and the Wnt family, are involved in the normal development of male external genitalia. Mutations in the genes of penile development (e.g., HOX, FGF, Shh) and testicular determination (e.g., WT1, SRY), luteinizing hormone receptor, and androgen receptor have also been proposed to be implicated in hypospadias. Here we review the recent advances in this field and discuss the potential genes that could determine the risk of hypospadias.

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Author NameID

Hassanian, Seyed Mahdi; ORCID: <http://orcid.org/0000-0002-5247-4043> Ghayour-Mobarhan, Majid; ORCID: <http://orcid.org/0000-0001-5947-8904>

Khazaei, Majid; ORCID: <http://orcid.org/0000-0002-7979-5699>

Avan, Amir; ORCID: <http://orcid.org/0000-0002-4968-0962>

Institution

(Joodi) Department of Pediatric Surgery, School of Medicine, Mashhad University of Medical Sciences, Mashhad, Iran, Islamic Republic of (Joodi) Sarvar Children's Hospital, Endoscopic and Minimally Invasive Surgery Research Center, Mashhad, Iran, Islamic Republic of (Amerizadeh, Hassanian, Ghayour-Mobarhan, Khazaei, Avan) Metabolic Syndrome Research Center, Mashhad University of Medical Sciences, Mashhad, Iran, Islamic Republic of

(Hassanian) Department of Medical Biochemistry, School of Medicine, Mashhad University of Medical Sciences, Mashhad, Iran, Islamic Republic of

(Erfani) Department of Neurology, Ghaem Hospital, Mashhad University of Medical Sciences, Mashhad, Iran, Islamic Republic of

(Ferns) Division of Medical Education, Brighton & Sussex Medical School Falmer, Brighton, United Kingdom

(Avan) Department of Modern Sciences and Technologies, Faculty of Medicine, Mashhad University of Medical Sciences, Mashhad, Iran, Islamic Republic of

(Avan) Cancer Research Center, Mashhad University of Medical Sciences, Mashhad, Iran, Islamic Republic of

Publisher

Wiley-Liss Inc. (E-mail: info@wiley.com)

Year of Publication

2019

245.

First data from the new, unified database of the Hungarian case-control surveillance of congenital abnormalities.

Acs N., Matrai A., Kaposi A.

Embase

Journal of Maternal-Fetal and Neonatal Medicine. (no pagination), 2019. Date of Publication: 2019.

[Article]

AN: 2003505097

The Hungarian Case-Control Surveillance of Congenital Abnormalities (HCCSCA) is one of the largest case-control data sets of CA-surveillance in the world. We unified all data collected in the HCCSCA between 1980 and 2009 into a new, validated single database that is now open for examination. The details of this unified database are given in this paper. The total number of cases and control newborns is 32,345 and 57,231, respectively. The overall prevalence of CAs recorded in the HCCSCA was 10.7/1000 live-births. Data available for each pregnancy are: CA(s), gender, birth year/month/date, birth weight, gestational age, area of mother's living, maternal age, paternal age, birth order, mother's and father's qualification, employment status

and type of employment, mother's marital status, outcome of previous pregnancies, maternal diseases during pregnancy (according to pregnancy months), drug intake during pregnancy (according to pregnancy months), folic acid and/or pregnancy vitamin supplement intake (according to pregnancy months), mother's smoking habits and alcohol consumption patterns. The most frequent anomalies detected were ventricular septal defect (2864), atrial septal defect (1895), polydactyly (1499), hypospadias (1083), and unilateral cleft lip +/- palate (961). According to ICD-10, 701 diseases have been found to affect case mothers during pregnancy. Eight hundred and sixteen drugs were identified that had been taken by mothers during pregnancy. The authors are absolutely open for any scientific cooperation based on this database.

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Status

Article-in-Press

Author NameID

Acs, Nandor; ORCID: <http://orcid.org/0000-0002-1919-1869>

Institution

(Acs, Matrai) Department of Obstetrics and Gynaecology, Semmelweis University School of Medicine, Budapest, Hungary (Kaposi) Department of Biophysics and Radiation Biology, Semmelweis University School of Medicine, Budapest, Hungary

Publisher

Taylor and Francis Ltd

Year of Publication

2019

246.

Challenges in managing proximal hypospadias: A 17-year single-center experience.

Misra D., Elbourne C., Vareli A., Banerjee D., Joshi A., Friedmacher F., Skeritt C.

Embase

Journal of Pediatric Surgery. 54 (10) (pp 2125-2129), 2019. Date of Publication: October 2019.

[Article]

AN: 2001941264

Background: There are only a few publications in the medical literature reporting on complication rates in proximal hypospadias surgery, particularly with regard to long-term follow-up.

Method(s): Over a 17.5-year period, we operated 100 patients with penoscrotal, scrotal and perineal hypospadias. Sixty-four had a single-stage repair, including 15 who received a buccal mucosa inlay "Snodgrass" repair. Thirty-six had a two-stage Bracka repair of which 19 received buccal or lower lip grafts and 17 had preputial grafts. Overall, 34 patients received buccal grafts. The median follow-up was eight years (range 1-16 years). Three patients were operated for residual chordee years later.

Result(s): Urethral fistulae occurred in a total of 26/100 (26.0%) cases, meatal stenosis in 16/100 (16.0%), wound breakdown in six (6.0%) and graft failure in one (1.0%). The fistula rate after the single-stage approach was 15/64 (23.4%), whereas it was 11/36 (30.6%) following two-stage repair ($P = 0.4811$).

Conclusion(s): Proximal hypospadias remains a challenging condition to treat. It is possible to perform a single-stage repair in 64.0% of cases. This brings down the median number of operations to only two. Lower lip grafts were used in 34.0% but are now used in redo-surgeries only. Our fistula rate was 26.0% but has decreased significantly in recent years.

Level of Evidence: Level III.

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Status

In-Process

Institution

(Misra, Elbourne, Vareli, Banerjee, Joshi, Friedmacher, Skerrett) Department of Paediatric Urology and Paediatric Surgery, The Royal London Hospital, London, United Kingdom

Publisher

W.B. Saunders

Year of Publication

2019

247.

Nonsecretory intestinocystoplasty: postoperative outcomes of 25 years.

Dantas R.A.F., Calisto F.C.F.S., Vilar F.O., Araujo L.A.P., Lima S.V.C.

Embase

International braz j urol : official journal of the Brazilian Society of Urology. 45 (4) (pp 681-685), 2019. Date of Publication: 01 Jul 2019.

[Article]

AN: 628258392

OBJECTIVE: The objective of bladder augmentation (BA) is to create a low-pressure reservoir with adequate capacity. Despite its benefits, the use of intestinal patches in bladder enlargement provides a high risk of developing complications and BA with demucosalised bowel represents a potential alternative. Therefore, this study evaluated urological parameters and long-term clinical follow-up of patients submitted to nonsecretory BA in a single center with 25 years of experience. **MATERIALS AND METHODS:** Patients treated with BA underwent urological evaluation, which included history, physical examination and urodynamic study. The main urodynamic parameters (bladder capacity and bladder compliance) were assessed in the pre and postoperative moments, and compared by the Wilcoxon Signed Rank test. The main long-term complications were described.

RESULT(S): 269 patients (mean age 14+/-13 years, 47% male) underwent BA with the use of demucolised intestinal segments. Among the patients in the sample, 187 (69.52%) had neurogenic bladder, 68 (25.28%) had bladder exstrophy, nine had tuberculosis (3.34%), four had a posterior urethral valve (1.49%) and one with hypospadias (0.37%). After the surgical procedure, a significant increment in both urodynamic parameters was found, with a 222% increase in bladder capacity and 604% in bladder compliance ($p < 0.001$ in both analyzes). Mean follow-up time ranged from 2 to 358 months, with a median of 72 months (IQR 74-247). Among all patients, 5 presented spontaneous perforation.

CONCLUSION(S): The study showed statistically significant increase in both compliance and bladder capacity after non-secretory BA, with a low rate of severe complications.

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Institution

(Dantas, Calisto) Programa de Pos-Graduacao em Cirurgia, Centro de Ciencias da Saude, Universidade Federal de Pernambuco-UFPE, PE, Brazil (Vilar) Departamento de Urologia, Hospital das Clinicas da Universidade Federal de Pernambuco-UFPE, PE, Brazil

(Araujo) Departamento de Cirurgia Pediatrica, Universidade Federal de Pernambuco-UFPE, PE, Brazil

(Lima) Departamento de Cirurgia, Centro de Ciencias da Saude da Universidade Federal de Pernambuco-UFPE, PE, Brazil

Publisher
NLM (Medline)
Year of Publication
2019

248.

Association Between Extra-Genital Congenital Anomalies and Hypospadias Outcome.

Al-Juraibah F., Lucas-Herald A., Nixon R., Toka C., Wang C., Flett M., O'Toole S., Ahmed S.F.

Embase

Sexual development : genetics, molecular biology, evolution, endocrinology, embryology, and pathology of sex determination and differentiation. 13 (2) (pp 67-73), 2019. Date of Publication: 2019.

[Article]

AN: 627083163

Extra-genital congenital anomalies are often present in cases of hypospadias, but it is unclear whether they have an association with the outcome of hypospadias surgery. The aim of this study was to review all hypospadias cases that had surgery between 2009 and 2015 at a single centre and identify clinical determinants of the surgical outcome. An extra-genital congenital anomaly was reported in 139 (22%) boys and 62 (10%) had more than 1 anomaly. Of the 626 boys, 54 (9%), including 44 with proximal hypospadias, had endocrine as well as limited genetic evaluation. Of these, 10 (19%) had a biochemical evidence of hypogonadism and 5 (9%) had a molecular genetic abnormality. At least 1 complication was reported in 167 (27%) patients, with 20% of complications (most frequently fistula) occurring after 2 years of surgery. The severity of hypospadias and the existence of other anomalies were clinical factors that were independently associated with an increased risk of complications ($p < 0.001$). In conclusion, complications following surgery are more likely in those cases that are proximal or who have additional extra-genital anomalies. To understand the biological basis of these complications, there is a greater need to understand the aetiology of such cases.

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249.

Comparison of short-term complications between Onlay and Duckett urethroplasty and the analysis of risk factors.

Lyu Y., Yu L., Xie H., Huang Y., Li X., Sun L., Bao J., Liang Y., Zhou X., Chen F.

Embase

International Urology and Nephrology. (no pagination), 2019. Date of Publication: 2019.

[Article]

AN: 626870251

Aims: To compare the short-term complications between Onlay and Duckett urethroplasty and to analyze the various risk factors cause the complications.

Method(s): The children with hypospadias who underwent treatment with Onlay or Duckett urethroplasty from November 2014 to June 2016 were followed up. The difference in complications between the two groups was analyzed. Moreover, a single-factor ANOVA was performed to analyze the length and curvature of the penis and the length of the urethral defect.

Result(s): 40 patients were treated by Duckett, while 54 by Onlay. In comparison to the Onlay group, the Duckett group showed the initial penile length was shorter ($P = 0.044$), the initial urethral defect ($P = 0.024$) and after the correction of chordee was longer ($P < 0.001$), and the initial penile curvature ($P < 0.001$) and after degloving ($P < 0.001$) was greater. Furthermore, the incidence of urethra percutaneous fistula ($P = 0.041$) and urethral stenosis ($P < 0.001$) in Duckett group was significantly higher. The analysis of risk factors showed that the age at the time of surgery, the initial penile curvature, the initial urethral defect, and the urethral defect after the correction of chordee were not correlated with the complications. The degree of penile curvature after skin degloving and urethra stenosis was significantly correlated ($P = 0.019$).

Conclusion(s): The incidence of complications of urethra percutaneous fistula and urethral stenosis after 1 year of Duckett was higher than that by Onlay approach. The greater the curvature of the penis after skin degloving, the more likely the occurrence of urethral stenosis after surgery.

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Author NameID

Chen, Fang; ORCID: <http://orcid.org/0000-0001-8635-2952>

Institution

(Lyu, Yu, Xie, Huang, Li, Sun, Bao, Liang, Chen) Department of Urology, Shanghai Children's Hospital, Shanghai Jiao Tong University, Shanghai 200040, China (Zhou) Department of Bioinformatics and Biostatistics, Center for Biomedical informatics, Shanghai Jiaotong University, Shanghai Children's Hospital, Shanghai, China

Publisher

Springer Netherlands (E-mail: rbk@louisiana.edu)

Year of Publication

2019

250.

Identification of gene variants in 130 Han Chinese patients with hypospadias by targeted next-generation sequencing.

Zhang W., Shi J., Zhang C., Jiang X., Wang J., Wang W., Wang D., Ni J., Chen L., Lu W., Xiao Y., Ye W., Dong Z.

Embase

Molecular genetics & genomic medicine. 7 (8) (pp e827), 2019. Date of Publication: 01 Aug 2019. [Article]

AN: 628424403

BACKGROUND: Hypospadias is a common congenital malformation of male external genitalia, which mainly manifests as an abnormal urethral opening on the ventral side of the penis. The etiology and clinical phenotype of hypospadias is highly heterogeneous, and its clinical diagnosis is challenging. Currently, over 70% of patients have an unknown etiology. Here, we performed a targeted analysis of gene mutations in 130 patients with hypospadias of unknown etiology to find the precise genetic cause.

METHOD(S): We developed a targeted next-generation sequencing (NGS) panel, encompassing the exon coding regions of 105 genes involved in external genitalia and urogenital tract development and performed sequencing analysis on 130 children with hypospadias of unknown etiology.

RESULT(S): In total, 25 patients with hypospadias (19.2%) were found to have 20 mutations among the nine genes involved in external genitalia and urogenital tract development, including 16 reported and four novel mutation sites. Twenty-two patients (16.9%) had diagnostic variants. Multiple genetic mutations were identified in three of the 25 patients. Hypospadias combined with micropenis was the most common phenotype (68%) in 25 patients.

CONCLUSION(S): Higher frequency mutations were identified in SRD5A2 (52%) and AR (24%) in our patient cohort. Middle or posterior hypospadias with micropenis may be significant indicators of genetic variations. Polygenic inheritance may be a rare genetic cause of hypospadias.

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PMID

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Author NameID

Xiao, Yuan; ORCID: <http://orcid.org/0000-0002-4927-8199> Dong, Zhiya; ORCID: <http://orcid.org/0000-0002-2436-7857>

Institution

(Zhang, Wang, Wang, Wang, Ni, Chen, Lu, Xiao, Dong) Department of Paediatrics, Ruijin Hospital, Shanghai Jiao Tong University School of Medicine, Shanghai, China (Shi, Zhang) Department of Genetics, Shanghai-MOST Key Laboratory of Health and Disease Genomics, Chinese National Human Genome Center and Shanghai Industrial Technology Institute (SITI), Shanghai, China

(Jiang, Ye) Department of Urology, Ruijin Hospital, Shanghai Jiao Tong University School of Medicine, Shanghai, China

Publisher

NLM (Medline)

Year of Publication

2019

251.

Midline incision of a graft in staged hypospadias repair-feasible and durable?.

Tonnhofer U., Hiess M., Metzelder M., Hebenstreit D., Springer A.

Embase

Frontiers in Pediatrics. 7 (MAR) (no pagination), 2019. Article Number: 60. Date of Publication: 2019.

[Article]

AN: 627528554

Purpose: In severe hypospadias staged repair is commonly used and it is regarded as feasible, safe, and durable. In this article we want to describe the results of a modification of the staged repair: a midline incision of the graft during the second stage.

Material(s) and Method(s): This is a consecutive single team (2 surgeons) retrospective series. Between 2014 and 2017, 250 patients underwent hypospadias repair, among them 35 patients that had primary staged hypospadias surgery with completed first and second stage repair. 24 (68.6%) cases received a preputial skin graft and 11 (31.4%) buccal mucosa graft. Median age at first stage was 1.5 (0.5-22.1) years, mean time between first and second stage operation was 0.72 (0.4-1.76) years. Follow up rate was 100%, mean follow up period was 1.50 (0.4-3.8) years. Result(s): The total complication rate was 22.9%. In buccal mucosa repair the complication rate was 36.4% and in preputial graft repair the complication rate was 16.7%, respectively. In 23 patients (65.7%) during second stage urethroplasty a midline incision was performed (8 glandular graft, 15 penile graft, 6 at level of urethral opening). Complication rate in non-incised urethroplasty was 8.3%, in incision at glandular level 37.5%, in incision at penile level 13.3% and in incision at urethral opening 16.7%, respectively.

Conclusion(s): Two stage repair is the method of choice in the correction of severe hypospadias. In selected cases a midline incision of the graft is feasible and can be applied if needed.

Randomized studies will be needed to evaluate the true benefit of incising the graft.

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Status

Embase

Institution

(Tonnhofer, Metzelder, Springer) Department of Pediatric Surgery, Medical University Vienna, Vienna, Austria (Hiess) Department of Urology, Medical University Vienna, Vienna, Austria (Hebenstreit) Department of Urology, Medical University Innsbruck, Innsbruck, Austria

Publisher

Frontiers Media S.A. (E-mail: info@frontiersin.org)

Year of Publication

2019

252.

Recent trends in the management of bladder exstrophy: The gordian knot has not yet been cut.

Promm M., Roesch W.H.

Embase

Frontiers in Pediatrics. 7 (MAR) (no pagination), 2019. Article Number: 110. Date of Publication: 2019.

[Short Survey]

AN: 627528410

Although enormous effort has been made to further improve the operative techniques worldwide, the management of bladder exstrophy (BE) remains one of the most significant challenges in pediatric urology. Today it is universally agreed that successful and gentle initial bladder closure is decisive for favorable long-term outcome with regard to bladder capacity, renal function and continence. Due to a number of reasons, including a lack of comparable multicenter studies, a range of concepts is currently used to achieve successful primary closure. We review the literature of the last 15 years on the current concepts of bladder exstrophy repair with regard to the time of primary closure (initial vs. delayed closure), the concepts of primary closure (single-stage vs. staged approach; without osteotomy vs. osteotomy) and their outcomes. There is a worldwide lack of multicenter outcome studies with adequate patient numbers and precisely defined outcome parameters, based on the use of validated instruments. The modern staged repair (MRSE) in different variations, the complete primary reconstruction of exstrophy (CPRE), and the radical soft-tissue mobilization (RSTM) had been the most extensively studied and reported procedures. These major concepts are obligatory stable now for more than 20 years. Nevertheless, there are still a lot of open-ended questions e.g., on the potential for development of the bladder template, on continence, on long-term orthopedic outcome, on sexuality and fertility and on quality of life. Management of BE remains difficult and controversial. Further, clinical research should focus on multi-institutional collaborative trials to determine the optimal approach.

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Status

Embase

Institution

(Promm, Roesch) Department of Pediatric Urology, Clinic St. Hedwig, University Medical Center of Regensburg, Regensburg, Germany

Publisher

Frontiers Media S.A. (E-mail: info@frontiersin.org)

Year of Publication

2019

253.

Prevalence and surgical management of pubic hypertrophy in hypospadias patients: results from a high-volume surgeon.

Bandini M., Sekulovic S., Stanojevic N., Spiridonescu B., Pesic V., Sansalone S., Slavkovic M., Briganti A., Salonia A., Montorsi F., Djinojic R.

Embase

International braz j urol : official journal of the Brazilian Society of Urology. 45 (6) (pp 1238-1248), 2019. Date of Publication: 01 Nov 2019.

[Article]

AN: 630094041

INTRODUCTION: Pubic hypertrophy, defined as an abnormal and abundant round mass of fatty tissue located over the pubic symphysis, is frequently underestimated in patients with hypospadias. We examined the prevalence of this condition, as well as the outcomes associated with its surgical treatment. MATERIAL AND METHODS: Within 266 hypospadias patients treated at our clinic, we assessed the prevalence of pubic hypertrophy, and we schematically described the surgical steps of pubic lipectomy. Multivariable logistic regression (MLR) tested for predictors of pubic hypertrophy. Finally, separate MLRs tested for predictors of fistula and any complications after pubic lipectomy.

RESULT(S): Of 266 hypospadias patients, 100 (37.6%) presented pubic hypertrophy and underwent pubic lipectomy. Patients with pubic hypertrophy more frequently had proximal hypospadias (44 vs. 7.8%), disorders of sex development (DSD) (10 vs. 0.6%), cryptorchidism (12 vs. 2.4%), and moderate (30degree-60degree) or severe (>60degree) penile curvature (33 vs. 4.2%). In MLR, the location of urethral meatus (proximal, Odds ratio [OR]: 10.1, $p < 0.001$) was the only significant predictor of pubic hypertrophy. Finally, pubic lipectomy was not

associated with increased risk of fistula (OR: 1.12, p=0.7) or any complications (OR: 1.37, 95% CI: 0.64-2.88, p=0.4) after multi-variable adjustment.

CONCLUSION(S): One out of three hypospadias patients, referred to our center, presented pubic hypertrophy and received pubic lipectomy. This rate was higher in patients with proximal hypospadias suggesting a correlation between pubic hypertrophy and severity of hypospadias. Noteworthy, pubic lipectomy was not associated with increased risk of fistula or any complications.

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Institution

(Bandini, Sekulovic, Stanojevic, Spiridonescu, Pesic, Slavkovic, Djinovic) Sava Perovic Foundation, Center for Genito-Urinary Reconstructive Surgery, BelMedic General Hospital, Belgrade, Serbia (Bandini, Briganti, Salonia, Montorsi) Division of Oncology and Unit of Urology, IRCCS Ospedale San Raffaele, Vita-Salute San Raffaele University, Milan, URI, Italy (Spiridonescu) Clinical Institute Fundeni, Center for Urology and Renal Transplantation, Bucharest, Romania

(Sansalone) Department of Experimental Medicine and Surgery, University of Tor Vergata, Rome, Italy

Publisher

NLM (Medline)

Year of Publication

2019

254.

Maternal rheumatoid arthritis and systemic lupus erythematosus and risk of cryptorchidism and hypospadias in boys: a Danish nationwide study.

Knudsen S.S., Bech B.H., Deleuran B.W., Ramlau-Hansen C.H., Arendt L.H.

Embase

Rheumatology (Oxford, England). (no pagination), 2019. Date of Publication: 18 Nov 2019.

[Article]

AN: 629888578

OBJECTIVES: RA and SLE are the most prevalent autoimmune rheumatic diseases affecting young women. Both diseases are characterized by systemic inflammation that may affect placental function and fetal development during pregnancy, and both diseases are associated with adverse pregnancy and child outcomes. We investigated the associations between maternal RA or SLE and the two genital malformations, cryptorchidism and hypospadias.

METHOD(S): In this nationwide register-based study including all male singleton live births in Denmark from 1995 to 2016, we assessed the occurrence of cryptorchidism and hypospadias according to the prenatal disease-state of the mothers. Using Cox proportional hazards models we calculated adjusted hazard ratios, accounting for varying age at diagnosis.

RESULT(S): Among 690 240 boys, 1026 had a mother with RA and 352 had a mother with SLE. We found adjusted hazard ratios of 1.72 (95% CI: 1.15; 2.57) for cryptorchidism among boys born to mothers with RA and 1.46 (95% CI: 0.69; 3.06) for boys born to mothers with SLE, compared with the general population. As the number of hypospadias cases was low, multivariate analysis was not feasible. The crude hazard ratios were 0.51 (95% CI: 0.16; 1.58) and 1.00 (95% CI: 0.25; 4.03) for RA and SLE, respectively.

CONCLUSION(S): Boys born to mothers with RA had higher risk of cryptorchidism, compared with unexposed boys. Boys born to mothers with SLE showed a similar tendency, however with less precision of the estimate. No conclusion could be reached on the risk of hypospadias, due to the low number of events.

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Institution

(Knudsen, Bech, Ramlau-Hansen) Department of Public Health, Aarhus University, Aarhus

(Knudsen, Deleuran) Department of Rheumatology, Aarhus University Hospital, Aarhus

(Deleuran) Department of Biomedicine, Aarhus University, Aarhus

(Arendt) Department of Gynaecology and Obstetrics, Horsens Regional Hospital, Horsens, Denmark

Publisher

NLM (Medline)

Year of Publication

2019

255.

Prevalence, assessment and surgical correction of penile curvature in hypospadias patients treated at one European Referral Center: description of the technique and surgical outcomes. Bandini M., Sekulovic S., Spiridonescu B., Krishnappa P., Dangi A.D., Slavkovic M., Pesic V., Salonia A., Briganti A., Montorsi F., Djinoovic R.

Embase

World journal of urology. (no pagination), 2019. Date of Publication: 25 Oct 2019.

[Article]

AN: 629704231

PURPOSE: Penile curvature (PC) is a common component of hypospadias, but its presence is inconstantly assessed. We aim to report prevalence of PC in hypospadias patients, as well as to report our method to assess and correct PC, with the associated postoperative outcomes.

METHOD(S): We scrutinized 303 pediatric hypospadias patients operated (2013-2018) at our referral center. PC was routinely assessed and eventually corrected with dorsal plications (DP) as one-stage procedure, or ventral tunica attenuations+/-DP as two-stage repair. PC severity and surgical treatment of PC were compared between primary and failed hypospadias. Finally, PC severity, failed repair and PC treatment were tested as predictors of perioperative complications.

RESULT(S): PC (>10degree) was identified in 274/303 (90.4%) patients, 86.1% with distal, 91.8% with midshaft, and 100% with proximal hypospadias, respectively. PC was found in 51/64 (79.7%) of failed hypospadias. One-stage and two-stage procedures were adopted in 211/274 (77%) and 63/274 (23%) children, respectively. PC severity ($p=0.1$) and PC treatment ($p=0.4$) did not differ between primary and failed hypospadias. PC severity (all $p>0.2$), failed repair ($p=0.8$), and PC treatment (all $p>0.09$) were not predictors of perioperative complications. 95.6% of patients achieved a straight penis.

CONCLUSION(S): Less than 1/10 patients did not require PC correction. High rate of residual PC in failed hypospadias and similar severity between failed and primary suggest that PC was usually under-corrected. It is possible to correct PC completely and the resulting complication would not be associated with PC severity, failed repair or treatment adopted.

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Author NameID

Bandini, Marco; ORCID: <http://orcid.org/0000-0002-1462-1698>

Institution

(Bandini, Sekulovic, Spiridonescu, Slavkovic, Pesic, DjinoVIC) Sava Perovic Foundation, Center for Genito-Urinary Reconstructive Surgery, Belgrade, Serbia (Bandini, Salonia, Briganti, Montorsi) Unit of Urology, Urological Research Institute (URI), IRCCS San Raffaele Hospital, Vita-Salute San Raffaele University, MI, Via Olgettina 60, Milan 20132, Italy
(Krishnappa) Department of Urology, NU Hospitals, Bangalore, India
(Dangi) Department of Urology, Christian Medical College and Hospital, Vellore, Tamil Nadu, India
Publisher
NLM (Medline)
Year of Publication
2019

256.

Recurrent Ventral Curvature with Long-Term Follow-up after Transverse Preputial Island Urethroplasty.

Wang C., Zhang W., Song H.

Embase

European journal of pediatric surgery : official journal of Austrian Association of Pediatric Surgery ... [et al] = Zeitschrift fur Kinderchirurgie. (no pagination), 2019. Date of Publication: 12 May 2019. [Article]

AN: 627822589

OBJECTIVE: This study was aimed to assess the long-term outcomes of recurrent ventral curvature (VC) repaired in early childhood after transverse preputial island flap urethroplasty.

MATERIALS AND METHODS: A total of 378 patients underwent transverse preputial island flap urethroplasty between January 2000 and January 2005 at our hospital. Of these patients, 43 were invited for assessment of VC after puberty. The age at surgery, types of hypospadias, degrees of recurrent VC, and surgical procedures were analyzed.

RESULT(S): The study included 43 patients with a mean age of 15.9 years (range, 12.3-17.9). The average age at the time of primary surgery was 1.9 years (range, 1.2-3.6). Of 43 patients, recurrent VC was identified in 14 (32.5%). In total, 8 out of 16 patients (50.0%) were successfully treated by urethral plate transection with skin release during the primary surgery, and 6 out of 27 patients (28.6%) underwent additional dorsal plication (DP; $p=0.093$). Severe recurrent VC was

observed in four, moderate curvature was observed in four, and mild curvature was observed in six cases. Recurrent VC was present more often in patients with complications (34.6 vs. 24.1%, $p=0.331$), especially in severe urethral strictures that required open surgical reconstruction ($p=0.039$).

CONCLUSION(S): Although the patients in our study represent only a small portion of the overall hypospadias population, it is notable that 32.5% of these patients showed recurrent VC, including 28.6% of patients with transection plus DP. We suggest long-term follow-up of hypospadias at least during adolescence or even into adulthood.

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Institution

(Wang, Zhang, Song) Department of Urology, Beijing Children's Hospital Affiliated to Capital Medical University, Beijing, China

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Year of Publication

2019

257.

Buccal mucosa graft for simultaneous correction of severe chordee and urethroplasty as a one-stage repair of scrotal hypospadias (watch technique).

Djordjevic M.L., Bizic M., Stojanovic B., Bencic M., Kojovic V., Korac G.

Embase

World journal of urology. 37 (4) (pp 613-618), 2019. Date of Publication: 01 Apr 2019.

[Article]

AN: 627453355

PURPOSE: Severe hypospadias repair still presents a great challenge. We evaluated a novel approach of using a specially shaped buccal mucosa graft for simultaneous ventral tunica grafting and new urethral plate creation, in combination with longitudinal dorsal island skin flap, as a one-stage repair of severe hypospadias.

METHOD(S): Between July 2014 and September 2017, 26 patients (aged from 12 to 22 months) underwent scrotal hypospadias repair. Short and non-elastic urethral plate is divided. Buccal mucosa graft is harvested from the inner cheek, and designed in a special "watch" shape, with the spherical part in the middle and two rectangular parts on both sides. Tunica albuginea is opened ventrally for penile straightening and grafted to the spherical part of the "watch-shaped" buccal mucosa with 6-8 "U-shape" stitches. The rectangular parts are fixed to the tip of the glans distally and native urethral meatus proximally. Longitudinal dorsal skin flap is harvested, button-holed ventrally and joined with buccal graft. Penile skin reconstruction is performed using available penile skin.

RESULT(S): The mean follow-up was 22 months (range from 9 to 46 months). Satisfactory results were achieved in 22 patients. Two urethral fistulas were successfully repaired by minor surgery after 3 months, while one meatal stenosis and one urethral diverticulum were successfully treated by temporary urethral dilation. There were no cases of residual curvature.

CONCLUSION(S): Specially shaped buccal mucosa graft for simultaneous curvature correction and urethroplasty could be a good choice for single-stage repair of scrotal hypospadias with severe curvature.

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Author NameID

Djordjevic, Miroslav L.; ORCID: <http://orcid.org/0000-0003-2298-3450>

Institution

(Djordjevic, Bizic, Kojovic) School of Medicine, University of Belgrade, Belgrade, Serbia

(Djordjevic, Bizic, Stojanovic, Bencic, Korac) Department of Urology, University Children's Hospital, Tirsova 10, Belgrade 11000, Serbia

Publisher

NLM (Medline)

Year of Publication

2019

258.

Interaction between CYP1A1/CYP17A1 polymorphisms and parental risk factors in the risk of hypospadias in a Chinese population.

Mao Y., Zhang K., Ma L., Yun X., Ou F., Liu G., Yang Y., Zhang Y., Pei X., Duan Z., Ma M.

Embase

Scientific reports. 9 (1) (pp 4123), 2019. Date of Publication: 11 Mar 2019.

[Article]

AN: 626757736

Hypospadias (HS) is a common congenital malformation of the genitourinary tract in males and its etiology is viewed as multifactorial, and studies about gene-environment interaction in the etiology of HS are rare. A total of 152 cases and 151 controls were selected in the present study. Information before and during pregnancy from questionnaires finished by mothers of subjects were extracted, and the relating data were analyzed to determine the risk factors of HS. Meanwhile, maternal genomic DNA was genotyped for the single nucleotide polymorphisms (SNPs) of CYP1A1 rs1048943 and CYP17A1 rs4919686. Results of multivariable logistic regression analyses showed that several factors were associated with hypospadias risk. Analysis of the distributions of SNPs in CYP1A1 and CYP17A1 genes showed that the mutant genotype CC (OR=4.87) of CYP1A1 rs1048943, and mutant genotype CC (OR=5.82), recessive genotype AC+CC (OR=2.17) and allele C (OR=1.77) of CYP17A1 rs4919686 significantly increased the risk of HS. In addition, the additive gene-environment interactions were also found in several models. Several maternal risk factors that are associated with HS risk can interact with CYP1A1/CYP17A1 polymorphisms, which lead to infants vulnerable to occurrence of HS in Chinese populations.

PMID

30858503 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=30858503>]

Author NameID

Ma, Mingyue; ORCID: <http://orcid.org/0000-0003-4440-1618>

Institution

(Mao, Zhang, Ma, Zhang, Pei, Duan, Ma) Department of Toxicology, School of Public Health, Shenyang Medical College, Shenyang, Liaoning Province 110034, China (Mao) Editorial

Department of Journal of Shenyang Medical College, Shenyang Medical College, Shenyang, Liaoning Province 110034, China

(Yun, Ou) Department of Clinical Nutrition, First Affiliated Hospital of China Medical University, Shenyang, Liaoning Province 110001, China

(Liu, Yang) Department of Pediatric Surgery, Shengjing Hospital of China Medical University, Shenyang, Liaoning Province 110004, China

Publisher

NLM (Medline)

Year of Publication

2019

259.

Surgical Outcomes of Plaque Excision and Grafting and Supplemental Tunica Albuginea Plication for Treatment of Peyronie's Disease With Severe Compound Curvature.

Chow AK; Sidelsky SA; Levine LA.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Journal of Sexual Medicine. 15(7):1021-1029, 2018 07.

[Journal Article]

UI: 29802005

BACKGROUND: There are limited data in the literature that describe the management of Peyronie's disease (PD) with severe compound curvature, which often requires additional straightening procedures after plaque excision and grafting (PEG) to achieve functional penile straightening (<20 degrees).

AIM: This study highlights the clinical distinction and our experience with men with PD and severe compound curvature treated with PEG and supplemental tunica albuginea plication (TAP).

METHODS: We performed a retrospective chart review of patients with PD and acute angulation who underwent PEG (group 1) and patients with compound curvature who underwent PEG with TAP (group 2) between 2007 and 2016.

OUTCOMES: Primary post-operative outcomes of interest include change in penile curvature, change in measured stretched penile length, and subjective report on penile sensation and sexually induced penile rigidity.

RESULTS: 240 Men with PD were included in the study, of which 79 (33%) patients in group 1 underwent PEG and 161 (67%) in group 2 underwent PEG and TAP. There was no difference in associated PD co-morbidities including age, hypertension, hyperlipidemia, hypogonadism, diabetes, or tobacco use. After artificial induction of erection with intracorporal trimix injection, the average primary curvature was 73 (range, 20-120) degrees for group 1 compared to 79 (range, 35-140) degrees for group 2 ($P = .01$). Group 2 had an average secondary curvature of 36 (20-80 degrees). After completion of PEG, men in group 2 had an average residual curvature of 30 (range, 20-50) degrees which required 1-6 TAPs to achieve functional straightness (<20 degrees). At an average follow-up of 61 months, there was no difference for group 1 and group 2, respectively, for recurrent curvature (11.4% vs 12.4%, $P = .33$), change in penile length (+0.57 vs

+0.36 cm, $P = .27$) or decreased penile sensation (6% vs 13%, $P = .12$). In all, 81% of group 1 and 79% of group 2 were able to engage in penetrative sex after penile straightening with or without pharmacotherapy ($P = .73$).

CLINICAL TRANSLATION: Our review shows promising surgical outcomes for the use of PEG and supplemental TAP for this subtype of complex PD.

STRENGTHS AND LIMITATIONS: This article reports the largest experience with treatment of PD with compound curvature to date. Limitations of this study include the retrospective nature of the analysis as well as the lack of a validated objective measurement of erectile function after penile straightening.

CONCLUSION: Our study found no baseline difference in underlying co-morbidities in men with severe compound curvature compared with men with acute severe angulated curvature. Men with severe compound curvature represent a severe and under-recognized population of men with PD who can be surgically corrected with PEG and supplemental TAP(s) when needed without an increased risk of loss of penile length, recurrent curvature, decreased penile sensation, or erectile dysfunction when compared to men treated with PEG alone. Chow AK, Sidelsky SA, Levine LA. Surgical Outcomes of Plaque Excision and Grafting and Supplemental Tunica Albuginea Plication for Treatment of Peyronie's Disease With Severe Compound Curvature. *J Sex Med* 2018;15:1021-1029.

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1

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MEDLINE

Authors Full Name

Chow, Alexander K; Sidelsky, Steven A; Levine, Laurence A.

Institution

Chow, Alexander K. Department of Urology, Rush University Medical Center, Chicago, IL, USA.
Electronic address: Alexander_k_chow@rush.edu. Sidelsky, Steven A. Department of Urology, Rush University Medical Center, Chicago, IL, USA.

Levine, Laurence A. Department of Urology, Rush University Medical Center, Chicago, IL, USA.

Year of Publication

2018

260.

Management of Post-traumatic and Iatrogenic urethrocuteaneous fistula in children (a case series of seven patients).

Khan I; Qureshi MA; Abbas SH; Shaukat M.

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JPMA - Journal of the Pakistan Medical Association. 68(6):955-958, 2018 Jun.

[Journal Article]

UI: 30323369

This is a retrospective study of seven patients with post-traumatic and iatrogenic urethrocuteaneous fistula of penile urethra, excluding complication of hypospadias surgery. It was conducted in the Paediatric Surgery Department, Jinnah Hospital Lahore from June 2014 to January 2017. The patients ages ranged from three to twelve years. All the patients were managed by repairing the fistula in three layers electively at 3 months from the date of initial presentation. They remain well with no complaints except one with a recurrence. The complications of circumcision can be avoided by preventing circumcision by non-doctors and quacks.

Version ID

1

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Authors Full Name

Khan, Ibrar; Qureshi, Muhammad Asif; Abbas, Syed Husnain; Shaukat, Mahmood.

Institution

Khan, Ibrar. Department of Pediatric Surgery, Allama Iqbal Medical College, Jinnah Hospital, Lahore. Qureshi, Muhammad Asif. Department of Pediatric Surgery, Allama Iqbal Medical College, Jinnah Hospital, Lahore.

Abbas, Syed Husnain. Department of Pediatric Surgery, Allama Iqbal Medical College, Jinnah Hospital, Lahore.

Shaukat, Mahmood. Department of Pediatric Surgery, Allama Iqbal Medical College, Jinnah Hospital, Lahore.

Year of Publication

2018

261.

Transitional Urology. [Review]

Kovell RC; Skokan AJ; Wood DN.

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MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Urologic Clinics of North America. 45(4):601-610, 2018 Nov.

[Journal Article. Review]

UI: 30316314

The field of transitional urology has taken on an increasing importance in recent years as more individuals with congenital urologic issues are living and thriving into adulthood. This article reviews the transitional process itself including barriers to successful transition and the consequences of failing to properly transition. Also provided is a broad overview of the urologic issues faced by patients who may benefit from lifelong care and the providers who will be helping them with transition and assuming their care.

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Authors Full Name

Kovell, Robert C; Skokan, Alexander J; Wood, Dan N.

Institution

Kovell, Robert C. Division of Urology, Department of Surgery, University of Pennsylvania Health System, Children's Hospital of Philadelphia, Perelman Center for Advanced Medicine, 3400 Civic Center Boulevard, 3 West, Philadelphia, PA 19104, USA. Electronic address:

robert.kovell@uphs.upenn.edu. Skokan, Alexander J. Division of Urology, Department of Surgery, University of Pennsylvania Health System, Perelman Center for Advanced Medicine, 3400 Civic Center Boulevard, 3 West, Philadelphia, PA 19104, USA.

Wood, Dan N. Department of Urology, University College London Hospitals, 16-18 Westmoreland Street, London W1H 6PL, UK.

Comments

Comment in (CIN)

Year of Publication

262.

Gender identity in patients with 5-alpha reductase deficiency raised as females.

Nascimento RLP; de Andrade Mesquita IM; Gondim R; Dos Apostolos RAAC; Toralles MB; de Oliveira LB; Cangucu-Campinho AK; Barroso U Jr.

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MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Journal of pediatric urology. 14(5):419.e1-419.e6, 2018 10.

[Journal Article]

UI: 30297225

BACKGROUND AND OBJECTIVE: 5-Alpha reductase type 2 deficiency (5-ARD) is a rare disorder of sex development. The lack of 5-alpha reductase, an enzyme that converts testosterone into dihydrotestosterone, results in external genitalia that may appear female, or predominantly male, albeit undervirilized, or, more often, ambiguous.

METHODS: This study describes a series of patients with 5-ARD raised as female, focusing on aspects related to gender identity. Following a retrospective chart review, patients with 5-ARD were invited to return to the clinic to enable their gender identity to be assessed using an 11-item structured in-house questionnaire. The Golombok-Rust Inventory of Sexual Satisfaction was applied to patients who had initiated their sexual life.

RESULTS: Six patients aged >15 years with 5-ARD and raised as female were included. Most patients were diagnosed late: two before and four after puberty. The mean length of the phallus was 2.8 cm (0.5-5.0). Reasons for seeing a doctor included genital appearance (n = 3), amenorrhea/absence of breast development (n = 2), and changes in gender role attitudes (n = 1). According to the gender identity assessment, 4 patients identified as female, 1 as male, and 1 as both genders. Only the patient identified as male requested gender re-assignment. Of the two patients who had initiated their sexual life, sexual satisfaction was found to be good in one and poor in the other due to vaginal discomfort during intercourse.

CONCLUSION: In the present series, the majority of undervirilized patients with a diagnosis of 5-ARD raised as female were in complete conformation with being female and described themselves as heterosexual. The more virilized patients were those least in conformity with their female-assigned gender.

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Authors Full Name

Nascimento, R L P; de Andrade Mesquita, I M; Gondim, R; Dos Apostolos, R A A C; Toralles, M B; de Oliveira, L B; Cangucu-Campinho, A K; Barroso, U Jr.

Institution

Nascimento, R L P. Disorders of Sexual Development Multidisciplinary Clinic, Federal University of Bahia, Salvador, Bahia, Brazil. de Andrade Mesquita, I M. Disorders of Sexual Development Multidisciplinary Clinic, Federal University of Bahia, Salvador, Bahia, Brazil.

Gondim, R. Disorders of Sexual Development Multidisciplinary Clinic, Federal University of Bahia, Salvador, Bahia, Brazil.

Dos Apostolos, R A A C. Disorders of Sexual Development Multidisciplinary Clinic, Federal University of Bahia, Salvador, Bahia, Brazil.

Toralles, M B. Disorders of Sexual Development Multidisciplinary Clinic, Federal University of Bahia, Salvador, Bahia, Brazil.

de Oliveira, L B. Disorders of Sexual Development Multidisciplinary Clinic, Federal University of Bahia, Salvador, Bahia, Brazil.

Cangucu-Campinho, A K. Disorders of Sexual Development Multidisciplinary Clinic, Federal University of Bahia, Salvador, Bahia, Brazil.

Barroso, U Jr. Disorders of Sexual Development Multidisciplinary Clinic, Federal University of Bahia, Salvador, Bahia, Brazil. Electronic address: ubarroso@uol.com.br.

Year of Publication

2018

263.

Novel mosaic SRY gene deletions in three newborn males with variable genitourinary malformations.

Roberts J; Lyalin D; Tosatto N; Rana P; Fadoul H; Welsh H; Zhang L; Cooley L; Repnikova E.

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American Journal of Medical Genetics. Part A. 176(9):2017-2023, 2018 09.

[Journal Article]

UI: 30055081

Ambiguous genitalia in the newborn can present a diagnostic challenge in medical practice. In most cases, the causes of genitourinary anomalies are not well understood; both genetic and environmental factors are thought to play a role. In this study, we report mosaic SRY gene deletion identified by fluorescence in situ hybridization (FISH) analysis in three unrelated newborn male patients with genital anomalies. G-banded chromosomes and microarray analysis were normal for all three patients. One patient had microphallus, hypospadias, bifid scrotum, exstrophic perineal tissue identified as a rectal duplication, lumbar vertebral anomalies, scoliosis, and a dysmorphic sacrum. The other two patients had isolated epispadias with the urethral meatus close to the penopubic junction. All three had bilateral palpable gonads in the scrotum. While this is the first report of mosaic SRY deletions, mosaic SRY sequence variants have been described in patients with variable genitourinary anomalies. This study identifies FISH analysis as a reliable method for mosaic SRY deletion detection. We suggest SRY FISH analysis should be used in the clinical workup of patients with genitourinary ambiguity.

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Author Initials

Repnikova, Elena; ORCID: <https://orcid.org/0000-0002-8797-5227>

Authors Full Name

Roberts, Jennifer; Lyalin, Dmitry; Tosatto, Norwood; Rana, Pratibha; Fadoul, Hiba; Welsh, Holly; Zhang, Lei; Cooley, Linda; Repnikova, Elena.

Institution

Roberts, Jennifer. Department of Pathology & Laboratory Medicine, Division of Clinical Laboratory Genetics & Genomics, The University of Missouri-Kansas City School of Medicine, Kansas City, Missouri. Lyalin, Dmitry. Department of Pathology & Laboratory Medicine, Division of Clinical Laboratory Genetics & Genomics, The University of Missouri-Kansas City School of Medicine, Kansas City, Missouri.

Tosatto, Norwood. Department of Pathology & Laboratory Medicine, Division of Clinical Laboratory Genetics & Genomics, The University of Missouri-Kansas City School of Medicine, Kansas City, Missouri.

Rana, Pratibha. Department of Pediatric Endocrinology, The University of Missouri-Kansas City School of Medicine, Kansas City, Missouri.

Rana, Pratibha. Children's Mercy Hospital, The University of Missouri-Kansas City School of Medicine, Kansas City, Missouri.

Fadoul, Hiba. Department of Pediatric Endocrinology, The University of Missouri-Kansas City School of Medicine, Kansas City, Missouri.

Fadoul, Hiba. Children's Mercy Hospital, The University of Missouri-Kansas City School of Medicine, Kansas City, Missouri.

Welsh, Holly. Children's Mercy Hospital, The University of Missouri-Kansas City School of Medicine, Kansas City, Missouri.

Welsh, Holly. Division of Clinical Genetics, The University of Missouri-Kansas City School of Medicine, Kansas City, Missouri.

Zhang, Lei. Department of Pathology & Laboratory Medicine, Division of Clinical Laboratory Genetics & Genomics, The University of Missouri-Kansas City School of Medicine, Kansas City, Missouri.

Zhang, Lei. Children's Mercy Hospital, The University of Missouri-Kansas City School of Medicine, Kansas City, Missouri.

Cooley, Linda. Department of Pathology & Laboratory Medicine, Division of Clinical Laboratory Genetics & Genomics, The University of Missouri-Kansas City School of Medicine, Kansas City, Missouri.

Cooley, Linda. Children's Mercy Hospital, The University of Missouri-Kansas City School of Medicine, Kansas City, Missouri.

Repnikova, Elena. Department of Pathology & Laboratory Medicine, Division of Clinical Laboratory Genetics & Genomics, The University of Missouri-Kansas City School of Medicine, Kansas City, Missouri.

Repnikova, Elena. Children's Mercy Hospital, The University of Missouri-Kansas City School of Medicine, Kansas City, Missouri.

Year of Publication

2018

5-alpha-Reductase type 2 deficiency: is there a genotype-phenotype correlation? A review.
[Review]

Avendano A; Paradisi I; Cammarata-Scalisi F; Callea M.

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MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present
Hormones. 17(2):197-204, 2018 Jun.

[Journal Article. Review]

UI: 29858846

5-alpha-Reductase type 2 enzyme catalyzes the conversion of testosterone into dihydrotestosterone, a potent androgen responsible for male sexual development during the fetal period and later during puberty. Its deficiency causes an autosomal recessive disorder of sex development characterized by a wide range of under-virilization of external genitalia in patients with a 46,XY karyotype. Mutations in the SRD5A2 gene cause 5-alpha-Reductase deficiency; although it is an infrequent disorder, it has been reported worldwide, with mutational heterogeneity. Furthermore, it has been proposed that there is no genotype-phenotype correlation, even in patients carrying the same mutation. The aim of this review was to perform an extensive search in various databases and to select those articles with a comprehensive genotype and phenotype description of the patients, classifying their phenotypes using the external masculinization score (EMS). Thus, it was possible to objectively compare the eventual genotype-phenotype correlation between them. The analysis showed that for most of the studied mutations no correlation can be established, although the specific location of the mutation in the protein has an effect on the severity of the phenotype. Nevertheless, even in patients carrying the same homozygous mutation, a variable phenotype was observed, suggesting that additional genetic factors might be influencing it. Due to the clinical variability of the disorder, an accurate diagnosis and adequate medical management might be difficult to carry out, as is highlighted in the review.

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Status

MEDLINE

Authors Full Name

Avendano, Andrea; Paradisi, Irene; Cammarata-Scalisi, Francisco; Callea, Michele.

Institution

Avendano, Andrea. Medicine Faculty, Medical Genetics Unit, Los Andes University, Merida, Venezuela. Paradisi, Irene. Human Genetics Laboratory, Venezuelan Institute for Scientific Research (IVIC), Caracas, Venezuela. ireneparadisi@hotmail.com.

Cammarata-Scalisi, Francisco. Medicine Faculty, Medical Genetics Unit, Los Andes University, Merida, Venezuela.

Callea, Michele. Unit of Dentistry, Bambino Gesù Children's Hospital, Rome, Italy.

Year of Publication

2018

265.

Trends in the Prevalences of Selected Birth Defects in Korea (2008-2014).

Ko JK; Lamichhane DK; Kim HC; Leem JH.

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International Journal of Environmental Research & Public Health [Electronic Resource]. 15(5), 2018 05 05.

[Journal Article. Research Support, Non-U.S. Gov't]

UI: 29734759

Little information is available on the prevalences of birth defects in Korea. The aims of this study were to estimate recent prevalences of selected birth defects and to analyze the prevalence trends of these defects during the period from 2008 to 2014. Prevalences were calculated for 69 major birth defects using health insurance claim data obtained from the Korea National Health Insurance Service (NHIS). Prevalence rate ratios were calculated using Poisson regression to analyze trends over the 7-year study period. The overall prevalence of a major birth defect was 446.3 per 10,000 births (95% CI: 444.0-448.6); 470.9 per 10,000 births (95% CI: 467.6-474.2) for males and 420.2 per 10,000 births (95% CI: 417-423.4) for females. The prevalence rates of the most common birth defects over the study period were; septal defect (138.2 per 10,000; 95% CI: 136.9-139.5), congenital hip dislocation (652 per 10,000; 95% CI: 64.1-65.9), and ventricular septal defect (62.62 per 10,000; 95% CI: 61.7-63.5). During the study period, a significant increase in the prevalence of a major birth defect was observed with a prevalence rate ratio (PRR) of 1.091. The strongest trend was observed for renal dysplasia, which had a PRR of 1.275 (95% CI: 1.211-1.343), and upward trends were observed for urogenital anomalies, such as, renal agenesis (PRR 1.102, 95% CI: 1.067-1.138), undescended testis (PRR 1.082, 95% CI: 1.072-1.093) and hypospadias (PRR 1.067, 95% CI: 1.044-1.090). This study shows an overall increase in the prevalences of birth defects, including hypospadias and undescended testis,

which are known to be associated with endocrine factors. In the future, standardized birth defect registries should be established to enable these trends to be monitored.

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1

Status

MEDLINE

Authors Full Name

Ko, Jung-Keun; Lamichhane, Dirga Kumar; Kim, Hwan-Cheol; Leem, Jong-Han.

Institution

Ko, Jung-Keun. Department of Social and Preventive Medicine, School of Medicine, Inha University, Incheon 22212, Korea. ecokjk@naver.com. Lamichhane, Dirga Kumar. Department of Social and Preventive Medicine, School of Medicine, Inha University, Incheon 22212, Korea. dirgalamichhane@gmail.com.

Kim, Hwan-Cheol. Department of Social and Preventive Medicine, School of Medicine, Inha University, Incheon 22212, Korea. cheol17@hanmail.net.

Kim, Hwan-Cheol. Department of Occupational and Environmental Medicine, School of Medicine, Inha University, Incheon 22212, Korea. cheol17@hanmail.net.

Leem, Jong-Han. Department of Social and Preventive Medicine, School of Medicine, Inha University, Incheon 22212, Korea. keeper21@naver.com.

Leem, Jong-Han. Department of Occupational and Environmental Medicine, School of Medicine, Inha University, Incheon 22212, Korea. keeper21@naver.com.

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Year of Publication

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266.

Bladder Re-augmentation in Classic Bladder Exstrophy: Risk Factors and Prevention.
Benz KS; Jayman J; Doersch K; Maruf M; Baumgartner T; Kasprenski M; Gearhart JP.
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MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present
Urology. 115:157-161, 2018 May.

[Journal Article]

UI: 29447946

OBJECTIVE: To characterize the causes of re-augmentation in patients with classic bladder exstrophy (CBE).

METHODS: A prospectively maintained institutional database of 1327 exstrophy-epispadias complex patients was reviewed for patients with CBE who underwent more than 1 augmentation cystoplasty (AC) procedure. Data regarding bladder capacities, complications following AC, and reasons for re-augmentation were evaluated.

RESULTS: A total of 166 patients with CBE underwent AC. Of these, 67 (40.4%) were included in the control group and 17 (10%) patients underwent a re-augmentation. There were several indications for re-augmentation including continued small bladder capacity (17 of 17), inadequate bladder necks (8 of 17), failed rattail augmentation (2 of 17), stomal incontinence (1 of 17), a urethrocutaneous fistula (1 of 17), and an hourglass augmentation (1 of 17). Of note, 5 of the 17 patients (29%) had a re-augmentation procedure with a ureteral reimplantation. The sigmoid colon was the most commonly used bowel segment in the failed initial AC (8 patients), whereas the ileum was the most commonly used segment during re-augmentation (12 patients). In the re-augmentation cohort, the mean amount of bowel used during the first AC procedure was 12 cm (standard deviation [SD] 3.6) compared with 19 cm (SD 5.0) during re-augmentation. The mean amount of bowel used for control group augmentations was 20.8 cm (SD 4). The mean re-augmentation preoperative bladder capacity of 100 mL (SD 60) immediately increased after re-augmentation to 180.8 mL (SD 56.4) ($P = .0001$).

CONCLUSION: Bladder re-augmentation is most commonly required in the setting of a small bladder capacity after an initial AC, when an insufficient amount of bowel is used during the first AC procedure.

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Status

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Authors Full Name

Benz, Karl S; Jayman, John; Doersch, Karen; Maruf, Mahir; Baumgartner, Timothy; Kasprenski, Matthew; Gearhart, John P.

Institution

Benz, Karl S. Robert D. Jeffs Division of Pediatric Urology, James Buchanan Brady Urological Institutions, Johns Hopkins Hospital, Johns Hopkins Medical Institutions, Charlotte Bloomberg Children's Hospital, Baltimore, MD. Jayman, John. Robert D. Jeffs Division of Pediatric Urology,

James Buchanan Brady Urological Institutions, Johns Hopkins Hospital, Johns Hopkins Medical Institutions, Charlotte Bloomberg Children's Hospital, Baltimore, MD.

Doersch, Karen. Robert D. Jeffs Division of Pediatric Urology, James Buchanan Brady Urological Institutions, Johns Hopkins Hospital, Johns Hopkins Medical Institutions, Charlotte Bloomberg Children's Hospital, Baltimore, MD; Texas A&M Health Science Center College of Medicine, Temple, TX.

Maruf, Mahir. Robert D. Jeffs Division of Pediatric Urology, James Buchanan Brady Urological Institutions, Johns Hopkins Hospital, Johns Hopkins Medical Institutions, Charlotte Bloomberg Children's Hospital, Baltimore, MD.

Baumgartner, Timothy. Robert D. Jeffs Division of Pediatric Urology, James Buchanan Brady Urological Institutions, Johns Hopkins Hospital, Johns Hopkins Medical Institutions, Charlotte Bloomberg Children's Hospital, Baltimore, MD.

Kasprenski, Matthew. Robert D. Jeffs Division of Pediatric Urology, James Buchanan Brady Urological Institutions, Johns Hopkins Hospital, Johns Hopkins Medical Institutions, Charlotte Bloomberg Children's Hospital, Baltimore, MD.

Gearhart, John P. Robert D. Jeffs Division of Pediatric Urology, James Buchanan Brady Urological Institutions, Johns Hopkins Hospital, Johns Hopkins Medical Institutions, Charlotte Bloomberg Children's Hospital, Baltimore, MD. Electronic address: Jgearha2@jhmi.edu.

Year of Publication

2018

267.

Cystoscopic-assisted laparoscopic excision of prostatic utricle.

Mostafa IA; Woodward MN; Shalaby MS.

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Journal of pediatric urology. 14(1):77-78, 2018 02.

[Journal Article]

UI: 29137943

We present a video of our technique for resection of a large prostatic utricle (PU) in a patient who presented initially with disordered sexual development. His karyotype was 46XY, and phenotypically had penoscrotal hypospadias, bifid scrotum, and retractile right testis. An initial

micturating cystourethrogram (MCUG) demonstrated the utricle but failed to cannulate the bladder. Being asymptomatic, we carried out staged repair of his hypospadias. Later, he started to have recurrent epididymo-orchitis with resistance to multiple antibiotics. Examination under anaesthesia was done and ruled out meatal or neo-urethral strictures. A subsequent MCUG demonstrated the large utricle and its relation to the bladder. We carried out a cystoscopic-assisted laparoscopic excision. There has been no consensus about the best surgical approach to resect a PU and most known procedures involved extensive pelvic dissection and carried a significant risk of damage to the pelvic nerves. The laparoscopic approach seems to be promising in this field as it provides proper view of the deep pelvis with reasonable magnification, less dissection and shorter postoperative pain and scarring. Cystoscopic assistance in this technique was a great addition to provide counter-traction movement and facilitate proper dissection. Copyright © 2017 Journal of Pediatric Urology Company. Published by Elsevier Ltd. All rights reserved.

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MEDLINE

Authors Full Name

Mostafa, Ibrahim A; Woodward, Mark N; Shalaby, Mohamed S.

Institution

Mostafa, Ibrahim A. Department of Paediatric Surgery, Bristol Royal Hospital for Children, Bristol, UK. Woodward, Mark N. Department of Paediatric Surgery, Bristol Royal Hospital for Children, Bristol, UK.

Shalaby, Mohamed S. Department of Paediatric Surgery, Bristol Royal Hospital for Children, Bristol, UK; Department of Paediatric Surgery, Ain Shams University, Cairo, Egypt. Electronic address: mshalaby@doctors.org.uk.

Year of Publication

2018

268.

Reconstructive surgery for recurrent penile curvature.

Takeda M; Seo S; Sueyoshi R; Nakamura H; Suda K; Lane GJ; Yamataka A.

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MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Pediatric Surgery International. 34(2):245-248, 2018 Feb.

[Journal Article]

UI: 29058072

PURPOSE: There are surprisingly few reports about reconstructive surgery for severe recurrent/persistent penile curvature (redo-PC). We present our experience.

METHODS: We reviewed 9 redo-PC cases we treated between 1998 and 2016.

RESULTS: Cases 1-3 and 5 were identified from 111 consecutive hypospadias patients we treated between 1998 and 2016 (4/111; 3.6%). Cases 4 and 6-9 had initial surgery elsewhere. Initial PC was severe ($> 45^\circ$; $n = 5$), moderate (30° - 45° ; $n = 1$), or unknown ($n = 3$), treated by dorsal plication (DP) in 4/9 (cases 1-4), chordectomy in 2/9 (cases 5, 6), and unknown in 3/9 (cases 7-9); no case had tunica albuginea incision (TAI). Straightening after initial surgery was confirmed by artificial erection (AE) in 4/9, not confirmed (2/9), and unknown (3/9). Cases 1, 2, 7 and 8 had had previous failed redos. Scarring of buccal mucosa used for urethroplasty caused worse PC in cases 7 and 9. After TAI ($n = 6$; cases 1, 3-6, and 8) or scar removal with DP ($n = 3$; cases 2, 7, and 9), AE confirmed successful straightening in all cases, without sequelae after a mean follow-up of 2.6 years.

CONCLUSION: TAI was most effective for redo-PC surgery. Preoperative AE and examination under anesthesia should be used to customize treatment.

Version ID

1

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Authors Full Name

Takeda, Masahiro; Seo, Shogo; Sueyoshi, Ryo; Nakamura, Hiroki; Suda, Kazuto; Lane, Geoffrey J; Yamataka, Atsuyuki.

Institution

Takeda, Masahiro. Department of Pediatric General and Urogenital Surgery, Juntendo University School of Medicine, 2-1-1 Hongo, Bunkyo-ku, Tokyo, 113-8421, Japan.

mstakeda@juntendo.ac.jp. Seo, Shogo. Department of Pediatric General and Urogenital Surgery, Juntendo University School of Medicine, 2-1-1 Hongo, Bunkyo-ku, Tokyo, 113-8421, Japan.

Sueyoshi, Ryo. Department of Pediatric General and Urogenital Surgery, Juntendo University School of Medicine, 2-1-1 Hongo, Bunkyo-ku, Tokyo, 113-8421, Japan.

Nakamura, Hiroki. Department of Pediatric General and Urogenital Surgery, Juntendo University School of Medicine, 2-1-1 Hongo, Bunkyo-ku, Tokyo, 113-8421, Japan.

Suda, Kazuto. Department of Pediatric General and Urogenital Surgery, Juntendo University School of Medicine, 2-1-1 Hongo, Bunkyo-ku, Tokyo, 113-8421, Japan.

Lane, Geoffrey J. Department of Pediatric General and Urogenital Surgery, Juntendo University School of Medicine, 2-1-1 Hongo, Bunkyo-ku, Tokyo, 113-8421, Japan.

Yamataka, Atsuyuki. Department of Pediatric General and Urogenital Surgery, Juntendo University School of Medicine, 2-1-1 Hongo, Bunkyo-ku, Tokyo, 113-8421, Japan.

Year of Publication

2018

269.

Prenatal exposure to glycol ethers and cryptorchidism and hypospadias: a nested case-control study.

Warembourg C; Botton J; Lelong N; Rouget F; Khoshnood B; Le Gleau F; Monfort C; Labat L; Pierre F; Heude B; Slama R; Multigner L; Charles MA; Cordier S; Garlantezec R.

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Occupational & Environmental Medicine. 75(1):59-65, 2018 01.

[Journal Article. Research Support, Non-U.S. Gov't]

UI: 29055888

OBJECTIVES: Glycol ethers (GE) are oxygenated solvents frequently found in occupational and consumer products. Some of them are well-known testicular and developmental animal toxicants. This study aims to evaluate the risk of male genital anomalies in association with prenatal exposure to GE using urinary biomarkers of exposure.

METHODS: We conducted a case-control study nested in two joint mother-child cohorts (5303 pregnant women). Cases of cryptorchidism and hypospadias were identified at birth and confirmed during a 2-year follow-up period (n=14 cryptorchidism and n=15 hypospadias). Each case was matched to three randomly selected controls within the cohorts for region of inclusion and gestational age at urine sampling. Concentrations of five GE acidic metabolites were measured in spot maternal urine samples collected during pregnancy. ORs were estimated with multivariate conditional logistic regressions including a Firth's penalisation.

RESULTS: Detection rates of urinary GE metabolites ranged from 8% to 93% and only two were sufficiently detected (>33%) in each cohort to be studied: methoxyacetic acid (MAA) and

phenoxyacetic acid (PhAA). A significantly higher risk of hypospadias was associated with the highest tertile of exposure to MAA: OR (95% CI) 4.5(1.4 to 23.4). No association were observed with urinary concentration of PhAA, nor with the risk of cryptorchidism.

CONCLUSIONS: In view of the toxicological plausibility of our results, this study, despite its small sample size, raises concern about the potential developmental toxicity of MAA on the male genital system and calls for thorough identification of current sources of exposure to MAA.

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MEDLINE

Authors Full Name

Warembourg, Charline; Botton, Jeremie; Lelong, Nathalie; Rouget, Florence; Khoshnood, Babak; Le Gleau, Florent; Monfort, Christine; Labat, Laurence; Pierre, Fabrice; Heude, Barbara; Slama, Remy; Multigner, Luc; Charles, Marie-Aline; Cordier, Sylvaine; Garlantezec, Ronan.

Institution

Warembourg, Charline. Exposure Assessment and Epidemiological Research on Environment, Reproduction and Development, INSERM UMR 1085 IRSET, Rennes, France. Warembourg, Charline. Universite de Rennes I, Rennes, France.

Botton, Jeremie. Center for Epidemiology and Statistics Sorbonne Paris Cite, INSERM UMR 1153, Team: Early Origin of the Child's Health and Development, Paris Descartes University, Villejuif, France.

Botton, Jeremie. Faculty of Pharmacy, University Paris Saclay, Malabry, France.

Lelong, Nathalie. Center for Epidemiology and Statistics Sorbonne Paris Cite, DHU Risks in Pregnancy, INSERM UMR 1153, Team: Obstetrical, Perinatal and Pediatric Epidemiology Research, Paris Descartes University, Paris, France.

Rouget, Florence. Exposure Assessment and Epidemiological Research on Environment, Reproduction and Development, INSERM UMR 1085 IRSET, Rennes, France.

Rouget, Florence. Universite de Rennes I, Rennes, France.

Rouget, Florence. Department of Pediatrics, University Hospital of Rennes, Brittany Registry of Congenital Malformations, Rennes, France.

Khoshnood, Babak. Center for Epidemiology and Statistics Sorbonne Paris Cite, DHU Risks in Pregnancy, INSERM UMR 1153, Team: Obstetrical, Perinatal and Pediatric Epidemiology Research, Paris Descartes University, Paris, France.

Le Gleau, Florent. LABOCEA Laboratory, Plouzane, France.

Monfort, Christine. Exposure Assessment and Epidemiological Research on Environment, Reproduction and Development, INSERM UMR 1085 IRSET, Rennes, France.

Monfort, Christine. Universite de Rennes I, Rennes, France.

Labat, Laurence. Laboratoire de Pharmacologie Toxicologie, Assistance Publique-Hopitaux de Paris, Groupe Cochin, Paris, France.

Pierre, Fabrice. Service de Gynecologie Obstetrique et Medecine de la Reproduction, Centre Hospitalier Universitaire de Poitiers, Poitiers, France.

Heude, Barbara. Center for Epidemiology and Statistics Sorbonne Paris Cite, INSERM UMR 1153, Team: Early Origin of the Child's Health and Development, Paris Descartes University, Villejuif, France.

Slama, Remy. Department of Environmental Epidemiology, INSERM, CNRS, University Grenoble-Alpes, IAB Institute of Advanced Biosciences Joint Research Center (U1209), Grenoble, France.

Multigner, Luc. Exposure Assessment and Epidemiological Research on Environment, Reproduction and Development, INSERM UMR 1085 IRSET, Rennes, France.

Multigner, Luc. Universite de Rennes I, Rennes, France.

Charles, Marie-Aline. Center for Epidemiology and Statistics Sorbonne Paris Cite, INSERM UMR 1153, Team: Early Origin of the Child's Health and Development, Paris Descartes University, Villejuif, France.

Cordier, Sylvaine. Exposure Assessment and Epidemiological Research on Environment, Reproduction and Development, INSERM UMR 1085 IRSET, Rennes, France.

Garlantezec, Ronan. Exposure Assessment and Epidemiological Research on Environment, Reproduction and Development, INSERM UMR 1085 IRSET, Rennes, France.

Garlantezec, Ronan. Universite de Rennes I, Rennes, France.

Garlantezec, Ronan. Service de Sante Publique et d'Epidemiologie, Centre Hospitalo-Universitaire de Rennes, Rennes, France.

Comments

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Year of Publication

2018

The role of preoperative intra muscular testosterone in improving functional and cosmetic outcomes following hypospadias repair: A prospective randomized study.

Babu R; Chakravarthi S.

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Journal of pediatric urology. 14(1):29.e1-29.e6, 2018 02.

[Journal Article. Randomized Controlled Trial]

UI: 28844754

BACKGROUND: There are very few well-designed studies on the role of preoperative testosterone in hypospadias repair. The aims of the current study are to ascertain the efficacy of intramuscular testosterone in increasing the glans width, reducing the complications and improving the functional and cosmetic outcomes.

MATERIALS AND METHODS: Between January 2013 and December 2016, a total of 200 patients with distal hypospadias were recruited to this prospective randomized study. Only patients with distal hypospadias eligible for a tubularized incised plate (TIP) repair were included. Group 1 did not receive any intervention, and group 2 received three doses of intramuscular testosterone enanthate 2 mg/kg at the age of 9, 10, and 11 months. Glans width was measured in millimeters in all patients using calipers at the time of recruitment and at the time of surgery. Within group 2, those who failed to have more than a 2 mm increase in size were considered non-responders (group 2a), and the remaining were taken to be responders (group 2b). TIP repair was performed by the same surgeon, and patients were followed up, looking for urethrocutaneous fistula, glans dehiscence, meatal stenosis, etc. Cosmetic evaluation was performed using the Parental Penile Perception Score (PPPS).

RESULTS: A total of eight patients dropped out in group 1 (n = 92), while six dropped out in group 2 (n = 94). In group 2, 17% patients did not respond to testosterone (group 2a; n = 16), whereas 83% showed significant increase in glans width (group 2b; n = 78). The table compares the functional and cosmetic outcomes between groups. Total complications were significantly less in group 2b (17.9%) than in group 2a (50%). The reoperation rate was significantly less in group 2b (11.5%) than in group 1 (23.1%). A significantly higher number of parents (p = 0.03) were satisfied with cosmesis of the glans/meatus in group 2b (71/78; 91%) compared with group 2a (11/16; 69%) or group 1 (72/92; 78%). The overall PPPS was significantly higher (p = 0.003) in group 2b (8.88 +/- 2.20) than in group 1 (8.03 +/- 1.55).

DISCUSSION: Two systematic reviews on the role of preoperative testosterone failed to reach any conclusion. One described a glans width <14 mm as a risk factor for urethral complications after hypospadias repair but another did not support this. Our findings reveal that preoperative testosterone significantly increases the glans width thereby reducing the complications and requirement for reoperation. It also improves the cosmetic outcomes and parental satisfaction.

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Version ID

1

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Authors Full Name

Babu, Ramesh; Chakravarthi, Sibi.

Institution

Babu, Ramesh. Department of Pediatric Urology, Sri Ramachandra Medical College and Research Institute, Porur, Chennai, India. Electronic address: drrameshbabu1@gmail.com.
Chakravarthi, Sibi. Department of Pediatric Urology, Sri Ramachandra Medical College and Research Institute, Porur, Chennai, India.

Comments

Comment in (CIN) Comment in (CIN)

Year of Publication

2018

271.

Association between diacylglycerol kinase kappa variants and hypospadias susceptibility in a Han Chinese population.

Xie H; Lin XL; Zhang S; Yu L; Li XX; Huang YC; Lyu YQ; Chen HT; Xu J; Chen F.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid

MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Asian Journal of Andrology. 20(1):85-89, 2018 Jan-Feb.

[Journal Article]

UI: 28597849

Previous genome-wide association studies have identified variants in the diacylglycerol kinase kappa (DGKK) gene associated with hypospadias in populations of European descent. However, no variants of DGKK were confirmed to be associated with hypospadias in a recent Han Chinese study population, likely due to the limited number of single-nucleotide polymorphisms (SNPs) included in the analysis. In this study, we aimed to address the inconsistent results and evaluate

the association between DGKK and hypospadias in the Han Chinese population through a more comprehensive analysis of DGKK variants. We conducted association analyses for 17 SNPs in or downstream of DGKK with hypospadias among 322 cases (58 mild, 113 moderate, 128 severe, and 23 unknown) and 1008 controls. Five SNPs (rs2211122, rs4554617, rs7058226, rs7063116, and rs5915254) in DGKK were significantly associated with hypospadias ($P < 0.05$), with odds ratios (ORs) of 1.64-1.76. When only mild and moderate cases were compared to controls, 10 SNPs in DGKK were significant ($P < 0.05$), with ORs of 1.56-2.13. No significant SNP was observed when only severe cases were compared to controls. This study successfully implicated DGKK variants in hypospadias risk among a Han Chinese population, especially for mild/moderate cases. Severe forms of hypospadias are likely due to other genetic factors.

Version ID

1

Status

MEDLINE

Authors Full Name

Xie, Hua; Lin, Xiao-Ling; Zhang, Song; Yu, Ling; Li, Xiao-Xi; Huang, Yi-Chen; Lyu, Yi-Qing; Chen, Hai-Tao; Xu, Jianfeng; Chen, Fang.

Institution

Xie, Hua. Department of Urology, Shanghai Children's Hospital, Shanghai Jiao Tong University, Shanghai 200062, China. Lin, Xiao-Ling. Fudan Institute of Urology, Huashan Hospital, Fudan University, Shanghai 200040, China.

Zhang, Song. State Key Laboratory of Genetic Engineering, School of Life Sciences, Fudan University, Shanghai 200433, China.

Yu, Ling. Department of Urology, Shanghai Children's Hospital, Shanghai Jiao Tong University, Shanghai 200062, China.

Li, Xiao-Xi. Department of Urology, Shanghai Children's Hospital, Shanghai Jiao Tong University, Shanghai 200062, China.

Huang, Yi-Chen. Department of Urology, Shanghai Children's Hospital, Shanghai Jiao Tong University, Shanghai 200062, China.

Lyu, Yi-Qing. Department of Urology, Shanghai Children's Hospital, Shanghai Jiao Tong University, Shanghai 200062, China.

Chen, Hai-Tao. Center for Genomic Translational Medicine and Prevention, School of Public Health, Fudan University, Shanghai 200433, China.

Xu, Jianfeng. Fudan Institute of Urology, Huashan Hospital, Fudan University, Shanghai 200040, China.

Xu, Jianfeng. Center for Genomic Translational Medicine and Prevention, School of Public Health, Fudan University, Shanghai 200433, China.

Xu, Jianfeng. Program for Personalized Cancer Care, NorthShore University HealthSystem, Evanston, IL 60201, USA.

Chen, Fang. Department of Urology, Shanghai Children's Hospital, Shanghai Jiao Tong University, Shanghai 200062, China.

PMID

<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5753560>

Year of Publication

2018

272.

A comparative study of surgical outcomes using a Colorado microdissection needle versus standard-size needle electrocautery in one-stage hypospadias repair using a transverse preputial island flap.

Liang WQ; Qiu YH; Ji CY; Chen YH; Zhang JQ; Yao YY; Zhang JM.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid

MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Minerva Pediatrica. 70(1):27-34, 2018 Feb.

[Comparative Study. Journal Article]

UI: 26154528

BACKGROUND: The purpose of this study was to compare the surgical outcomes of a Colorado microdissection needle (CMN) with that of a standard-size electrocautery needle in one-stage hypospadias repair using a transverse preputial island flap (TPIF).

METHODS: The records of patients who received hypospadias repair from September 2012 to October 2013 were retrospectively reviewed. Patients were divided into a group that received repair using a CMN and those in which a standard-size electrocautery needle was used. Data collected and compared included age, types of hypospadias, duration of surgery, intraoperative blood loss, and postoperative edema and complications.

RESULTS: There were 51 patients in the CMN group and 44 in the standard needle group, and the groups were similar with respect to age and type of hypospadias. The median surgery time for the CMN group was significantly shorter than that of the standard group (15.7 minutes vs. 20.6 minutes, respectively, $P < 0.001$). At postoperative day 7 and day 30, the CMN group had significantly less patients with edema than the standard needle group (31.4% vs. 65.9%, $P < 0.01$;

and 37.3% vs. 79.5%, $P < 0.001$, respectively). The overall complication rate has no significant difference between two groups.

CONCLUSIONS: The use of CMN for tissue dissection and separation in hypospadias repair can facilitate foreskin degloving, shape the flap in a more efficient way, and help maintain adequate blood supply for the new urethra and its skin coverage.

Version ID

1

Status

MEDLINE

Authors Full Name

Liang, Wei-Qiang; Qiu, Yi-Hong; Ji, Chen-Yang; Chen, Yu-Hong; Zhang, Jia-Qi; Yao, Yuan-Yuan; Zhang, Jin-Ming.

Institution

Liang, Wei-Qiang. Department of Plastic and Reconstructive Surgery, Sun Yat-Sen Memorial Hospital of Sun Yat-Sen University, Guangzhou, Guangdong Province, China. Qiu, Yi-Hong. Department of Plastic and Reconstructive Surgery, Sun Yat-Sen Memorial Hospital of Sun Yat-Sen University, Guangzhou, Guangdong Province, China.

Ji, Chen-Yang. Department of Plastic and Reconstructive Surgery, Sun Yat-Sen Memorial Hospital of Sun Yat-Sen University, Guangzhou, Guangdong Province, China.

Chen, Yu-Hong. Department of Plastic and Reconstructive Surgery, Sun Yat-Sen Memorial Hospital of Sun Yat-Sen University, Guangzhou, Guangdong Province, China.

Zhang, Jia-Qi. Department of Plastic and Reconstructive Surgery, Sun Yat-Sen Memorial Hospital of Sun Yat-Sen University, Guangzhou, Guangdong Province, China.

Yao, Yuan-Yuan. Department of Plastic and Reconstructive Surgery, Sun Yat-Sen Memorial Hospital of Sun Yat-Sen University, Guangzhou, Guangdong Province, China.

Zhang, Jin-Ming. Department of Plastic and Reconstructive Surgery, Sun Yat-Sen Memorial Hospital of Sun Yat-Sen University, Guangzhou, Guangdong Province, China -

40381594@qq.com.

Year of Publication

2018

Needs Assessment in Care of Adults With Anorectal Malformations and Exstrophy-Epispadias Complex in Germany.

Ebert AK; Zwink N; Schwarzer N; Brunner L; Reutter H; Jenetzky E; Huber J; Ludwikowski B.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid

MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Frontiers in Pediatrics. 6:392, 2018.

[Journal Article]

UI: 30619788

Introduction: Medical needs of adults with anorectal malformations (ARM) and the exstrophy-epispadias complex (EEC) are not fully understood. Therefore, the aim of the study was to evaluate how affected individuals get along with the current national medical care and what their medical or social long-term requirements are. Patients and Methods: Between 11/2014-07/2016 all adult members (≥ 18 years, ARM $n = 113$, EEC $n = 126$) of the German self-help organizations SoMA e.V. and Blasenexstrophie/Epispadie e.V. were contacted via email or post and asked to fill out an anonymous online questionnaire regarding medical requirements, treatment satisfaction, daily life impairment and expectations regarding physicians soft skills. The results were compared between both groups and male and female participants.

Results: 56 participants with ARM (median age 26 years, IQR 19-38) and 52 participants with EEC (median age 31 years, IQR 22-37) filled in the questionnaire completely. Forty-five percent of the ARM and 67% of the EEC participants contacted an urologist. A general surgeon was visited by 23% of the ARM individuals, a paediatric surgeon by 20%. Although 60% of the females with ARM and 82% of the females with EEC assessed gynecological counseling as helpful or neutral, a small subgroup of ARM females ($n = 6$, 16%; 70% non-isolated ARM or ARM with Hirschsprung disease and additional associated anomalies) were not satisfied. The majority of both groups reported no or only minor daily life impairment ($p = 0.38$). Professional knowledge, paying attention to patients' concerns, having empathy and taking enough time was important for over 90% of all participants. Thirty-eight percent of the ARM and 27% of the EEC individuals needed psychological support. Most medical consultations were judged to be helpful. Conclusion: Although adults with ARM and EEC being a self-help organization member and thus well informed and generally cope well, participants expressed their wish for expert counseling regarding family planning, reconstructive procedures, continence management, urological care and social welfare issues. Furthermore, specific expert consultations for gynecological issues in a subgroup of ARM females, mainly non-isolated, might be required. Actual needs of adults with rare conditions must be better clarified to improve medical care beyond childhood and adolescence.

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1

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PubMed-not-MEDLINE

Authors Full Name

Ebert, Anne Karoline; Zwink, Nadine; Schwarzer, Nicole; Brunner, Lilian; Reutter, Heiko;

Jenetzky, Ekkehart; Huber, Johannes; Ludwikowski, Barbara.

Institution

Ebert, Anne Karoline. Department of Pediatric Urology, University Hospital for Urology and Pediatric Urology Ulm, Ulm, Germany. Zwink, Nadine. Department of Pediatric and Adolescent Psychiatry and Psychotherapy, University Medical Centre, Johannes Gutenberg University Mainz, Mainz, Germany.

Schwarzer, Nicole. SoMA e.V., Self-Help Organisation for People With Anorectal Malformation, Munich, Germany.

Brunner, Lilian. Self-Help Organisation for People With Bladder exstrophy-epispadias complex, Freital, Germany.

Reutter, Heiko. Institute of Human Genetics, University of Bonn, Bonn, Germany.

Reutter, Heiko. Department of Neonatology and Pediatric Intensive Care, Children's Hospital, University of Bonn, Bonn, Germany.

Jenetzky, Ekkehart. Department of Pediatric and Adolescent Psychiatry and Psychotherapy, University Medical Centre, Johannes Gutenberg University Mainz, Mainz, Germany.

Jenetzky, Ekkehart. Child Center Maulbronn GmbH, Hospital for Pediatric Neurology and Social Pediatrics, Maulbronn, Germany.

Huber, Johannes. University Hospital for Urology, University Medical Center Carl Gustav Carus, Dresden, Germany.

Ludwikowski, Barbara. Department of Pediatric Surgery and Pediatric Urology, Medical Center for Children and Adolescents AUF DER BULT, Hannover, Germany.

PMID

<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC6306024>

Year of Publication

2018

274.

Stricture Urethra in Children: An Indian Perspective.

Herle K; Jehangir S; Thomas RJ.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid

MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Journal of Indian Association of Pediatric Surgeons. 23(4):192-197, 2018 Oct-Dec.

[Journal Article]

UI: 30443113

Background: Pediatric urethral stricture and its treatment have functional implications in the growing child.

Subjects and Methods: A retrospective study of records on urethral strictures encountered in our institution between January 2005 and May 2016 yielded 23 boys against a backdrop of 19,250 admissions during the same period; stenosis and strictures after hypospadias repair were not included in this study. Demographic data were collected from the charts, and the success of repair was assessed clinically by success of repair was assessed clinically by observing for presence or absence of symptoms such as dribbling, straining at voiding, adequacy of urinary stream and radiologically by assessing the micturition phase of voiding cystourethrogram. Success was defined as successful initiation, flow, and completion of voiding with radiological evidence of reestablishment of urethral continuity.

Results: The most common cause of urethral stricture was perineal or pelvic trauma (56.5%). Three after surgery for anorectal malformation (13.04%) and 2 (8.6%) followed otherwise unspecified urethritis. Transperineal and transpubic anastomotic routes were used for surgery. Redo surgery was required in 47.8%. The overall success rate was 82%. A self-catheterizable mitrofanoff channel was created as part of the primary procedure in 63.6% (7/11) or after the failure of the first procedure in 36.3% (4/11).

Conclusion: The majority of urethral strictures are long-segment strictures or those with complete disruption not amenable to endoscopic techniques. The aim of the surgery is to obtain end-to-end opposition of healthy proximal and distal urethra. The route - transperineal or transpubic - which will give the best access to the ends of the urethra is determined by the location and extent of the stricture and the alteration in anatomy as a consequence of the pelvic fracture. Even after the introduction of laser and endoscopic techniques, surgical repair is required to tackle the majority of urethral strictures in children.

Version ID

1

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Authors Full Name

Herle, Koushik; Jehangir, Susan; Thomas, Reju J.

Institution

Herle, Koushik. Department of Pediatric Surgery and Paediatric Urology, Christian Medical College Hospital, Vellore, Tamil Nadu, India. Jehangir, Susan. Department of Pediatric Surgery and Paediatric Urology, Christian Medical College Hospital, Vellore, Tamil Nadu, India.

Thomas, Reju J. Department of Pediatric Surgery and Paediatric Urology, Christian Medical College Hospital, Vellore, Tamil Nadu, India.

PMID

<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC6182950>

Year of Publication

2018

275.

A rare variety of congenital adrenal hyperplasia with mosaic Klinefelter syndrome: a unique combination presenting with ambiguous genitalia and sexual precocity.

Shehab MA; Mahmood T; Hasanat MA; Fariduddin M; Ahsan N; Hossain MS; Hossain MS; Jahan S.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Endocrinology, Diabetes & Metabolism Case Reports. 2018(1):18-0108, 2018 Oct 13.

[Journal Article]

UI: 30328339

Congenital adrenal hyperplasia (CAH) due to the three-beta-hydroxysteroid-dehydrogenase (3beta-HSD) enzyme deficiency is a rare autosomal recessive disorder presenting with sexual precocity in a phenotypic male. Klinefelter syndrome (KS) is the most common sex chromosome aneuploidy presenting with hypergonadotropic hypogonadism in a male. However, only a handful of cases of mosaic KS have been described in the literature. The co-existence of mosaic KS with CAH due to 3beta-HSD enzyme deficiency portrays a unique diagnostic paradox where features of gonadal androgen deficiency are masked by simultaneous adrenal androgen excess. Here, we report a 7-year-old phenotypic male boy who, at birth presented with ambiguous genitalia, probably a microphallus with penoscrotal hypospadias. Later on, he developed accelerated growth with advanced bone age, premature pubarche, phallic enlargement and hyperpigmentation. Biochemically, the patient was proven to have CAH due to 3beta-HSD deficiency. However, the co-existence of bilateral cryptorchidism made us to consider the

possibility of hypogonadism as well, and it was further explained by concurrent existence of mosaic KS (47,XXY/46,XX). He was started on glucocorticoid and mineralocorticoid replacement and underwent right-sided orchidopexy on a later date. He showed significant clinical and biochemical improvement on subsequent follow-up. However, the declining value of serum testosterone was accompanied by rising level of FSH thereby unmasking hypergonadotropic hypogonadism due to mosaic KS. In future, we are planning to place him on androgen replacement as well. Learning points: ** Ambiguous genitalia with subsequent development of sexual precocity in a phenotypic male points towards some unusual varieties of CAH. ** High level of serum testosterone, adrenal androgen, plasma ACTH and low basal cortisol are proof of CAH, whereas elevated level of 17-OH pregnenolone is biochemical marker of 3beta-HSD enzyme deficiency. ** Final diagnosis can be obtained with sequencing of HSD3B2 gene showing various mutations. ** Presence of bilateral cryptorchidism in such a patient may be due to underlying hypogonadism. ** Karyotyping in such patient may rarely show mosaic KS (47,XXY/46,XX) and there might be unmasking of hypergonadotropic hypogonadism resulting from adrenal androgen suppression from glucocorticoid treatment.

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Version ID

1

Status

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Authors Full Name

Shehab, M A; Mahmood, Tahseen; Hasanat, M A; Fariduddin, Md; Ahsan, Nazmul; Hossain, Mohammad Shahnoor; Hossain, Md Shahdat; Jahan, Sharmin.

Institution

Shehab, M A. Department of Endocrinology, BSMMU, Dhaka, Bangladesh. Mahmood, Tahseen. Department of Endocrinology, BSMMU, Dhaka, Bangladesh.

Hasanat, M A. Department of Endocrinology, BSMMU, Dhaka, Bangladesh.

Fariduddin, Md. Department of Endocrinology, BSMMU, Dhaka, Bangladesh.

Ahsan, Nazmul. Department of Genetic Engineering & Biotechnology, University of Dhaka, Dhaka, Bangladesh.

Hossain, Mohammad Shahnoor. Department of Genetic Engineering & Biotechnology, University of Dhaka, Dhaka, Bangladesh.

Hossain, Md Shahdat. Department of Genetic Engineering & Biotechnology, University of Dhaka, Dhaka, Bangladesh.

Jahan, Sharmin. Department of Endocrinology, BSMMU, Dhaka, Bangladesh.

PMID

<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC6198180>

Year of Publication

2018

276.

Caudal Analgesia for Hypospadias in Pediatrics: Comparative Evaluation of Adjuvants Dexamethasone and Dexmedetomidine Combination versus Dexamethasone or Dexmedetomidine to Bupivacaine: A Prospective, Double-Blinded, Randomized Comparative Study.

Hassan PF; Hassan AS; Elmetwally SA.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Albang Maqalat Wa Abhat Fi Altahdar Waalinas. 12(3):644-650, 2018 Jul-Sep.

[Journal Article]

UI: 30283169

Background: Caudal block is the most commonly used regional anesthetic technique in pediatric surgeries; different additives have been used for better and safer outcome.

Aim: The aim of this study is to compare the combination of dexamethasone and dexmedetomidine as adjuvants to bupivacaine versus using each agent solely with bupivacaine in pediatric caudal block as regards their efficiency in pain relief (the duration of postoperative analgesia, first time to request analgesia, and modified objective pain score [MOPS]).

Study Design: This was a prospective, double-blinded, randomized study.

Patients and Methods: Patients and Methods: Sixty-three children scheduled for hypospadias surgery wererandomized into three groupsaccording to the adjuvant drug added to caudal bupivacaine : Group I (n = 21): dexamethasone 0.1 mg/kg + 0.5 mg/kg bupivacaine 0.25%, Group II (n = 21): dexmedetomidine0.01 mug/kg + 0.5 mg/kg bupivacaine 0.25% and Group III (n = 21): dexamethasone 0.1 mg/kg + dexmedetomidine 0.01 mug/kg + 0.5 mg/kgbupivacaine 0.25%.

Intraoperative and postoperative hemodynamics were recorded. In postoperative anesthesia care unit and then the ward, MOPS and sedation score were recorded at 30 min and 1, 2, 3, 6 and 12 h. Further, the time of first analgesic request and side effects were recorded.

Statistical Analysis: Categorical data were presented as frequencies (%) and analyzed using Chi-square test. Continuous data were presented as mean (standard deviation) and median

(quartiles). Continuous data were analyzed using one-way analysis of variance for single measures and two-way mixed model for repeated measures. Kaplan-Meier analysis was performed for the duration of analgesia.

Results: In Group III, MOPS was lower than Groups I and II at the study times. Further, Group III had prolonged time for first request of analgesic. Sedation scores were prolonged in Group III and Group II than in Group I. There was a reduction in heart rates in Group III more than Group I and Group II but with no significant difference. However, there was a significant reduction in mean arterial blood pressure 30 min intraoperatively and postoperatively in Group III as compared to Groups I and II.

Conclusion: The addition of combined dexmedetomidine at a dose of 1 mug/kg and dexamethasone 0.1 mg/kg to caudal bupivacaine seemed to be an attractive alternative to each drug if used alone with more prolonged analgesia and almost no adverse effects.

Version ID

1

Status

PubMed-not-MEDLINE

Authors Full Name

Hassan, Passaint Fahim; Hassan, Amany Saleh; Elmetwally, Sarah Abdelsalam.

Institution

Hassan, Passaint Fahim. Department of Anesthesia and Surgical Intensive Care Unit, Kasr Alaini Hospital, Cairo University, El Haram, Giza, Egypt. Hassan, Amany Saleh. Department of Anesthesia and Surgical Intensive Care Unit, Kasr Alaini Hospital, Cairo University, El Haram, Giza, Egypt.

Elmetwally, Sarah Abdelsalam. Department of Anesthesia and Surgical Intensive Care Unit, Kasr Alaini Hospital, Cairo University, El Haram, Giza, Egypt.

PMID

<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC6157238>

Year of Publication

2018

Comparison of Anatomical Landmarks and Dimensions in a Hypospadiac Glans with Those of a Normal Glans.

Dhua AK; Anand S; Agarwala S; Bhatnagar V.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid

MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Journal of Indian Association of Pediatric Surgeons. 23(3):144-147, 2018 Jul-Sep.

[Journal Article]

UI: 30050263

Aim of the Study: The aim of this study is to establish ventral glans length (VGL), meatus (M) size, and their ratio VGL/M (R) in normal Indian boys and to compare these with the respective equivalent dimensions in boys with distal and mid-penile hypospadias using anatomic landmarks. **Methods:** Normal boys were designated as Group A (n = 108), and the boys with hypospadias were designated as Group B (n = 81). The anatomical landmarks marked on the glans were measured using a digital camera and appropriate software.

Results: The mean age of the boys in Groups A and B was 4.26 +/- 3.59 (range 0.5-12) and 3.82 +/- 2.85 (0.7-11) years, respectively. The observed mean values in Group A for VGL-A, M-A, and R-A were 3.05 +/- 1.27 (range 1.19-8.09), 4.3 +/- 1.23 (range 1.61-7.04), and 0.8 +/- 0.45 (range 0.27-2.1) mm, respectively. The observed mean values in Group B for VGL-B, M-B, and R-B were 3.77 +/- 0.81 (range 12.41-5.2), 3.27 +/- 0.71 (range 2.25-5.34), and 1.17 +/- 0.26 (range 0.84-1.86) mm, respectively. Comparison of R-A and R-B showed a statistically significant difference ($P < 0.0001$).

Conclusions: The dimensions and the studied ratio between a hypospadiac glans and a normal glans differed significantly. The ratio between VGL and M was 0.8 in normal boys. In comparison, the ratio between the equivalent VGL and M in the hypospadiac boys was 1.17. During glansplasty in surgery for hypospadias, this factor should be taken into consideration to avoid the creation of long and tight ventral glans closure.

Version ID

1

Status

PubMed-not-MEDLINE

Authors Full Name

Dhua, Anjan Kumar; Anand, Sachit; Agarwala, Sandeep; Bhatnagar, Veereshwar.

Institution

Dhua, Anjan Kumar. Department of Pediatric Surgery, AIIMS, New Delhi, India. Anand, Sachit.

Department of Pediatric Surgery, AIIMS, New Delhi, India.

Agarwala, Sandeep. Department of Pediatric Surgery, AIIMS, New Delhi, India.

Bhatnagar, Veereshwar. Department of Pediatric Surgery, AIIMS, New Delhi, India.

PMID

<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC6042169>

Year of Publication

2018

278.

Best of the 2017 Society for Pediatric Urology Fall Congress.

Anonymous.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid
MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present
Reviews in Urology. 20(1):28-30, 2018.

[Journal Article]

UI: 29942199

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PubMed-not-MEDLINE

PMID

<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC6003301>

Year of Publication

2018

279.

What is the pathogenesis of proximal hypospadias?.

Acimi S.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid
MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Turkish Journal of Urology. 44(4):357-361, 2018 Jul.

[Journal Article]

UI: 29799398

OBJECTIVE: To report the information concerning the pathogenesis of proximal hypospadias and causes of curvature associated with it.

MATERIAL AND METHODS: From January 2009 to December 2015, 74 patients underwent repair of proximal hypospadias. In 70 patients we performed a systematic biopsies in the lateral areas of the urethral plate, as well as under this plate. The study of the histological structure of these areas was performed using routine staining with hematoxylin and eosin, and the Masson's trichrome which color the collagen fibers in blue and monoclonal antibody against alpha-smooth muscle actin.

RESULTS: This prospective study shows that the fibrotic tissue abnormally present on the ventral side of the penis consists of a mixture of fibrous connective tissue, nerve nets, short vessels, and smooth muscle fibers. In contrast to the scrotal dartos, penile dartos fascia does not contain smooth muscle fibers. Therefore, these fibers may come from a blood vessel or spongy tissue which existed during neonatal period in the distal part of the penis before disappearing. In addition, in 13 cases, the presence downstream of the urethral meatus, of a bifurcation of corpus spongiosum into two branches supposes that the corpus spongiosum is form by fusion around the urethra of two mesenchymal bodies. The arterial supply of this purely masculine formation originates from a new vascularization and it is probably, developed under secretion of androgens (angiogenic substances in target tissues).

CONCLUSION: These findings allow us to suggest that the proximal hypospadias is due to avascular necrosis of the distal part ie. poorly vascularized part of the corpus spongiosum.

Version ID

1

Status

PubMed-not-MEDLINE

Author Initials

Acimi, Smail; ORCID: <https://orcid.org/0000-0002-2764-0302>

Authors Full Name

Acimi, Smail.

Institution

Acimi, Smail. Department of Visceral Surgery, Children hospital Canastel, Faculty of Medicine, University of Oran, Oran, Algeria.

PMID

<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC6016654>

Year of Publication

2018

280.

Hernia uteri inguinalis in ovotesticular disorder of sexual differentiation: A rare complication and role of imaging.

Ponnapatapura J.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Indian Journal of Radiology & Imaging. 28(1):41-44, 2018 Jan-Mar.

[Journal Article]

UI: 29692525

Neonate with ambiguous genitalia can cause great apprehension for the family as well as for healthcare providers. We report a rare complication of delayed diagnosis of hernia uteri inguinalis in ovotesticular disorder of sexual differentiation (DSD) in 20-year-old male patient who presented with pain and swelling in left inguinal region since 1 month. He had a past surgical history of repair of hypospadias 10 years back. On imaging, the left inguinal hernia sac contained nonfunctioning uterus and one ovary in the left scrotal sac and one testis in the right scrotal sac. Further investigation confirmed genotypically female (46XX) with negative sex determining region-Y gene on fluorescence in situ hybridization. The patient was given psychiatric counseling and wished to remain as male. The left inguinal hernia was repaired with excision of nonfunctioning uterus, ovary, and fallopian tube. Hernia uteri inguinalis is rare complication seen in DSD with only three cases being reported worldwide thus far, including our case.

Version ID

1

Status

PubMed-not-MEDLINE

Authors Full Name

Ponnapatapura, Janardhana.

Institution

Ponnapatapura, Janardhana. Department of Radiodiagnosis, Kempegowda Institute of Medical Sciences, Bengaluru, Karnataka, India.

PMID

<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5894317>

Year of Publication

2018

281.

Transitioning patients with hypospadias and other penile abnormalities to adulthood: What to expect?. [Review]

Rourke K; Braga LH.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Canadian Urological Association Journal. 12(4 Suppl 1):S27-S33, 2018 Apr.

[Journal Article. Review]

UI: 29681271

Hypospadias patients presenting to adult urologists do so with a wide range of symptoms and problems, including urethral stricture (45-72%), lower urinary tract symptoms (with or without stricture) (50-82%), urethrocutaneous fistula (16-30%), persisting hypospadias (14-43%), micturition spraying (24%), ventral curvature (14-24%), urinary tract infection (15-25%), or lichen sclerosus (13%; range 8-43). Many of these men have concurrent complications as the result of multiple operations and a variety of techniques. Patients with childhood repairs performed by a pediatric urologist are often lost to followup during adolescence and will reemerge in adulthood after what appeared to be a successful pediatric single-stage repair, stressing the need for long-term followup and transitional care. One of the major challenges in successful transitional care is that patients can feel traumatized with feelings of hopelessness surrounding their defects, leaving them hesitant to seek care. As well, these patients often have little knowledge regarding the type of repair or original location of the meatus. Urethral stricture is the most common presenting complication and could be related to various factors, with the clear etiology still under debate. These strictures can fall under four categories based on length, location, and previous surgeries. To lessen the difficulties in transitioning hypospadias patients from pediatric to adult practitioners, followup throughout childhood and adolescence for physical examination, as well as uroflowmetry, is mandatory.

Version ID

1

Status

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Authors Full Name

Rourke, Keith; Braga, Luis H.

Institution

Rourke, Keith. Division of Urology, Department of Surgery, University of Alberta, Edmonton, AB.
Braga, Luis H. Department of Surgery, Division of Urology, McMaster University, Hamilton, ON;
Canada.

PMID

<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5926912>

Year of Publication

2018

282.

Long term follow up of proximal hypospadias repair-urethral stricture should be excluded in adults who present with epididymo-orchitis.

Bhandarkar KP; Kittur DH; Patil SV; Jadhav SS.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid

MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Turkish Journal of Urology. 44(2):162-165, 2018 Mar.

[Journal Article]

UI: 29511587

Objective: Epididymo-orchitis (EO) is a well-known complication of urinary tract infections (UTI) in children. This is commonly seen in children who had hypospadias repair or in those who had surgery for anorectal malformation especially when it results in urethral stricture. However EO occurring as a complication of urethral stricture in adults operated for hypospadias in childhood is not well documented in the literature.

Material and methods: This is a retrospective review of four adults who had proximal hypospadias repair in childhood. All four men presented to us with EO. They were thoroughly investigated to rule out presence of urethral stricture.

Results: Three patients had urethral stricture of which two responded well to dilatation and one required cystoscopy and visual internal urethrotomy. The fourth patient did not have urethral

stricture. EO in this patient is thought to be due to excessive straining during micturition causing reflux into seminal vesicles.

Conclusion: Urethral stricture should be excluded in any adult who had a hypospadias repair and presents with EO. Urethral strictures after hypospadias surgery respond well to dilatation and to endoscopic urethrotomy.

Version ID

1

Status

PubMed-not-MEDLINE

Authors Full Name

Bhandarkar, Kailas P; Kittur, Dinesh H; Patil, Santosh V; Jadhav, Sudhakar S.

Institution

Bhandarkar, Kailas P. Department of Urology, Ege University School of Medicine, Izmir, Turkey.

Kittur, Dinesh H. Department of Urology, Ege University School of Medicine, Izmir, Turkey.

Patil, Santosh V. Department of Urology, Ege University School of Medicine, Izmir, Turkey.

Jadhav, Sudhakar S. Department of Urology, Ege University School of Medicine, Izmir, Turkey.

PMID

<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5832379>

Year of Publication

2018

283.

Results of Onlay Preputial Flap Urethroplasty for the Single-Stage Repair of Mid- and Proximal Hypospadias.

Gonzalez R; Lingnau A; Ludwikowski BM.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid

MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Frontiers in Pediatrics. 6:19, 2018.

[Journal Article]

UI: 29473028

Aims: To report current results of preputial flap onlay urethroplasty using the principle of the total preputial flap (TPF) for the one-stage repair of mid- and proximal hypospadias.

Methods: This study was a retrospective chart review of patients in a prospectively kept database of all hypospadias operations performed at two institutions from January 1 2011 to August 2017.

Inclusion criteria: all patients who underwent hypospadias repair using a preputial only flap urethroplasty based on the principle of the TPF. Demographic data, duration of follow-up, complications, and reoperations were recorded. A successful result was considered to be a straight penis, a glanular meatus, and absence of voiding symptoms. Whenever possible an uroflow was obtained during the follow-up visits.

Results: Forty-nine children met the inclusion criteria. All patients had marked penile curvature. Three patients had chromosomal abnormalities. The mean age at the time of surgery was 22 months (11-110) and the mean duration of follow-up 23.4 months (1-79). In 48 cases, the urethral plate could be preserved without dividing it. The penile curvature was corrected with chordectomy alone in 10 patients, 38 required a dorsal plication of the tunica albuginea, and 1 required an additional ventral dermal graft. In 38 patients (77.5%), the initial operation was successful, and no further operations were needed. There were eight urethrocutaneous fistulas, three dehiscences of the glans approximation. One patient suffered a wound infection and partial loss of the flap.

Conclusion: One-stage repair of mid- and proximal hypospadias preserving the urethral plate and using a TPF for the urethroplasty and coverage of the ventral penis is successful in 77.5% of cases. Complications in the remaining patients were easily managed or did not require treatment. Compared to a planned two-stage approach, the technique described in this report resulted in significantly fewer procedures till complete resolution of the problem.

Version ID

1

Status

PubMed-not-MEDLINE

Authors Full Name

Gonzalez, Ricardo; Lingnau, Anja; Ludwikowski, Barbara Magda.

Institution

Gonzalez, Ricardo. Pediatric Surgery and Urology, Kinder- und Jugendkrankenhaus AUF DER BULT, Hanover, Germany. Lingnau, Anja. Department of Urology, Charite Universitätsmedizin Berlin, Berlin, Germany.

Ludwikowski, Barbara Magda. Pediatric Surgery and Urology, Kinder- und Jugendkrankenhaus AUF DER BULT, Hanover, Germany.

Comments

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PMID

<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5809423>

Year of Publication

284.

Human 3beta-hydroxysteroid dehydrogenase deficiency associated with normal spermatic numeration despite a severe enzyme deficit.

Donadille B; Houang M; Netchine I; Siffroi JP; Christin-Maitre S.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Endocrine Connections. 7(3):395-402, 2018 Mar.

[Journal Article]

UI: 29420188

Human 3 beta-hydroxysteroid dehydrogenase deficiency (3b-HSD) is a very rare form of congenital adrenal hyperplasia resulting from HSD3B2 gene mutations. The estimated prevalence is less than 1/1,000,000 at birth. It leads to steroidogenesis impairment in both adrenals and gonads. Few data are available concerning adult testicular function in such patients. We had the opportunity to study gonadal axis and testicular function in a 46,XY adult patient, carrying a HSD3B2 mutation. He presented at birth a neonatal salt-wasting syndrome. He had a micropenis, a perineal hypospadias and two intrascrotal testes. HSD3B2 gene sequencing revealed a 687del27 homozygous mutation. The patient achieved normal puberty at the age of 15 years. Transition from the paediatric department occurred at the age of 19 years. His hormonal profile under hydrocortisone and fludrocortisone treatments revealed normal serum levels of 17OH-pregnenolone, as well as SDHEA, ACTH, total testosterone, inhibin B and AMH. Pelvic ultrasound identified two scrotal testes of 21 mL each, without any testicular adrenal rest tumours. His adult spermatic characteristics were normal, according to WHO 2010 criteria, with a sperm concentration of 57.6 million/mL (N > 15), 21% of typical forms (N > 4%). Sperm vitality was subnormal (41%; N > 58%). This patient, in contrast to previous reports, presents subnormal sperm parameters and therefore potential male fertility in a 24-years-old patient with severe 3b-HSD deficiency. This case should improve counselling about fertility of male patients carrying HSD3B2 mutation.

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Version ID

Status

PubMed-not-MEDLINE

Authors Full Name

Donadille, Bruno; Houang, Muriel; Netchine, Irene; Siffroi, Jean-Pierre; Christin-Maitre, Sophie.

Institution

Donadille, Bruno. Service d'Endocrinologie et Medecine de la ReproductionCentre de Reference des Maladies Endocrines Rares de la Croissance, Hopital Saint Antoine, Groupe Hospitalier Universitaire Est, AP-HP, Paris, France bruno.donadille@aphp.fr. Houang, Muriel. Service d'Explorations Fonctionnelles EndocriniennesCentre de Reference des Maladies Endocrines Rares de la Croissance, Hopital Trousseau, Groupe Hospitalier Universitaire Est, AP-HP, Paris, France.

Netchine, Irene. Service d'Explorations Fonctionnelles EndocriniennesCentre de Reference des Maladies Endocrines Rares de la Croissance, Hopital Trousseau, Groupe Hospitalier Universitaire Est, AP-HP, Paris, France.

Netchine, Irene. Universite Pierre et Marie CurieSorbonne Universite, Paris, France.

Siffroi, Jean-Pierre. Universite Pierre et Marie CurieSorbonne Universite, Paris, France.

Siffroi, Jean-Pierre. INSERM UMR_S933Paris, France.

Christin-Maitre, Sophie. Service d'Endocrinologie et Medecine de la ReproductionCentre de Reference des Maladies Endocrines Rares de la Croissance, Hopital Saint Antoine, Groupe Hospitalier Universitaire Est, AP-HP, Paris, France.

Christin-Maitre, Sophie. Universite Pierre et Marie CurieSorbonne Universite, Paris, France.

Christin-Maitre, Sophie. INSERM UMR_S933Paris, France.

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Year of Publication

2018

285.

The relation between isolated micropenis in childhood with CAG and GGN repeat polymorphisms in the androgen receptor gene.

Tug E., Guntekin Ergun S., Ergun M.A., Dilek F.N., Percin E.F.

Embase

Turkish Journal of Medical Sciences. 48 (2) (pp 430-434), 2018. Date of Publication: 2018.

[Article]

AN: 621966988

Background/aim: In micropenis cases accompanied by external genital abnormalities such as hypospadias and cryptorchidism, infertility and spermatogenic failures have been reported to correlate with androgen receptor (AR) gene CAG and GGN repeat polymorphisms. While there is one study on isolated micropenis and CAG repeats, no study related to GGN repeats has been reported. We investigated the relation between CAG and GGN repeats in the AR gene with development of penis length in boys with isolated micropenis.

Material(s) and Method(s): A total of 24 Turkish boys with isolated micropenis (<-2.5 SD) and 64 healthy controls who had normal basal serum gonadotropin levels were examined. Genotyping was performed by DNA sequencing of the patients and controls.

Result(s): The distribution of CAG and GGN repeat lengths in our patients and controls was within the normal range and did not significantly differ between the patients and the controls.

Conclusion(s): CAG repeat length in the AR constitutes one of multiple genetic factors relevant to the development of isolated micropenis, and the expansion of this repeat can be detected as a likely modifying factor. Moreover, the interactions of other genes that may be involved in the etiology of isolated micropenis with CAG and GGN repeats have to be taken into consideration.

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Status

Embase

Institution

(Tug, Guntekin Ergun, Ergun, Percin) Department of Medical Genetics, School of Medicine, Gazi University, Ankara, Turkey (Dilek) Dr. Sami Ulus Obstetrics Child Health and Illness Training and Research Hospital, Ankara, Turkey

Publisher

Turkiye Klinikleri (Talapapa Bulvary no. 102, Hamammonu 1 06230, Turkey)

Year of Publication

2018

Dexamethasone as an adjuvant for caudal blockade in pediatric surgical patients: A systematic review and meta-analysis.

Chong M.A., Szoke D.J., Berbenetz N.M., Lin C.

Embase

Anesthesia and Analgesia. 127 (2) (pp 520-528), 2018. Date of Publication: 2018.

[Article]

AN: 624837730

BACKGROUND: Caudal block is commonly used to provide postoperative analgesia after pediatric surgery in the lower abdomen. Typically administered as a single-shot technique, 1 limitation of this block is the short duration of analgesia. To overcome this, dexamethasone has been used as an adjuvant to prolong block duration. However, there are concerns about steroid-related morbidity and the optimal route of dexamethasone administration (eg, caudal or intravenous) is unknown.

METHOD(S): We conducted a systematic review and random-effects meta-analysis of randomized controlled trials recruiting pediatric surgical patients receiving a caudal block for surgical anesthesia or postoperative analgesia. Included studies compared dexamethasone (caudal, intravenous, or both) to control. Duration of analgesia was the primary outcome. Database sources were Medline, Embase, the Cochrane Library, and Google Scholar searched up to August 18, 2017, without language restriction. Screening of studies, data extraction, and risk of bias assessment were performed independently and in duplicate by 2 authors. Risk of bias was assessed using Cochrane methodology and the strength of evidence was scored using the Grading of Recommendations Assessment, Development, and Evaluation (GRADE) system.

RESULT(S): The initial search retrieved 93 articles. Fourteen randomized controlled trials that comprised 1315 pediatric patients met the inclusion criteria. All but 1 study involved lower abdominal operations (orchidopexy, inguinal hernia repair, and hypospadias repair). The caudal and intravenous dose of dexamethasone ranged from 0.1 to 0.2 mg/kg and 0.5 to 1.5 mg/kg, respectively, and all studies were pooled in the main analysis. Dexamethasone prolonged the duration of analgesia by both the caudal route (5.43 hours, 95% confidence interval [CI], 3.52-7.35; $P < .001$; $I^2 = 99.3\%$; $N = 9$; $n = 620$; GRADE quality = moderate) and intravenous route (5.51 hours; 95% CI, 3.56-7.46; $P < .001$; $I^2 = 98.9\%$; $N = 5$; $n = 364$; GRADE quality = moderate) versus control. Secondary benefits of dexamethasone included reduced narcotic rescue analgesia requirement in the postanesthetic care unit (relative risk [RR], 0.30; 95% CI, 0.18-0.51; $P < .001$; $I^2 = 0.0\%$; $N = 5$; number needed to treat for benefit [NNTB] = 5; 95% CI, 4-7), less subsequent postoperative rescue analgesia requirement (RR, 0.46; 95% CI, 0.23-0.92; $P = .03$; $I^2 = 96.0\%$; $N = 9$; $n = 629$; NNTB = 3; 95% CI, 2-20; $n = 310$), and lower rates of postoperative

nausea and vomiting (RR, 0.47; 95% CI, 0.30-0.73; P = .001; I2 = 0.0%; NNTB = 11; 95% CI, 8-21; N = 9; n = 628). Adverse events linked to the dexamethasone were rare.

CONCLUSION(S): Caudal and intravenous dexamethasone are similarly effective for prolonging the duration of analgesia from caudal blockade, resulting in a doubled to tripled duration. Given the off-label status of caudal dexamethasone, intravenous administration is recommended - although only high intravenous doses (0.5 mg/kg up to 10 mg) have been studied.

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Status

Embase

Institution

(Chong, Szoke, Lin) Department of Anesthesia and Perioperative Medicine, Western University, University Hospital (London Health Sciences Centre), 339 Windermere Rd., C3-108, London, ON N6A 5A5, Canada (Berbenetz) Division of Cardiology, Department of Medicine, Queen's University, Kingston, ON, Canada

Publisher

Lippincott Williams and Wilkins (E-mail: kathiest.clai@apta.org)

Year of Publication

2018

287.

Association of In Utero Exposure to Polybrominated Diphenyl Ethers With the Risk of Hypospadias.

Poon S., Koren G., Carnevale A., Aleksa K., Ling J., Ozsarfati J., Kapur B.M., Bagli D.

Embase

JAMA Pediatrics. 172 (9) (pp 851-856), 2018. Date of Publication: September 2018.

[Article]

AN: 623752290

IMPORTANCE Polybrominated diphenyl ethers (PBDEs) are added to many consumer products as flame retardants, and their endocrine-disrupting properties are a growing health concern in pregnancy. OBJECTIVE To investigate whether in utero PBDE exposure as measured in

maternal hair is associated with increased risk for hypospadias. **DESIGN, SETTING, AND PARTICIPANTS** In this case-control study, the setting was the urology clinic of a tertiary pediatric hospital between January 3, 2011, and April 1, 2013. Participants were children diagnosed as having hypospadias and their mothers and a control group of children without hypospadias and their mothers. Dates of data analysis were September 3, 2017, to December 28, 2017. **EXPOSURES** Gestational exposure to 8 PBDEs as measured in the 3-cm segment closest to the skull of maternal hair by gas chromatography-mass spectroscopy as a proxy for in utero exposure. The mothers resided in the same household for the duration of their pregnancy. **MAIN OUTCOMES AND MEASURES** Difference in total maternal hair PBDE levels between the hypospadias and control groups. **RESULTS** Total PBDE levels were significantly higher among mothers of infants with hypospadias (n = 152) (total PBDE level, 51.4 pg/mg; interquartile range, 35.8-78.5 pg/mg) than among controls (n = 64) (total PBDE level, 35.8 pg/mg; interquartile range, 18.1-69.9 pg/mg) (P = .02). Of the 152 women with sufficient hair samples for analysis in the case group, 89 completed a questionnaire and were included in a multivariable analysis, and of the 64 women with sufficient hair samples for analysis in the control group, 54 completed a questionnaire and were included in a multivariable analysis. Adjusting for potential confounders, hypospadias was associated with a relative 48.2% (95% CI, 23.2%-65.4%) higher maternal level of total PBDE levels in the multivariable analysis. **CONCLUSIONS AND RELEVANCE** In this analysis, mothers of children with hypospadias were exposed during pregnancy to significantly higher levels of PBDEs. The results of this study suggest that level of exposure to PBDEs during gestation may have a role in the etiology of hypospadias.

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Status

Embase

Institution

(Koren) Division of Clinical Pharmacology, Maccabi Research Institute, 4 Koifman St, Eighth Floor, Tel Aviv 6812509, Israel (Poon, Carnevale, Aleksa, Ozsarfati, Kapur) Department of Pediatrics, Hospital for Sick Children, Toronto, ON, Canada

(Poon, Carnevale, Ling, Kapur) Department of Pharmacology and Toxicology, University of Toronto, Toronto, ON, Canada

(Koren) Western University, London, ON, Canada

(Koren) Division of Clinical Pharmacology, Maccabi Research Institute Tel Aviv, Tel Aviv, Israel

(Aleksa) Leslie Dan Faculty of Pharmacy, University of Toronto, Toronto, ON, Canada

(Aleksa) School of Pharmacy, University of Waterloo, Waterloo, ON, Canada

(Kapur) Department of Clinical Pathology, Sunnybrook Health Sciences Center, Toronto, ON, Canada

(Kapur) Department of Laboratory Medicine and Pathobiology, Faculty of Medicine, University of Toronto, Toronto, ON, Canada

(Bagli) Department of Surgery, Hospital for Sick Children, Toronto, ON, Canada

Publisher

American Medical Association (E-mail: smcleod@itsa.ucsf.edu)

Year of Publication

2018

288.

Architecture of structures in the urogenital triangle of young adult males; comparison with females.

Wu Y., Dabhoiwala N.F., Hagoort J., Hikspoors J.P.J.M., Tan L.-W., Mommen G., Hu X., Zhang S.-X., Lamers W.H.

Embase

Journal of Anatomy. 233 (4) (pp 447-459), 2018. Date of Publication: October 2018.

[Article]

AN: 623405788

The fibro-muscular architecture of the urogenital triangle remains contentious. Reasons are small size of the constituting structures and poor visibility with most imaging methods. We reinvestigated the area in serial sections of three males (21-38 years old) of the American and Chinese Visible Human Projects and two 26-week-old male fetuses, and compared the findings with earlier observations in females. The mass of the levator ani muscle was approximately twofold smaller and its funnel shape steeper in males than females. In the levator hiatus, a strand of the smooth longitudinal muscle layer of the rectum, the 'rectourethral (RU) muscle', extended anteriorly from the anorectal bend to the penile bulb. Fibrous tissue that formed in the inferior reach of the fetal RU muscle identified the location of the developing perineal body (PB) and divided the muscle into posterior 'rectoperineal' and anterior 'deep perineal' portions. In males, the PB remained small and bipartite, so that the RU muscle presented as an undivided midline structure. The well-developed female PB, instead, intertwined with the deep perineal muscle and both structures passed the vagina bilaterally to form the perineal membrane in the posterior

portion of the urogenital triangle. The urethral rhabdosphincter extended in the anterior portion of the urogenital triangle between the penile bulb inferiorly and the bladder neck superiorly, and consisted of a well-developed circular 'membranous' portion with bilateral posteroinferior 'wings' and a thinner 'prostatic' portion on the prostate anterior side. In men, muscles occupy the urogenital triangle, but additional tightening of the locally fibrous adipose tissue by the superficial transverse perineal muscle appears necessary to generate functional support in women. An interactive 3D pdf file with these anatomical details (available online) should allow more accurate interpretation of ultrasound, computed tomography and magnetic resonance images.

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Status

Embase

Author NameID

Lamers, Wouter H.; ORCID: <http://orcid.org/0000-0003-3032-7824> Zhang, Shao-Xiang; ORCID: <http://orcid.org/0000-0002-9930-4332>

Institution

(Wu, Dabhoiwala, Lamers) Tytgat Institute for Liver and Intestinal Research, Academic Medical Center, University of Amsterdam, Amsterdam, Netherlands (Wu, Tan, Hu, Zhang) Institute of Digital Medicine, College of Biomedical Engineering, Third Military Medical University, Chongqing, China

(Hagoort) Department of Anatomy & Embryology, Academic Medical Center, University of Amsterdam, Amsterdam, Netherlands

(Hikspoors, Mommen, Lamers) Department of Anatomy & Embryology, Maastricht University, Maastricht, Netherlands

Publisher

Blackwell Publishing Ltd

Year of Publication

2018

A model for sustained collaboration to address the unmet global burden of bladder exstrophy-epispadias complex and penopubic epispadias the international bladder exstrophy consortium. Joshi R.S., Shrivastava D., Grady R., Kundu A., Ramji J., Reddy P.P., Pippi-Salle J.L., Frazier J.R., Canning D.A., Shukla A.R.

Embase

JAMA Surgery. 153 (7) (pp 618-624), 2018. Date of Publication: July 2018.

[Article]

AN: 623384549

IMPORTANCE International collaboration to alleviate the massive burden of surgical disease is recognized by World Health Organization as an urgent need, yet the surgical mission model to treat reconstructive surgical challenges is often constrained in ensuring adequate patient follow-up, optimal outcomes, and sustainability. **OBJECTIVE** To determine whether a collaboration predicated on long-term commitment by surgeons returning to the same institution annually combined with an experienced host surgical team and infrastructure to ensure sustained patient follow-up could provide surgical care with acceptable outcomes to treat bladder exstrophy-epispadias complex (BE) and penopubic epispadias (PE). **DESIGN, SETTING, AND PARTICIPANTS** In this prospective, observational study, long-term collaboration was created and based at a public hospital in Ahmedabad, India, between January 2009 and January 2015. The entire postoperative cohort was recalled in January 2016 for comprehensive examination, measurement of continence outcomes, and assessment of surgical complications. Seventy-six percent of patients (n = 57) who underwent complete primary repair of exstrophy during the study interval returned for annual follow-up in 2016 and formed the study cohort: 23 patients with primary BE, 19 patients with redo BE, and 11 patients with PE repair. **MAIN OUTCOMES AND MEASURES** Demographics, operative techniques, and perioperative complications were recorded. A postoperative protocol outlining procedures to ensure monitoring of study participants was followed including removal of ureteral stents, urethral catheter, external fixators, imaging, and patient discharge. **RESULTS** Of the 57 patients, 4 were excluded because they underwent ureterosigmoid-ostomy. Median age at time of surgery was 3 years (primary BE), 7 years (redo BE), and 10 years (PE), with median follow-up of 3 years, 5 years and 3 years, respectively; boys made up more than 70% of each cohort (n = 17 for primary BE, n = 15 for redo BE, and n = 9 for PE). All BE and 3 PE repairs (27%) were completed with concurrent anterior pubic osteotomies. Seventeen of 53 patients (32%) experienced complications. Only 1 patient with BE (4%) had a bladder dehiscence and was repaired the following year. **CONCLUSIONS AND RELEVANCE** A unique surgical mission model consisting of an international collaborative focused on treating the complex diagnoses of BE and PE offers outcomes comparable with those in high-income countries, demonstrating a significant patient retention rate and an opportunity to rigorously study outcomes over an accelerated interval owing to the high burden of disease in India. Postoperative

care following a systematized algorithm and rigorous follow-up is mandatory to ensure safety and optimal outcomes.

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29516095 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=29516095>]

Status

Embase

Institution

(Joshi, Ramji) B.J. Medical College and Civil Hospital, Ahmedabad, Gujarat, India (Shrivastava)

Gandhi Medical College, Bhopal, India

(Grady) Seattle Children's Hospital, Seattle, WA, United States

(Kundu) Dayton Children's Hospital, Dayton, OH, United States

(Reddy) Cincinnati Children's Hospital, Cincinnati, OH, United States

(Pippi-Salle) Sidra Medical and Research Center, Doha, Qatar, United States

(Frazier, Canning, Shukla) Children's Hospital of Philadelphia, Philadelphia, PA, United States

Publisher

American Medical Association (E-mail: smcleod@itsa.ucsf.edu)

Year of Publication

2018

290.

Male Sexuality, Fertility, and Urinary Continence in Bladder Exstrophy-Epispadias Complex.

Reynaud N., Courtois F., Mouriquand P., Morel-Journel N., Charvier K., Gerard M., Ruffion A., Terrier J.-E.

Embase

Journal of Sexual Medicine. 15 (3) (pp 314-323), 2018. Date of Publication: March 2018.

[Article]

AN: 621015555

Background: The bladder exstrophy-epispadias complex is a rare congenital malformation associated with severe dysfunction of the genital and urinary tracts and requiring a staged surgical reconstruction.

Aim(s): The primary aims of this study were to report the sexuality, infertility, and urinary incontinence outcomes in a cohort of men born with bladder exstrophy-epispadias complex. The secondary aim was to highlight some predictive factors of infertility in this population.

Method(s): We conducted a descriptive, cross-sectional study of men diagnosed with classic presentations of bladder exstrophy or epispadias.

Outcome(s): Patients were asked to complete 4 validated questionnaires: the International Index of Erectile Function (IIEF)-5, the Erection Hardness Score (EHS), the Self-Esteem and Relationship, and the International Consultation Incontinence modular Questionnaire-Short Form. Fertility potential was assessed with semen analysis and a non-validated questionnaire.

Result(s): 38 Patients 18-64 years old (M [mean] = 32.2) completed the questionnaires. The average IIEF-5 score was 18.1/25 (ranging from 3-25; SE = 7.62), with results indicating that 55% of the sample had normal erectile function. Results also showed higher scores for patients with normal spermatozoa concentration (M = 22.75, SE = 1.89, P = .08) than for those with oligospermia (M = 17.30, SE = 8.53, P = .08). Results on the IIEF-5 also indicated higher scores for patients who conceived children without assisted reproductive technologies (ART) (M = 22.83, SE = 2.317, P = .02) than for patients without children (M = 15.76, SE = 8.342, P = .02). The average EHS was 3.43/4 (ranging from 1-4, SE = 0.9). EHS was higher for patients who had reconstruction than for patients who had cystectomy (M = 3.88, SE = 1.07 and 2.78, SE = 1.09, P = .02). The average total Self-Esteem and Relationship score was 67.04/100 (ranging from 10.71-96.43, SE = 22.11). The average total International Consultation Incontinence modular Questionnaire-Short Form score was 4.97/21 (ranging from 0-18, SE = 5.44), higher score indicating more urinary incontinence. Among the patients surveyed, 31.6% were parents at the time of study and 50% of them benefited from ART. With regards to the 14 semen analyses performed, only 7.1% produced normal results and 44.7% indicated that ejaculation was weak and dribbling.

Clinical Translation: Erectile function appears to be decreased and psychological aspects of sexuality indicate low self-esteem about sexual relationship. Although ethical problems could not allow prospective spermograms, our cohort is large enough to provide significant data.

Conclusion(s): Early sperm storage for future ART, sexual medicine management, and complementary genital reconstruction in adulthood constitute potential treatment options for this population. Reynaud N, Courtois F, Mouriquand P, et al. Male Sexuality, Fertility, and Urinary Continence in Bladder Exstrophy-Epispadias Complex. J Sex Med 2017;15:314-323.

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Status

Embase

Institution

(Reynaud, Morel-Journal, Ruffion, Terrier) Service d'urologie, Centre Hospitalier Lyon Sud, Hospices Civils de Lyon, Universite Claude-Bernard-Lyon 1-France, France (Courtois, Gerard) Department of Sexology, Universite du Quebec, Montreal, Quebec, Canada (Mouriquand) Service de Chirurgie Uro-Viscerale de l'Enfant-Hopital Mere Enfant, Hospices Civils de Lyon, Universite Claude-Bernard-Lyon 1-France, France (Charvier) Service de reeducation neuro-perineale et sexologique, Hopital Henry Gabrielle, Hospices Civils de Lyon, France (Ruffion) Universite Claude Bernard-Lyon 1-Centre de Recherche en Cancerologie de Lyon-Institut National de la Sante Et de la Recherche Medicale, Centre National de la Recherche Scientifique-Lyon 1-France, France

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291.

Placental Weight and Risk of Cryptorchidism and Hypospadias in the Collaborative Perinatal Project.

Ghazarian A.A., Trabert B., Graubard B.I., Longnecker M.P., Klebanoff M.A., McGlynn K.A.

Embase

American Journal of Epidemiology. 187 (7) (pp 1354-1361), 2018. Date of Publication: 01 Jul 2018.

[Article]

AN: 623483853

Cryptorchidism and hypospadias are the most common congenital anomalies of the genitourinary tract in males, but their etiology remains unclear. Placental insufficiency has been suggested to be linked to both conditions. Placental weight is a commonly used proxy measure for placental insufficiency; thus, we examined placental weight and other placental characteristics in relation to cryptorchidism and hypospadias in the Collaborative Perinatal Project, a US mother-child cohort study. Pregnant women were recruited between 1959 and 1965. The analysis contrasted boys with cryptorchidism (n = 413) and boys with hypospadias (n = 145) with boys without

cryptorchidism (n = 23,799) and boys without hypospadias (n = 22,326). Odds ratios and 95% confidence intervals were calculated using unconditional logistic regression. In categorical analyses in which the middle tertile was the referent, cryptorchidism was inversely associated with placental weight (odds ratio = 0.66, 95% confidence interval: 0.46, 0.95) among white boys and positively associated with the lowest tertile of placental weight among black boys (odds ratio = 1.70, 95% confidence interval: 1.11, 2.59). We conclude that lower placental weight may be related to risk of cryptorchidism. Further investigation of placental functioning may offer insights into the etiology of cryptorchidism.

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Status

Embase

Institution

(McGlynn) National Cancer Institute, 9609 Medical Center Drive, Room 6-E446, Bethesda, MD 20892, United States (Ghazarian, Trabert, Graubard, McGlynn) Division of Cancer Epidemiology and Genetics, National Cancer Institute, Bethesda, MD, United States

(Ghazarian) Division of Cancer Control and Population Sciences, National Cancer Institute, Bethesda, MD, United States

(Longnecker) Epidemiology Branch, National Institute of Environmental Health Sciences, Research Triangle Park, NC, United States

(Klebanoff) Ohio Perinatal Research Network, Department of Pediatrics, College of Medicine, Ohio State University, Nationwide Children's Hospital, Columbus, OH, United States

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292.

Evaluation of the ISL1 gene in the pathogenesis of bladder exstrophy in a Swedish cohort.

Arkani S., Cao J., Lundin J., Nilsson D., Kallman T., Barker G., Holmdahl G., Clementsson Kockum C., Matsson H., Nordenskjold A.

Embase

Human Genome Variation. 5 (no pagination), 2018. Article Number: 18009. Date of Publication: 29 Mar 2018.

[Article]

AN: 621429195

Bladder exstrophy is a congenital closure defect of the urinary bladder with a profound effect on morbidity. Although the malformation is usually sporadic, a genetic background is supported by an increased recurrence risk in relatives, higher concordance rates in monozygotic twins and several associated chromosomal aberrations. Recently, the ISL1 gene was presented as a candidate gene for bladder exstrophy and epispadias complex (BEEC) development in two different studies. In our study, we screened for genetic variants in the ISL1 gene in DNA from 125 Swedish patients using Sanger sequencing and array-CGH analysis. In addition, we evaluated ISL1 expression in RNA of human bladder during embryonic and fetal weeks 5-10 relative to that in lung tissue (week 9). In total, 21 single-nucleotide variants were identified, including a potentially novel missense variant, c.137C>G p.(Ala46Gly), substituting a conserved amino acid. This variant was inherited from an unaffected mother. No structural variants were identified. RNA sequencing revealed ISL1 mRNA expression during the critical time frame of human bladder development. In conclusion, we did not detect any known or likely pathogenic variants in the ISL1 gene in 125 Swedish BEEC patients, indicating that variation in the ISL1 gene is not a common genetic mechanism of BEEC development in the Swedish population.

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Status

Embase

Author NameID

Arkani, Samara; ORCID: <http://orcid.org/0000-0002-8339-4783>

Institution

(Arkani, Cao, Lundin, Matsson, Nordenskjold) Department of Women's and Children's Health, Center for Molecular Medicine, Karolinska Institutet, Stockholm, Sweden (Lundin) Department of Clinical Genetics, Karolinska University Hospital, Stockholm, Sweden

(Lundin, Nilsson) Department of Molecular Medicine and Surgery, Center for Molecular Medicine, Karolinska Institutet, Stockholm, Sweden

(Nilsson) Science for Life Laboratory, Karolinska Institutet Science Park, Stockholm, Sweden

(Kallman) Science for Life Laboratory, Department of Medical Biochemistry and Microbiology, Uppsala University, Uppsala, Sweden

(Kallman) NBIS-National Bioinformatics Infrastructure Sweden, Uppsala, Sweden

(Barker) Department of Women's and Children's Health, Uppsala Academic Children Hospital, Uppsala, Sweden

(Holmdahl) Department of Pediatric Surgery, Queen Silvia Children's Hospital, Gothenburg, Sweden

(Clementsson Kockum) Department of Pediatric Surgery, University Hospital Lund, Lund, Sweden

(Nordenskjold) Department of Pediatric Surgery, Astrid Lindgren Children Hospital, Karolinska University Hospital, Stockholm, Sweden

Publisher

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293.

The Double Dutch technique: A new way of creating an ileocystoplasty with a lengthy catheterizable ileal tube.

Polm P.D., Beyerlein S., Klijn A.J., de Jong T.P.V.M., Dik P.

Embase

Journal of Pediatric Urology. 14 (3) (pp 255.e1-255.e6), 2018. Date of Publication: June 2018.

[Article]

AN: 620896854

Introduction: A tubularized conduit from an open 2-cm vascularized ileal segment is a frequently used technique to create a continent catheterizable channel in cases of an inappropriate or absent appendix. In the long term, many patients experience catheterization problems with the classic ileal segment tube, and even more with spiral or double-segment tubes.

Objective(s): The objective of this paper was to introduce an ileocystoplasty modification combined with a long ileal segment tube that has better support by surrounding tissue than other lengthy ileal segment tubes. Briefly summarized, this newly introduced method begins with isolating approximately 30 cm of ileum and dividing it into two parts. Two strips are then created and closed as a double-length tube. The ileal segments are opened antimesenterically and closed over the tube in the middle. The lower part of the tube is implanted with a submucosal tunnel in the bladder wall, and the ileal patch is then anastomosed with the bladder. The tube is

anastomosed to the umbilicus in an ordinary way without any traction (see Summary Fig.). Study design: Between May 2005 and November 2012 the new technique was used at the current institution in nine children who needed an ileocystoplasty (mean age: 9 years and 3 months).

Underlying etiology was neurogenic bladder in seven cases and epispadias in two.

Result(s): All patients ultimately had stomas without leakage or strictures. During follow-up, three of nine tubes developed stenoses that were corrected; four stomas in total had some sort of surgical revision. Median follow-up was 93 months. Intermittent catheterization was uncomplicated in all at this time.

Discussion(s): With this modification of the standard technique it seemed to be possible to create a more stable channel. The blood supply of the tube was secured by completely embedding the mesentery of the tube. Limitations included the small number of patients treated so far.

Conclusion(s): The lengthy tubes appeared to be straight and well supported by the surrounding tissues, which prevented kinking and sacculation. It is hoped that this technique will have better results and fewer complications at long-term follow-up.[Figure presented]

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Author NameID

Polm P.D.; ORCID: <http://orcid.org/0000-0002-8403-5977>

Institution

(Polm, Beyerlein, Klijn, de Jong, Dik) Department of Pediatric Urology, Wilhelmina Children's Hospital Utrecht, Utrecht, Netherlands (Beyerlein) Department of Pediatric Surgery, Pediatric Urology and Pediatric Orthopedics, DRK Kinderklinik, Siegen, Germany

(Polm) Department of Urology, University Medical Centre Utrecht, Utrecht, Netherlands

(de Jong) Department of Paediatric Urology, University Children's Hospital, AMC, Amsterdam, Netherlands

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294.

The effect of assisted reproductive technology on the incidence of birth defects among livebirths.

Shechter-Maor G., Czuzoj-Shulman N., Spence A.R., Abenhaim H.A.

Embase

Archives of Gynecology and Obstetrics. 297 (6) (pp 1397-1403), 2018. Date of Publication: 01 Jun 2018.

[Article]

AN: 620741610

Purpose: Our study objective is to examine the association between births conceived with assisted reproductive technology (ART) and birth defects using a large database from the United States.

Method(s): Using the Centers for Disease Control and Prevention's Period-linked birth-infant death data files and fetal death database for 2011-2013, we conducted a retrospective cohort study comprised of live births that occurred in the USA during that time. Multivariate logistic regression was used to estimate the association between ART and birth defects, both overall and by specific defects.

Result(s): There were 11,862,780 live births between 2011 and 2013. Of these births, 11,791,730 were spontaneous pregnancies and 71,050 were conceived by ART, with an increasing trend in incidence of ART during the study period and an overall increasing trend of birth defects. Overall, infants conceived by ART had a greater risk of having birth defects than did infants conceived spontaneously (77/10,000 vs 25/10,000, respectively, OR 2.14, 95% CI 1.94-2.35). The malformations most commonly associated with ART were cyanotic heart defects (OR 2.74, 95% CI 2.42-3.09), cleft lip and/or palate (OR 1.47, 95% CI 1.14-1.89), and hypospadias (OR 1.77, 95% CI 1.42-2.19). There were no differences in risk of omphalocele or neural tube defects between the two groups.

Conclusion(s): There is an overall and type-specific increased risk of birth defects in the ART population. Appropriate counseling and specialized ultrasound evaluations should be considered in pregnancies conceived by ART.

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Status

Embase

Institution

(Shechter-Maor, Abenhaim) Department of Obstetrics and Gynecology, Jewish General Hospital, McGill University, Pav H, Room 325, 5790 Cote-Des-Neiges Road, Montreal, QC H3S 1Y9,

Canada (Czuzoj-Shulman, Spence, Abenhaim) Centre for Clinical Epidemiology and Community Studies, Jewish General Hospital, Montreal, QC, Canada

Publisher

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295.

Association between male genital anomalies and adult male reproductive disorders: a population-based data linkage study spanning more than 40 years.

Schneuer F.J., Milne E., Jamieson S.E., Pereira G., Hansen M., Barker A., Holland A.J.A., Bower C., Nassar N.

Embase

The Lancet Child and Adolescent Health. 2 (10) (pp 736-743), 2018. Date of Publication: October 2018.

[Article]

AN: 2001193103

Background: The male genital anomalies hypospadias and undescended testes have been linked to adult male reproductive disorders, testicular cancer, and decreased fertility. Few population-based studies have evaluated their effects on adult fertility outcomes and, in the case of undescended testes, the importance of early corrective surgery (orchidopexy).

Method(s): We did a population-based cohort study of all liveborn boys in Western Australia in 1970-99, and followed them up until 2016 via data linkage to registries for hospital admissions, congenital anomalies, cancer, and assisted reproductive technologies (ART). Study factors were hypospadias or undescended testes, and study outcomes were testicular cancer, paternity, and use of ART for male infertility. Cox regression was used to calculate hazard ratios (HRs) for the associations between genital anomalies and testicular cancer or paternity, and log-binomial regression was used to calculate relative risks (RRs) for the associations between genital anomalies and use of ART.

Finding(s): The cohort comprised 350 835 boys, of whom 2484 (0.7%) had been diagnosed with hypospadias and 7499 (2.1%) with undescended testes. There were 505 (0.1%) cases of testicular cancer, 109 471 (31.2%) men had fathered children, and 2682 (0.8%) had undergone

fertility treatment with ART. Undescended testes was associated with a more than two times increase in risk of testicular cancer (HR 2.43, 95% CI 1.65-3.58) and hypospadias with an almost 40% increase (1.37, 0.51-3.67), although this increase was not significant. Both hypospadias and undescended testes were associated with a 21% reduction in paternity (adjusted HR 0.79 [95% CI 0.71-0.89] for hypospadias and 0.79 [0.74-0.85] for undescended testes). Undescended testes was associated with a two times increase in use of ART (adjusted RR 2.26, 95% CI 1.58-3.25). For every 6 months' delay in orchidopexy, there was a 6% increase in risk of testicular cancer (HR 1.06, 95% CI 1.03-1.08), a 5% increase in risk of future use of ART (1.05, 1.03-1.08), and a 1% reduction in paternity (RR 0.99, 95% CI 0.98-0.99).

Interpretation(s): Undescended testes is associated with an increased risk of testicular cancer and male infertility, and decreased paternity. We provide new evidence to support current guidelines for orchidopexy before age 18 months to decrease the risk of future testicular cancer and infertility.

Funding(s): National Health and Medical Research Council and Sydney Medical School Foundation.

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Status

Embase

Institution

(Schneuer, Nassar) Child Population and Translational Health Research, The Children's Hospital at Westmead Clinical School, The University of Sydney, NSW, Australia (Holland) Discipline of Child and Adolescent Health, The Children's Hospital at Westmead Clinical School, The University of Sydney, NSW, Australia

(Milne, Jamieson, Pereira, Hansen, Bower) Telethon Kids Institute, University of Western Australia, Perth, WA, Australia

(Pereira) School of Public Health, Curtin University, Perth, WA, Australia

(Barker) Department of Paediatric Surgery, Princess Margaret Hospital for Children, Perth, WA, Australia

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Birth prevalence of hypospadias and hypospadias risk factors in newborn males in the United States from 1997 to 2012.

Chen M.J., Karaviti L.P., Roth D.R., Schlomer B.J.

Embase

Journal of Pediatric Urology. 14 (5) (pp 425.e1-425.e7), 2018. Date of Publication: October 2018.

[Article]

AN: 2001178834

Background: Hypospadias is a common genitourinary malformation and there are conflicting data on whether its prevalence is increasing. Previous studies have described associations with risk factors including small for gestational age (SGA), multiple gestation birth, environmental influences, and maternal factors.

Objective(s): The objective of this study was to examine birth prevalence of hypospadias and hypospadias risk factors in a large national dataset and to evaluate for changes from 1997 to 2012. We hypothesized that any increase in the birth prevalence of hypospadias would be associated with an increase in risk factors such as SGA, prematurity, or multiple gestation birth.

Study design: The Kids' Inpatient Database was used to generate national estimates for prevalence of males born with hypospadias, SGA, prematurity, or to a multiple gestation and then prevalences were evaluated for association with time. Multivariable logistic regression was used to evaluate whether birth prevalence of hypospadias was associated with increasing year, SGA, prematurity, and multiple gestation birth.

Result(s): The estimated birth prevalence of hypospadias increased from 6.1 per 1000 births (95% confidence interval [CI] 5.9 to 6.3) to 6.8 per 1000 births (95% CI 6.7 to 7.0), an 11.5% increase from 1997-2012 ($P = 0.014$). Among male births, the prevalence of SGA increased 74%, multiple gestation increased 25%, and prematurity increased 20% ($P < 0.001$ for all) (Summary Figure). A risk factor was seen in around 20% of males born with hypospadias. Hypospadias birth prevalence also increased in males without risk factors but was not statistically significant (9.1% increase, $P = 0.5$). On multivariable logistic regression, being born SGA (odds ratio [OR] = 3.3), to a multiple gestation (OR = 1.1), or premature (OR 1.9) were associated with increased odds of hypospadias ($P < 0.01$ for all), whereas increasing year was not ($P = 0.3$).

Conclusion(s): The estimated birth prevalence of hypospadias in the United States increased from 6.1 to 6.8 per 1000 births from 1997 to 2012. Known hypospadias risk factors of SGA birth, multiple gestation birth, and premature birth also increased over this time to a higher degree. About 20% of males born with hypospadias had one of these risk factors. The birth prevalence of

hypospadias in males without any studied risk factors also increased, but this was not statistically significant. More studies are needed to evaluate whether this increase in hypospadias prevalence is due to increases in known hypospadias risk factors, new environmental exposures, improved diagnosis at birth, some combination, or unrelated causes.[Figure presented]

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Status

Embase

Institution

(Chen, Karaviti) Section of Pediatric Diabetes and Endocrinology, Department of Pediatrics, Baylor College of Medicine and Texas Children's Hospital, 6701 Fannin St, Suite 1020, Houston, TX 77030, United States (Roth) Division of Pediatric Urology, Department of Surgery, Baylor College of Medicine and Texas Children's Hospital, 6701 Fannin St, Suite 620, Houston, TX 77030, United States

(Schlomer) Department of Urology, University of Texas Southwestern, 2305 Stemmons Freeway, Suite D-4300, MC F4.04, Dallas, TX 75207, United States

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297.

Perineal hypospadias: back to the future Chordee Excision & Distal Urethroplasty.

Hadidi A.T.

Embase

Journal of Pediatric Urology. 14 (5) (pp 424.e1-424.e9), 2018. Date of Publication: October 2018. [Article]

AN: 2001161878

Purpose: Perineal hypospadias correction has swung from two-stage repair in the 1960s to one-stage repair in the 1980s and back to two-stage repair in the 2000s. The author's experience with a technique in which Chordee Excision and Distal Urethroplasty (CEDU) was performed at the

first operation leaving a 1-cm segment as perineal urethrotomy to be reconstructed at the second operation is presented.

Patients and Methods: Between January 2013 and December 2016, the CEDU technique was performed in 63 patients with perineal hypospadias. The records of 59 patients who maintained regular follow-up were reviewed. The principle is to excise the hypoplastic urethral plate, atretic corpus spongiosum, and longitudinal layer of tunica albuginea; split the glans in the midline; and reconstruct a healthy urethral plate using preputial and lateral skin flaps to the tip of the glans. Distal urethroplasty was performed leaving 1 cm at the proximal end to be reconstructed 3-6 months later. This principle was used by Duplay in the 1880s. Patient age ranged between 6 months and 2 years (mean 8 months). All the patients had perineal hypospadias and bifid scrotum with severe deep chordee. Follow-up period ranged from 17 to 53 months (mean 36). A transurethral Silastic catheter was inserted for 4 days. Three months later, the remaining 1 cm of the new urethra was reconstructed, and final adjustment of the glans and foreskin was performed. **Result(s):** Satisfactory results were obtained in 54 patients (90%). Three children experienced glans dehiscence that was corrected in the second stage, one child developed fistula after the second operation, and one developed diverticulum. The fistula and diverticulum were corrected at the third operation successfully.

Discussion(s): It was necessary in this series to divide the urethral plate and excise the hypoplastic corpus spongiosum and the outer longitudinal layer of tunica albuginea to correct the associated deep chordee. The lateral skin flaps receive double blood supply from the base of the penis and the preputial vessels. This natural urinary diversion allows early removal of the catheter, reduces the discomfort of the patient, and allows the new urethra to heal without urine irritation for 3 months. Long-term follow-up for 15 years is necessary to assess the technique objectively.

Conclusion(s): The CEDU technique diverts urine away from the site of urethroplasty for 3 months without a catheter. It reduces the hospital stay and patient discomfort. It produces satisfactory results and has become a the standard technique in perineal hypospadias.[Figure presented]

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Status

Embase

Author NameID

Hadidi A.T.; ORCID: <http://orcid.org/0000-0001-6436-9905>

Institution

(Hadidi) Hypospadias Centre, Department of Pediatric Surgery, Emma and Sana Klinikum Offenbach Hospitals, Germany

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298.

Stent-related complications after hypospadias repair: a prospective trial comparing Silastic tubing and Koyle urethral stents.

Lee L.C., Schroder A., Bagli D.J., Lorenzo A.J., Farhat W.A., Koyle M.A.

Embase

Journal of Pediatric Urology. 14 (5) (pp 423.e1-423.e5), 2018. Date of Publication: October 2018.

[Article]

AN: 2001121145

Introduction: There is a paucity of data comparing urethral stents after hypospadias repair. The aim of this study is to compare Silastic tubing vs Koyle stents (Cook Medical), addressing outcomes related to stent-related complications, added visits to healthcare providers in the early postoperative period, and postoperative complications at clinic follow-up.

Material(s) and Method(s): Following an alternate week allocation, 150 patients were prospectively assigned to have Silastic tubes (n = 76) and Koyle stents (n = 74) after hypospadias repair. Exclusion criteria included fistula repairs, drainage via alternative catheter, or stentless repairs. Silastic tubes were secured with 5-0 Prolene and removed during a planned clinic visit. Koyle stents were secured with 7-0 PDS and left to fall out spontaneously. Questionnaires capturing postoperative outcomes were completed. Results and discussion: Median age was 13 and 11 months in the Silastic and Koyle stent groups, respectively (P = 0.48). There was no statistically significant difference in hypospadias location. Blockage/kinking of stents occurred in 8% (n = 6) of the Silastic and 9% (n = 7) Koyle stent groups, P = 0.78. Although follow-up was short, there was no difference in fistula rate among the Silastic (21%, n = 14) versus Koyle stent group (17%, n = 11), P = 0.66. There was a twofold higher rate of emergency department (ED) visits in the Silastic (32%, n = 24) versus Koyle stent group (16%, n = 12), P = 0.03. Half of ED visits in the Silastic group were related to stents falling out before planned removal. The authors

propose that Silastic stents falling out before the removal date may have led to increased parental anxiety and thus a visit to the ED. With improved parental education, the authors propose that many of these visits may have been preventable.

Conclusion(s): There were no significant differences in stent-related complications or fistula rate between the Silastic and Koyle stent groups. Although there were a twofold higher number of visits to the ED in the Silastic stent group, the authors propose that this was due to parental education rather than the stent itself. [Table presented]

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Status

Embase

Author NameID

Schroder A.; ORCID: <http://orcid.org/0000-0002-6508-2364>

Institution

(Lee, Schroder, Bagli, Lorenzo, Farhat, Koyle) Division of Urology, Department of Surgery, The Hospital for Sick Children, University of Toronto, 555 University Avenue, Rm M299, Toronto, Ontario M5G 1X8, Canada

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Year of Publication

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299.

Endoscopic injection of bulking agent around the ejaculatory ducts at the verumontanum for recurrent paediatric epididymitis.

Faure A., Haddad M., Hery G., Merrot T., Guys J.-M.

Embase

Journal of Pediatric Urology. 14 (5) (pp 476-482), 2018. Date of Publication: October 2018.

[Article]

AN: 2001055947

Introduction: Paediatric recurrent epididymitis is frequently observed in several urogenital conditions, and may result in deterioration of long-term fertility. The management of recurrent epididymitis is still a therapeutic challenge for paediatric urologists, and as yet there is no consensus for treatment.

Objective(s): To present a minimally invasive endoscopic approach for the treatment of recurrent epididymitis (more than three episodes).

Patients and Methods: Eleven boys were referred with a history of recurrent epididymitis in a context of urogenital malformations. All children underwent endoscopic transurethral injection. Dextranomer/hyaluronic acid was injected around the ejaculatory ducts at the verumontanum (Summary Fig.). The medical records and outcomes of the patients were retrospectively reviewed.

Result(s): Of the 11 boys, two (18%) had a history of bladder exstrophy, three (27%) anorectal malformation, two (18%) peno-scrotal hypospadias, two (18%) posterior urethral valves, one (9%) seminal vesicle cyst, and one (9%) urethral stricture. The median age at injection was 3.75 years (range 8 months-14.7 years). Endoscopic injection effectively prevented recurrence in eight patients (73%) with a mean follow-up of 3 years (range 6 months-8.8 years). The mean injected volume was 0.7 ml/session. No perioperative complications were recorded. Vas clipping was performed in three patients after unsuccessful injections.

Discussion(s): The current discussion for management of recurrent epididymitis is mainly based on vas clipping. Endoscopic injection in the verumontanum could offer several potential advantages over vas clipping; moreover, it is easy to perform for an urologist who usually uses endourological approaches. It is believed that only Kajbafzadeh et al. have reported their experience with endoscopic injection in the verumontanum in seven patients with structural anomalies, and they had a 42% success rate. Similarly, the current study did not observe perioperative or postoperative complications.

Conclusion(s): In this series, endoscopic injection of the verumontanum was considered to be a safe and effective treatment in almost 73% of children with recurrent epididymitis. It did not result in perioperative complications and not contraindicate a subsequent surgical procedure such as vas clipping.

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Status

Embase

Author NameID

Faure A.; ORCID: <http://orcid.org/0000-0002-3327-6391>

Institution

(Faure, Haddad, Hery, Merrot, Guys) Department of Paediatric Surgery, Aix-Marseille University, APHM, CHU Hopital La Timone Enfants, Marseille, France

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300.

Family History is Underestimated in Children with Isolated Hypospadias: A French Multicenter Report of 88 Families.

Ollivier M., Paris F., Philibert P., Garnier S., Coffy A., Fauconnet-Servant N., Haddad M., Guys J.M., Reynaud R., Faure A., Merrot T., Wagner K., Breaud J., Valla J.S., Dobremez E., Gaspari L., Daures J.-P., Sultan C., Kalfa N.

Embase

Journal of Urology. 200 (4) (pp 890-894), 2018. Date of Publication: October 2018.

[Article]

AN: 2001035432

Purpose: While familial forms of complex disorders/differences of sex development have been widely reported, data regarding isolated hypospadias are sparse and a family history is thought to be less frequent. We aimed to determine the frequency of hypospadias in families of boys with hypospadias, to establish whether these familial forms exhibit a particular phenotype and to evaluate the prevalence of genetic defects of the main candidate genes.

Material(s) and Method(s): A total of 395 boys with hypospadias were prospectively screened for a family history with a standardized questionnaire, extensive clinical description, family tree and sequencing of AR, SF1, SRD5A2 and MAMLD1.

Result(s): Family history of hypospadias was more frequent than expected (88 patients, 22.3%). In 17 instances (19.3%) familial hypospadias cases were multiple. Familial hypospadias was related to the paternal side in 59.1% of cases, consisting of the father himself (30.7%) as well as paternal uncles and cousins. Premature birth, assisted reproductive techniques, other congenital abnormalities and growth retardation were not more frequent in familial hypospadias than in sporadic cases. The severity of phenotype was similar in both groups. The results of genetic analysis combined with previous data on androgen receptor sequencing revealed that familial

cases more frequently tend to demonstrate genetic defects than sporadic cases (5.68% vs 1.63%, $p = 0.048$).

Conclusion(s): Familial forms of hypospadias are far more frequent than previously reported. Even minor and isolated forms justify a full clinical investigation of the family history. Detecting these hereditary forms may help to determine the underlying genetic defects, and may improve followup and counseling of these patients.

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Institution

(Ollivier, Garnier, Kalfa) Department of Pediatric Surgery and Urology, Lapeyronie Hospital, CHU Montpellier, Montpellier University, Montpellier, France (Paris, Philibert, Fauconnet-Servant, Kalfa) National Reference Center of Genital Development (Constitutive South), Arnaud de Villeneuve Hospital, CHU Montpellier, Montpellier University, Montpellier, France (Paris, Gaspari, Sultan, Kalfa) Pediatric Endocrinology and Gynecology Unit, Department of Pediatrics, Arnaud de Villeneuve Hospital, CHU Montpellier, Montpellier University, Montpellier, France

(Paris, Philibert, Fauconnet-Servant, Kalfa) Genetic Genital Development Unit, University Institute of Clinical Research, CHU Montpellier, Montpellier University, Montpellier, France

(Coffy, Daures) Laboratory of Biostatistics and Epidemiology, University Institute of Clinical Research, Montpellier University, Montpellier, France

(Haddad, Guys) Department of Pediatric Surgery and Urology, Timone Hospital, AP-HM, Marseille, France

(Reynaud) Pediatric Endocrinology and Diabetology Unit, Timone Hospital, AP-HM, Marseille, France

(Faure, Merrot) Department of Pediatric Surgery and Urology, Nord Hospital, AP-HM, Marseille, France

(Wagner) Pediatric Service, Lénval Hospital, CHU de Nice, Nice, France

(Breaud, Valla) Department of Pediatric Surgery and Urology, Lénval Hospital, CHU de Nice, Nice, France

(Dobremez) Department of Pediatric Surgery, Pellegrin Children's Hospital, Bordeaux, France

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301.

The Cologne pouch procedure for continent anal urinary diversion in children with bladder exstrophy-epispadias complex.

Klein T., Winkler A., Vahdad R.M., Ekamp A., Boemers T.M.

Embase

Journal of Pediatric Urology. 14 (5) (pp 431.e1-431.e6), 2018. Date of Publication: October 2018.

[Article]

AN: 2000957070

Introduction: In children who remain incontinent after reconstruction of bladder exstrophy-epispadias complex (BEEC), continent anal urinary diversion (CAD) is one option to achieve continence. Known problems after CAD are an increased stool frequency and ureterointestinal stenosis. We devised a new surgical technique of CAD that we named the "Cologne pouch procedure" (CPP) that renders the possibility of separate evacuation of urine and feces. Furthermore, we connect the bladder plate to the rectosigmoid pouch instead of performing a ureterosigmoidostomy to reduce the rate of ureterointestinal stenosis. In this study, we want to introduce the CCP and critically evaluate our results. Study design: In CPP a detubularized sigmoid-bladder pouch is created, which is naturally connected to the rectum. A retrospective study was performed including all patients with BEEC and CPP treated in our hospital between January 1, 2007, and December 31, 2016. Epidemiological and surgical key data, complications, and the need for alkaline supplementation were assessed. At follow-up examinations, we evaluated continence, ability of independent urine and feces evacuation, need for bicarbonate supplementation, status of the upper urinary tract, and complications such as urinary tract infections or urolithiasis.

Result(s): In total, 29 patients with BEEC and CPP were included. The mean age at surgery was 4.2 +/- 3.3 years (range 0.1-12.7 years). Overall, 14 short-term complications occurred in nine patients. Postoperatively, all patients were continent for urine and feces during daytime and only one child occasionally lost small portions of urine at night. An independent evacuation of urine and feces was accomplished in 22 patients (81.5%). Continued bicarbonate supplementation was necessary in 15 patients (55.6%). During the follow-up period six patients (22.2%) had a single urinary tract infection and four patients (14.8%) calculi of the urinary tract. No urinary tract abnormalities-especially no vesicoureteral reflux (VUR) or stenosis-were detected during follow-

up ultrasound examination. In two children, a preoperatively known hydronephrosis decreased after CPP.

Conclusion(s): CPP is a novel technique that yields excellent results concerning continence. In contrast to other forms of rectosigmoid urinary diversion, functional separation of defecation and urination can be achieved in most patients.[Figure presented]

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Embase

Author NameID

Klein, Tobias; ORCID: <http://orcid.org/0000-0003-0987-7514>

Institution

(Klein, Winkler, Vahdad, Ekamp, Boemers) Department of Pediatric Surgery and Pediatric Urology, Children's Hospital of Cologne, Cologne, Germany

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302.

Description of mutation spectrum and polymorphism of Wilms' tumor 1 (WT1) gene in hypospadias patients in the Indonesian population.

Diposarosa R., Pamungkas K.O., Sribudiani Y., Herman H., Suciati L.P., Rahayu N.S., Effendy S.H.

Embase

Journal of Pediatric Urology. 14 (3) (pp 237.e1-237.e7), 2018. Date of Publication: June 2018.

[Article]

AN: 2000887745

Introduction: Hypospadias is one of the most common congenital anomalies of the penis.

Previous studies reported mutation of the Wilms' tumor 1 (WT1) gene as a cause of hypospadias.

The aim of this study is to describe the WT1 mutation spectrum and polymorphism in hypospadias patients in Indonesia.

Material(s) and Method(s): DNA was isolated from 74 hypospadias patients at the Division of Pediatric Surgery, Department of Surgery Hasan Sadikin Hospital. All exons in the WT1 gene were amplified by a PCR method, followed by Sanger sequencing. Mutation analysis was performed using BioEdit software and in silico analysis using Mutation Taster, Polymorphism Phenotyping-2 (PolyPhen-2), and Sorting Intolerant from Tolerant (SIFT).

Result(s): DNA analysis results showed two types of heterozygous mutations in five subjects (Table), hence the frequency of WT1 mutations was 6.7% (10/148 allele). The first mutation was a missense mutation identified in twin boys. The second was a novel heterozygous alteration in the non-coding region nine bp upstream of exon 6 (c.366-9T>C), which was identified in three patients. One heterozygous polymorphism in the coding region of exon 7 (c.471A>G/rs16754) was identified in 10 subjects. This variant did not cause any change in amino acid products (silence polymorphism). Allele frequency for the G allele (mutant allele) and A allele (wild type) was 13.5% and 86.5%, respectively.

Discussion(s): WT1 is one of the best known hypospadias genes. The WT1 gene is involved in male genital development in the early and late periods of sex determination, and hence is known as a long-term expression gene in genitalia development. Mutation analysis of WT1 in a Chinese population identified that the WT1 mutation frequency was 4.4%. The WT1 mutation frequency identified in the present study was higher, at 6.7%. Coincidentally, research subjects with p.R158H variants were monozygotic twin siblings with midshaft hypospadias accompanied by undescended testis in one and penoscrotal hypospadias with micro penis in the other. The incidence of familial hypospadias in male siblings suffering from hypospadias was reported to be 9.6% in a study conducted by Sorensen et al. Moreover, in the present study polymorphism c.471A>G(rs16754) at exon 7 was identified heterozygously in 10 research subjects (minor allele frequency 13.5%).

Conclusion(s): WT1 mutations were identified in only a few cases of hypospadias and most of these were syndromic. This result implies that mutation of WT1 is not a common cause of hypospadias in the Indonesian population. [Table presented]

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Embase

Institution

(Diposarosa, Pamungkas) Department of Surgery, Pediatric Surgery Division, Hasan Sadikin Hospital, Faculty of Medicine, Padjadjaran University, Bandung, Indonesia (Sribudiani)

Department of Biochemistry and Molecular Biology, Faculty of Medicine, Padjadjaran University, Bandung, Indonesia

(Sribudiani, Effendy) Clinical Genetics Working Group, Faculty of Medicine, Padjadjaran University, Bandung, Indonesia

(Herman) Department of Surgery, Orthopedic Surgery Division, Hasan Sadikin Hospital, Faculty of Medicine, Padjadjaran University, Bandung, Indonesia

(Suciati, Rahayu) Laboratory of Genetic and Biology Molecular, Faculty of Medicine, Padjadjaran University, Bandung, Indonesia

(Effendy) Department of Pediatric, Hasan Sadikin Hospital, Faculty of Medicine, Padjadjaran University, Bandung, Indonesia

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303.

Predictors and outcomes of perioperative blood transfusions in classic bladder exstrophy repair: A single institution study.

Maruf M., Jayman J., Kasprenski M., Benz K., Feng Z., Friedlander D., Baumgartner T., Trock B.J., Di Carlo H., Sponseller P.D., Gearhart J.P.

Embase

Journal of Pediatric Urology. 14 (5) (pp 430.e1-430.e6), 2018. Date of Publication: October 2018.

[Article]

AN: 2000859294

Background: Primary bladder closure of classic bladder exstrophy (CBE) is a major operation that occasionally requires intraoperative or postoperative (within 72 h) blood transfusions.

Objective(s): This study reported perioperative transfusion rates, risk factors for transfusion, and outcomes from a high-volume exstrophy center in primary bladder closure of CBE patients. Study design: A prospectively maintained, institutional exstrophy-epispadias complex database of 1305 patients was reviewed for primary CBE closures performed at the authors' institution (Johns Hopkins Hospital) between 1993 and 2017. Patient and surgical factors were analyzed to

determine transfusion rates, risk factors for transfusions, and outcomes. Patients were subdivided into two groups based upon the time of closure: neonatal and delayed closure.

Result(s): A total of 116 patients had a primary bladder closure during 1993-2017. Seventy-three patients were closed in the neonatal period, and 43 were delayed closures. In total, 64 (55%) patients received perioperative transfusions. No transfusion reactions were observed. Twenty-five transfusions were in the neonatal closure group, yielding a transfusion rate of 34%. In comparison, 39 patients were transfused in the delayed closure group, giving a transfusion rate of 91%. Pelvic osteotomy, delayed bladder closure, higher estimated blood loss (EBL), larger pubic diastasis, and longer operative time were all associated with blood transfusion. In multivariable logistic regression, pelvic osteotomy (OR 5.4; 95% CI 1.3-22.8; $P < 0.001$), higher EBL-to-weight ratio (OR 1.3; 95% CI 1.1-1.6; $P = 0.029$), and more recent years of primary closure (OR 1.1; 95% CI 1.0-1.2; $P = 0.018$) remained independent predictors of receiving a transfusion (Summary Table). No adverse transfusion reactions or complications were observed.

Discussion(s): This was the first study from a single high-volume exstrophy center to explore factors that contribute to perioperative blood transfusions. Pelvic osteotomy as a risk factor was unsurprising, as the osteotomy may bleed both during and immediately after closure. However, it is important to use osteotomy for successful closure, despite the increased transfusion risk. The risks accompanying contemporary transfusions are minimal and osteotomies are imperative for successful bladder closure.

Conclusion(s): More than half of CBE patients undergoing primary closure at a single institution received perioperative blood transfusions. While there was an association between transfusions and osteotomy, delayed primary closure, larger diastasis, increased operative time, and increased length of stay, only the use of pelvic osteotomy, higher EBL-to-weight ratio, and recent year of closure independently increased the odds of receiving a transfusion on multivariate analysis.[Table presented]

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Author NameID

Kasprenski M.; ORCID: <http://orcid.org/0000-0002-8638-6068> Friedlander D.; ORCID: <http://orcid.org/0000-0002-4823-7803>

Di Carlo H.; ORCID: <http://orcid.org/0000-0003-2396-4796>

Gearhart J.P.; ORCID: <http://orcid.org/0000-0002-0177-6121>

Institution

(Maruf, Jayman, Kasprenski, Benz, Friedlander, Baumgartner, Di Carlo, Gearhart) Robert D. Jeffs
Division of Pediatric Urology, The James Buchanan Brady Urological Institutions, Johns Hopkins
Hospital, Johns Hopkins Medical Institutions, Charlotte Bloomberg Children's Hospital, Baltimore,
MD, United States (Feng, Trock) The James Buchanan Brady Urological Institute and
Department of Urology, Johns Hopkins University School of Medicine, Baltimore, Maryland MD,
United States

(Sponseller) Department of Orthopedic Surgery, The Johns Hopkins University, Baltimore, MD,
United States

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304.

The inadequate bladder template: Its effect on outcomes in classic bladder exstrophy.

Di Carlo H.N., Maruf M., Jayman J., Benz K., Kasprenski M., Gearhart J.P.

Embase

Journal of Pediatric Urology. 14 (5) (pp 427.e1-427.e7), 2018. Date of Publication: October 2018.

[Article]

AN: 2000842049

Introduction: Newborns with classic bladder exstrophy (CBE) may present with a bladder template that is inadequate for closure in the neonatal period (figure). In these cases, a delayed primary closure (DPC) is conducted to permit growth of the bladder template. This study reports the surgical and long-term urinary continence outcomes of poor template CBE patients undergoing DPC and compares them to patients who underwent DPC for reasons unrelated to bladder quality (i.e., prematurity, comorbidities, or a late referral).

Method(s): An institutionally approved, prospectively maintained database of 1330 exstrophy-epispadias complex patients was reviewed for CBE patients who underwent DPC at the authors' institution. A bladder template was considered inadequate for neonatal closure if found to be inelastic, <3 cm in diameter, and/or covered in hamartomatous polyps.

Result(s): In total, 63 patients (53 male and 10 female) undergoing DPC were identified. Of these, 36 had poor bladder templates (group 1). The remaining 27 patients (group 2) had adequate

templates and their bladder closure was delayed for reasons unrelated to bladder quality. At the time of DPC, those in group 1 were relatively than those in group 2 (median of 229 vs. 128 days, $p = 0.094$). All 36 group 1 patients and 26 (96%) group 2 patients underwent pelvic osteotomy during DPC ($p = 0.429$). All patients in this study had a successful primary closure. There was little difference in longitudinal bladder capacities between group 1 and group 2 ($p = 0.518$). Also, there was minimal difference in the median number of continence procedures between groups, with both groups having 1 (IQR 1-1) continence procedure ($p = 0.880$). Eight patients in group 1, and three patients in group 2 underwent a bladder neck transection with urinary diversion. Of the 13 and 16 patients who have undergone a continence procedure in group 1 and 2, respectively, 11 (84.6%) and 13 (81.3%) are continent of urine. The age of first continence procedure was different between groups 1 and 2 at 8.0 years (5.8-9.9 years) and 4.8 (3.5-6.0 years), respectively $p = 0.009$. The majority of patients in group 1 established continence at a relatively later age when compared to those in group 2, at 11.4 (8.0-14.8) years and 7.9 (2.6-13.2) years of age respectively $p = 0.087$.

Discussion(s): In the authors' view, neonatal bladder closure is ideal for CBE patients as it minimizes potential damage to exposed bladder mucosa. However, prior studies indicate that the rate of bladder growth for patients undergoing a delayed primary closure does not differ from patients with a neonatal closure. Results from this study show continued evidence that patients with poor templates who undergo delayed closure have excellent primary closure outcomes, which is critical for further management. Furthermore, this study shows that an inadequate bladder does not affect DPC outcomes or the continence outcomes in DPC patients. However, the inadequate template does affect the type of continence procedure available to a DPC patient, the age of first continence procedure, and the age of continence.

Conclusion(s): DPC of the exstrophic bladder has a high rate of success when pelvic osteotomy is utilized as an adjunct. Patients having a DPC for reasons of an inadequate bladder template have comparable rates of bladder growth when compared to DPC of an adequate bladder template. The inadequate bladder template affects the type of continence procedure, with the majority of patients requiring urinary diversion for continence. Patients with an inadequate bladder template have a later age of first continence procedure and a relatively later age of continence, because of an inherently smaller bladder template at birth. The inadequate bladder template patients require a longer period of surveillance to assess bladder growth and capacity in preparation of a continence procedure. Furthermore, as the majority of inadequate bladder template patients require a catheterizable channel for continence, the age of continence is also likely influenced by the patient's preparation as they transition from volitional voiding to catheterization.

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Author NameID

Di Carlo, Heather N.; ORCID: <http://orcid.org/0000-0003-2396-4796> Gearhart, John P.; ORCID: <http://orcid.org/0000-0002-0177-6121>

Institution

(Di Carlo, Maruf, Jayman, Benz, Kasprinski, Gearhart) Robert D. Jeffs Division of Pediatric Urology, James Buchanan Brady Urological Institutions, Johns Hopkins Hospital, Johns Hopkins Medical Institutions, Charlotte Bloomberg Children's Hospital, Baltimore, MD, United States

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305.

Salvaging the dehiscent glans penis.

White C.M., Hanna M.K.

Embase

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[Article]

AN: 2000839104

Objective: The glans penis may show a deep groove (surgically favorable), or may appear flat with an absent sulcus (unfavorable). Glans dehiscence following hypospadias repair, especially after multiple surgeries, frequently results in a scarred, obliterated, or absent urethral plate. The glans penis appears to be flat and grooveless. This study reported on the outcome of a two-stage salvage repair for glans dehiscence in 49 consecutive patients.

Material(s) and Method(s): Retrospective chart review was performed for all patients who underwent repair for glans dehiscence following hypospadias repair.

Result(s): Between January 2009 and April 2015, 49 children aged 16 months to 18 years presented with glans dehiscence following hypospadias repair. The prior number of operations ranged from one to six. Eleven children had urethral fistulas, and seven had chordee. In the first

stage, the flat glans was incised deeply to visualize, but spare, the corpora. Thereafter, a free graft of oral mucosa harvested either from the lower lip or cheek, or the residual preputial skin, was sutured to the glans cleft. The grafts were fenestrated, quilted in the midline, and a tie-over dressing was applied. Any fistula or chordee was repaired during the first stage. The neo-plate was tubularized 6-12 months later, and urine drainage with a catheter was maintained for 10-14 days. In 11 patients, skin flaps appeared dusky, and nitroglycerine ointment 2% was applied for 24 h to enhance the blood supply of the tissues. Subsequently, six of these children received nine or ten 90-min hyperbaric oxygen therapy sessions. Following the first stage, two patients developed hypertrophy of the mucosal grafts, and one skin graft contracted. These three patients underwent revision using a second buccal mucosal graft harvested from the cheek. One recurrent fistula was closed during the second stage. Following the second stage two patients developed a urethral fistula, and the distal sutures broke down in one patient, resulting in an over-sized meatus. None developed meatal stenosis or glans dehiscence.

Conclusion(s): Graft initial take and subsequent behavior were unpredictable, but the two stage approach optimized the process of take and healing. Glans dehiscence was repaired safely and successfully by developing a deep groove, with creation of a new urethral plate followed by tubularization in two stages.

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Status

Embase

Institution

(White) New Jersey Medical School, Newark, NJ, United States (Hanna) RWJ Saint Barnabas Medical Center, West Orange, NJ, United States

(Hanna) New York-Presbyterian Weill Cornell Hospital, New York, United States

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Tunica vaginalis flap for urethrocutaneous fistula repair after proximal and mid-shaft hypospadias surgery: A 12-year experience.

Pescheloché P., Parmentier B., Hor T., Chamond O., Chabaud M., Irtan S., Audry G.

Embase

Journal of Pediatric Urology. 14 (5) (pp 421.e1-421.e6), 2018. Date of Publication: October 2018.

[Article]

AN: 2000810910

Introduction: Fistulas are a common complication of hypospadias surgery; they are more frequent after mid-shaft and posterior hypospadias repair. Surgical treatment of fistula still remains challenging with a significant failure rate. The basic principle is to add layers between skin and neourethra in order to decrease the incidence of recurrent urethrocutaneous fistula (UCF). We report our experience of UCF repair using a vascularized tunica vaginalis flap (TVF) after posterior and mid-shaft hypospadias surgery.

Material(s) and Method(s): A retrospective review of all patients operated on using TVF for UCF in our institution between December 2005 and July 2017 was performed.

Result(s): Among 36 cases, TVF was used at a first attempt in 22 patients; 14 children had a prior attempt to close the fistula, and four of them had two surgeries before TVF repair. UCF was respectively penoscrotal (n = 3, 8%), posterior (n = 19, 53%), midshaft (n = 9, 25%) and anterior (n = 5, 14%). The size of the fistula was more than 5 mm in 26 patients. The UCF was treated successfully in every case after one single procedure. In the three children with two fistulas, both fistulas were successfully treated by the same TVF. After an average follow-up time of 45 months there was no recurrence of the initial UCF. In four cases of undescended testis, it was possible to dissect the flap through an inguinal incision and perform an orchidopexy in the same time. One patient presented a testicular atrophy after undescended testis surgery.

Discussion(s): Area review of published series shows excellent results in UCF repair including recurrent fistula (Table). TVF can aspire to some advantages with regard to a dartos flap (DF). First of all, a nearly 2.5-fold lower incidence of fistula after fistula repair with TVF than with DF (5.1% vs. 12.2%) has been shown. Secondly, TVF allows treating multiple fistulas, and can also be brought to the anterior part of the penis until the balano preputial furrow, allowing curing anterior fistula. Furthermore, it doesn't lead to aesthetic complications such as penile rotation or distal skin necrosis, which can occur during DF procedures.

Conclusion(s): TVF is a simple and reproductive technique for UCF repair, with a high success rate. The risk of testicular atrophy has to be considered in case of associated undescended testis surgery, and careful attention must be given to the TVF dissection. This technique should be considered as first choice treatment for any UCF. [Table presented]

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Status

Embase

Institution

(Pescheloche, Parmentier, Hor, Chamond, Chabaud, Irtan, Audry) Department of Pediatric Surgery, APHP Hopital Armand Trousseau, Sorbonne Universites, Paris, France

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307.

Effectiveness of caudal epidural block on intraoperative blood loss during hypospadias repair: A randomized clinical trial.

Alizadeh F., Heydari S.M., Nejadgashti R.

Embase

Journal of Pediatric Urology. 14 (5) (pp 420.e1-420.e5), 2018. Date of Publication: October 2018.

[Article]

AN: 2000805854

Background: Intraoperative blood loss is considered to be an important issue in hypospadias surgery. Some studies have demonstrated the utility of caudal epidural block (CEB) in this regard among pediatric patients with hypospadias. Though there is evidence in favor of the use of CEB as the only anesthetic method for pediatric surgeries, it is usually used in combination with general anesthesia. In this form of use, it could have more favorable outcomes for both intra- and postoperative periods. There are few studies regarding the effectiveness of CEB on intraoperative blood loss.

Objective(s): We aimed to evaluate the utility of CEB on blood loss, operation time, and analgesic use during hypospadias repairs of pediatric population referred to our university hospitals. Study design: In this randomized clinical trial, consecutive patients with hypospadias who were candidate for surgery were enrolled and randomly allocated to one of the two groups: group A received caudal epidural block (CEB) plus general anesthesia (GA) before surgery and group B received only GA. Intraoperative blood loss, operation time, and dose of fentanyl used during the

procedure were recorded and compared. All surgeries were performed by a single pediatric urologist in two different university hospitals that was not blinded to the study groups.

Result(s): In this study, 57 pediatric patients with hypospadias who underwent surgical repair were studied, from which 29 and 28 patients were allocated to receive or not to receive preoperative CEB, respectively. The patients' age, weight, ASA, score and severity of hypospadias were not significantly different between the two groups. The operation time was significantly lower in the CEB before surgery group ($p < 0.05$). The mean dose of fentanyl and volume of blood loss during procedure were lower in the CEB group, although for fentanyl it approached but did not reach statistical significance.

Conclusion(s): The findings of current study indicated that caudal epidural anesthesia in addition to general anesthesia has a favorable effect on reducing blood loss during operation, operation time, and analgesic use. Our data confirm the findings of previous studies in this field. Further studies are recommended to evaluate the effect of this type of analgesia in other outcomes of hypospadias repair surgery. Our results could be used for revising existing surgical guidelines for better management of hypospadias.[Figure presented]

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Author NameID

Alizadeh, Farshid; ORCID: <http://orcid.org/0000-0001-6855-2061>

Institution

(Alizadeh, Nejadgashti) Kidney Transplantation Research Center, Isfahan University of Medical Sciences, Department of Urology, Isfahan, Iran, Islamic Republic of (Heydari) Department of Anesthesiology and Critical Care Research Center, Isfahan University of Medical Sciences, Isfahan, Iran, Islamic Republic of

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Histological and morphological characteristics of the prepuce of penis skin structure in different age groups.

Dossanova A., Lozovoy V., Manekenova K., Lozovaya Y., Seidakhmetov M., Dossanov B., Omarov T., Botabaeva A., Shakeeva A., Baubekov Z.

Embase

Journal of Pediatric Urology. 14 (3) (pp 280.e1-280.e6), 2018. Date of Publication: June 2018.

[Article]

AN: 2000704849

Background: Hypospadias is one of the most common congenital abnormalities in childhood. The number of cases has rapidly grown in recent years.

Objective(s): The purpose of this research was to analyze the histological and morphological differences of the foreskin samples taken from boys in three age groups. Study design: A total of 30 Asian patients participated in the research. Clinical materials obtained via biopsy were divided into three age groups. The first group included 10 biopsy materials of preputial skin taken from boys aged <3 years. The second included 10 similar biopsy materials from boys aged 3-5 years. The third included 10 biopsy materials taken from boys aged 5-7 years. The skin areas were taken from the dorsal, two lateral and the ventral surfaces (closer to the bridge) with dimensions of 1.0 x 1.0 cm. All removed foreskins underwent histological examination.

Result(s): Obtained results showed that the number of vein clusters in the prepuce and the cases of vessel wall fibrosis grew with age. It is worth noting that no such discoveries were made in younger boys (aged <3 years). Sample analysis showed that the number of nerve, vessel, and collagen fibers increased with age.

Discussion(s): It is believed that it is important to continue investigating the prepuce in hypospadias, in order to gain a better understanding of the abnormality depending on type.

Conclusion(s): Peculiarities of prepuce in hypospadias discovered in different age groups allowed a full understanding of the pathology development processes.

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Institution

(Dossanova, Lozovoy, Manekenova, Lozovaya, Seidakhmetov, Dossanov, Omarov, Botabaeva, Shakeeva) JCS Astana Medical University, Astana, Kazakhstan (Baubekov) West Kazakhstan Medical University, Aktobe, Kazakhstan

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309.

Age-related changes in urinary flow following dorsal inlay graft urethroplasty for hypospadias in early childhood: Potential improvement over 11 years of age.

Kim W.J., Hayashi C., Yamazaki Y.

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[Article]

AN: 2000639922

Introduction: One of the goals of hypospadias repair is to create a neourethra with normal urinary stream and normal growth. Several studies have reported that dorsal inlay graft urethroplasty (DIG) has wide indications for various clinical phenotypes of hypospadias, with good short-term outcomes and few complications. However, there have been no reports that evaluated both short-term and long-term functional outcomes using uroflowmetry in patients with DIG.

Objective(s): The aim was to investigate whether uroflowmetry parameters change with time following DIG for hypospadias in early childhood.

Material(s) and Method(s): Uroflowmetry parameters after DIG for hypospadias in childhood were retrospectively evaluated and compared between two defined ages at follow-up: 4-6 years and 11-13 years. Maximum urinary flow (Qmax) under the 25th percentile on the Miskolc nomogram was defined as obstruction. To evaluate the shapes of uroflow quantitatively, the flow index (FI) was used. The FI cut-offs for the flow shapes were defined as tower >1.253, plateaus <0.659; bells were between these two values.

Result(s): Thirty-four patients met the inclusion criteria. The median operative age was 1.8 years (range 1.0-4.9 years). The median follow-up time was 10.3 years (range 7.2-12.3 years). The mean $\pm$ SD Qmax at the two follow-up times increased with time, from 9.2 $\pm$ 3.7 to 18.8 $\pm$ 7.8. The mean $\pm$ SD FI changed from 0.53 $\pm$ 0.19 to 0.85 $\pm$ 0.31. Both Qmax and the FI were significantly improved ($p < 0.001$, $p < 0.001$, respectively). There were significantly fewer patients with obstruction evaluated by the Miskolc nomogram at 11-13 years of age ($n = 11$, 32.4%) than at 4-6 years ($n = 31$, 91.2%) ($p < 0.001$). Plateau shapes assessed by the FI were seen in 24

(70.6%) patients at 4-6 years and 10 (29.4%) patients at 11-13 years (Table). The number of patients with plateau shapes was significantly decreased ($p = 0.001$).

Discussion(s): Only one report noted long-term outcomes and spontaneous uroflowmetry resolution after tubularized incised plate urethroplasty. The current report is the first to present the short-term and long-term uroflowmetry outcomes after DIG. In addition, the FI was used for the first time to evaluate uroflowmetry after hypospadias repair. It was found that the uroflowmetry parameters improved spontaneously over 11 years of age following DIG for hypospadias in early childhood.

Conclusion(s): Uroflowmetry parameters improved spontaneously over 11 years of age following DIG for hypospadias in early childhood. [Table presented]

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Institution

(Kim, Hayashi, Yamazaki) Department of Urology, Kanagawa Children's Medical Center, Yokohama, Japan

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310.

Surgical Approaches and Long-Term Outcomes in Adults with Complex Reoperative Hypospadias Repair.

Morrison C.D., Cina D.P., Gonzalez C.M., Hofer M.D.

Embase

Journal of Urology. 199 (5) (pp 1296-1301), 2018. Date of Publication: May 2018.

[Article]

AN: 2000606477

Purpose: Patients with failed hypospadias repair are a challenging population for pediatric and reconstructive urologists. We describe our long-term outcomes and factors associated with complications of repeat hypospadias repair.

Material(s) and Method(s): We retrospectively reviewed the records of 32 adult patients with a history of hypospadias repair who required subsequent urethroplasty between 2002 and 2012. Data on the presenting complaint, past medical and surgical history, demographic data, surgical approach, intraoperative findings and complications were collected and analyzed.

Result(s): Median patient age at urethroplasty was 32 years. Stricture of the penile urethra was the most common presentation. Urethroplasty was done in 30 patients as stricture treatment, 1 underwent perineal urethrostomy and 1 underwent diverticulectomy. Two-stage repair was performed in 90% of the men who underwent urethroplasty. The initial success rate was 83% in patients who underwent 1 or 2-stage urethroplasty. At a median followup of 9.5 years complications included 4 recurrent strictures and 1 fistula. Patient age, previous interventions, stricture length, hair present at the time of repair, the need to excise the urethral plate and the number of stages were not associated with complications or recurrence. If a graft was required, skin grafts were significantly associated with recurrence compared to buccal mucosa grafts.

Conclusion(s): Excellent outcomes can be achieved using a 2-stage approach with replacement or augmentation of the urethral plate in adults with failed hypospadias repair. In our experience buccal mucosa appears to be associated with fewer complications and less stricture recurrence than skin grafts.

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Embase

Institution

(Morrison, Cina, Gonzalez, Hofer) Department of Urology, Northwestern University Feinberg School of Medicine, Chicago, Illinois; Urology Institute, University Hospitals Cleveland Medical Center, Case Western Reserve Medical School, Cleveland, Ohio

Publisher

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Proximal hypospadias treated with a transverse preputial island tube: long-term functional, sexual, and cosmetic outcomes.

Rynja S.P., de Jong T.P.V.M., Bosch J.L.H.R., de Kort L.M.O.

Embase

BJU International. 122 (3) (pp 463-471), 2018. Date of Publication: September 2018.

[Article]

AN: 622125382

Objective: To study long-term urinary and sexual function, and cosmetic outcomes in adult patients who underwent single-stage transverse preputial island tube (TPIT) for proximal hypospadias repair in childhood. Long-term data on outcomes of patients with proximal hypospadias with severe coexisting curvature and insufficient urethral plate are scarce, but are necessary to decide which repair technique is most beneficial. **Patients, Subjects and Methods:** Patients with proximal hypospadias operated with TPIT (TPIT Group) were compared to patients with distal hypospadias repair (Distal Group) and to a control group of male medical students (Control Group). Participants completed the International Prostate Symptom Score, the International Index of Erectile Function (IIEF-15), additional non-validated questions, and performed uroflowmetry. Cosmesis was assessed in the patients with hypospadias using the Pediatric Penile Perception Score (PPPS); stretched penile length was also measured.

Result(s): Of the 121 eligible patients with hypospadias, 54 with either TPIT or distal hypospadias repairs participated. The TPIT Group comprised 12 patients (median age of 20.0 years) and the Distal Group comprised 42 patients (median age of 19.6 years). The complication rates were similar, at 8 of the 12 patients in the TPIT Group vs 26/42 (62%) in the Distal Group ($P = 0.76$). Urinary outcomes were similar in the TPIT, Distal, and the Control (comprised of 148 medical students with a median age of 21.0 years) groups. The TPIT Group had a lower maximum urinary flow rate compared to the Control Group, at 24.1 vs 28.6 mL/s ($P < 0.05$). IIEF-15 scores were similar in the TPIT, Distal and Control groups, except for 'Orgasmic Function' (7.5 vs 10.0 vs 10.0, respectively; $P < 0.01$). Although the TPIT Group had a smaller penile length compared to the Distal Group (10.1 vs 12.9 cm, $P < 0.01$), PPPS outcomes were similar.

Conclusion(s): In these TPIT patients, long-term urinary, sexual and cosmetic outcomes were similar to those in patients with distal hypospadias repairs and controls.

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Status

Embase

Author NameID

Rynja, Sybren P.; ORCID: <http://orcid.org/0000-0002-8155-0885>

Institution

(Rynja, Bosch, de Kort) University Medical Center (UMC), Utrecht, Netherlands (de Jong)

University Children's Hospital UMC Utrecht, Utrecht, Netherlands

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312.

Contemporary outcomes of hypospadias retrieval surgery in adults.

Aldamanhori R.B., Osman N.I., Inman R.D., Chapple C.R.

Embase

BJU International. 122 (4) (pp 673-679), 2018. Date of Publication: October 2018.

[Article]

AN: 622113210

Objective: To describe the surgical approach and outcomes in the treatment of adult patients with complications of childhood hypospadias surgery, as such patients present a significant reconstructive challenge due to the combination of anatomical and cosmetic deformity, which often results in major functional and psychosexual sequelae.

Patients and Methods: We analysed prospectively collected data on 79 adults with complications of childhood hypospadias surgery, who were operated on between 2004 and 2016. Of the 79 patients, 48 underwent a two-stage urethroplasty using a buccal mucosa graft, and 31 underwent a one-stage distal urethroplasty.

Result(s): Patients were followed up using flexible cystoscopy (every 6-9 months). The mean (range) follow-up was 48 (12-96) months. Of the 48 patients who underwent a two-stage repair, eight (16%) needed a revision of the first-stage graft. In total, nine of the 48 patients (16%) developed fistula requiring closure after the second stage; all but one was closed successfully on the first attempt, whilst one required two attempts before closure. Only two of the 48 patients that

underwent a two-stage procedure required a re-do urethroplasty within 3 years. Of the 31 patients who underwent a one-stage repair, six (19%) needed fistula closure, all of which were successful. No patient required a further urethroplasty during follow-up.

Conclusion(s): Despite the significant surgical challenges found in this patient group, excellent long-term functional outcomes can be achieved. As expected there is a need for additional intervention, either for revision of the first stage or to close fistulae and less commonly for further reconstruction for stricture recurrence.

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Status

Embase

Author NameID

Aldamanhori, Reem B.; ORCID: <http://orcid.org/0000-0003-3676-6860>

Institution

(Aldamanhori, Osman, Inman, Chapple) Section of Functional and Reconstructive Urology, Department of Urology, Royal Hallamshire Hospital, Sheffield, United Kingdom (Aldamanhori) University of Dammam, Dammam, Saudi Arabia

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2018

313.

Congenital Abnormalities of the Male Reproductive System and Risk of Autism Spectrum Disorders.

Rotem R.S., Chodick G., Davidovitch M., Hauser R., Coull B.A., Weisskopf M.G.

Embase

American Journal of Epidemiology. 187 (4) (pp 656-663), 2018. Date of Publication: 01 Apr 2018.

[Article]

AN: 621707034

Androgens have an extensive influence on brain development in regions of the brain that are relevant for autism spectrum disorder (ASD), yet their etiological involvement remains unclear. Hypospadias (abnormal positioning of the urethral opening) and cryptorchidism (undescended testes) are 2 relatively common male birth defects that are strongly associated with prenatal androgen deficiencies. Having either disorder is a proxy indicator of atypical gestational androgen exposure, yet the association between these disorders and autism has not been extensively studied. We analyzed male singleton live births ($n = 224,598$) occurring from January 1, 1999, through December 31, 2013, in a large Israeli health-care organization. Boys with autism, cryptorchidism, and hypospadias were identified via International Classification of Diseases, Ninth Revision, codes, with further verification of autism case status by review of medical records. In multivariable-adjusted analyses, the odds ratio for ASD among boys with either condition was 1.62 (95% confidence interval (CI): 1.44, 1.82). The odds ratio for boys with cryptorchidism only was 1.55 (95% CI: 1.34, 1.78), and that for boys with hypospadias only was 1.65 (95% CI: 1.38, 1.98). ASD risk was not elevated among unaffected brothers of hypospadias or cryptorchidism cases, despite familial aggregation of all 3 conditions, providing some indication for the possibility of pregnancy-specific risk factors driving the observed associations. Results suggest that in-utero hypoandrogenicity could play a role in ASD etiology.

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Status

Embase

Institution

(Rotem, Hauser, Coull, Weisskopf) Department of Environmental Health, Harvard T.H. Chan School of Public Health, 401 Park Drive, Landmark Center, P.O. Box 15697, Boston, MA 02215, United States (Hauser, Weisskopf) Department of Epidemiology, Harvard T.H. Chan School of Public Health, Boston, MA, United States

(Coull) Department of Biostatistics, Harvard T.H. Chan School of Public Health, Boston, MA, United States

(Chodick) Maccabi Institute for Research and Innovation, Maccabi Healthcare Services, Tel Aviv, Israel

(Davidovitch) Child Development Department, Maccabi Healthcare Services, Tel Aviv, Israel

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314.

Primary female epispadias: Perineal approach or Kelly repair?.

Leclair M.-D., Faraj S., Villemagne T., Carrouget J., Arnaud A., Heloury Y.

Embase

Journal of Pediatric Urology. 14 (1) (pp 33-39), 2018. Date of Publication: February 2018.

[Article]

AN: 621216991

Objectives: Primary female epispadias encompasses a spectrum of disease, presenting with a variable degree of incontinence. We hypothesized that although perineal urethrocervicoplasty can be a successful first-line procedure in patients with normal bladder, a more radical reconstruction was necessary to achieve continence in cases lying at the most severe end of the spectrum. Our aim was to assess the results of a surgical management using perineal approach in girls with normal bladder capacity, and Kelly radical soft-tissue mobilization (RSTM) in patients with inadequate bladder, based on the assumption that bladder capacity (BC) is a reliable marker of epispadias severity. Study design: Prospective inclusion of incontinent girls with female epispadias referred to a single institution. Patients with normal BC were treated with perineal urethrocervicoplasty (PUCP, group 1). Patients with small bladder underwent RSTM (group 2). Follow-up was at 1, 3, 6, and 12 months postoperatively, then annually, including physical examination, renal ultrasound at each visit, continence status, and estimation of functional/maximal BC. The main study outcome was continence status at the age of 5 years or later, if postoperative follow-up was >12 months.

Result(s): From 2006 to 2017, 16 consecutive children were prospectively included in this study, at a median age of 39 months (5-102 months). Seven girls were included in group 1 and underwent PUCP; at the last follow-up, five out of seven were dry by day (4/5 day and night), although three out of five required bladder-neck injection after perineal reconstruction due to stress incontinence. Two patients with persistent incontinence and absence of BC increase after PUCP subsequently underwent RSTM. Eleven patients with low BC (56% [10-94%] of expected BC) were included in group 2 (9 without prior surgery, 2 after PUCP failure). Among the eight evaluable patients, eight out of eight achieved diurnal continence, and 3/8 were fully continent. One girl with obstructive micturition required clean intermittent catheterization.

Discussion(s): The traditional approach of female epispadias based on staged reconstruction (urethroplasty followed by bladder-neck reconstruction) raised concerns regarding the risk of non-physiological obstructive micturition. The perineal approach was suggested as an alternative, with reported diurnal continence rates of 60-80%, but less than 50% of nocturnal continence, presumably in relation with limited bladder capacity. In cases selected within the most severe end of the epispadias spectrum, the Kelly RSTM seems to offer excellent continence rates.

Conclusion(s): A tailored approach to female epispadias, based on perineal reconstruction in favorable cases, and radical soft-tissue mobilization in severe cases, seems to yield good continence outcomes in the long term.

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Status

Embase

Author NameID

Leclair, Marc-David; ORCID: <http://orcid.org/0000-0002-1771-9025> Heloury, Yves; ORCID: <http://orcid.org/0000-0003-0843-3970>

Institution

(Leclair, Faraj, Villemagne, Carrouget, Arnaud, Heloury) Pediatric Urology Department, University Children Hospital, Nantes, France (Villemagne) Pediatric Surgery Department, University Hospital, Tours, France

(Carrouget) Pediatric Surgery Department, University Hospital, Angers, France

(Arnaud) Pediatric Surgery Department, University Hospital, Rennes, France

(Heloury) Pediatric Urology Department, Royal Children's Hospital, Melbourne, Australia

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315.

Postoperative pain and analgesia administration in children after urological outpatient procedures. Schroder A., Campbell F.A., Farhat W.A., Salle J.L.P., Bagli D.J., Lorenzo A.J., Koyle M.A.

Embase

Journal of Pediatric Urology. 14 (2) (pp 171.e1-171.e6), 2018. Date of Publication: April 2018.

[Article]

AN: 620707033

Introduction: There are limited data about pain patterns, analgesic requirements and factors predicting opioid requirements of children undergoing outpatient urologic surgery. This prospective study aimed to assess recovery profiles and pain medication requirements.

Method(s): Patients between 6 months and 12 years of age were recruited prospectively between December 2013 and June 2014. Demographic and operative characteristics were collected.

Following discharge home, the parents were asked to administer both acetaminophen and ibuprofen Q6H at a weight-adjusted dose, based on a schedule, until the end of postoperative day 2, and to administer the medication as required on postoperative day 3. Pain severity was recorded using validated pain scores (Face, Legs, Activity, Cry, Consolability/Parents' Postoperative Pain Measurement). A morphine prescription was provided for breakthrough pain. A Likert scale was used to assess parent's satisfaction with the pain management.

Result(s): A total of 249 patients were recruited, 111 patients (45%) returned appropriately completed surveys and were included in the final analysis. Mean age was 44.1 months (SD = 37.3). The performed procedures were orchidopexy (31), hypospadias repair (26), hernia/hydrocele repair (15), Fowler-Stephens procedure (13), meatoplasty (7), phalloplasty (4), scrotoplasty (1), circumcision (7), and diagnostic laparoscopy (5). After discharge home 17 patients (15.3%) received morphine. Mean utilization of non-opioid analgesia was 79% on postoperative day 1, 67% on day 2, 36% on day 3, and 2% on day 4. Parental satisfaction was high (92.0% satisfied/very satisfied). No patient, anaesthetic or surgical factors were associated with opioid use or prolonged need for postoperative analgesia.

Conclusion(s): The combination of scheduled non-opioid medications for maintenance and opioids for breakthrough pain provided satisfactory pain control after outpatient urologic surgery in children. There were no specific patient, anesthetic or surgical factors that predicted postoperative opioid requirements.[Figure presented]

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Institution

(Schroder, Farhat, Salle, Bagli, Lorenzo, Koyle) Division of Urology, Hospital for Sick Children and University of Toronto, Toronto, Canada (Campbell) Department of Anesthesiology and Pain Management, Hospital for Sick Children and University of Toronto, Toronto, Canada

(Schroder) Division of Pediatric Urology, Department of Urology, University of Mainz, Medical School, Germany

(Salle) Division of Pediatric Urology, Sidra Medical and Research Center, Qatar

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316.

Preliminary report: Surgical outcomes following genitoplasty in children with moderate to severe genital atypia.

Bernabe K.J., Nokoff N.J., Galan D., Felsen D., Aston C.E., Austin P., Baskin L., Chan Y.-M., Cheng E.Y., Diamond D.A., Ellens R., Fried A., Greenfield S., Kolon T., Kropp B., Lakshmanan Y., Meyer S., Meyer T., Delozier A.M., Mullins L.L., Palmer B., Paradis A., Reddy P., Reyes K.J.S., Schulte M., Swartz J.M., Yerkes E., Wolfe-Christensen C., Wisniewski A.B., Poppas D.P.
Embase

Journal of Pediatric Urology. 14 (2) (pp 157.e1-157.e8), 2018. Date of Publication: April 2018.

[Article]

AN: 620534986

Introduction: Prior studies of outcomes following genitoplasty have reported high rates of surgical complications among children with atypical genitalia. Few studies have prospectively assessed outcomes after contemporary surgical approaches.

Objective(s): The current study reported the occurrence of early postoperative complications and of cosmetic outcomes (as rated by surgeons and parents) at 12 months following contemporary genitoplasty procedures in children born with atypical genitalia. Study design: This 11-site, prospective study included children aged ≤ 2 years, with Prader 3-5 or Quigley 3-6 external genitalia, with no prior genitoplasty and non-urogenital malformations at the time of enrollment. Genital appearance was rated on a 4-point Likert scale. Paired t-tests evaluated differences in cosmesis ratings.

Result(s): Out of 27 children, 10 were 46,XY patients with the following diagnoses: gonadal dysgenesis, PAIS or testosterone biosynthetic defect, severe hypospadias and microphallus, who were reared male. Sixteen 46,XX congenital adrenal hyperplasia patients were reared female and

one child with sex chromosome mosaicism was reared male. Eleven children had masculinizing genitoplasty for penoscrotal or perineal hypospadias (one-stage, three; two-stage, eight). Among one-stage surgeries, one child had meatal stenosis (minor) and one developed both urinary retention (minor) and urethrocutaneous fistula (major). Among two-stage surgeries, three children developed a major complication: penoscrotal fistula, glans dehiscence or urethral dehiscence. Among 16 children who had feminizing genitoplasty, vaginoplasty was performed in all, clitoroplasty in nine, external genitoplasty in 13, urethroplasty in four, perineoplasty in five, and total urogenital sinus mobilization in two. Two children had minor complications: one had a UTI, and one had both a mucosal skin tag and vaginal mucosal polyp. Two additional children developed a major complication: vaginal stenosis. Cosmesis scores revealed sustained improvements from 6 months post-genitoplasty, as previously reported, with all scores reported as good or satisfied.

Discussion(s): In these preliminary data from a multi-site, observational study, parents and surgeons were equally satisfied with the cosmetic outcomes 12 months after genitoplasty. A small number of patients had major complications in both feminizing and masculinizing surgeries; two-stage hypospadias repair had the most major complications. Long-term follow-up of patients at post-puberty will provide a better assessment of outcomes in this population.

Conclusion(s): In this cohort of children with moderate to severe atypical genitalia, preliminary data on both surgical and cosmetic outcomes were presented. Findings from this study, and from following these children in long-term studies, will help guide practitioners in their discussions with families about surgical management.[Figure presented]

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Author NameID

Chan Y.-M.; ORCID: <http://orcid.org/0000-0003-0554-8502> Kolon T.; ORCID: <http://orcid.org/0000-0002-8740-5021>

Institution

(Bernabe, Galan, Felsen, Poppas) The Comprehensive Center for Congenital Adrenal Hyperplasia, New York-Presbyterian Hospital/Weill Cornell Medicine, New York, NY, United States (Nokoff) Children's Hospital Colorado, Aurora, CO, United States (Aston) University of Oklahoma Health Sciences Center, Department of Pediatrics, Oklahoma City, OK, United States (Austin, Paradis) St. Louis Children's Hospital, St. Louis, Missouri, United States (Baskin) University of California San Francisco, San Francisco, CA, United States

(Chan, Diamond, Swartz) Children's Hospital Boston, Department of Urology, Boston, MA, United States

(Cheng, Meyer, Yerkes) Ann & Robert H. Lurie Children's Hospital of Chicago, Chicago, IL, United States

(Ellens, Lakshmanan, Wolfe-Christensen) Children's Hospital of Michigan, Detroit, MI, United States

(Fried, Greenfield, Meyer) Women and Children's Hospital of Buffalo, Buffalo, NY, United States

(Kolon) Children's Hospital of Philadelphia, Philadelphia, PA, United States

(Kropp, Palmer, Reyes, Wolfe-Christensen, Wisniewski) Genitourinary Institute, Cook Children's Hospital, Fort Worth, TX, United States

(Delozier, Mullins) Oklahoma State University, College of Arts and Sciences, Stillwater, OK, United States

(Reddy, Schulte) Cincinnati Children's Hospital Medical Center, Cincinnati, OH, United States

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317.

Parental decisional regret and views about optimal timing of female genital restoration surgery in congenital adrenal hyperplasia.

Szymanski K.M., Whittam B., Kaefer M., Frady H., Casey J.T., Tran V.T., Cain M.P., Rink R.C.

Embase

Journal of Pediatric Urology. 14 (2) (pp 156.e1-156.e7), 2018. Date of Publication: April 2018.

[Article]

AN: 620148422

Purpose: The role of female genital restoration surgery (FGRS) in girls with congenital adrenal hyperplasia (CAH) is controversial, with no long-term parent-reported outcomes available.

Decisional regret (DR) affects most parents after their children's treatment of pediatric conditions, including hypospadias. We aimed to assess parental DR after FGRS in infancy or toddlerhood and explore optimal timing for surgery.

Material(s) and Method(s): One-hundred and six parents of females with CAH undergoing FGRS before 3 years old and followed at our institution (1999-2017) were invited to enroll online. Higher Decision Regret Scale (DRS) scores indicated greater DR (range 0-100). Participants also reported preferred FGRS timing relative to their surgery (earlier, same, later/delayed). Non-parametric statistical tests were used.

Result(s): Thirty-nine parents (median 4.4 years after FGRS) participated (36.8% response rate). Median age at FGRS was 9 months. Median DRS score was 0 (mean: 5.0). Overall, 20.5% of parents reported some regret (all mild-moderate) (Figure). Fewer parents reported DR after FGRS compared with published DR after hypospadias repair (50-92%, $p \leq 0.001$) or adenotonsillectomy (41-45%, $p \leq 0.03$). No parent preferred delayed FGRS. Seven parents (18.1%) preferred earlier surgery, especially when performed after birthday (80.0% vs. 8.8%, $p = 0.004$).

Discussion(s): We present the first report of validated long-term parent-reported outcomes after FGRS in infant and toddler girls with CAH. One limitation is that this is largely a single surgeon series. Reasons for the observed low levels of DR are likely multifactorial. Far from a definitive study, we aimed to provide parents willing to share about their experience an opportunity to do so. For that reason, selection bias may exist in our study. While parents with higher DR were potentially less likely to participate because of mistrust of the medical establishment, those with a negative experience may in fact be more likely to voice their opinions. A low participation rate was likely a result of the sensitive nature of FGRS, a desire for privacy, and inability to locate parents. A larger study will be required to assess how DR is affected by sexual function, genital appearance and complications, and DR among women with CAH.

Conclusion(s): Parents of females with CAH report low levels of DR after FGRS in infancy and toddlerhood. This appears to be lower than after other genital and non-genital pediatric procedures. When present, parental DR is usually mild. No parents preferred delayed surgery, even among those with DR. Some preferred earlier surgery.

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Status

Embase

Institution

(Szymanski, Whittam, Kaefer, Frady, Casey, Tran, Cain, Rink) Division of Pediatric Urology, Riley Hospital for Children at IU Health, IN, United States

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318.

Distal urethral plate adhesions: New anatomical perspective in hypospadias.

Drlik M., Kocvara R., Sedlacek J., Dite Z., Vitkova I., Komarc M.

Embase

Journal of Pediatric Urology. 14 (2) (pp 164.e1-164.e5), 2018. Date of Publication: April 2018.

[Article]

AN: 619406526

Introduction: We found midline epithelial adhesions in the glandar urethral plate in patients with hypospadias. After dissolution, a blind epithelized channel becomes visualized inside of the plate pointing to immature embryonic luminization. In addition it reveals that the epithelized surface of the distal urethral plate is larger than previously considered.

Objective(s): To determine the incidence and extent of these new anatomical details of urethral plate in hypospadias patients.

Method(s): We prospectively assessed the detailed anatomy of the urethral plate in 72 consecutive patients with hypospadias. We recorded the presence of adhesions in the middle of the glandar urethral groove that can be easily dissolved (dissolution line - D-line). We recorded the plate width before and after D-line dissolution, the presence of the hidden blind channel at continuation of D-line (channel type-A) and of the visible blind channel between D-line and urethral hypospadiac meatus (type-B) (Figure). In 62 patients, where the urethral plate tubularization was considered (Duplay, TIP), septa between channels were opened in the midline and a final width of the plate was measured by rolling the plate around a tube.

Result(s): Midline adhesions (D-line) were found in all 72 patients. Mean length of D-line was 5.13 +/- 0.17 mm. Mean plate width before dissolution was 5.9 +/- 0.15 mm, and after dissolution 7.8 +/- 0.16 mm. A blind channel of type A was detected in 22 patients (31%), type B in 24 (33%), type A and B in 16 (22%), and none in 10 patients (14%). Mean final plate width after D-line dissolution and opening of septa between channels in 62 patients with urethral plate tubularization was 8.7 +/- 0.15 mm.

Discussion(s): The main contribution of our study is a new perspective of distal urethral plate anatomy that enables enlargement of the epithelized surface of the distal urethral plate by

dissolution of the preexisting epithelized groove and opening of epithelized channels within the plate. To the best of our knowledge, this anatomical anomaly has not been described previously. Conclusion(s): The distal urethral plate of all hypospadias patients is partially "folded" in the midline by epithelial adhesions of different depth and extent that may be easily dissolved. In half of the patients (53%) the "folded" part of the plate continues proximally as a blind channel inside the urethral plate (type A channel). Opening of these structures together with the well-known urethral plate pits (type B channel) helps augment the width and the overall epithelized surface of the distal urethral plate.[Figure presented]

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Status

Embase

Institution

(Drlik, Kocvara, Sedlacek, Dite) Division of Paediatric Urology, Department of Urology, General Teaching Hospital, Charles University First Faculty of Medicine, Prague, Czechia (Vitkova)
Department of Pathology, General Teaching Hospital, Charles University First Faculty of Medicine, Prague, Czechia

(Komarc) Department of Methodology, Faculty of Physical Education and Sport, Charles University, Prague, Czechia

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2018

319.

Testosterone prior to hypospadias repair: Postoperative complication rates and long-term cosmetic results, penile length and body height.

Rynja S.P., de Jong T.P.V.M., Bosch J.L.H.R., de Kort L.M.O.

Embase

Journal of Pediatric Urology. 14 (1) (pp 31.e1-31.e8), 2018. Date of Publication: February 2018.

[Article]

AN: 619389043

Introduction: The use of hormonal therapy was first described in 1971 before hypospadias surgery, and it has been debated ever since. The long-term outcomes after puberty of patients treated with pre-operative testosterone in childhood are lacking.

Objective(s): Possible long-term effects of testosterone are often asked about in daily practice. The current study investigated the long-term outcomes regarding height, penile length and penile cosmesis in adult men after hypospadias surgery in childhood with and without pre-operative testosterone.

Method(s): Adult men ($n = 121$) who underwent primary hypospadias repair in childhood were included. Pre-operative penile appearance, judged by a paediatric urologist, determined the use of pre-operative testosterone. Data on hypospadias characteristics, healing complications, surgical repair, and testosterone use were collected retrospectively. At adult age, stretched penile length and body height were measured, and penile cosmesis was evaluated using the Pediatric Penile Perception Score (PPPS).

Result(s): Postoperative complication rates in patients ($n = 121$) with and without testosterone were similar (50% vs. 43%; $P = 0.54$). Sixty adult patients (50%) with a median age of 19.8 years and follow-up time of 18.3 years were examined at the outpatient clinic at adult age. Of this group, testosterone was applied in 12/43 patients with distal, 3/6 patients with midshaft, and 9/11 patients with proximal hypospadias. Adult stretched penile length (12.0 cm vs. 12.4 cm; $P = 0.47$) and adult height (180.1 cm vs. 179.0 cm $P = 0.65$) showed no difference between patients with and without testosterone treatment. Penile cosmesis was (very) satisfactory in all PPPS domains, and showed no difference between the testosterone group and the non-testosterone group. Univariate and multivariate analysis was conducted to determine if the hypospadias type or pre-operative testosterone therapy had more influence on the long-term outcomes. None of the long-term outcomes were significantly associated with pre-operative testosterone therapy on multivariate analysis.

Discussion(s): This was the first study reporting long-term outcomes of hypospadias patients after puberty who received pre-operative hormonal therapy. Validated instruments were used as much as possible. Shortcomings of this study were the 50% response rate, the retrospective design, and the lack of objective inclusion criteria reported to indicate pre-operative testosterone therapy.

Conclusion(s): This study suggested that the long-term results of patients receiving pre-operative testosterone treatment, who often had more challenging hypospadias, were similar to those who did not. However, a randomised controlled study is needed to confirm these results. [Table presented]

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PMID

29174377 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=29174377>]

Status

Embase

Institution

(Rynja, de Jong, Bosch, de Kort) Department of Urology, University Medical Center Utrecht, Utrecht, Netherlands (de Jong) Department of Pediatric Urology, University Children's Hospital UMC Utrecht and Academic Medical Center Amsterdam, Utrecht, Netherlands

Publisher

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Year of Publication

2018

320.

Posterior hypospadias: Evaluation of a paradigm shift from single to staged repair.

Badawy H., Orabi S., Hanno A., Abdelhamid H.

Embase

Journal of Pediatric Urology. 14 (1) (pp 28.e1-28.e8), 2018. Date of Publication: February 2018.

[Article]

AN: 618065686

Introduction: Repair of posterior hypospadias is a current dilemma. Single versus staged repair is the main question to answer. The answer is not easily reached due to lack of comparative studies. Hence such studies are not available, the reports of a change from one approach to the other one are important to point out the results of each procedure in the same hands and in the same center. Herein, we report our results of the repair of posterior hypospadias shifting from single stage to staged repair.

Patients and Methods: 65 children were operated in a single Centre in the period from 2011-2016 using single stage repair by dorsal island flap in the first 40 children and then a shift to staged repair involved 25 children repaired using Bracka procedure, children are evaluated for the outcome and for the development of complications during the period of follow up.

Result(s): The mean age of children operated using single stage technique was 2.8 years (0.83-12.0), Onlay repair was performed in 29 cases (72.5%), while a tube was performed in 11 cases (27.5%). The success rate was 55% with 45% complication rate, in the form of infection in (2.5%), partial dehiscence in (10%), urethrocutaneous fistula in (15%), meatal stenosis in (12.5%),

urethral diverticulum in (5%). 25 children were operated using staged repair according to Bracka using inner preputial graft in fresh cases and buccal graft in redo cases. Mean age of 4.5 years (7 months-18 years), 15 primary cases and 10 redo cases, 12 penoscrotal, 11 scrotal and 2 perineal cases, preputial graft in 17, buccal graft in 8, 25 children completed their second stage, tunica vaginalis cover was used in 23 children, localized penile skin dartos was used in 2 children, the overall success after second stage was 80%, complications were in the form of 4 fistulas (16%), hematoma and complete disruption in a redo case (4%). There is a significant statistical difference in the incidence of complications between both groups in favor of lower complication rate (20%) in the staged group versus the single stage group (45%) with a $P = 0.0419$.

Conclusion(s): Staged repair considerably improves complication rate of posterior hypospadias reconstruction compared to single stage repair using pedicled island flap. More follow up and continuous reporting of honest complication rate is needed to improve the outcome of a complex pathology and to help the choice of the best procedure.

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Status

Embase

Institution

(Badawy, Orabi, Hanno) Pediatric Urology, University of Alexandria, Alexandria, Egypt

(Abdelhamid) Pediatric Urology, Alexandria Insurance Children's Hospital, Alexandria, Egypt

Publisher

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Year of Publication

2018

321.

Incidence and diagnoses of disorders of sex development in proximal hypospadias.

Wong Y.S., Tam Y.H., Pang K.K.Y., Yau H.C.

Embase

Journal of Pediatric Surgery. 53 (12) (pp 2498-2501), 2018. Date of Publication: December 2018.

[Article]

AN: 2001103715

Background: Evidence-based guidelines on evaluation of boys with proximal hypospadias for the possibility of a disorder of sex development (DSD) have yet to be developed. We aimed to investigate the incidence and diagnoses of DSD in patients with proximal hypospadias.

Method(s): We retrospectively reviewed the records of consecutive boys who underwent proximal hypospadias repairs from 2006 to Sept 2017. Data collected included scrotal anomaly, testes position/palpability, micropenis, DSD investigations, and surgical techniques.

Result(s): 165 patients were eligible for the study. 14 (8.5%) were diagnosed to have DSD. The diagnoses were 46,XX testicular DSD [n = 1], 46,XY DSD [n = 7; partial gonadal dysgenesis (PGD) = 3; 5alpha-reductase type 2 deficiency = 3; 17alpha-hydroxylase deficiency = 1], Sex Chromosome DSD [n = 6; 45,X/46,XY PGD = 4; Klinefelter = 2]. 3/7 (43%) patients with PGD had gonadal germ cell neoplasms. Of the DSD patients, 6/14 (43%), 11/14 (79%) and 11/14 (79%) had undescended/impalpable testes, micropenis and penoscrotal transposition/bifid scrotum, respectively, significantly higher prevalence rates than those without DSD diagnosis (p-values < 0.05). 10/14 (71.4%) DSD patients underwent 2-stage repair compared with 57/151 (37.7%) of others without DSD diagnosis (p = 0.01).

Conclusion(s): Patients presenting with proximal hypospadias and one or more of the coexisting anomalies of micropenis, undescended/impalpable testes, and penoscrotal transposition/bifid scrotum should warrant DSD evaluation. Presence of bilaterally descended testes in scrotum does not preclude the possibility of DSD.

Level of Evidence: IV.

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Status

Embase

Institution

(Wong, Tam, Pang) Division of Paediatric Surgery and Paediatric Urology, Department of Surgery, Prince of Wales Hospital, The Chinese University of Hong Kong, Hong Kong (Yau) Department of Paediatrics, Prince of Wales Hospital, The Chinese University of Hong Kong, Hong Kong

Publisher

W.B. Saunders

Year of Publication

2018

322.

Endocrine disruptors and testicular function.

Lymperi S., Giwercman A.

Embase

Metabolism: Clinical and Experimental. 86 (pp 79-90), 2018. Date of Publication: September 2018.

[Article]

AN: 2000719871

Despite concerns of the scientific community regarding the adverse effects of human exposure to exogenous man-made chemical substances or mixtures that interfere with normal hormonal balance, the so called "endocrine disruptors (EDs)", their production has been increased during the last few decades. EDs' extensive use has been implicated in the increasing incidence of male reproductive disorders including poor semen quality, testicular malignancies and congenital developmental defects such as hypospadias and cryptorchidism. Several animal studies have demonstrated that exposure to EDs during fetal, neonatal and adult life has deleterious consequences on male reproductive system; however, the evidence on humans remains ambiguous. The complexity of their mode of action, the differential effect according to the developmental stage that exposure occurs, the latency from exposure and the influence of the genetic background in the manifestation of their toxic effects are all responsible factors for the contradictory outcomes. Furthermore, the heterogeneity in the published human studies has hampered agreement in the field. Interventional studies to establish causality would be desirable, but unfortunately the nature of the field excludes this possibility. Therefore, future studies based on standardized guidelines are necessary, in order to estimate human health risks and implement policies to limit public exposure.

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Status

Embase

Institution

(Lymperi) Unit of Reproductive Endocrinology, First Department of Obstetrics and Gynecology, Aristotle University of Thessaloniki, Thessaloniki, Greece (Giwercman) Department of Translational Medicine, Lund University, Malmo, Sweden

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Year of Publication
2018

323.

Anogenital distance is associated with genital measures and seminal parameters but not anthropometrics in a large cohort of young adult men.

Foresta C., Valente U., Di Nisio A., Cacco N., Magagna S., Cosci I., Presciutti A., Garolla A.
Embase

Human Reproduction. 33 (9) (pp 1628-1635), 2018. Date of Publication: 2018.

[Article]

AN: 624416397

STUDY QUESTION: Is the anogenital distance (AGD) correlated to anthropometric, genital and sperm parameters in young adult men? SUMMARY ANSWER: We observed that reduced AGD is strongly associated with altered semen parameters and reduced testicular volume. WHAT IS KNOWN ALREADY: Abnormalities in the foetal development of the testis have been suggested as causative of common male reproductive disorders, such as cryptorchidism, hypospadias, reduced semen quality and testicular germ cell tumour, collectively defined as 'testicular dysgenesis syndrome'. In human epidemiological studies, alterations in AGD have been frequently associated with clinically relevant outcomes of reproductive health, suggesting AGD as a marker of foetal testicular development. STUDY DESIGN, SIZE, DURATION: This study was performed within the annual screening protocol to evaluate male reproductive health in the high schools of Padua and surroundings (Veneto Region, the North-East of Italy). Here we report the findings of 794 subjects who completed the study protocol between October 2016 and May 2017. PARTICIPANTS/MATERIALS, SETTING, METHODS: We evaluated 794 students aged 18-19 years recording the following parameters: height, weight, BMI, waist circumference, arm span, pubis-to-floor and crown-to-pubis length, penile length and circumference, testicular volumes, semen parameters and AGD (measured from the posterior base of the scrotum to the centre of the anus). MAIN RESULTS AND THE ROLE OF CHANCE: Of the subjects, 49% had an abnormal arm span-height difference (>3 cm) and 63.4% had an altered ratio of crown-to-pubis/pubis-to-floor length (≤ 0.92). The rate of subjects with reduced testicular volume was

23%. Median sperm concentration was $51.0 \times 10^6/\text{ml}$ and total sperm count was 122.5×10^6 . AGD showed a direct positive relation with testicular volume and penile length and circumference ($R = 0.265, 0.176$ and 0.095 , respectively, all $P < 0.05$). No significant relation was observed between AGD and anthropometric parameters. Sperm concentration, total sperm count, progressive motility and normal morphology showed a significant and positive correlation with AGD ($R = 0.205, 0.210, 0.216$ and 0.117 , respectively, all $P < 0.05$). LIMITATIONS, REASONS FOR CAUTION: Our cohort of young adults is not representative of the general population. Hormonal evaluation was missing. WIDER IMPLICATIONS OF THE FINDINGS: Our findings show that AGD is associated with testicular volumes, penile measures and seminal parameters in young adult men. Because AGD is hormonally determined during foetal life, the reported high incidence of reduced semen quality and reduced testicular volume could be related to a reduced androgenic exposure in utero. AGD could represent a simple and useful method to evaluate testicular and penile development in adult men. STUDY FUNDING/COMPETING INTEREST(S): The authors have no potential conflict of interest to declare. No external funding was obtained for this study. TRIAL REGISTRATION NUMBER: N/A.

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Status

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Institution

(Foresta, Valente, Di Nisio, Cacco, Magagna, Cosci, Presciutti, Garolla) Department of Medicine, Operative Unit of Andrology and Medicine of Human Reproduction, University of Padova, Via Giustiniani, 2, Padova 35128, Italy

Publisher

Oxford University Press

Year of Publication

2018

324.

Genetic Polymorphism in the RYR1 C6487T is Associated with Severity of Hypospadias in Chinese Han Children.

Zhang H., Zhang Z., Jia L., Ji W., Li H.

Embase

BioMed Research International. 2018 (no pagination), 2018. Article Number: 7397839. Date of Publication: 2018.

[Article]

AN: 623822957

Objective. Hypospadias is a common congenital malformation of the male external genitalia. Most cases have an unknown etiology, which is probably a mix of monogenic and multifactorial forms, implicating both genetic and environmental factors. Ryanodine receptor 1 (RYR1) mutations are a common cause of congenital diseases associated with both dominant and recessive inheritance in humans. Herein, we evaluated the correlations of RYR1 C6487T polymorphism with the risk and severity of hypospadias. **Methods.** 263 congenital hypospadias children and 312 healthy children were recruited. The polymorphism of RYR1 C6487T in the peripheral blood was detected by polymerase chain reaction-restriction fragment length polymorphism, and different genotypes and allelic genes were analyzed to explore their associations with the risk of congenital hypospadias. **Results.** The distribution frequencies of CC/CT/TT genotypes and two alleles (C and T) at RYR1 C6487T showed significant differences between the case and control groups ($P < 0.05$). The frequency of C allele in the case and control groups was 46.95% and 54.94%, respectively, and of T allele was 53.05% and 45.06% ($P < 0.05$). In addition, the distribution frequency of CC/CT/TT genotypes exhibited significant difference between patients with mild hypospadias and those with moderate or severe hypospadias (all $P > 0.05$), suggesting that RYR1 C6487T polymorphism is correlated with the severity of congenital hypospadias ($X^2 = 13.722$, $P = 0.001$). **Conclusion.** Our study demonstrated that RYR1 C6487T polymorphism might be associated with an increased risk of congenital hypospadias in Chinese Han children. Our findings highlight the heterogeneous nature of hypospadias genetic susceptibility.

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Status

Embase

Author NameID

Li, Hai; ORCID: <http://orcid.org/0000-0002-0485-4981> Jia, Linpei; ORCID: <http://orcid.org/0000-0003-0348-7814>

Institution

(Zhang) Department of Gastrointestinal Surgery, China-Japan Union Hospital of Jilin University, Changchun 130033, China (Zhang, Li) Department of Urology, China-Japan Union Hospital of Jilin University, Changchun 130033, China

(Jia) Department of Nephrology, Xuanwu Hospital of Capital Medical University, Beijing 100053, China

(Ji) Department of Vascular Surgery, Jilin Provincial People's Hospital, Changchun 130000, China

Publisher

Hindawi Limited (410 Park Avenue, 15th Floor, 287 pmb, New York NY 10022, United States)

Year of Publication

2018

325.

Congenital genital abnormalities detected during routine circumcision at a South African institution: A retrospective record review.

Spencer K., Mokhele I., Firnhaber C.

Embase

African Health Sciences. 18 (2) (pp 352-358), 2018. Date of Publication: June 2018.

[Article]

AN: 622793468

Background: Due to the reduction in HIV transmission through male medical circumcisions (MMC), numerous clinics throughout South Africa offer a voluntary free service to boys from the age of ten years and above. An examination prior to the procedure may detect congenital abnormalities missed after birth.

Objective(s): The aim of this study was to measure the incidence of these abnormalities, determine the demographic and clinical characteristics of this group and determine what referral systems, interventions, and follow-up is available to them.

Method(s): The study was a descriptive, observational, retrospective analysis of de-identified medical records at a routine MMC service at a Johannesburg clinic in 2015. The participants were male patients between the ages of 10 - 49.

Result(s): Out of 1548 participants, 91.0% (n=1409) had a normal genital examination while 3.7% (n=57) had an abnormal examination and 5.1% (n=79) had no examination recorded. Thirty five congenital anomalies were detected and only 2 patients (diagnosed with hypospadias) were seen at the urology out-patient's department.

Conclusion(s): The incidence of congenital genital abnormalities of males presenting for routine circumcision is low. Despite the low incidence the effect on fertility, sexuality, ability to urinate and on psychological wellbeing is significant. Referral services to the urology department should be restructured to improve all outcomes.

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Status

Embase

Institution

(Spencer) University of the Witwatersrand, Urology, South Africa (Mokhele) Right to Care, South Africa

(Firnhaber) University of the Witwatersrand, Clinical HIV Research unit, South Africa

Publisher

Makerere University, Medical School (E-mail: pic@infocom.co.ug)

Year of Publication

2018

326.

Maternal Diabetes Mellitus and Genital Anomalies in Male Offspring: A Nationwide Cohort Study in 2 Nordic Countries.

Arendt L.H., Lindhard M.S., Henriksen T.B., Olsen J., Cnattingius S., Petersson G., Parner E.T., Ramlau-Hansen C.H.

Embase

Epidemiology. 29 (2) (pp 280-289), 2018. Date of Publication: 01 Mar 2018.

[Article]

AN: 622578379

Background: Pre-existing diabetes has been associated with an increased risk of congenital malformations overall, but studies on genital anomalies in boys are conflicting and possible causal mechanisms are not well understood. Previous studies have mainly assessed pregestational and gestational diabetes in combination. Yet considering the vulnerable time

windows for the genital anomalies, associations could well differ between types of diabetes and between the 2 genital anomalies and we therefore aimed to study this further.

Method(s): A population-based cohort study of 2,416,246 singleton live-born boys from Denmark (1978-2012) and Sweden (1987-2012) was carried out using Danish and Swedish register-based data. Using Cox regression models, we estimated hazard ratios for hypospadias and cryptorchidism according to maternal diabetes. We considered type and severity of diabetes, as well as timing of diagnosis in relation to birth.

Result(s): Pregestational type 1 diabetes was associated with a higher risk of both genital anomalies. The highest risks were seen for boys of mothers with diabetic complications (hazard ratio for hypospadias = 2.33 [95% confidence interval, 1.48, 3.66] and hazard ratio for cryptorchidism = 1.92 [95% confidence interval, 1.39, 2.65]). Gestational diabetes was associated with slightly increased risks of both genital anomalies.

Conclusion(s): These results are consistent with the hypothesis that poor glycemic control may interfere with fetal genital development in the critical early period of organogenesis. Given the widespread and increasing occurrence of diabetes, these results are of public health importance.

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Embase

Institution

(Arendt, Ramlau-Hansen) Department of Public Health, Section for Epidemiology, Aarhus University, Bartholins Alle 2, Aarhus C 8000, Denmark (Arendt, Lindhard, Henriksen) Perinatal Epidemiology Research Unit, Department of Paediatric and Adolescent Medicine, Aarhus University Hospital, Aarhus, Denmark

(Olsen) Department of Clinical Epidemiology, Aarhus University Hospital, Aarhus, Denmark

(Cnattingius, Petersson) Clinical Epidemiology Unit, Department of Medicine, Karolinska University Hospital, Karolinska Institute, Stockholm, Sweden

(Parner) Department of Public Health, Section for Biostatistics, Aarhus University, Aarhus, Denmark

Publisher

Lippincott Williams and Wilkins (E-mail: kathiest.clai@apta.org)

Year of Publication

2018

327.

Prevalence and risk factors of testicular microlithiasis in patients with hypospadias: A retrospective study.

Nakamura M., Moriya K., Nishimura Y., Nishida M., Kudo Y., Kanno Y., Kitta T., Kon M., Shinohara N.

Embase

BMC Pediatrics. 18 (1) (no pagination), 2018. Article Number: 179. Date of Publication: 29 May 2018.

[Article]

AN: 622334304

Background: It has been described that the incidence of testicular microlithiasis is high in several congenital disorders which may be associated with testicular impairment and infertility. Several reports have shown that a prepubertal or pubertal hormonal abnormality in the pituitary-gonadal axis was identified in some patients with hypospadias that is one of the most common disorders of sex development. However, exact prevalence or risk factors of testicular microlithiasis in patients with hypospadias have not reported so far. In the present study, to clarify the prevalence and risk factors of testicular microlithiasis in patients with hypospadias, a retrospective chart review was performed.

Method(s): Children with hypospadias who underwent testicular ultrasonography between January 2010 and April 2016 were enrolled in the present study. Severity of hypospadias was divided into mild and severe. The prevalence and risk factors of testicular microlithiasis or classic testicular microlithiasis were examined.

Result(s): Of 121 children, mild and severe hypospadias were identified in 66 and 55, respectively. Sixteen children had undescended testis. Median age at ultrasonography evaluation was 1.7 years old. Testicular microlithiasis and classic testicular microlithiasis were documented in 17 children (14.0%) and 8 (6.6%), respectively. Logistic regression analysis revealed that presence of undescended testis was only a significant factor for testicular microlithiasis and classic testicular microlithiasis. The prevalence of testicular microlithiasis or classic testicular microlithiasis was significantly higher in children with undescended testis compared to those without undescended testis (testicular microlithiasis; 43.8% versus 9.5% ($p=0.002$), classic testicular microlithiasis; 37.5% versus 1.9% ($p<0.001$)).

Conclusion(s): The current study demonstrated that the presence of undescended testis was only a significant risk factor for testicular microlithiasis or classic testicular microlithiasis in patients with hypospadias. As co-existing undescended testis has been reported as a risk factor for

testicular dysfunction among patients with hypospadias, the current findings suggest that testicular microlithiasis in children with hypospadias may be associated with impaired testicular function. Conversely, patients with isolated HS seem to have lower risks for testicular impairment. Further investigation with longer follow-up will be needed to clarify these findings.

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Status

Embase

Institution

(Nakamura, Moriya, Nishimura, Kanno, Kitta, Kon, Shinohara) Hokkaido University Graduate School of Medicine, Department of Renal and Genitourinary Surgery, North-15, West-7, Kita-Ku, Sapporo 060-8638, Japan (Nishida, Kudo) Hokkaido University Hospital, Diagnostic Center for Sonography, Sapporo, Japan

(Nishida, Kudo) Hokkaido University Hospital, Division of Laboratory and Transfusion Medicine, Sapporo, Japan

Publisher

BioMed Central Ltd. (E-mail: info@biomedcentral.com)

Year of Publication

2018

328.

Width proportion of the urethral plate to the glans can serve as an appraisal index of the urethral plate in hypospadias repair.

Ru W., Shen J., Tang D., Xu S., Wu D., Tao C., Chen G., Gao L., Wang X., Shen Y.

Embase

International Journal of Urology. 25 (7) (pp 649-653), 2018. Date of Publication: July 2018.

[Article]

AN: 621955366

Objectives: To find a new appropriate evaluation for urethral plate quality in hypospadias repair, with particular interest in the width proportion of the urethral plate to the glans, serving as an appraisal index.

Method(s): Data were prospectively collected from prepubertal boys who underwent primary tubularized incised plate hypospadias repair between January 2014 and April 2016 in one center. Intrinsic parameters of the penis (meatal location, glans width, urethral plate width and curvature degree) were measured during the operation. Urethroplasty complications were recorded during follow up. The correlation between width proportion of the urethral plate to the glans and urethroplasty complications was analyzed.

Result(s): Primary tubularized incised plate repair was carried out in 442 patients (mean age 2.8 years, range 0.5-12 years). At mean follow up of 26 months (range 12-38 months), urethroplasty complications occurred in 59 (13.3%) patients. The width proportion of the urethral plate to the glans was weakly correlated to both the glans width and meatal location. The width proportion of the urethral plate to the glans ranged from 0.18 to 0.73, with a mean of 0.39. The cut-off value of width proportion of the urethral plate to the glans was determined to be 0.36 by the receiver operating characteristic curve. Urethroplasty complications occurred in 17 out of 254 patients (6.7%) with width proportion of the urethral plate to the glans >0.36 , and 42 out of 188 patients (22.3%) with width proportion of the urethral plate to the glans ≤ 0.36 . The width proportion of the urethral plate to the glans ≤ 0.36 showed an increased odds of 4.819-fold (95% confidence interval 2.548-9.112, $P < 0.001$) risk of urethroplasty complications compared with width proportion of the urethral plate to the glans >0.36 . Midshaft and proximal meatal location also increased the risk of urethroplasty complications.

Conclusion(s): The present study highlights the value of the width proportion of the urethral plate to the glans for objectivity and accuracy in urethral plate evaluation, which in turn serves as an independent factor influencing outcomes in tubularized incised plate repair.

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Embase

Institution

(Ru, Tang, Xu, Wu, Tao, Chen, Gao, Wang, Shen) Zhejiang University School of Medicine Children's Hospital, Hangzhou, China (Shen) Amcare Women's and Children's Hospital, Hangzhou, China

Publisher

Blackwell Publishing

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2018

329.

Periconception Exposure to Air Pollution and Risk of Congenital Malformations.

Ren S., Haynes E., Hall E., Hossain M., Chen A., Muglia L., Lu L., DeFranco E.

Embase

Journal of Pediatrics. 193 (pp 76-84.e6), 2018. Date of Publication: February 2018.

[Article]

AN: 619622722

Objective: To evaluate the association between increased exposure to airborne fine particulate matter (PM2.5) during the periconception period with risk of congenital anomalies. Study design: Using birth certificate data from the Ohio Department of Health (2006-2010) and PM2.5 data from the US Environmental Protection Agency's 57 monitoring stations located throughout Ohio, the geographic coordinates of the mother's residence for each birth were linked to the nearest PM2.5 monitoring station and monthly exposure averages were calculated. The association between congenital anomalies and increased PM2.5 levels was estimated, with adjustment for coexistent risk factors.

Result(s): After adjustment for coexisting risk factors, exposure to increased levels of PM2.5 in the air during the periconception period was modestly associated with risk of congenital anomalies. Compared with other periconception exposure windows, increased exposure during the 1 month before conception was associated with the highest risk increase at lesser distances from monitoring stations. The strongest influences of PM2.5 on individual malformations were found with abdominal wall defects and hypospadias, especially during the 1-month preconception.

Conclusion(s): Increased exposure to PM2.5 in the periconception period is associated with some modest risk increases for congenital malformations. The most susceptible time of exposure appears to be the 1 month before and after conception. Although the increased risk with PM2.5 exposure is modest, the potential impact on a population basis is noteworthy because all pregnant women have some degree of exposure.

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Status

Embase

Author NameID

DeFranco, Emily; ORCID: <http://orcid.org/0000-0002-0164-5703>

Institution

(Ren) Department of Mathematics, University of Cincinnati, Cincinnati, OH, United States (Ren, Lu) Division of Biomedical Informatics, Cincinnati Children's Hospital Medical Center, Cincinnati, OH, United States

(Haynes, Chen) Department of Environmental Health, University of Cincinnati College of Medicine, Cincinnati, OH, United States

(Hall) Perinatal Institute, Cincinnati Children's Hospital Medical Center, Cincinnati, OH, United States

(Hossain, Muglia, DeFranco) Division of Biostatistics and Epidemiology, Cincinnati Children's Hospital Medical Center, Cincinnati, OH, United States

(Muglia, DeFranco) Division of Maternal-Fetal Medicine, Department of Obstetrics and Gynecology, University of Cincinnati College of Medicine, Cincinnati, OH, United States

Publisher

Mosby Inc. (E-mail: customerservice@mosby.com)

Year of Publication

2018

330.

Anatomic findings associated with epispadias in boys: Implications for surgical management and urinary continence.

Cendron M., Cho P.S., Pennison M., Rosoklija I., Diamond D.A., Borer J.G.

Embase

Journal of Pediatric Urology. 14 (1) (pp 42-46), 2018. Date of Publication: February 2018.

[Article]

AN: 619456684

Introduction: Pre-operative physical examination of male epispadias allows for classification of epispadias level as glanular (GE), penile (PE) or penopubic (PPE), and for delineation of anatomic anomalies. The incidence of associated extragenital abnormalities, such as vesicoureteral reflux (VUR), bladder neck (BN) abnormality and abnormal pubic diastasis (PD), and their impact on urinary continence has not yet been systematically studied.

Objective(s): The goal of this study was to evaluate whether the more proximal level of epispadias correlated with associated extragenital anatomic anomalies seen on initial imaging or endoscopic evaluation, and whether these pre-operative findings contributed to subsequent surgical management and impacted on achieving urinary continence. It was hypothesized that the more severe forms of epispadias may be associated with a higher frequency of associated anomalies. **Study design:** The study was an IRB-approved, retrospective case study of all male patients treated initially for isolated epispadias at the current institution between 1994 and 2011. Data collection was achieved by chart and radiology review evaluating PD, BN appearance, presence of VUR, surgical treatment, and urinary continence.

Result(s): A total of 26 patients were identified and divided into three groups based on appearance at physical examination: four glanular (GE), eight penile (PE), and 14 penopubic (PPE); 17 patients had an abnormal BN. Reflux was noted in nine of 20 patients who had a voiding cystourethrogram (VCUG), two of which had an episode of pyelonephritis. Of the 22 patients past the age of toilet training, 17 were continent (64% (9/14) penopubic, 63% (5/8) penile, and 75% (3/4) glanular).

Discussion(s): Anatomic classification for male epispadias did not provide sufficient information regarding extragenital findings. This study provided new information regarding PD, BN appearance, presence of reflux, and ultimate urinary continence. Pubic diastasis and BN abnormalities were more frequently seen in more severe forms of epispadias, whereas VUR seemed more prevalent in less severe forms. A template for pre-operative evaluation was outlined. Limitations of the study were its retrospective design and relatively small cohort of patients, which reflected the rarity of the condition.

Conclusion(s): Based on the information generated, additional anatomic information was generated regarding boys with epispadias. This information will help guide the evaluation and the management of these patients in the future.

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Status

Embase

Institution

(Cendron, Cho, Pennison, Rosoklija, Diamond, Borer) Department of Urology, Boston Children's Hospital, Boston, MA, United States

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331.

Phthalate exposure and male reproductive outcomes: A systematic review of the human epidemiological evidence.

Radke E.G., Braun J.M., Meeker J.D., Cooper G.S.

Embase

Environment International. Part 1. 121 (pp 764-793), 2018. Date of Publication: December 2018.

[Review]

AN: 2001188162

Objective: We performed a systematic review of the epidemiology literature to identify the male reproductive effects associated with phthalate exposure. Data sources and study eligibility criteria: Six phthalates were included in the review: di(2-ethylhexyl) phthalate (DEHP), diisononyl phthalate (DINP), dibutyl phthalate (DBP), diisobutyl phthalate (DIBP), butyl benzyl phthalate (BBP), and diethyl phthalate (DEP). The initial literature search (of PubMed, Web of Science, and Toxline) included all studies of male reproductive effects in humans, and outcomes were selected for full systematic review based on data availability. Study evaluation and synthesis methods: For each outcome, studies were evaluated using criteria defined a priori for risk of bias and sensitivity by two reviewers using a domain-based approach. Evidence was synthesized by outcome and phthalate and strength of evidence was summarized using a structured framework.

Result(s): The primary outcomes reviewed here are (number of included/excluded studies in parentheses): anogenital distance (6/1), semen parameters (15/9), time to pregnancy (3/5), testosterone (13/8), timing of pubertal development (5/15), and hypospadias/cryptorchidism (4/10). Looking at the overall hazard, there was robust evidence of an association between DEHP and DBP exposure and male reproductive outcomes; this was based primarily on studies of anogenital distance, semen parameters, and testosterone for DEHP and semen parameters and time to pregnancy for DBP. There was moderate evidence of an association between DINP and BBP exposure and male reproductive outcomes based on testosterone and semen parameters for DINP and semen parameters and time to pregnancy for BBP. DIBP and DEP were considered to have slight evidence of an association. For DIBP, the less conclusive evidence was attributed to a more limited literature base (i.e., fewer studies) and lower exposure levels in the population, decreasing the ability to observe an effect. For DEP, the findings were consistent with experimental animal data that suggest DEP does not have as strong an anti-androgenic effect

as other phthalates. Conclusions and implications of key findings: Overall, despite some inconsistencies across phthalates in the specific outcomes associated with exposure, these results support that phthalate exposure at levels seen in human populations may have male reproductive effects, particularly DEHP and DBP. The relative strength of the evidence reflects differing levels of toxicity as well as differences in the range of exposures studied and the number of available studies. The views expressed are those of the authors and do not necessarily represent the views or policies of the U.S. EPA.

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Status

Embase

Institution

(Radke, Cooper) U.S. Environmental Protection Agency, National Center for Environmental Assessment, United States (Braun) Brown University, School of Public Health, United States (Meeker) University of Michigan, School of Public Health, United States (Cooper) The Innocence Project, United States

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332.

Improving surgical training by identifying the most common feedback trainees receive for index cases.

Maizels M., Liu D., Yerkes E., Meade P., Biondi M., Sutherland R.

Embase

Journal of Pediatric Urology. 14 (6) (pp 577-583), 2018. Date of Publication: December 2018.

[Article]

AN: 2001137358

Background: To make surgical training more effective, a proven method is needed to provide feedback to residents on their surgeries. Residency programs may make up for limited training time in the operating room by improving feedback that trainees receive about cases.

Objective(s): The goals of this study were (1) to determine if an online tool to communicate feedback for attendings and trainees shows face validity and (2) to use an online tool to identify the most common feedback trainees receive after performing orchiopexy and hypospadias repair by survey.

Method(s): In 2016, determining whether an online tool to provide pediatric urology trainees feedback after surgery shows face validity begun. The tool was launched at the authors' institutions. Then, attendings, fellows, and postgraduate year 4-5 trainees of 65 resident training programs were surveyed for their observations on preparing for and performing orchiopexy and hypospadias repair using the study tool to identify common feedback.

Result(s): The results of using the tool to provide feedback shows face validity are as follows: feedback was exchanged between attendings and trainees on orchiopexy (n = 28) and hypospadias (n = 22). Anecdotally, the tool was easy to use. The results of using the tool to identify the most common feedback trainees receive by survey are as follows: from a pool of 65 institutions, 37 attendings and 28 trainees were enrolled who made 219 observations. Most trainees prepare using undocumented online resources (17/28, 67%) instead of speaking with their attendings or cotrainees (11/28, 33%). For orchiopexy, most respondents reported that trainees need to improve skills for hernia ligation (observations: attending = 28/45, 62%; and trainee = 17/26, 65%) and strategies for hernia exposure (observations: attending = 17/27, 62%; and trainee = 7/12, 58%). For hypospadias, most respondents reported that trainees need to improve skills for neourethroplasty (observations: attending = 31/53, 58%; and trainee = 10/16, 62%) and strategies for repair choice (observations: attending = 15/22, 68%; and trainee = 12/18, 67%) (chi-squared, all P = NS).

Discussion(s): It was shown that both trainees and attendings agree on the areas of surgical strategy and execution which require improvement. With this study, it is also shown that the online feedback tool developed shows face validity in allowing attendings and trainees to communicate before and after surgery.

Conclusion(s): The most common feedback pediatric urology trainees receive for routine pediatric urology surgery is identified. Online tools that emphasize remediations to address a trainee's specific feedback needs are to be built, so that they will be able to improve their skills at their next case.[Figure presented]

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Institution

(Maizels, Liu, Yerkes, Meade) Ann and Robert H. Lurie Children's Hospital of Chicago, Chicago, IL, United States (Biondi, Sutherland) UNC School of Medicine, Chapel Hill, NC, United States

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333.

Secular trends in the incidence and timing of surgical intervention for congenital undescended testis and surgically treated hypospadias in Ontario, Canada between 1997 and 2007.

Dave S., Liu K., Garg A.X., Shariff S.Z.

Embase

Journal of Pediatric Urology. 14 (6) (pp 552.e1-552.e7), 2018. Date of Publication: December 2018.

[Article]

AN: 2000986583

Introduction: Recent studies have suggested contradictory trends in the incidence of undescended testis (UDT) and hypospadias (HYP), partly because of methodological issues and ascertainment bias. The recently described association of "testicular dysgenesis syndrome" links concomitant UDT and HYP, with decreasing sperm counts and testicular cancer. Current guidelines suggest that orchidopexy for UDT should be performed by 18 months of age.

Objective(s): We conducted a retrospective population-based cohort study to estimate the incidence of UDT, HYP, and concomitant UDT and HYP in Ontario, based on a surgical procedure performed in the 5 years after birth. We hypothesized that the incidence of UDT and HYP are stable in the province of Ontario, Canada, over an 11-year time period. Study design: Linked administrative databases held at the Institute of Clinical Evaluative Sciences (ICES) in the province of Ontario, were used to identify all live male newborns between 1997 and 2007.

Incidence rates of UDT, HYP and concomitant UDT and HYP were calculated by identifying a surgical procedure for these anomalies, within 5 years of birth. Incidence trends were analyzed

using the Cochrane Armitage test for trend. Age at surgery for surgical intervention for an orchidopexy or HYP repair was determined.

Result(s): The incidence of UDT, defined by an orchidopexy within 5 years of birth, has remained stable in Ontario, Canada (8.2/1000 male live births, p-value for trend 0.9, 95% CI 8.0-8.4). The incidence of hypospadias has similarly remained stable (3.8/1000 male live births, p-value for trend 0.8, 95% CI 3.7-3.9). The incidence of concomitant UDT and HYP repair showed a significant increase over the 11-year period (0.2/1000 male live births, p-value for trend 0.03, 95% CI 0.2-0.3). The median age at orchidopexy (23 months, IQR 16-34 months) was beyond guideline recommendations, with earlier orchidopexy in recent years. The median age at hypospadias repair was 17 months (IQR 12-26 months).

Discussion(s): The variable rates of incidence for UDT and HYP can be explained by variations in study methodology and differing data sources utilized. The current study uses a surgical procedure to minimize information bias to correctly identify index cases of UDT and HYP.

Conclusion(s): The incidence of undescended testis and hypospadias, over 5 years after birth, has remained stable in the province of Ontario between 1997 and 2007 (Summary Table).

Concomitant UDT and HYP incidence showed a significant increase over this time period. Most boys in Ontario, Canada, undergo orchidopexy beyond 18 months of age. [Table presented]

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Status

Embase

Author NameID

Dave, Sumit; ORCID: <http://orcid.org/0000-0003-1462-647X>

Institution

(Dave) Department of Surgery and Paediatrics, Division of Urology, Western University, London, Ontario, Canada (Dave, Liu, Garg, Shariff) Institute for Clinical Evaluative Sciences, London, Ontario, Canada

(Garg, Shariff) Arthur Labatt Family School of Nursing, Western University, London, Ontario, Canada

(Garg, Shariff) Lawson Health Research Institute, London, Ontario, Canada

(Garg, Shariff) Department of Medicine, Division of Nephrology, Western University, London, Ontario, Canada

(Garg, Shariff) Department of Epidemiology and Biostatistics, Western University, London, Ontario, Canada

Publisher

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2018

334.

Combined Bladder Neck Reconstruction and Continent Stoma Creation as a Suitable Alternative for Continence in Bladder Exstrophy: A Preliminary Report.

Kasprenski M., Benz K., Jayman J., Lue K., Maruf M., Baumgartner T., Gearhart J.P.

Embase

Urology. 119 (pp 133-136), 2018. Date of Publication: September 2018.

[Article]

AN: 2000870805

Objective: To explore the use of concomitant bladder neck reconstruction (BNR) and creation of a continent stoma (CS) in patients who are not quite eligible for BNR but still strongly desire volitional voiding.

Method(s): The authors retrospectively reviewed an institutional database of patients with exstrophy-epispadias complex who underwent BNR-CS between 2000 and 2015. Indications for a BNR-CS, perioperative outcomes, and continence status were evaluated. Method of voiding and continence status were analyzed for patients with greater than 6 months of follow-up after the BNR-CS.

Result(s): A total of 24 patients with exstrophy-epispadias complex (15 male and 9 female) underwent BNR-CS at a median age of 8.9 years (range 5.4-17.4). This included 18 patients with classic bladder exstrophy, 5 with epispadias, and 1 with a cloacal exstrophy variant. There were 5 surgical complications (20.1%) following the BNR-CS, including 3 febrile urinary tract infections, 1 superficial wound infection, and 1 urethrocutaneous fistula. The median follow-up time from the time of BNR-CS was 1.1 years (range 0.1-14.1). Seventeen of 24 patients (71%) had a follow-up greater than 6 months and were evaluated for continence. Twelve patients (71%) were completely dry for intervals greater than 3 hours following BNR-CS. Five (29%) did not achieve continence with BNR-CS. Of those 5 patients, 3 (60%) underwent subsequent bladder neck transection.

Conclusion(s): Combined BNR and CS is a suitable alternative to achieve urinary continence in patients who are not ideal candidates for BNR alone. This approach can offer a select group of patients the opportunity for volitional voiding.

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Status

Embase

Author NameID

Gearhart, John P.; ORCID: <http://orcid.org/0000-0002-0177-6121>

Institution

(Kasprenski, Benz, Jayman, Lue, Maruf, Baumgartner, Gearhart) Robert D. Jeffs Division of Pediatric Urology, James Buchanan Brady Urological Institutions, Johns Hopkins Hospital, Johns Hopkins Medical Institutions, Charlotte Bloomberg Children's Hospital, Baltimore, MD, United States

Publisher

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Year of Publication

2018

335.

Early readmission and reoperation characteristics of ambulatory hypospadias repair.

Roth J.D., Whittam B.M., Carroll A.E., Szymanski K.M., Misseri R., Cain M.P., Bennett W.E.

Embase

Journal of Pediatric Urology. 14 (6) (pp 532.e1-532.e7), 2018. Date of Publication: December 2018.

[Article]

AN: 2000870097

Introduction and Background: Hypospadias is a common birth defect. It is present in about 34.2 in 10,000 live births in North America. However, few studies have evaluated the immediate reoperation and readmission rates following elective repair.

Objective(s): This study aimed to define the rates of readmission and reoperation following elective hypospadias repair, to improve pre-operative counseling on risks of the procedure. Study design: The Pediatric Health Information System (PHIS) was interrogated from 2004 to 2015 for all elective hypospadias repairs (ICD-9-CM code 58.45) performed in the ambulatory surgical

setting. The following were then determined: age at initial operation, insurance status, race, presence or absence of readmission or reoperation within 30 days, presence of repeat hypospadias repair (same ICD-9 code), and presence or absence of another urethral operation (ICD-9 CM code 58.XX, excluding 58.45). Mixed effects logistic regression were then performed with dependent variables of 30-day repeat encounter, 30-day emergency department (ED) visit, 30-day readmission, or 30-day reoperation; and independent variables of age, race, ethnicity, and insurance status.

Result(s): The study identified 45,264 hypospadias repairs during 2004-2015 performed in 43 hospitals. Within 30 days of the procedures, 2826 (6.2%) had additional encounters in the ED at the same facilities, and 546 (1.2%) had readmissions. A total of 105 (0.2%) underwent second anesthetic within the first 30 days. With regards to a 30-day repeat encounter, odds of repeat encounter were significantly increased in patients aged <5 years, Black and Asian patients, and those with Medicaid. Of the 4882 repeat encounters, 954 (19.5%) had discharge ICD-9 codes related to the penis, or to postoperative complications in general.

Discussion(s): This study described the epidemiology of clinical events occurring at the same tertiary children's hospital within the first 30 days following more than 45,000 hypospadias repairs. Limitations included a cohort generated from a single set of ICD-9 codes.

Conclusion(s): Elective hypospadias repair had a low rate of readmission (1.2%) and reoperation (0.2%) within the first 30 days. Patients aged <5 years, of non-white race, Hispanic ethnicity, and on Medicaid had significantly higher odds of 30-day repeat encounters.

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Embase

Author NameID

Roth J.D.; ORCID: <http://orcid.org/0000-0002-2990-2978>

Institution

(Roth, Whittam, Szymanski, Misseri, Cain, Bennett) Division of Pediatric Urology, Riley Hospital for Children, Indiana University School of Medicine, Indianapolis, IN, United States (Carroll, Bennett) Department of Pediatrics, Section of Pediatric and Adolescent Comparative Effectiveness Research, Indiana University School of Medicine, Indianapolis, IN, United States

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336.

Isolated hypospadias: The impact of prenatal exposure to pesticides, as determined by meconium analysis.

Haraux E., Tourneux P., Kouakam C., Stephan-Blanchard E., Boudailliez B., Leke A., Klein C., Chardon K.

Embase

Environment International. 119 (pp 20-25), 2018. Date of Publication: October 2018.

[Article]

AN: 2000869422

Although endocrine-disrupting chemicals (EDCs, including pesticides) are thought to increase the risk of hypospadias, no compounds have been formally identified in this context. Human studies may now be possible via the assessment of meconium as a marker of chronic prenatal exposure. The objective of the present study was to determine whether or not prenatal exposure to pesticides (as detected in meconium) constitutes a risk factor for isolated hypospadias. In a case-control study performed between 2011 and 2014 in northern France, male newborns with isolated hypospadias (n = 25) were matched at birth with controls (n = 58). Newborns with obvious genetic or hormonal anomalies, undescended testis, micropenis, a congenital syndrome or a family history of hypospadias were not included. Neonatal and parental data were collected. Foetal exposure was assessed by determining the meconium concentrations of the pesticides or metabolites (organophosphates, carbamates, phenylurea, and phenoxyherbicides) most commonly used in the region. Risk factors were assessed in a multivariate analysis. The pesticides most commonly detected in meconium were organophosphates (in up to 98.6% of samples, depending on the substance) and phenylurea (>85.5%). A multivariate analysis revealed an association between isolated hypospadias and the presence in meconium of the phenylurea herbicide isoproturon and of the phenoxyherbicide 2-methyl-4-chlorophenoxyacetic acid (odds ratio [95% confidence interval]: 5.94 [1.03-34.11] and 4.75 [1.20-18.76]) respectively). We conclude that prenatal exposure to these two herbicides (as assessed by meconium analysis) was correlated with the occurrence of isolated hypospadias. The results of our case-control study (i) suggest that prenatal exposure to pesticides interferes with the development of the male genitalia, and (ii) emphasize the importance of preventing pregnant women from being exposed to EDCs in general and pesticides in particular.

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Status

Embase

Institution

(Haraux, Klein) Paediatric Surgery Unit, Amiens University Hospital, Amiens F-80054, France

(Haraux, Tourneux, Kouakam, Stephan-Blanchard, Leke, Chardon) PeriTox - UMI 01, UFR de Medecine, Jules Verne University of Picardy, Amiens F-80054, France

(Tourneux) Paediatric Intensive Care Unit, Amiens University Hospital, Amiens F-80054, France

(Boudailliez) Department of Paediatrics, Amiens University Hospital, Amiens F-80054, France

(Leke) Neonatal Intensive Care Unit, Amiens University Hospital, Amiens F-80054, France

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337.

Practice variation on use of antibiotics: An international survey among pediatric urologists.

Kim J.K., Chua M.E., Ming J.M., Braga L.H., Smith G.H.H., Driver C., Koyle M.A.

Embase

Journal of Pediatric Urology. 14 (6) (pp 520-524), 2018. Date of Publication: December 2018.

[Article]

AN: 2000802607

Introduction and background: Although there is abundance in literature focusing on the use of prophylactic antibiotics for adult urological procedures, the evidence for using antibiotics following common pediatric urological procedures is limited with no specific guidelines for use.

Consequently, current practices on antibiotic usage for common interventions may be variable among practicing pediatric urologists, lacking evidence-based support.

Objective(s): The aim was to evaluate the current practice pattern on antibiotic usage for common interventions amongst pediatric urologists (PU) practicing in four English-speaking sectors of the world.

Material(s) and Method(s): An anonymous survey of five scenarios with multiple choice options was disseminated to all active practicing members of the Pediatric Urologist of Canada (PUC) and Society of Pediatric Urology of Australia and New Zealand (SPUNZA), as well as all those attending the 2016 British Association of Pediatric Urology (BAPU) and 2017 American Association of Pediatric Urology (AAPU) meetings. The response for each scenario was summarized for overall practice pattern variation and the pattern for each sector was compared using the Fisher exact test.

Result(s): A total of 126 respondents completed the survey (68.5% response rate) with at least a 65% response rate for each of the four sectors. The majority of respondents do not use antibiotics for indwelling urethral (46.8%) and suprapubic catheters (53.4%); however, they do give antibiotics for J-J stent placement (65.1%) and hypospadias surgery (84.9%), and use antibiotics after hypospadias surgery where catheters or stents are left indwelling (80.9%, 84.2%, respectively). Among those surveyed, the PUC members and AAPU PU demonstrated similar practice patterns which often significantly differed from that of SPUNZA members and BAPU attendees. Specifically, a significantly larger proportion of the North American pediatric urologists do not use antibiotics for common procedures compared with Australia, New Zealand, and the UK (Table).

Discussion(s): In the absence of prospective studies in antibiotic use for pediatric patients to guide clinicians, there is a clear variability among sectors in the use of antibiotics for most clinical scenarios investigated. With increasing resistance patterns and possible adverse effects of antibiotics, it is important that the international pediatric urology community engage in discussions and collaborations to address this issue.

Conclusion(s): Practice patterns in antibiotic usage amongst PU varies widely, some of which may be associated with their local "culture." There is a need to understand these differences and begin to standardize treatment in the hopes of increasing appropriate use of antibiotics internationally. [Table presented]

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Status

Embase

Author NameID

Kim, Jin K.; ORCID: <http://orcid.org/0000-0003-0964-9503> Ming, Jessica M.; ORCID: <http://orcid.org/0000-0002-9049-3536>

Smith, Grahame H.H.; ORCID: <http://orcid.org/0000-0002-2911-6520>

Institution

(Kim) Faculty of Medicine, University of Toronto, Toronto, Canada (Kim, Chua, Ming, Koyle)
Division of Urology, The Hospital for Sick Children, Toronto, Canada
(Braga) McMaster Children's Hospital and McMaster University, Hamilton, Canada
(Smith) Department of Urology, The Sydney Children's Hospital Network, Sydney, Australia
(Driver) Department of Surgical Paediatrics, Royal Aberdeen Children's Hospital, Aberdeen,
United Kingdom
(Koyle) Department of Surgery, University of Toronto, Toronto, Canada
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338.

The Role of Human Acellular Dermis in Preventing Fistulas After Bladder Neck Transection in the Exstrophy-epispadias Complex.

Benz K.S., Jayman J., Maruf M., Joice G., Kasprenski M., Sopko N., Di Carlo H., Gearhart J.P.

Embase

Urology. 117 (pp 137-141), 2018. Date of Publication: July 2018.

[Article]

AN: 2000754807

Objective: To evaluate human acellular dermis (HAD) as an adjunct during bladder neck transection (BNT) by comparing surgical outcomes with other types of tissue interposition.

Method(s): A prospectively maintained institutional database of exstrophy-epispadias complex (EEC) patients was reviewed for those who underwent a BNT with at least 6 months follow-up. The primary outcome was the occurrence of BNT-related fistulas.

Result(s): In total, 147 EEC patients underwent a BNT with a mean follow-up time of 6.9 years (range 0.52-23.35 years). There were 124 (84.4%) classic exstrophy patients, 22 (15.0%) cloacal exstrophy patients, and 1 (0.7%) penopubic epispadias patient. A total of 12 (8.2%) BNTs resulted in fistulization, including 4 vesicoperineal fistulas, 7 vesicourethral fistulas, and 1 vesicovaginal fistula. There were 5 (22.7%) fistulas in the cloacal exstrophy cohort and 7 (5.6%) fistulas in the classic bladder exstrophy cohort ($P = .019$). Using either HAD or native tissue flaps resulted in a lower fistulization rate than using no interposed layers (5.8% vs 20.8%; $P = .039$). Of

those with HAD, the use of a fibrin sealant did not decrease fistulization rates when compared to HAD alone (6.5% vs 8.8%, $P = .695$). There was no statistical difference in surgical complications between the use of HAD and native flaps (8.6% vs 5%, $P = .716$).

Conclusion(s): Use of soft tissue flaps and HAD is associated with decreased fistulization rates after BNT. HAD is a simple option and an effective adjunct that does not require harvesting of tissues in patients where a native flap is not feasible.

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Status

Embase

Author NameID

Sopko, Nikolai; ORCID: <http://orcid.org/0000-0003-4913-0570> Di Carlo, Heather; ORCID: <http://orcid.org/0000-0003-2396-4796>

Gearhart, John P.; ORCID: <http://orcid.org/0000-0002-0177-6121>

Institution

(Benz, Jayman, Maruf, Joice, Kasprenski, Sopko, Di Carlo, Gearhart) Robert D. Jeffs Division of Pediatric Urology, James Buchanan Brady Urological Institute, Johns Hopkins Hospital, Baltimore, MD, United States

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339.

Birth defects in children of men exposed in utero to diethylstilbestrol (DES).

Tournaire M., Devouche E., Epelboin S., Cabau A., Dunbavand A., Levadou A.

Embase

Therapie. 73 (5) (pp 399-407), 2018. Date of Publication: October 2018.

[Article]

AN: 2000614968

Objective: Prenatal exposure to diethylstilbestrol (DES) is associated with adverse effects, including genital anomalies and cancers in men and women. Animal studies showed birth defects and tumors in the offspring of mice prenatally exposed to DES. In humans, birth defects, such as hypospadias were observed in children of prenatally exposed women. The aim of this research was to assess the birth defects in children of prenatally exposed men.

Method(s): In a retrospective study conceived by a patients' association (Reseau DES France), the reports of men prenatally exposed to DES on adverse health effects in their children were compared with those of unexposed controls and general population.

Result(s): An increased incidence of two genital anomalies, cryptorchidism (OR = 5.72; 95% CI 1.51-21.71), and hypoplasia of the penis (OR = 22.92; 95% CI 3.81-137.90), was observed in the 209 sons of prenatally exposed men compared with controls, but hypospadias incidence was not increased in comparison with either the controls or the general population. No increase of genital anomalies was observed in daughters.

Conclusion(s): With caution due to the methods and to the small numbers of defects observed, this work suggests an increased incidence of two male genital tract defects in sons of men prenatally exposed to DES. This transgenerational effect, already observed in animals and in the offspring of women prenatally exposed to DES, could be the result of epigenetic changes transmitted to the subsequent generation through men.

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Status

Embase

Author NameID

Devouche, Emmanuel; ORCID: <http://orcid.org/0000-0003-3987-1842> Dunbavand, Annabel;

ORCID: <http://orcid.org/0000-0002-2879-0820>

Institution

(Tournaire, Cabau) Assistance publique-Hopitaux de Paris, Paris 75014, France (Tournaire, Devouche) Universite Paris Descartes, Paris 75006, France

(Devouche) Laboratoire psychopathologie et processus de sante EA4057, Boulogne-Billancourt 92100, France

(Epelboin) Assistance publique-Hopitaux de Paris, Hopital Bichat-Claude Bernard, Paris 75018, France

(Epelboin) Universite Paris 7 Diderot, Paris 75013, France

(Dunbavand) Mutualite francaise, Paris 75015, France

(Levadou) Association reseau DES France, Mont-de-Marsan 40000, France

Publisher

Elsevier Masson SAS (62 rue Camille Desmoulins, Issy les Moulineaux Cedex 92442, France)

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340.

Novel NR5A1 mutations found in Chinese patients with 46, XY disorders of sex development.

Yu B., Liu Z., Gao Y., Mao J., Wang X., Hao M., Ma W., Huang Q., Zhang R., Nie M., Wu X.

Embase

Clinical Endocrinology. 89 (5) (pp 613-620), 2018. Date of Publication: November 2018.

[Article]

AN: 623963580

Objective: To analyze nuclear receptor subfamily 5 group A member 1 (NR5A1) gene mutations in a cohort of Chinese patients with 46, XY Disorders of Sex Development (DSD).

Method(s): Sixty 46, XY DSD patients were recruited at Peking Union Medical College Hospital. Targeted next-generation and Sanger sequencing were performed to investigate pathogenic gene variants and validate NR5A1 gene variants, respectively. In silico tools and in vitro function studies were used to analyze the pathogenicity of rare variants. The clinical and endocrinological characteristics of patients with NR5A1 variants were retrospectively analyzed.

Result(s): A total of four novel and three recurrent NR5A1 variants were identified in seven 46, XY DSD patients. These variants widely spread almost all the functional domains. Functional studies showed that novel mutations including p.S32N, p.N44del and p.G91D reduced transactivation of CYP11A1, while the other missense variant p.A168E did not impact protein function. All patients with NR5A1 rare variants had normal adrenal function and showed genital defects. Results of the genitalia examination showed female external genitalia (three patients), ambiguous external genitalia (two patients), female external genitalia with clitoromegaly (one patient), and hypospadias (one patient). All seven patients had bilateral testis and five of seven patients lacked Mullerian structures.

Conclusion(s): Four novel mutations in the NR5A1 gene were identified in our cohort with 46, XY DSD, expanding the spectrum of NR5A1 gene mutations. All patients with NR5A1 rare variants had normal adrenal function and showed genital defects.

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Embase

Author NameID

Yu, Bingqing; ORCID: <http://orcid.org/0000-0002-0459-1923> Mao, Jiangfeng; ORCID: <http://orcid.org/0000-0003-0157-1545>

Nie, Min; ORCID: <http://orcid.org/0000-0001-7179-0044>

Institution

(Yu, Liu, Gao, Mao, Wang, Hao, Ma, Huang, Zhang, Nie, Wu) NHC Key laboratory of Endocrinology, Peking Union Medical College Hospital, Beijing, China (Yu, Liu, Gao, Mao, Wang, Hao, Ma, Huang, Zhang, Nie, Wu) Department of Endocrinology, Peking Union Medical College Hospital, Peking Union Medical College, Chinese Academy of Medical Sciences, Beijing, China

Publisher

Blackwell Publishing Ltd

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2018

341.

Mutant NR5A1/SF-1 in patients with disorders of sex development shows defective activation of the SOX9 TESCO enhancer.

Sreenivasan R., Ludbrook L., Fisher B., Declosmenil F., Knowler K.C., Croft B., Bird A.D., Ryan J., Bashamboo A., Sinclair A.H., Koopman P., McElreavey K., Poulat F., Harley V.R.

Embase

Human Mutation. 39 (12) (pp 1861-1874), 2018. Date of Publication: December 2018.

[Article]

AN: 623692635

Nuclear receptor subfamily 5 group A member 1/Steroidogenic factor 1 (NR5A1; SF-1; Ad4BP) mutations cause 46,XY disorders of sex development (DSD), with phenotypes ranging from developmentally mild (e.g., hypospadias) to severe (e.g., complete gonadal dysgenesis). The molecular mechanism underlying this spectrum is unclear. During sex determination, SF-1 regulates SOX9 (SRY [sex determining region Y]-box 9) expression. We hypothesized that SF-1

mutations in 46,XY DSD patients affect SOX9 expression via the Testis-specific Enhancer of Sox9 core element, TESCO. Our objective was to assess the ability of 20 SF-1 mutants found in 46,XY DSD patients to activate TESCO. Patient DNA was sequenced for SF-1 mutations and mutant SF-1 proteins were examined for transcriptional activity, protein expression, sub-cellular localization and in silico structural defects. Fifteen of the 20 mutants showed reduced SF-1 activation on TESCO, 11 with atypical sub-cellular localization. Fourteen SF-1 mutants were predicted in silico to alter DNA, ligand or cofactor interactions. Our study may implicate aberrant SF-1-mediated transcriptional regulation of SOX9 in 46,XY DSDs.

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Status

Embase

Author NameID

Harley, Vincent R.; ORCID: <http://orcid.org/0000-0002-2405-1262>

Institution

(Sreenivasan, Ludbrook, Fisher, Knowler, Croft, Bird, Ryan, Harley) Hudson Institute of Medical Research, VIC, Australia (Sreenivasan) Department of Anatomy and Neuroscience, University of Melbourne, VIC, Australia

(Ludbrook, Fisher, Harley) Department of Biochemistry and Molecular Biology, Monash University, VIC, Australia

(Declosmenil) Institut de Genetique Humaine, Montpellier, France

(Croft, Poulat) Department of Molecular Translational Science, Monash University, VIC, Australia

(Bashamboo, McElreavey) Institut Pasteur, Paris, France

(Sinclair) Murdoch Children's Research Institute, Royal Children's Hospital and Department of Paediatrics, The University of Melbourne, Melbourne, VIC, Australia

(Koopman) Institute for Molecular Bioscience, The University of Queensland, Brisbane, Australia

(Harley) Department of Anatomy and Developmental Biology, Monash University, VIC, Australia

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Year of Publication

2018

342.

Management of Post-traumatic and iatrogenic urethrocuteaneous fistula in children (a case series of seven patients).

Khan I., Qureshi M.A., Abbas S.H., Shaukat M.

Embase

Journal of the Pakistan Medical Association. 68 (6) (pp 956-958), 2018. Date of Publication: June 2018.

[Article]

AN: 622161357

This is a retrospective study of seven patients with post-traumatic and iatrogenic urethrocuteaneous fistula of penile urethra, excluding complication of hypospadias surgery. It was conducted in the Paediatric Surgery Department, Jinnah Hospital Lahore from June 2014 to January 2017. The patients ages ranged from three to twelve years. All the patients were managed by repairing the fistula in three layers electively at 3 months from the date of initial presentation. They remain well with no complaints except one with a recurrence. The complications of circumcision can be avoided by preventing circumcision by non-doctors and quacks.

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Status

Embase

Institution

(Khan, Qureshi, Abbas, Shaukat) Department of Pediatric Surgery, Allama Iqbal Medical College, Jinnah Hospital, Lahore, Pakistan

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2018

343.

Differences in early career operative experiences among pediatric urologists.

Suson K.D., Wolfe-Christensen C., Elder J.S., Lakshmanan Y.

Embase

Journal of Pediatric Urology. 14 (4) (pp 333.e1-333.e7), 2018. Date of Publication: August 2018.

[Article]

AN: 2000935872

Introduction: Previous research suggests that pediatric urologists feel well trained by their fellowship for cases encountered early in their career. We questioned the complexity and diversity of cases new pediatric urologists were actually performing.

Objective(s): The aim was to identify the frequency with which newly trained pediatric urologists are performing various procedures, investigate which factors are associated with case complexity and diversity, and evaluate for differences between male and female surgeons. Study design: Case logs of urologists from July 30, 2007, to June 30, 2013, initially applying for the certifying examination who self-identified as pediatric urologists were reviewed. Data points included cases/dates, and surgeon demographics. An in-depth analysis was performed on 51 index cases from the 71 included pediatric urologists, for which a level of complexity was assigned.

Result(s): Compared with the bottom volume quartile, surgeons in the top quartile performed more cases of minimal (115.9 +/- 8.7 vs. 51.7 +/- 8.7, $p < 0.001$), moderate (31.1 +/- 2.7 vs. 10.1 +/- 1.0, $p < 0.001$) and significant (10.8 +/- 1.9 vs. 2.0 +/- 0.4, $p < 0.001$) complexity. More than 90% logged circumcisions, orchiopexies, and inguinal hernia repairs, while less than 1.5% logged open nephroureterectomies or complete male epispadias repair. Surgeons submitted at least one of 17.2 +/- 0.5 (range 5-28) unique codes. The figure presents the percentage of current procedural terminology (CPT) codes performed by each urologist. Surgeons with the least case diversity performed a higher percentage of low-complexity cases, and lower percentages of moderate and complex cases ($p < 0.001$). Males, comprising 60.6% of urologists, performed more cases than females (342.9 +/- 30.9 vs. 229.1 +/- 18.1, $p = 0.007$), averaging more cases of minimal (95.0 +/- 6.6 vs. 73.3 +/- 4.6, $p = 0.018$) and significant (6.7 +/- 1.0 vs. 2.8 +/- 0.5, $p = 0.005$) complexity. There was no difference in cases of moderate complexity (22.0 +/- 1.9 vs. 18.1 +/- 2.1, $p = 0.201$).

Discussion(s): In general, pediatric urologists should expect to perform many minor cases when they enter practice. Women are entering urology in increasing numbers. In our study, female urologists performed fewer cases. This could have implications for the workforce, which in urology in general is expected to decrease.

Conclusion(s): Case diversity and degree of complexity vary among newly trained pediatric urologists. The urologist with the greatest case diversity never performed 45% of the 51 analyzed CPT codes, while the one with the least case diversity never performed 90% of the codes. Male surgeons performed more operations, particularly those of minimal and significant complexity.

The variability in operative experience reinforces the importance of continuing education and mentorship after completion of fellowship.[Figure presented]

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Status

Embase

Institution

(Suson, Lakshmanan) Children's Hospital of Michigan, Detroit, MI, United States (Wolfe-Christensen) Cook Children's Health Care System, Fort Worth, TX, United States (Elder) Massachusetts General Hospital for Children, Boston, MA, United States

Publisher

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2018

344.

Abnormal dartos fascia in buried penis and hypospadias: Evidence from histopathology.

Atmoko W., Shalmont G., Situmorang G.R., Wahyudi I., Tanurahardja B., Rodjani A.

Embase

Journal of Pediatric Urology. 14 (6) (pp 536.e1-536.e7), 2018. Date of Publication: December 2018.

[Article]

AN: 2000896515

Introduction: The importance of the pathology of the dartos fascia in hypospadias and buried penis is still debatable. Understanding the properties of connective tissue of dartos fascia in hypospadias and buried penis may give a clue to the underlying mechanism.

Objective(s): This study aimed to compare connective tissue and vascularization of dartos fascia between normal penis, buried penis, and hypospadias. Study design: We conducted this prospective study from May 2013 to November 2016. We collected dartos fascia specimens from three groups: buried penis, hypospadias, and normal penis as control. All of the patients underwent primary surgery in all groups. Patients with penile abnormalities, such as phimosis or

Balanitis Xerotica Obliterans (BXO) were excluded from the normal penis group. We compared the fibers between these groups using Masson trichrome histochemical staining, Gomori's silver impregnation staining, Weigert resorcin-fuchsin staining, and CD31 immunohistochemistry staining for evaluation of collagen fibers, reticulin fibers, elastin fibers, and endothelial cells of blood vessels, respectively. The collagen fibers, reticular fibers, elastic fibers, and vascular vessels were counted with ImageJ and manually calibrated and counted and were analyzed using the one-way ANOVA test. The assessment conducted by two pathologists was blinded, without knowing the clinical diagnosis of patients.

Result(s): There was a total of 60 patients with 20 patients in each group. Collagen fibers for most cases of buried penis and hypospadias showed thicker but fewer collagen fibers than the normal penis. There was a reduction of total collagen and elastin of dartos fascia in hypospadias and buried penis cases. On the other hand, the ratio of reticulin fibers, which represents collagen type III to total collagen, was increased compared to normal penis.

Discussion(s): Although the dartos fascia in buried penis and hypospadias is thick and inelastic when palpated or during traction/counter traction, it is well-vascularized tissue. This inelastic dartos fascia tissue is an abnormal tissue, but its characteristics are not similar to fibrotic tissue. However, further study with a larger sample is warranted and should differentiate the degree of chordee in patients with hypospadias and buried penis.

Conclusion(s): There was a difference between connective the tissue of dartos fascia in buried penis and patients with hypospadias compared with normal penis. Inelastic dartos fascia tissue in patients diagnosed with buried penis and hypospadias is an abnormal tissue. Therefore, it is suggested that this tissue is excised during reconstructive surgery. Further research is needed to unveil the pathophysiology of the condition.

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Status

Embase

Author NameID

Atmoko, Widi; ORCID: <http://orcid.org/0000-0002-7793-7083>

Institution

(Atmoko, Situmorang, Wahyudi, Rodjani) Department of Urology, Dr. Cipto Mangunkusumo National General Hospital, Jakarta Pusat, Indonesia (Shalmont, Tanurahardja) Department of Pathology Anatomy, Dr. Cipto Mangunkusumo National General Hospital, Jakarta Pusat, Indonesia

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345.

Back to the future: The Cecil-Culp technique for salvage penile reconstructive procedures.

Weiss D.A., Long C.J., Frazier J.R., Shukla A.R., Srinivasan A.K., Kolon T.F., DiCarlo H.,
Gearhart J.P., Canning D.A.

Embase

Journal of Pediatric Urology. 14 (4) (pp 328.e1-328.e7), 2018. Date of Publication: August 2018.

[Article]

AN: 2000835682

Introduction: Re-operative penile reconstruction is challenging and requires tension-free skin closure. The repair popularized by Cecil and Culp in the 1940s, using the scrotum to provide a temporary vascularized bed for complex hypospadias repairs, fell out of favor due to temporal trends towards single-stage repairs and concern for utilizing hair-bearing skin on the penile shaft. Objective(s): It was hypothesized that a modified Cecil-Culp (CC) concept of penile scrotalization, leaving the penis attached to the scrotum for 1 year rather than 6 weeks as originally described, improves outcomes with this reconstruction for ventral skin deficiency or poor vascular support. Method(s): Institutional Review Board-approved registries were reviewed to identify patients who underwent a CC repair during 1987-2016 at two institutions. Cecil-Culp technique was utilized in multi-stage hypospadias complication repairs or for insufficient ventral penile shaft skin coverage. Anatomic abnormality, number and type of prior surgeries, and complications before and after CC were recorded.

Result(s): Thirty-nine patients underwent CC: 23 failed hypospadias repairs, three hypospadias after bladder exstrophy, 10 penile curvature following circumcision, and three with skin loss from trauma. Mean age at CC was 61.8 months (hypospadias), and 59.8 months (non-hypospadias). Hypospadias patients underwent a mean of 3.6 surgeries (range 1-9) prior to CC. Four of the 39 patients (10.3%) had perioperative complications after CC, including scrotal abscess, skin infections, and difficulty removing the urethral stent. Eight of 37 (21.6%) patients had longer-term complications related to their hypospadias repair, including fistulae, diverticula, dehiscence, and stricture. Mean time from CC placement to release was 345 and 473 days for hypospadias and

non-hypospadias cases, respectively. There was no apparent scrotal skin transferred to the penile shaft at the final take-down. Mean follow-up was 22.3 months.

Discussion(s): Embedding the penis into the scrotum for added vascularity and ventral skin coverage has been used effectively in cases of the most tenacious fistulas and for significant skin loss and trauma. Limitations of this study were its retrospective approach at two institutions over an extended period of time by multiple surgeons, so patient selection and procedure may have varied.

Conclusion(s): Modification of CC repair by delaying 9-12 months before CC take-down enhanced the benefits of a robust vascular bed for wound healing, and helped to avoid transfer of hair-bearing scrotal skin to the penile shaft. The CC technique is an important tool for penile reconstructive surgery of complex hypospadias repairs with inadequate skin, and for traumatic injuries.

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Author NameID

Kolon T.F.; ORCID: <http://orcid.org/0000-0002-8740-5021> Gearhart J.P.; ORCID: <http://orcid.org/0000-0002-0177-6121>

Institution

(Weiss, Long, Frazier, Shukla, Srinivasan, Kolon, Canning) Division of Urology, Children's Hospital of Philadelphia, Philadelphia, PA, United States (DiCarlo, Gearhart) Jeffs Division of Pediatric Urology, James Buchanan Brady Urological Institute, The Johns Hopkins University School of Medicine, Baltimore, MD, United States

Publisher

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Year of Publication

2018

346.

Description of a novel variant in the MAMLD1 gene in isolated hypospadias.

Fernandez N., Pabon J., Ayala P., Perez J., Ortiz A.M., Zarante I.

Embase

Urologia Colombiana. 27 (1) (pp 105-109), 2018. Date of Publication: 2018.

[Article]

AN: 628630392

Introduction Hypospadias is a multifactorial malformation. Among previously described genes, the MAMLD1 has recently been identified in association with the development of hypospadias. So far, there are no studies in Latin America addressing this gene. The aim of this paper is to describe the presence of variants in exon 3 of the MAMLD1 in our population. Materials and Methods Using the Bio-Repository's database of the Human Genetics Institute, we identified 51 patients between 2008 and 2012 with varying degrees of isolated hypospadias. Exon 3 was sequenced looking for polymorphisms. These were assessed with in silico prediction algorithms to describe the effect of these changes on the function and structure of the protein. Results Four patients with 3 variants were identified. The p.S364P variant has not been previously described and has a deleterious effect. The other two, rs41313406 and rs61740566, have been previously described and are not considered variations with harmful effect on the protein function. The genotype-phenotype correlation varies in severity between the variables described. Conclusions For the first time a Latin American population is molecularly analyzed. The p. S364P variation has never been described before. Variations with different degrees of hypospadias are described.

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Status

Embase

Institution

(Fernandez, Perez, Ortiz) Unidad de Urologia, Hospital Universitario San Ignacio, Colombia

(Fernandez, Perez, Ortiz) Pontificia Universidad Javeriana, Colombia

(Pabon, Ayala, Zarante) Instituto de Genetica Humana, Facultad de Medicina, Pontificia Universidad Javeriana, Colombia

Publisher

Thieme Medical Publishers, Inc. (E-mail: custserv@thieme.com)

Year of Publication

2018

Impact of changes in human reproduction on the incidence of endocrine-related diseases.

Swaen G.M.H., Boffetta P., Zeegers M.

Embase

Critical Reviews in Toxicology. 48 (9) (pp 789-795), 2018. Date of Publication: 21 Oct 2018.

[Review]

AN: 626192972

The incidence rates of a wide range of diseases and conditions have increased over the last decades. There is controversy over the origin of these increases, whether they are caused by exposure to compounds thought to have an effect on the endocrine system, the "endocrine disruption theory", or whether some other factor is responsible. In this analysis, the authors take a closer look at the role that changes in reproductive factors have played in this respect. They apply the relative risks of age at first pregnancy and parity or family size to a set of Dutch demographic data from 1955 and 2015 and calculate the percentage of disease increase explained. The decrease in parity over the last decades explains an increase of 26% in testicular cancer. The combination of decrease in parity and increase in maternal age at first pregnancy explains an increase of 34% in hypospadias prevalence. This combination of decreased parity and increased maternal age at pregnancy explains an increase of 24% in childhood obesity prevalence. The authors further point to a perhaps even more profound effect of the trend toward smaller families. This trend has led to an estimated doubling of the proportion of children born from subfertile couples. Since children born from subfertile couples are more likely to be preterm or of low birth weight, the incidence of these conditions must have increased as well. Low birth weight and preterm delivery are risk factors for a wide range of diseases and conditions. The changes in human reproduction over the last decades have had a profound impact on the incidence of a range of diseases and conditions in the next generation and thus provide a sound explanation for a substantial portion of the reported increases. Key messages The incidence rates of a wide range of diseases and conditions have increased in the Western societies over the last decades. Many have argued that these increases are attributable to compounds thought to have effects on the human endocrine system: the endocrine disruption theory This analysis shows, however, that human reproductive factors such as maternal age at first pregnancy and parity explain substantial proportions of the reported increases.

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Status

Embase

Institution

(Swaen, Zeegers) CAPHRI Research Institute and Department of Complex Genetics, Maastricht University, Maastricht, Netherlands (Boffetta) Tisch Cancer Institute, Icahn School of Medicine at Mount Sinai, New York City, NY, United States

Publisher

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Year of Publication

2018

348.

Gonadal function and pubertal development in patients with Silver-Russell syndrome.

Goedegebuure W.J., Smeets C.C.J., Renes J.S., De Rijke Y.B., Hokken-Koelega A.C.S.

Embase

Human Reproduction. 33 (11) (pp 2122-2130), 2018. Date of Publication: 2018.

[Article]

AN: 625128439

STUDY QUESTION: Is gonadal function affected in males and females with Silver-Russell Syndrome (SRS)? SUMMARY ANSWER: Sertoli cell dysfunction is more common in males with SRS, with 11p15 LOM, but gonadal function seems to be unaffected in females with SRS. WHAT IS KNOWN ALREADY: Males with SRS have an increased risk for genital abnormalities such as cryptorchidism and hypospadias, which could be associated with reproductive problems in later life. In SRS females, an association has been described with Mayer-Rokitansky-Kuster-Hauser syndrome, which might compromise their reproductive function. STUDY DESIGN, SIZE, DURATION: Longitudinal follow-up study, involving 154 subjects, over a time period of 20 years. PARTICIPANTS/MATERIALS, SETTING, METHODS: Thirty-one SRS patients (14 males) and 123 non-SRS patients born at same gestational age (SGA; 65 males). All received growth hormone and 27.3% received additional gonadotropin-releasing hormone analog treatment (GnRHa). MAIN RESULTS AND THE ROLE OF CHANCE: Mean age at onset of puberty was 11.5 years in SRS males versus 11.6 years in non-SRS males ($P = 0.51$), and 10.5 years in SRS females versus 10.7 years in non-SRS females ($P = 0.50$). Four of the 14 SRS males had a post-pubertal inhibin-B level below the fifth percentile compared to healthy controls, and two of them an FSH above the 95th percentile, indicating Sertoli cell dysfunction. One of them had a history of

bilateral cryptorchidism and orchiopexy. All SRS females had AMH, LH and FSH levels within the reference range. Pubertal duration to Tanner stage five was similar in SRS and non-SRS. Pubertal height gain was better in SRS patients who additionally received GnRHa ($P < 0.01$). Mean age at menarche was 13.1 years in SRS versus 13.3 years in non-SRS ($P = 0.62$). One SRS female had primary amenorrhea due to Mullerian agenesis. LIMITATIONS, REASONS FOR CAUTION: As this is a rare syndrome, the SRS group had a small size. WIDER IMPLICATIONS OF THE FINDINGS: As gonadal function is not affected in females with SRS, it is likely that reproductive function is also not affected. Sertoli cell dysfunction in males with SRS could cause impaired reproductive function and should be assessed during pubertal development. STUDY FUNDING/COMPETING INTEREST(S): No external funding was used for the study. The authors have no conflicts of interest.

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Status

Embase

Institution

(Goedegebuure, Hokken-Koelega) Dutch Growth Research Foundation, Rotterdam 3016AH, Netherlands (De Rijke) Department of Clinical Chemistry, Erasmus University Medical Centre, Rotterdam 3015GD, Netherlands

(Goedegebuure, Smeets, Renes, Hokken-Koelega) Department of Pediatrics, Subdivision of Endocrinology, Erasmus University Medical Centre, Wytemaweg 80, Rotterdam 3015 CN, Netherlands

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349.

Penile Median Raphe Anomalies as an Indicator of Megameatus Intact Prepuce Anomaly in Children Undergoing Routine Circumcision.

Fahmy M.A.B., Shenawy A.A.E., Altramsy A., Samahy O.A., Yehya A., Othman D.

Embase

Urology. 121 (pp 164-167), 2018. Date of Publication: November 2018.

[Article]

AN: 2001103173

Objective: To arouse the suspicious for early diagnosis and hence, proper management of megameatus with an intact prepuce (MIP), as there is no external clue for detection of such cases, which usually come to light for the first time in a boy who is about to retract his prepuce or during neonatal circumcision.

Material(s) and Method(s): Examination of neonates and infants coming to circumcision clinic to detect congenital genitourinary anomalies. Evaluation of 12,518 neonates and infants coming for ritual circumcision from 2006-2017, who were examined thoroughly to detect any incidental congenital genitourinary anomalies. Fifteen of them were diagnosed to have a MIP anomaly.

They were investigated to perceive any associated median raphe (MR) anomalies. Sensitivity, positive predictive value, specificity, and negative predictive value of MR anomalies in cases of MIP were estimated and compared with other children who had a normally positioned meatus.

Result(s): Overall incidence of MIP in this group of babies was 0.12%. Twelve of 15 cases (80%) with MIP had 19 forms of MR anomalies; mainly raphe deviation in 6 cases, hyperpigmented raphe in 6, prominent raphe in 4, and bifurcation in 3 cases. Three cases had a redundant long prepuce, and 1 had paraphimosis after preputial retraction, otherwise no other genitourinary anomalies could be detected in those cases.

Conclusion(s): MR anomalies, mainly deviation and hyperpigmented prominent raphe, are significant indicators for the presence of an invisible MIP anomaly. Abnormally redundant long prepuce may be seen in such cases, but this is not common.

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Status

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Author NameID

Fahmy, Mohamed A. Baky; ORCID: <http://orcid.org/0000-0002-3570-1742>

Institution

(Fahmy, Samahy, Yehya, Othman) AL-Azhar University, Faculty of Medicine, Pediatric Surgery Department, Cairo, Egypt (Shenawy) National Institute of Urology & Nephrology, Urology Department, Cairo, Egypt

(Altramsy) AL-Azhar University, Faculty of Medicine, Plastic Surgery Department, Cairo, Egypt

Publisher

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Year of Publication

2018

350.

Associations between hypospadias, cryptorchidism and anogenital distance: Systematic review and meta-analysis.

Hua X.-G., Hu R., Hu C.-Y., Li F.-L., Jiang W., Zhang X.-J.

Embase

Andrologia. 50 (10) (no pagination), 2018. Article Number: e13152. Date of Publication: December 2018.

[Review]

AN: 624023859

Hypospadias and cryptorchidism are potential manifestations of testicular dysgenesis syndrome (TDS) at birth. Anogenital distance (AGD) has been presumed as an indicator related to endocrine disruptors proposed as one of the pathogenetic mechanisms underlying male reproductive disorders. In humans, recent studies have correlated AGD in boys to testicular anomalies. However, the associations between hypospadias, cryptorchidism and AGD remain inconsistent and have not been combined. Hence, we conducted a meta-analysis to assess gradations in the severity of the endocrine disruption in cryptorchidism or hypospadias by using AGD. A total of 2,119 boys from five birth cohort studies and two cross-sectional studies were subjected to meta-analysis. Random-effect model was used to calculate the standardised mean difference (SMD) of AGD. Our results reveal that boys with hypospadias or cryptorchidism have shorter AGD ([SMD, -2.63; 95% CI, -4.65 to -0.62] and [SMD, -0.69; 95% CI, -1.36 to -0.02]) respectively. There was no indication of a publication bias either from the result of Egger's test or Begg's test for hypospadias and cryptorchidism.

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Status

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Author NameID

Zhang, Xiu-Jun; ORCID: <http://orcid.org/0000-0002-8123-185X>

Institution

(Hua, Hu, Li, Jiang, Zhang) Department of Epidemiology and Biostatistics, School of Public Health, Anhui Medical University, Hefei, Anhui, China (Hu) School of Health Services Management, Anhui Medical University, Hefei, Anhui, China

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351.

Psychosocial and Sexual Outcomes in Adolescents following Surgery for Proximal Hypospadias in Childhood.

Andersson M., Sjostrom S., Wangqvist M., Ortqvist L., Nordenskjold A., Holmdahl G.

Embase

Journal of Urology. 200 (6) (pp 1362-1370), 2018. Date of Publication: December 2018.

[Article]

AN: 2001193637

Purpose: We evaluated psychosocial outcomes, psychosexual development and sexual function in adolescents who had undergone surgery for proximal hypospadias. We hypothesized that these outcomes would be impaired compared to peers.

Material(s) and Method(s): We identified 55 males age 14 years or older who underwent surgery for penoscrotal to perineal (intraoperatively defined) hypospadias between 1996 and 2005. A total of 33 patients with a median age of 17.5 years (range 14 to 25) answered a Web based questionnaire with self-constructed questions, completed the validated Psychological General Well-Being Index, Body-Esteem Scale for Adolescents and Adults and Penile Perception Score, and underwent clinical evaluation. A total of 31 patients with distal hypospadias (median age 19 years, range 14 to 35) and 25 age matched healthy men (17.5 years, range 14 to 25) served as controls.

Result(s): Interest in sex, age at sexarche and satisfaction with sexual experiences were comparable between patients and controls. Three patients with proximal hypospadias (10%) and 1 control (4%) reported occasional erectile problems. Three patients with proximal hypospadias

(11%), 1 patient with distal hypospadias (3%) and 1 control (4%) affirmed anejaculation. There were no differences in results between validated questionnaires. Patients with proximal hypospadias were more dissatisfied with penile length (39%) compared to controls (12%, $p = 0.049$). Concerning physical contact, 10 patients (38%) expressed uncertainty. Extra support in school was more frequent among patients with proximal hypospadias ($p = 0.024$ vs distal hypospadias, $p = 0.068$ vs control group).

Conclusion(s): Despite concerns regarding penile length, sexual experiences were comparable to those of other adolescents, although more than a third of patients with proximal hypospadias demonstrated uncertainty on questions relating to desire for physical contact. Specialized tutoring in school was more common in patients with proximal hypospadias. Continuous followup throughout childhood allowing extra time for age adequate information and support is warranted.

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Status

Embase

Institution

(Andersson, Sjostrom, Holmdahl) Sahlgrenska Academy, Women's and Children's Health, Gothenburg, Sweden (Andersson, Sjostrom, Holmdahl) Pediatric Uro-Nephrologic Center, Department of Pediatric Surgery, Queen Silvia's Children's Hospital, Gothenburg, Sweden (Wangqvist) Department of Psychology, University of Gothenburg, Gothenburg, Sweden (Ortqvist, Nordenskjold) Department of Women's and Children's Health, Center for Molecular Medicine, Karolinska Institutet, Stockholm, Sweden (Ortqvist, Nordenskjold) Department of Pediatric Surgery, Astrid Lindgren Children's Hospital, Karolinska University Hospital, Stockholm, Sweden

Publisher

Elsevier Inc. (E-mail: usjcs@elsevier.com)

Year of Publication

2018

Parental subfertility and hypospadias and cryptorchidism in boys: results from two Danish birth cohorts.

Arendt L.H., Lindhard M.S., Kjersgaard C., Henriksen T.B., Olsen J., Ramlau-Hansen C.H.

Embase

Fertility and Sterility. 110 (5) (pp 826-832), 2018. Date of Publication: October 2018.

[Article]

AN: 2001168398

Objective: To study if parental subfertility is related to the occurrence of the male genital anomalies, cryptorchidism and hypospadias.

Design(s): Population-based cohort study.

Setting(s): Not applicable. Patient(s): A total of 80,220 singleton boys and their mothers from the Danish National Birth Cohort and the Aarhus Birth Cohort. Intervention(s): None. Main Outcome Measure(s): The two congenital anomalies; cryptorchidism and hypospadias, registered within the Danish National Patient Register up until December 31, 2012. Result(s): By means of Cox regression analyses, we found no associations between waiting time-to-pregnancy (TTP) and cryptorchidism or hypospadias among those who conceived spontaneously. The highest hazard ratio for cryptorchidism was seen among boys of couples with a TTP>12 months who conceived after fertility treatment (adjusted hazard ratio [aHR] 1.19, 95% confidence interval 0.92-1.55). For hypospadias, we found that boys of couples with a TTP>12 months who conceived after fertility treatment, had a 71% higher risk of hypospadias (aHR 1.71, [95% confidence interval 1.24-3.36]) as compared with boys of couples with a TTP<5 months. Conclusion(s): The findings from this study showed that boys of couples with TTP>12 months who conceived after fertility treatment, had a higher occurrence of hypospadias than boys conceived spontaneously of couples with a short TTP. Among those who conceived spontaneously, TTP was not associated with hypospadias or cryptorchidism. These findings indicate that fertility treatment or severity of subfertility is related to hypospadias.

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Status

Embase

Institution

(Arendt, Kjersgaard, Ramlau-Hansen) Department of Public Health, Section for Epidemiology, Aarhus University, Aarhus University Hospital, Aarhus, Denmark (Arendt, Lindhard, Henriksen) Perinatal Epidemiology Research Unit, Department of Paediatric and Adolescent Medicine, Aarhus University Hospital, Aarhus, Denmark

(Olsen) Department of Clinical Epidemiology, Aarhus University Hospital, Aarhus, Denmark

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Year of Publication

2018

353.

Is there a shortening of the urethral plate in complete penile disassembly used in epispadias repair, and what is its impact on the final outcomes?.

Acimi S., Debbous L., Acimi M.A., Khelil A.L.

Embase

Journal of Plastic, Reconstructive and Aesthetic Surgery. 71 (11) (pp 1637-1643), 2018. Date of Publication: November 2018.

[Article]

AN: 2001055024

Purpose: To assess the importance of shortening of the urethral plate that occurred with complete penile disassembly technique in epispadias repair and its impact on cosmetic and functional results (on urinary incontinence).

Method(s): From January 2009 to December 2016, 26 boys underwent complete penile disassembly technique for proximal epispadias repair. Twenty-one patients had epispadias after primary repair of bladder exstrophy, and 5 patients had isolated penopubic epispadias. The age of the patients ranged from 11 months to 6 years (median 3 years).

Result(s): After disassembling the penis in three parts, the shortening and narrowing of urethral plate were found in all patients; the shortening varied between 2 and 16 mm. However, in isolated epispadias, the urethral plate is easily extensible. The cosmetic results (after dehiscence and fistulas repair) were found to be satisfactory in 24 patients with conical glans and meatus in the orthotopic position without any necrosis of the glans. However, 18 patients (81.8% of cases) who initially had a bladder exstrophy presented a dehiscence or fistula. The urinary continence ≥ 1 h was observed in 5 patients (19% of cases), and only 3 patients (11.5% of cases) had a urinary continence ≥ 3 h.

Conclusion(s): The complete penile disassembly procedure restores the normal anatomy of the penis. Despite the shortening and narrowing of the urethral plate, the cosmetic results were good

in the majority of patients. However, its functional outcomes on urinary incontinence, particularly for epispadias with bladder exstrophy, remain uncertain.

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Status

Embase

Author NameID

Acimi, Smail; ORCID: <http://orcid.org/0000-0002-2764-0302>

Institution

(Acimi, Debbous, Acimi) Department of Visceral Surgery, Children's Hospital Canastel, University Of Oran, Oran, Algeria (Khelil) Department of Pediatric Emergency, Children's Hospital Canastel, University Of Oran, Oran, Algeria

Publisher

Churchill Livingstone

Year of Publication

2018

354.

Regional anesthesia to ameliorate postoperative analgesia outcomes in pediatric surgical patients: An updated systematic review of randomized controlled trials.

Kendall M.C., Alves L.J.C., Suh E.I., McCormick Z.L., De Oliveira G.S.

Embase

Local and Regional Anesthesia. 11 (pp 91-109), 2018. Date of Publication: 2018.

[Review]

AN: 625571197

Regional anesthesia is becoming increasingly popular among anesthesiologists in the management of postoperative analgesia following pediatric surgery. The main objective of this review was to systematically evaluate the last 5 years of randomized controlled trials on the role of regional anesthesia techniques in alleviating postoperative pain associated with various pediatric surgical procedures. Forty studies on 2,408 pediatric patients were evaluated. The majority of the articles published from 2013 to 2017 reported that the use of regional anesthesia

minimized postoperative pain and reduced opioid consumption. Only a few surgical procedures (cholecystectomy, inguinal hernia repair, and non-laparoscopic major abdominal surgery) reported no significant difference in the postoperative pain relief compared with the standard anesthetic management. The growing number of randomized controlled trials in the pediatric literature is very promising; however, additional confirmation is needed to reinforce the use of specific regional anesthesia techniques to provide optimal postoperative pain relief for a few surgical procedures (reconstructive ear surgery, chest wall deformity, hypospadias, umbilical hernia, cleft palate repair) in pediatric patients. More randomized controlled trials are needed to establish regional anesthesia as an essential component of postoperative analgesia management in children.

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Institution

(Kendall, Suh, De Oliveira) Department of Anesthesiology, Rhode Island Hospital, The Warren Alpert Medical School of Brown University, Providence, RI, United States (Alves) Department of Anesthesiology, Sao Paulo State University, Sao Paulo, Brazil

(McCormick) Division of Physical Medicine and Rehabilitation, University of Utah, Salt Lake City, UT, United States

Publisher

Dove Medical Press Ltd (PO Box 300-008, Albany, 44 Corinthian Drive, Albany, Auckland 0752, New Zealand. E-mail: angela@dovepress.com)

Year of Publication

2018

355.

Investigation of Azoospermia Factor (AZF) microdeletion of hypospadias patients in Indonesian population.

Juniarto A.Z., Yasari N.A.L., Santosa A., Faradz S.M.H.

Embase

Pakistan Journal of Medical and Health Sciences. 12 (3) (pp 1350-1353), 2018. Date of Publication: July-September 2018.

[Article]

AN: 625436554

Background: Hypospadias, a midline fusion defect of the male ventral urethra, is disorder of male external genital development occurring 0.7 - 4.5 per 10.000 live births. Hypospadias patients might have fertility problem and genetic factor could be involved on this aspect. Microdeletion of the Y chromosome, notably in Azoospermia factor region (AZF) have been observed in some patients with cryptorchidism and severe defects of spermatogenesis.

Aim(s): This study aimed to investigate microdeletions of AZF region in patients with hypospadias as a potential predictor factor for infertility. AZF a microdeletion was associated to Sertoli cell syndrome, while AZF b microdeletion lead to maturation arrest at the spermatocyte stage and AZF c microdeletion caused defect in sperm production.

Method(s): Total of 60 isolated hypospadias patients who admitted to CEBIOR were analyzed for AZF microdeletions during period of 2008 - 2016. DNA samples were analyzed by PCR-screening using several sequences-tagged-site (STS) markers from different region of the AZFa, AZFb, AZFc on Yq chromosome and SRY on Yp as internal control.

Result(s): Out of 60 analyzed cases (mean age 5.66 years), 3 (5%) patients showed microdeletion of AZF regions and only detected in AZFa region. No deletion was observed in AZFb and AZFc region. In addition, used as internal control, there no SRY gene microdeletion was found.

Conclusion(s): AZF microdeletions analysis can be used as an infertility potential prognostic predictor in hypospadias patients and become important leading of genetic counseling related to possibility of infertility in the future.

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Status

Embase

Institution

(Juniarto, Faradz) Division of Human Genetics Center for Biomedical Research (CEBIOR), Faculty of Medicine, Diponegoro University, Sexual Adjustment Team, Dr. Kariadi Hospital, Semarang, Indonesia (Yasari) Faculty of Medicine, Diponegoro University, Indonesia (Santosa) Division of Urology, Sexual Adjustment Team, Dr. Kariadi Hospital, Semarang, Indonesia

Publisher

Lahore Medical And Dental College (Lahore Medical & Dental College, Lahore x, Pakistan. E-mail: nayyar_salam@yahoo.com)

Year of Publication

2018

356.

Hypertensive disorders of pregnancy and genital anomalies in boys: A danish nationwide cohort study.

Arendt L.H., Henriksen T.B., Lindhard M.S., Parner E.T., Olsen J., Ramlau-Hansen C.H.

Embase

Epidemiology. 29 (5) (pp 739-748), 2018. Date of Publication: 2018.

[Article]

AN: 624836760

Background: Although congenital abnormalities in the male reproductive tract are common, their causes remain poorly understood. We studied associations between hypertensive disorders of pregnancy (pregestational hypertension, gestational hypertension, and preeclampsia) and the genital anomalies, cryptorchidism (undescended testes), and hypospadias (ventrally displaced urethral meatus).

Method(s): We established a population of 1,073,026 Danish boys born alive between 1 January 1978 and 31 December 2012. By means of Cox regression analyses, we estimated hazard ratios with 95% confidence intervals for cryptorchidism and hypospadias according to type and severity of hypertensive disorder. Further, we used restricted cubic spline analyses to investigate the association between gestational age at onset of severe and moderate preeclampsia and the two genital anomalies.

Result(s): We found associations between pregestational hypertension and cryptorchidism (HR: 1.3; 95% CI = 1.1, 1.6) and hypospadias (HR: 1.7; 95% CI = 1.3, 2.3), whereas gestational hypertension was only associated with cryptorchidism (HR: 1.2; 95% CI = 1.1, 1.4). Boys of mothers with preeclampsia had the highest occurrence of cryptorchidism and hypospadias, increasing with preeclampsia severity. Women with HELLP syndrome faced the highest risk of having a child with both cryptorchidism (HR: 2.1; 95% CI = 1.4, 3.2) and hypospadias (HR: 3.9; 95% CI = 2.5, 6.1). Further, the occurrence increased with early onset of preeclampsia diagnosis.

Conclusion(s): These findings support the hypotheses that preeclampsia and genital anomalies share common etiologic factors and that placental dysfunction and androgen deficiency in early pregnancy are important in the etiology of male genital anomalies.

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Status

Embase

Institution

(Arendt, Ramlau-Hansen) Department of Public Health, Section for Epidemiology, Aarhus University, Bartholins Alle 2, Aarhus 8000, Denmark (Arendt, Henriksen, Lindhard) Perinatal Epidemiology Research Unit, Department of Paediatric and Adolescent Medicine, Aarhus University Hospital, Aarhus, Denmark

(Parner) Department of Public Health, Section for Biostatistics, Aarhus University, Aarhus, Denmark

(Olsen) Department of Clinical Epidemiology, Aarhus University Hospital, Aarhus, Denmark

Publisher

Lippincott Williams and Wilkins (E-mail: kathiest.clai@apta.org)

Year of Publication

2018

357.

Management of failed hypospadias: choosing the right method and achieving optimal results.

Wu M., Chen F., Xie H., Lv Y., Huang Y., Liu Y., Ye W.

Embase

International Urology and Nephrology. 50 (10) (pp 1795-1800), 2018. Date of Publication: 01 Oct 2018.

[Article]

AN: 623564434

Objective: Because of the complexity of the abnormalities and limited options for reconstruction of failed hypospadias, creating a neourethra presents a challenge to surgeons. We reviewed our experiences with staged urethroplasty strategies to repair the penis of failed hypospadias.

Material(s) and Method(s): We retrospectively reviewed 56 consecutives patients following multiple unsuccessful hypospadias repairs from 2010 to 2016. Patients were divided into the following two groups based on their penile conditions and urethroplasty procedures: staged buccal mucosa graft Bracka urethroplasty (group1) and two-stage urethroplasty with additional buccal mucosa graft augmentation of the dorsal urethral plate (group2).

Result(s): Median follow-ups were 26.5 months (12-59 months) and 28.6 months (14-59 months) in the group 1 and group 2. After the second stage, three patients (11.1%) in group 1 and two patients (6.89%) in group 2 did not have a meatal opening at the top of the glans. Three patients (11.1%) in group 1 and 4 patients (13.79%) in group 2 had urethrocutaneous fistulas. One patient (3.70%) in group 1 and no patients in group 2 had meatal stenosis. Two patients (6.89%) in group 2 and no patients in group 1 had urethral strictures; all patients with strictures were cured using dilations, so follow-up surgeries were not required. No patients in either group had signs of diverticulum or residual chordee. Three patients (11.1%) in group 1 and 4 patients (13.79%) in group 2 needed reoperations.

Conclusion(s): Failed hypospadias repairs were often due to the underestimation of the penile conditions at the prior surgery. The results indicated that two-staged strategies were preferred for treating complex situations during the intermediate period of our study. Staged buccal mucosa graft Bracka urethroplasty and two-stage urethroplasty with additional buccal mucosa graft augmentation of the dorsal urethral plate seemed as reliable approaches in complex hypospadias cases and could improve the overall success rate.

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Status

Embase

Institution

(Wu, Chen, Xie, Lv, Huang) Department of Urology, Children's Hospital of Shanghai, School of Medicine, Shanghai Jiao Tong University, Shanghai, China (Liu, Ye) Department of Urology, Ren Ji Hospital, School of Medicine, Shanghai Jiao Tong University, Shanghai, China (Chen) Department of Urology, Sixth People's Hospital; Oriental Urethral Reconstructive Center, Jiao Tong University of Shanghai, Shanghai, China

Publisher

Springer Netherlands (E-mail: rbk@louisiana.edu)

Year of Publication

2018

Parental perception of terminology of disorders of sex development in Western Turkey.

Tiryaki S., Tekin A., Yagmur I., Ozen S., Ozbaran B., Goksen D., Darcan S., Ulman I., Avanoglu A.

Embase

JCRPE Journal of Clinical Research in Pediatric Endocrinology. 10 (3) (pp 216-222), 2018. Date of Publication: September 2018.

[Article]

AN: 623401103

Objective: Disorders of sex development (DSD) is a nomenclature intended to defeat the discomfort of families and patients and has found worldwide usage. The aim of this study was to address the perception and usage of terminology among the parents of DSD patients in a tertiary center in western Turkey.

Method(s): The records of the DSD council (multidisciplinary team where each patient with DSD is discussed) between years 2008-2015 were reviewed retrospectively. Data including details of the management process, patient characteristics and follow-up details were noted. Then inquiries reflecting parental perception about terminology were implemented during clinical visits.

Result(s): In total, 121 patients were evaluated in monthly meetings of the DSD council and 79 inquiries were completed. Fifty-one percent of the families admitted knowing the terms DSD, ambiguous genitalia, "dubious genitals" and intersex. However, only 2% preferred using DSD, 6% intersex and 14% ambiguous genitalia. Fifty-two percent of the parents used a disease name in Latin (mostly hypospadias) addressing the disorder. The offspring of 69% of the parents who were familiar with the name "dubious genitals" were diagnosed in the neonatal period. The preferred terminology used by parents was strongly associated with the terminology used most commonly in the medical speciality their child most often attended.

Conclusion(s): Each country has its own social norms. We suggest therefore that local committees including medical professionals, patients and families should be employed to develop proper terminology.

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Author NameID

Tiryaki, Sibel; ORCID: <http://orcid.org/0000-0003-4087-1911>

Institution

(Tiryaki, Tekin, Yagmur, Ulman, Avanoglu) Ege University, Faculty of Medicine, Department of Pediatric Surgery, Division of Pediatric Urology, Izmir, Turkey (Ozen, Goksen, Darcan) Ege

University, Faculty of Medicine, Department of Pediatrics, Division of Pediatric Endocrinology,
Izmir, Turkey

(Ozbaran) Ege University, Faculty of Medicine, Department of Child and Adolescent Psychiatry,
Izmir, Turkey

Publisher

Galenos Yayıncılık, (Molla Gurani Cad. 21, Fındıkzade-Istanbul 34093, Turkey)

Year of Publication

2018

359.

The risk factors of Urethrocuteaneous fistula after hypospadias surgery in the youth population.

Sheng X., Xu D., Wu Y., Yu Y., Chen J., Qi J.

Embase

BMC Urology. 18 (1) (no pagination), 2018. Article Number: 64. Date of Publication: 24 Jul 2018.

[Article]

AN: 623158928

Background: The current research aims to evaluate the risk factors of urethrocuteaneous fistula after hypospadias surgery among the youth in China.

Method(s): One hundred twenty hypospadias patients were enrolled in our study. All of them were defined as Tanner 4 or 5. The information collected from the participants include age, urethral operation history, urinary comorbidities before operation, urine test before operation, body temperature before and after operation, type of surgical repair, chordee degree, urethral defect length and whether received vesicostomy after surgery or not. Independent t test, chi-square test and multivariate logistic regression were performed to evaluate the risk factor of urethrocuteaneous fistula.

Result(s): Among the enrolled patients, 39 patients (32.5%) developed urethrocuteaneous fistula after hypospadias repair. Our result showed significant association between the group with urethrocuteaneous fistula and the group without urethrocuteaneous fistula with respect to age, pyuria before operation, urethral defect length and the urethral operation history. The following logistic regression showed that urethral defect length and the urethral operation history were the risk factors of urethrocuteaneous fistula.

Conclusion(s): Urethral defect length and urethral operation history should be taken into consideration before undergoing hypospadias surgery since our study discovered that the risk of developing urethrocutaneous fistula after hypospadias repair is associated with urethral defect length and urethral operation history. Age, surgical procedure, type of surgical repair, chordee degree and other factors were not obviously related to the development of urethrocutaneous fistula.

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Status

Embase

Institution

(Sheng, Xu, Wu, Yu, Chen, Qi) Department of Urology, School of Medicine, Xinhua Hospital, Shanghai Jiao Tong University, 1665 Kongjiang Rd, XinHua Hospital, Shanghai 200092, China

Publisher

BioMed Central Ltd. (E-mail: info@biomedcentral.com)

Year of Publication

2018

360.

Hypospadias surgery in children: Improved service model of enhanced recovery pathway and dedicated surgical team.

Wong Y.S., Pang K.K.Y., Tam Y.H.

Embase

Hong Kong Medical Journal. 24 (3) (pp 238-244), 2018. Date of Publication: June 2018.

[Article]

AN: 622706788

Introduction: Children in Hong Kong are generally hospitalised for 1 to 2 weeks after hypospadias repairs. In July 2013, we introduced a new service model that featured an enhanced recovery pathway and a dedicated surgical team responsible for all perioperative services. In this study, we investigated the outcomes of hypospadias repair after the introduction of the new service model.

Method(s): We conducted a retrospective study on consecutive children who underwent primary hypospadias repair from January 2006 to August 2016, comparing patients under the old service with those under the new service. Outcome measures included early morbidity, operative success, and completion of enhanced recovery pathway.

Result(s): The old service and new service cohorts comprised 176 and 126 cases, respectively. There was no difference between the two cohorts in types of hypospadias and surgical procedures performed. The median hospital stay was 2 days in the new service cohort compared with 10 days in the old service cohort ($P<0.001$). Patients experienced less early morbidity (5.6% vs 15.9%; $P=0.006$) and had a lower operative failure rate (20.2% vs 44.2%; $P<0.001$) under the new service than the old service. Multivariable analysis revealed that the new service significantly reduced the odds of early morbidity (odds ratio=0.35, 95% confidence interval=0.15-0.85; $P=0.02$) and operative failure (odds ratio=0.32, 95% confidence interval=0.17-0.59; $P<0.001$) in comparison with the old service. Of the new service cohort, 111(88.1%) patients successfully completed the enhanced recovery pathway.

Conclusion(s): The enhanced recovery pathway can be implemented safely and effectively to primary hypospadias repair. A dedicated surgical team may play an important role in successful implementation of the enhanced recovery pathway and optimisation of surgical outcomes.

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Status

Embase

Institution

(Wong, Pang, Tam) Division of Paediatric Surgery and Paediatric Urology, Department of Surgery, Prince of Wales Hospital, The Chinese University of Hong Kong, Shatin, Hong Kong

Publisher

Hong Kong Academy of Medicine Press (Room 901, 9/F, HKAM Jockey Club Build, 99 Wong Chuk Hang Road, Aberdeen, Hong Kong, Hong Kong)

Year of Publication

2018

Outcomes and morbidities of patients who survive haemoglobin Bart's hydrops fetalis syndrome: 20-year retrospective review.

Chan W.Y.K., Leung A.W.K., Luk C.W., Li R.C.H., Ling A.S.C., Ha S.Y.

Embase

Hong Kong Medical Journal. 24 (2) (pp 107-118), 2018. Date of Publication: April 2018.

[Review]

AN: 621731527

Introduction: Haemoglobin Bart's hydrops fetalis syndrome was once considered a fatal condition. However, advances over the past two decades have enabled survival of affected patients. Data relating to their morbidities and outcomes will help medical specialists formulate a management plan and parental counselling.

Method(s): All babies with the syndrome who survived beyond the neonatal period and were subsequently managed long-term in eight public hospitals in Hong Kong from 1 January 1996 to 31 December 2015 were included. Patient and parent characteristics, antenatal care, reasons for continuation of pregnancy, intrauterine interventions, perinatal course, presence of congenital malformations, stem-cell transplantation details, and long-term neurodevelopmental outcomes were reviewed.

Result(s): A total of nine patients were identified, of whom five were female and four male. The median follow-up duration was 7 years. All were Chinese and were homozygous for the Southeast Asian alpha-thalassaemia deletion. Five of the nine mothers received antenatal care at a public hospital and opted to continue the pregnancy after antenatal diagnosis and counselling. Despite intrauterine transfusions, all babies were born with respiratory depression and required intubation and mechanical ventilation during the neonatal period. Hypospadias was identified in all four male infants. Growth retardation, global developmental delay, and residual neurological deficits were noted in two-thirds of the patients. Haematopoietic stem-cell transplantation was performed in two patients, who became transfusion-independent.

Conclusion(s): Survival of patients with Bart's hydrops fetalis syndrome is possible but not without short and long-term complications; local epidemiology is comparable to that documented for an international registry. Detailed antenatal counselling of parents with a non-judgemental attitude and cautious optimism are imperative.

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Status

Embase

Institution

(Chan, Ha) Department of Paediatrics and Adolescent Medicine, Queen Mary Hospital, Pokfulam, Hong Kong (Leung) Department of Paediatrics and Adolescent Medicine, Prince of Wales Hospital, Shatin, Hong Kong

(Luk) Department of Paediatrics and Adolescent Medicine, Queen Elizabeth Hospital, Kowloon, Hong Kong

(Li) Department of Paediatrics and Adolescent Medicine, Tuen Mun Hospital, Tuen Mun, Hong Kong

(Ling) Department of Paediatrics and Adolescent Medicine, Princess Margaret Hospital, Laichikok, Hong Kong

Publisher

Hong Kong Academy of Medicine Press (Room 901, 9/F, HKAM Jockey Club Build, 99 Wong Chuk Hang Road, Aberdeen, Hong Kong, Hong Kong)

Year of Publication

2018

362.

Pelvic and lower extremity immobilization for cloacal exstrophy bladder and abdominal closure in neonates and older children.

Benz K.S., Jayman J., Maruf M., Baumgartner T., Kasprenski M.C., Friedlander D.A., Di Carlo H.N., Sponseller P.D., Gearhart J.P.

Embase

Journal of Pediatric Surgery. 53 (11) (pp 2160-2163), 2018. Date of Publication: November 2018.

[Article]

AN: 620258158

Introduction: Successful bladder closure in cloacal exstrophy (CE) is best accomplished through a multidisciplinary team and attention to pre- and postoperative technique. This study from a high volume exstrophy center investigates outcomes and complications of primary and reoperative bladder closures in patients immobilized with spica cast or patients with external fixation (EF) and skin traction.

Method(s): The authors reviewed an institutionally approved and daily updated database of 1311 patients with exstrophy-epispadias complex and identified patients with cloacal exstrophy born between 1975 and 2015 who had undergone primary or reoperative bladder closures. Only the

closures that used spica casting or external fixation were included for analysis. Demographic, operative, and outcomes data were compared between patients with spica cast only and patients with external fixation and skin traction.

Result(s): Out of 140 patients with CE or a CE variant, a total of 71 patients with 94 bladder closures (66 primary and 28 reoperative) met inclusion criteria. Median follow-up time was 8.8 years (range 1.5-29.1). There were 37 closures performed at the authors' institution and 58 from outside hospitals. Pelvic osteotomy was undertaken in 66 (70.2%) of all closures, and in 36 (97.3%) of closures at the authors' institution. Postoperative immobilization was achieved with spica cast alone in 46 (48.9%) closures, external fixation and skin traction in 43 (45.7%), and spica cast and external fixation in 5 (5.3%) closures. For all closures, there were 33 failures (71.7%) among those immobilized with spica cast alone versus 4 failures (9.3%) for those immobilized with external fixation and skin traction ($p < 0.001$). When restricted to closures performed with osteotomy, the failure rates were 50.0% and 9.3% respectively ($p = 0.002$). There was minimal differences in complication rates between spica and external fixation groups (8.7% versus 23.3%, $p = 0.059$).

Conclusion(s): Failure of CE closure can occur with any form of pelvic and lower extremity immobilization. This study, however, provides continued evidence that external fixation with skin traction is an optimal, secure technique (3.8% failure rate) for postoperative management in an older child (1-2 years).

Level of Evidence: Level III, Retrospective comparative study Study Type: Therapeutic study

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Status

Embase

Institution

(Benz, Jayman, Maruf, Baumgartner, Kasprenski, Friedlander, Di Carlo, Gearhart) Robert D. Jeffs
Division of Pediatric Urology, James Buchanan Brady Urological Institutions, Johns Hopkins
Hospital, Johns Hopkins Medical Institutions, Charlotte Bloomberg Children's Hospital, Baltimore,
MD, United States (Sponseller) Department of Orthopedic Surgery, The Johns Hopkins
University, Baltimore, MD, United States

Publisher

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2018

363.

Elsahy-Waters syndrome is caused by biallelic mutations in CDH11.

Harms F.L., Nampoothiri S., Anazi S., Yesodharan D., Alawi M., Kutsche K., Alkuraya F.S.

Embase

American Journal of Medical Genetics, Part A. 176 (2) (pp 477-482), 2018. Date of Publication: Februaryy 2018.

[Article]

AN: 619937203

Elsahy-Waters syndrome (EWS), also known as branchial-skeletal-genital syndrome, is a distinct dysmorphology syndrome characterized by facial asymmetry, broad forehead, marked hypertelorism with proptosis, short and broad nose, midface hypoplasia, intellectual disability, and hypospadias. We have recently published a homozygous potential loss of function variant in CDH11 in a boy with a striking resemblance to EWS. More recently, another homozygous truncating variant in CDH11 was reported in two siblings with suspected EWS. Here, we describe in detail the clinical phenotype of the original CDH11-related patient with EWS as well as a previously unreported EWS-affected girl who was also found to have a novel homozygous truncating variant in CDH11, which confirms that EWS is caused by biallelic CDH11 loss of function mutations. Clinical features in the four CDH11 mutation-positive individuals confirm the established core phenotype of EWS. Additionally, we identify upper eyelid coloboma as a new, though infrequent clinical feature. The pathomechanism underlying EWS remains unclear, although the limited phenotypic data on the *Cdh11*^{-/-} mouse suggest that this is a potentially helpful model to explore the craniofacial and brain development in EWS-affected individuals.

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Status

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Author NameID

Kutsche, Kerstin; ORCID: <http://orcid.org/0000-0001-8494-8963> Alkuraya, Fowzan S.; ORCID: <http://orcid.org/0000-0003-4158-341X>

Institution

(Harms, Kutsche) Institute of Human Genetics, University Medical Center Hamburg-Eppendorf, Hamburg, Germany (Nampoothiri, Yesodharan) Department of Pediatric Genetics, Amrita Institute of Medical Sciences & Research Centre, Cochin, Kerala, India
(Anazi, Alkuraya) Department of Genetics, King Faisal Specialist Hospital and Research Center, Riyadh, Saudi Arabia
(Alawi) University Medical Center Hamburg-Eppendorf, Bioinformatics Core, Hamburg, Germany
(Alkuraya) Department of Anatomy and Cell Biology, College of Medicine, Alfaisal University, Riyadh, Saudi Arabia
Publisher
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Year of Publication
2018

364.

External Genital Development, Urethra Formation, and Hypospadias Induction in Guinea Pig: A Double Zipper Model for Human Urethral Development.

Wang S., Shi M., Zhu D., Mathews R., Zheng Z.

Embase

Urology. 113 (pp 179-186), 2018. Date of Publication: March 2018.

[Article]

AN: 619857385

Objective: To determine whether the guinea pig phallus would be an appropriate model of human penile development, we characterized the embryology and sexual differentiation of guinea pig external genitalia and attempted to induce hypospadias in males and tubular urethra formation in females pharmacologically.

Material(s) and Method(s): The external genitalia of guinea pig were collected from genital swelling initiation to newborn stages, and scanning electronic microscopy and histology were performed to visualize the morphology and structure. Immunohistochemistry was used to determine the androgen receptor localization. Bicalutamide and methyltestosterone were given to pregnant dams to reveal the role and timing of androgen in guinea pig penile masculinization.

Result(s): Canalization and dorsal-to-ventral movement of the urethral canal develops the urethral groove in both sexes, and then the males perform distal-opening-proximal-closing to form tubular

urethra. More nuclear-localized androgen receptor is found in proximal genital tubercles of males than in females at (E) 29. Antiandrogen treatment at E26-E30 can cause hypospadias, and methyltestosterone administration at E27-E31 can induce tubular urethra formation in females. Conclusion(s): Fetal development of the guinea pig phallus is homologous to that of humans. Although guinea pig has structures similar to mouse, the urethral groove and the tubular urethra formation are more similar to humans. Antiandrogen treatment causes hypospadias in males and additional androgen induces tubular urethra formation in females. Thus, guinea pig is an appropriate model for further study of cellular and molecular mechanisms involved in distal-opening-proximal-closing in tubular urethra formation and the evaluation of the pathophysiological processes of hypospadias.

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Status

Embase

Author NameID

Zheng, Zhengui; ORCID: <http://orcid.org/0000-0003-0997-2976>

Institution

(Wang, Shi, Zhu, Zheng) Department of Physiology, School of Medicine, Southern Illinois University Carbondale, Carbondale, IL, United States (Zhu) Department of Pharmacy, Jiangsu Food & Pharmaceutical Science College, Huai'an, Jiangsu Province, China (Mathews) Department of Urology, School of Medicine, Southern Illinois University, Springfield, IL, United States

Publisher

Elsevier Inc. (E-mail: usjcs@elsevier.com)

Year of Publication

2018

365.

Sevoflurane Affects Oxidative Stress and Alters Apoptosis Status in Children and Cultured Neural Stem Cells.

Zhou X., Lu D., Li W.-D., Chen X.-H., Yang X.-Y., Chen X., Zhou Z.-B., Ye J.-H., Feng X.

Embase

Neurotoxicity Research. 33 (4) (pp 790-800), 2018. Date of Publication: 01 May 2018.

[Article]

AN: 618962423

Anesthesia-induced neurotoxicity in immature animals has raised concerns about similar effects occurring in young children. Our study investigated two commonly used anesthetics-sevoflurane and propofol-for neurotoxicity in young children. Forty-seven children (aged 12-36 months) undergoing hypospadias repair surgery were randomized to receive sevoflurane (SG, n = 24) or propofol (PG, n = 23) general anesthesia. Venous blood was collected at three different times-immediately after induction, 2 h, and 3 days after surgery. The cellular portion was assessed for antioxidant defense and DNA damage, using enzyme assay kits and qRT-PCR, respectively, while serum was used to treat cultured neural stem cells (NSCs). MTT assay and TUNEL staining were performed, and the mRNA levels of antioxidant enzymes and apoptosis indicators were evaluated by qRT-PCR. Antioxidant defense and apoptosis status in the SG group were significantly higher than in the PG group at 2 h after surgery. Additionally, exposure of NSCs to postoperative serum of the SG group resulted in decreased cell density and viability, increased TUNEL-positive cells, elevated mRNA levels of antioxidant enzymes, and cleaved caspase-3 expression. Our data shows for the first time that in young children, administration of sevoflurane, but not propofol, leads to temporally increased antioxidant defense and apoptosis status as well as damage of NSCs.

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Status

Embase

Author NameID

Feng, Xia; ORCID: <http://orcid.org/0000-0002-5263-6233>

Institution

(Zhou, Lu, Yang, Chen, Zhou, Feng) Department of Anesthesiology, The First Affiliated Hospital, Sun Yat-sen University, No. 2nd Zhongshan Road, Guangzhou, Guangdong 510080, China (Li)

Department of Hepatobiliary Surgery, The Sun Yat-sen Memorial Hospital, Sun Yat-sen University, No. 107 Yanjiang West Road, Guangzhou, Guangdong 510120, China

(Chen) Department of Anesthesiology, Fujian Provincial Hospital, Fuzhou 350001, China

(Ye) Department of Anesthesiology, Rutgers, the State University of New Jersey, New Jersey Medical School, 185 South Orange Avenue, Newark, NJ 07103, United States

Publisher

Springer New York LLC (E-mail: barbara.b.bertram@gsk.com)

Year of Publication

2018

366.

Frequency of Antibiotic-Associated Diarrhea and Related Complications in Pediatric Patients Who Underwent Hypospadias Repair: a Comparative Study Using Probiotics vs Placebo.

Esposito C., Roberti A., Turra F., Cerulo M., Severino G., Settimi A., Escolino M.

Embase

Probiotics and Antimicrobial Proteins. 10 (2) (pp 323-328), 2018. Date of Publication: 01 Jun 2018.

[Article]

AN: 618186818

This study aimed to evaluate the effectiveness of probiotics (*Lactobacillus rhamnosus* GG), as a preventive measure of antibiotic-associated diarrhea (AAD) in children who underwent hypospadias repair and its clinical consequences on postoperative outcome, comparing the group treated with probiotics + antibiotics with two control groups (only antibiotics and antibiotics + placebo). We performed a prospective, randomized, placebo-controlled study with three groups of patients (30 boys for each group) who underwent hypospadias repair in our unit from March 2016 to December 2016. G1 received antibiotics + probiotics (*L. rhamnosus* GG), while G2 and G3 respectively received only antibiotics or antibiotics + placebo (glucose solution at 5%) for the same period. The patients were evaluated in regard to the number of evacuations/day, stool consistency, and the number of dressings/day. The overall incidence of postoperative AAD was 33.3% (30/90), and it was statistically lower in G1 patients compared to G2 and G3 ones ($p = 0.002$). The duration of AAD was significantly longer in G2 and G3 compared to G1 ($p = 0.001$). In G1, the frequency of dressing change was significantly lower compared to G2 and G3 ($p = 0.001$). The incidence of postoperative complications (fistula and dehiscence) was significantly higher in G2 and G3 compared to G1 ($p = 0.001$). Our study confirmed that the use of probiotic *L. rhamnosus* GG associated with antibiotics significantly reduced the incidence and the duration of postoperative AAD. In addition, the use of probiotics LGG reduced the frequency of dressing changes and the incidence of postoperative complications, such as urethral fistula and foreskin dehiscence.

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Status

Embase

Institution

(Esposito, Roberti, Turra, Cerulo, Severino, Settimi, Escolino) Pediatric Surgery Unit, Department of Translational Medical Sciences, Federico II University of Naples, Via Pansini 5, Naples 80131, Italy

Publisher

Springer New York LLC (E-mail: barbara.b.bertram@gsk.com)

Year of Publication

2018

367.

Interet de la technique de Koyanagi dans le traitement de l'hypospadias posterieur chez l'enfant.
Youssef S.B., Ksia A., Fredj M.B., Messaoud M., Laamiri R., Belhassen S., Mosbahi S.,
Bouzzaffara B., Sahnoun L., Mekki M., Belguith M., Nouri A.

Embase

African Journal of Urology. 24 (4) (pp 331-335), 2018. Date of Publication: December 2018.

[Article]

AN: 2001132974

Introduction: The koyanagi technique because of its satisfying results and less risk of postoperative complications for the patient has generated an increasing interest during recent years.

Objective(s): To report the indications and the results of the koyanagi technique. Patients an methods: This is a descriptive retrospective study of patients admitted in the pediatric surgery's department of the EPS Monastir, Tunisia for proximal hypospadias and operated on according to Koyanagi's technique during the period from 02 january 2006 to 30 june 2013, with subsequent follow-up of at least 02 years.

Result(s): The data of total 35 patients were collected. The average age at the first consultation in pediatric surgery was 12.2 months and the age at surgery was 28.8 months with extremes ranging from 12-55 months. The mean operative time was 166.1 min. The average stay at

hospital was 7.9 days. Only 4 patients (11.4%) presented immediate complications. Reoperation was indicated for 23 cases and indications were by decreasing order of frequency: dehiscence (34.3%), fistula (11.4%) and dehiscence with persistent penile-scrotal transposition (2.9%). The final clinical evaluation of the surgery found that meatus was distal in the majority of cases (85.7%), two patients kept a residual curvature estimated as being minimal and not requiring any further surgery and none of our patients presented abnormalities of the urinary stream.

Conclusion(s): Koyanagi's technique in repairing proximal hypospadias seems to be an excellent option especially in cases of severe curvature but with a high rate of complications.

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Embase

Institution

(Youssef, Ksia, Fredj, Messaoud, Laamiri, Belhassen, Mosbahi, Bouzzaffara, Sahnoun, Mekki, Belguith, Nouri) Service de Chirurgie Pédiatrique, EPS Fattouma Bourguiba, Monastir, 5000, Tunisie; Laboratoire de recherche LR12SP13

Publisher

Pan African Urological Surgeons Association(PAUSA) (E-mail: sunnydoodu@yahoo.com)

Year of Publication

2018

368.

Can sacral development as a marker for caudal regression help identify associated urologic anomalies in patients with anorectal malformation?.

Vilanova-Sanchez A., Reck C.A., Sebastiao Y.V., Fuchs M., Halleran D.R., Weaver L., Gregory Bates D., Gasior A.C., Maloof T., Hoover E.J., Jaggars J., Gagnon R., Ching C.C., Dajusta D., Jayanthi V.R., Levitt M.A., Wood R.J.

Embase

Journal of Pediatric Surgery. 53 (11) (pp 2178-2182), 2018. Date of Publication: November 2018.

[Article]

AN: 2000673797

Background and aim: The sacral ratio (SR) is a well-established tool to quantify sacral development in patients with anorectal malformations (ARM) and can be used as a predictor of

fecal continence. We hypothesized that a lower SR correlated with the presence of urologic and renal malformations.

Method(s): We retrospectively reviewed the medical records of patients with ARM treated at our center from 2014 to 2016. We measured the lateral SR as a marker for sacral development and assessed the spine for the presence of tethered cord (TC). Urological and renal anomalies, including single kidney, hydronephrosis, hypospadias, vesicoureteral reflux (VUR), ectopic ureter, and penoscrotal transposition were assessed. Analysis of variance (ANOVA), t-tests, and multivariable linear regression were used to test for differences in SR with consideration of associated urologic malformations and tethered cord.

Result(s): 283 patients with ARM were included for analysis (156 females). The median age was 39 months (10-90). Among these, 178 (55.6%) had 1 or more urologic malformations, and 81 (25.3%) had a TC. Hydronephrosis, high-grade VUR (3-5), solitary kidney, and tethered cord were significantly associated with lower SR ($p < 0.01$). In multivariable regression models, the presence of urologic abnormalities remained significantly associated with lower a SR despite the presence or absence of TC ($p < 0.001$).

Conclusion(s): SR is a potentially useful indicator of certain urologic anomalies including hydronephrosis, high grade VUR, and solitary kidney in patients with ARM. This association is independent of the presence of TC. A sacral ratio as a part of the VACTERL screening can help the surgeon identify which patients need closer urologic follow up.

Level of Evidence: IV

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Status

Embase

Institution

(Vilanova-Sanchez, Reck, Halleran, Weaver, Gasior, Maloof, Hoover, Jagers, Gagnon, Levitt, Wood) Center for Colorectal and Pelvic Reconstruction, Nationwide Children's Hospital, Columbus, OH, United States (Sebastiao) Center for Surgical Outcomes Research, Center for Innovation in Pediatric Practice, Nationwide Children's Hospital, Columbus, OH, United States (Fuchs, Ching, Dajusta, Jayanthi) Pediatric Urology Department, Nationwide Children's Hospital, Columbus, OH, United States

(Gregory Bates) Children's Radiological Institute, The Ohio State College of Medicine and Public Health, Columbus, OH, United States

Publisher

W.B. Saunders

Year of Publication

369.

Ultrasound-guided Pudendal Block Is a Viable Alternative to Caudal Block for Hypospadias Surgery: A Single-Surgeon Pilot Study.

Hecht S., Pineda J., Bayne A.

Embase

Urology. 113 (pp 192-196), 2018. Date of Publication: March 2018.

[Article]

AN: 2000555976

Objective: To evaluate pudendal nerve block as an alternative to caudal block for hypospadias surgery.

Method(s): Data were obtained by chart review. Children who underwent hypospadias repair between 2012 and 2016 by a single surgeon at an academic institution were included. Patients received ultrasound-guided pudendal block (n = 21) or caudal block (n = 19) as a regional adjunct to general anesthesia. Primary outcomes included analgesic requirement and postoperative length of stay in the recovery unit.

Result(s): The pudendal block cohort was slightly older (27.6 vs 18.5 months, $P = .017$) and had more severe hypospadias than the caudal block cohort (53% vs 35% proximal hypospadias, respectively). We detected no statistically significant difference in intraoperative opioid, postoperative opioid, or nonopioid analgesic requirement (17.9 vs 12.9 mcg fentanyl, $P = .267$; 0.3 vs 0.3 doses, $P = .92$; 0.2 vs 0.1 doses, $P = .46$, respectively). Postoperative length of stay was significantly shorter in the pudendal block cohort (96 vs 128 minutes, $P = .016$).

Discussion(s): We are the first to report the use of ultrasound-guided pudendal block for hypospadias repair. This appears to be a safe and effective alternative to caudal block with no perioperative delays. Pudendal block has several advantages over caudal block. It avoids the risks of urinary retention and lower extremity weakness and can be administered to older patients and children with spinal anomalies.

Conclusion(s): Compared with caudal block, ultrasound-guided pudendal nerve block is safe, provides equivalent pain control for hypospadias repair, and results in a shorter time to discharge.

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Status

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Author NameID

Hecht, Sarah; ORCID: <http://orcid.org/0000-0003-1104-1484>

Institution

(Hecht, Bayne) Division of Pediatric Urology, Doernbecher Children's Hospital, Portland, OR, United States (Pineda) Division of Pediatric Anesthesiology, Doernbecher Children's Hospital, Portland, OR, United States

Publisher

Elsevier Inc. (E-mail: usjcs@elsevier.com)

Year of Publication

2018

370.

Achieving urinary continence in cloacal exstrophy: The surgical cost.

Maruf M., Kasprenski M., Jayman J., Goldstein S.D., Benz K., Baumgartner T., Gearhart J.P.

Embase

Journal of Pediatric Surgery. 53 (10) (pp 1937-1941), 2018. Date of Publication: October 2018.

[Article]

AN: 2000546662

Introduction: Cloacal exstrophy (CE) is a severe midline congenital abnormality that requires numerous surgical corrections to achieve an acceptable quality of life. Candidates for urinary continence undergo multiple procedures, most often continent bladder diversions, to become socially dry. Here, the authors investigate the number of genitourinary interventions that patients with CE undergo to attain urinary continence.

Material(s) and Method(s): A retrospective review of a prospectively maintained database of 1311 exstrophy epispadias complex patients was performed. Patients with CE who have had at least one continence procedure were included. A continence procedure was defined as bladder neck reconstruction with or without augmentation, bladder neck transection with continent urinary diversion, augmentation cystoplasty, or use of injectable bulking agents. Continence was defined as a dry interval greater than 3 hours without leakage at night.

Result(s): In total, 140 CE and CE variant patients have been managed at the authors' institution. Of the 116 CE patients, 59 received at least one continence procedure, 14 were excluded for incontinent diversion or cystectomy, and the remaining 43 patients are awaiting a continence procedure. At the time of analysis, 42 (71%) patients who underwent a continence procedure were dry. The median number of total urologic procedures to reach urinary continence was 4 (range 2-10). This included 1 bladder closure (range 1-3), 2 urinary continence procedures (range 1-4), and 1 (range 0-4) "other" genitourinary procedures. The median time to urinary continence was 11.0 years (95% CI [9.2-14.2]).

Conclusion(s): A majority of CE patients who undergo a diversion procedure can achieve urinary continence. However multiple continence procedures are likely necessary. Of patients who are candidates for a continence procedure, half will be continent by the age of 11.

Level of Evidence: Level IV, Case series with no comparison group.

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Embase

Institution

(Maruf, Kasprenski, Jayman, Benz, Baumgartner, Gearhart) Robert D. Jeffs Division of Pediatric Urology, The James Buchanan Brady Urological Institutions, The Johns Hopkins Hospital, Johns Hopkins Medical Institutions, Charlotte Bloomberg Children's Hospital, Baltimore, MD, United States (Goldstein) Division of General Pediatric Surgery, Department of Surgery, The Johns Hopkins Hospital, Baltimore, MD, United States

Publisher

W.B. Saunders

Year of Publication

2018

371.

Molecular and clinical analyses of two patients with UPD(16)mat detected by screening 94 patients with Silver-Russell syndrome phenotype of unknown aetiology.

Inoue T., Yagasaki H., Nishioka J., Nakamura A., Matsubara K., Narumi S., Nakabayashi K., Yamazawa K., Fuke T., Oka A., Ogata T., Fukami M., Kagami M.

Embase

Journal of Medical Genetics. (no pagination), 2018. Date of Publication: 2018.

[Article In Press]

AN: 624045587

Background: Recently, a patient with maternal uniparental disomy of chromosome 16 (UPD(16)mat) presenting with Silver-Russell syndrome (SRS) phenotype was reported. SRS is characterised by growth failure and dysmorphic features.

Objective(s): To clarify the prevalence of UPD(16)mat in aetiology-unknown patients with SRS phenotype and phenotypic differences between UPD(16)mat and SRS.

Method(s): We studied 94 patients with SRS phenotype of unknown aetiology. Sixty-three satisfied the Netchine-Harbison clinical scoring system (NH-CSS) criteria, and 25 out of 63 patients showed both protruding forehead and relative macrocephaly (clinical SRS). The remaining 31 patients met only three NH-CSS criteria, but were clinically suspected as having SRS. To detect UPD(16)mat, we performed methylation analysis for the ZNF597:TSS-differentially methylated region (DMR) on chromosome 16 and subsequently performed microsatellite, SNP array and exome analyses in the patients with hypomethylated ZNF597:TSS-DMR.

Result(s): We identified two patients (2.1%) with a mixture of maternal isodisomy and heterodisomy of chromosome 16 in 94 aetiology-unknown patients with SRS phenotype. Both patients exhibited preterm birth and prenatal and postnatal growth failure. The male patient had ventricular septal defect and hypospadias. Whole-exome sequencing detected no gene mutations related to their phenotypes.

Conclusion(s): We suggest considering genetic testing for UPD(16)mat in SRS phenotypic patients without known aetiology.

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Article-in-Press

Institution

(Inoue, Nakamura, Matsubara, Narumi, Yamazawa, Fuke, Ogata, Fukami, Kagami) Department of Molecular Endocrinology, National Research Institute for Child Health and Development, Tokyo 157-8535, Japan (Inoue, Oka) Department of Pediatrics, University of Tokyo, Tokyo, Japan (Yagasaki) Department of Pediatrics, Faculty of Medicine, University of Yamanashi, Chuo, Japan

(Nishioka) Department of Pediatrics and Child Health, Kurume University School of Medicine, Kurume, Japan

(Nakamura) Department of Pediatrics, Hokkaido University Graduate School of Medicine, Sapporo, Japan

(Nakabayashi) Department of Maternal-Fetal Biology, National Research Institute for Child Health and Development, Tokyo, Japan

(Ogata) Department of Pediatrics, Hamamatsu University School of Medicine, Hamamatsu, Japan

Publisher

BMJ Publishing Group (E-mail: subscriptions@bmjgroup.com)

Year of Publication

2018

372.

Modeled exposure to tetrachloroethylene-contaminated drinking water and the occurrence of birth defects: A case-control study from Massachusetts and Rhode Island.

Aschengrau A., Gallagher L.G., Winter M., Butler L., Patricia Fabian M., Vieira V.M.

Embase

Environmental Health: A Global Access Science Source. 17 (1) (no pagination), 2018. Article Number: 75. Date of Publication: 06 Nov 2018.

[Article]

AN: 624753266

Background: Residents of Massachusetts and Rhode Island were exposed to tetrachloroethylene-contaminated drinking water from 1968 through the early 1990s when it leached from the vinyl lining of asbestos cement water distribution pipes. While occupational exposure to solvents during pregnancy has consistently been linked to an increased risk of certain birth defects, mixed results have been observed for environmental sources of exposure, including contaminated drinking water. The present case-control study was undertaken to examine further the association between prenatal exposure to tetrachloroethylene-contaminated drinking water and the risk of central nervous system defects, oral clefts and hypospadias.

Method(s): Cases were comprised of live- and stillborn infants delivered between 1968 and 1995 to mothers who resided in 28 Massachusetts and Rhode Island cities and towns with some PCE-

contaminated water supplies. Infants with central nervous system defects (N = 268), oral clefts (N = 112) and hypospadias (N = 94) were included. Controls were randomly selected live-born, non-malformed infants who were delivered during the same period and geographic area as cases (N = 771). Vital records and self-administered questionnaires were used to gather identifying information, birth defect diagnoses, and other relevant data. PCE exposure during the first trimester was estimated using water distribution system modeling software that incorporated a leaching and transport model. Prenatal PCE exposure was dichotomized as "high" or "low" exposure at the level corresponding to an estimated average concentration of 40 mug/L, the criterion for remediation when PCE contamination was discovered in 1980.

Result(s): Mothers with "high" levels of exposure to PCE-contaminated drinking water during the first trimester (> 40 mug/L) had increased odds of having a child with spina bifida (OR: 2.0, 95% CI: 0.8-5.4), cleft lip with or without cleft palate (OR: 3.8, 95% CI: 1.2-12.3) and hypospadias (OR: 2.1, 95% CI: 0.5-8.3). No increases in the odds of other defects were observed in relation to "high" exposure levels.

Conclusion(s): The results of the present study suggest that mothers with "high" PCE exposure levels during the first trimester have increased odds of having a child with spina bifida, cleft lip with or without cleft palate, and hypospadias. These findings support several prior studies that observed an increased risk of selected birth defects following prenatal exposure to solvents in occupational and environmental settings. Even though PCE contamination from vinyl lined pipes was remediated many years ago, it remains a widespread contaminant across the U.S and so environmental regulations must be guided by a precautionary perspective that safeguards pregnant women and their offspring.

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Status

Embase

Author NameID

Aschengrau, Ann; ORCID: <http://orcid.org/0000-0001-8153-7712>

Institution

(Aschengrau, Gallagher) Department of Epidemiology, Boston University, School of Public Health, 715 Albany Street, Talbot 3 East, Boston, MA 02118, United States (Winter) Biostatistics and Epidemiology Data Analytics Center, Boston University, School of Public Health, 715 Albany Street, Fuller-9, Boston, MA 02118, United States

(Butler, Patricia Fabian) Department of Environmental Health, Boston University, School of Public Health, 715 Albany Street, Talbot 4 West, Boston, MA 02118, United States

(Vieira) University of California, Irvine, Program in Public Health, 653 East Peltason Drive, Irvine, CA 92697, United States

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Year of Publication

2018

373.

Clinical features of children with multicystic dysplastic kidney.

Kara A., Gurgoze M.K., Aydin M., Koc Z.P.

Embase

Pediatrics International. 60 (8) (pp 750-754), 2018. Date of Publication: August 2018.

[Article]

AN: 623724109

Background: To evaluate the clinical features of patients with multicystic dysplastic kidney (MCDK).

Method(s): The medical files of children diagnosed with MCDK between January 2008 and November 2015 were retrospectively reviewed. The demographic, clinical, laboratory and radiological data were evaluated.

Result(s): Of 128 children with MCDK enrolled in the study, 82 (64.1%) were male, and 46 (35.9%) were female ($P < 0.05$). MCDK were located on left and right sides in 66 (51.6%) and 62 children (48.4%), respectively ($P > 0.05$). Antenatal diagnosis was present in 64 patients (50%). The mean age at diagnosis was 2.8 ± 2.7 years (range, 0-8 years), and follow-up duration was 4.5 years. Fifteen patients (20.8%) had vesicoureteral reflux. Of these, four underwent endoscopic surgical correction. Other associated urological anomalies were ureteropelvic junction obstruction ($n = 6$), hypospadias ($n = 1$), and kidney stones ($n = 1$). On technetium-99 m dimercaptosuccinic acid scintigraphy, which was performed in all patients, no significant association between grade of reflux and presence of scarring was seen. Hypertension was diagnosed only in one child (0.8%) who required antihypertensive treatment. The prevalence of unilateral undescended testicle in children aged <1 year in the 82 male patients was 4.9%. Seventy-six patients (59.4%) developed compensatory hypertrophy in the contralateral kidney

during a 1 year follow-up period. Of the total, only seven children (5.5%) had undergone nephrectomy.

Conclusion(s): MCDK follows a benign course with relatively few sequelae, and therefore these patients should be closely followed up and conservatively managed.

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Embase

Author NameID

Kara, Aslihan; ORCID: <http://orcid.org/0000-0003-4410-0444>

Institution

(Kara, Gurgoze) Department of Pediatric Nephrology, Firat University School of Medicine, Elazig, Turkey (Aydin) Department of Neonatology, Firat University School of Medicine, Elazig, Turkey (Koc) Department of Nuclear Medicine, Firat University School of Medicine, Elazig, Turkey

Publisher

Blackwell Publishing

Year of Publication

2018

374.

Klinefelter syndrome in childhood: Variability in clinical and molecular findings.

Akcan N., Poyrazoglu S., Bas F., Bundak R., Darendeliler F.

Embase

JCRPE Journal of Clinical Research in Pediatric Endocrinology. 10 (2) (pp 100-107), 2018. Date of Publication: June 2018.

[Article]

AN: 622263853

Objective: Klinefelter syndrome (KS) is the most common (1/500-1/1000) chromosomal disorder in males, but only 10% of cases are identified in childhood. This study aimed to review the data of children with KS to assess the age and presenting symptoms for diagnosis, clinical and laboratory findings, together with the presence of comorbidities.

Method(s): Twenty-three KS patients were analyzed retrospectively. Age at admission, presenting symptoms, comorbid problems, height, weight, pubertal status, biochemical findings, hormone profiles, bone mineral density and karyotype were evaluated. Molecular analysis was also conducted in patients with ambiguous genitalia.

Result(s): The median age of patients at presentation was 3.0 (0.04-16.3) years. Most of the cases were diagnosed prenatally (n=15, 65.2%). Other reasons for admission were scrotal hypospadias (n=3, 14.3%), undescended testis (n=2, 9.5%), short stature (n=1, 4.8%), isolated micropenis (n=1, 4.8%) and a speech disorder (n=1, 4.8%). The most frequent clinical findings were neurocognitive disorders, speech impairment, social and behavioral problems and undescended testes. All except two patients were prepubertal at admission. Most of the patients (n=20, 86.9%) showed the classic 47,XXY karyotype. Steroid 5 alpha-reductase 2 gene and androgen receptor gene mutations were detected in two of the three cases with genital ambiguity.

Conclusion(s): Given the large number of underdiagnosed KS patients before adolescence, pediatricians need to be aware of the phenotypic variability of KS in childhood. Genetic analysis in KS patients may reveal mutations associated with other forms of disorders of sex development besides KS.

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Status

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Author NameID

Akcan, Nese; ORCID: <http://orcid.org/0000-0003-2583-5736>

Institution

(Akcan) Near East University Faculty of Medicine, Department of Pediatric Endocrinology, Nicosia, Cyprus (Poyrazoglu, Bas, Darendeliler) Istanbul University Istanbul Faculty of Medicine, Department of Pediatric Endocrinology, Istanbul, Turkey

(Bundak) University of Kyrenia Faculty of Medicine, Department of Pediatric Endocrinology, Kyrenia, Cyprus

Publisher

Galenos Yayincilik, (Molla Gurani Cad. 21, Findikzade-Istanul 34093, Turkey)

Year of Publication

2018

375.

Different managements for prepubertal epididymitis based on a preexisting genitourinary anomaly diagnosis.

Lee Y.S., Kim S.W., Han S.W.

Embase

PLoS ONE. 13 (4) (no pagination), 2018. Article Number: e0194761. Date of Publication: April 2018.

[Article]

AN: 621718307

There is no clear consensus regarding investigating for accompanying genitourinary anomalies (GUAs) in patients with prepubertal acute epididymitis (AE). Moreover, risk factors for the recurrence and the need for a surgical intervention have never been discussed. The purpose of this study was to evaluate the different clinical courses of prepubertal AE based on knowledge of preexisting GUAs. Between January 2005 and December 2014, AE was diagnosed in 189 pediatric patients <10 years old. Clinical characteristics and treatments were retrospectively analyzed. The median age at first AE was 64.3 months. A GUA was detected prior to the development of AE in 49 patients (known GUA group) including 34 with hypospadias. Among the other 140 patients (unknown GUA status group), six patients were diagnosed with a GUA after the first AE episode. In the known GUA group, 35 patients (71.4%) experienced recurrence and the only risk factor associated with recurrence was the presence of cystic dilated prostatic utricle ($p = 0.013$). In the unknown GUA status group, the risk factors for an existing GUA were being <1-year-old ($p < 0.001$) and positive urine culture ($p = 0.015$). Only nine patients (6.4%) in this group experienced recurrence. Vasectomy was recommended for patients with recurrent AE with an accompanying GUA and performed in 19 patients (10.1%). Most GUAs are diagnosed prior to AE development. Clinicians should consider different treatment approaches based on whether the AE patient has been diagnosed with a GUA previously, because the clinical characteristics and the recurrence rate are significantly different.

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Status

Embase

Institution

(Lee, Kim, Han) Department of Urology, Urological Science Institute, Yonsei University College of Medicine, Seoul, South Korea

Publisher

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Year of Publication

2018

376.

Multicentre cross-sectional clinical evaluation study about quality of life in adults with disorders/differences of sex development (DSD) compared to country specific reference populations (dsd-LIFE).

Rapp M., Mueller-Godeffroy E., Lee P., Roehle R., Kreukels B.P.C., Kohler B., Nordenstrom A., Bouvattier C., Thyen U.

Embase

Health and Quality of Life Outcomes. 16 (1) (no pagination), 2018. Article Number: 54. Date of Publication: 03 Apr 2018.

[Article]

AN: 621506829

Background: Previous studies in quality of life (QOL) in individuals with disorders/differences of sex development (DSD) have been restricted to subpopulations of the condition. We describe QOL in adult persons with DSD compared to country specific references and assess the impact of diagnosis.

Method(s): The multicentre cross-sectional clinical evaluation (dsd-LIFE) took place in 14 specialized clinics in six European countries. Adolescents (≥ 16 years) and adults having a DSD condition were included from 02/2014 to 09/2015. The main outcome QOL was measured by the WHOQOL-BREF (domains of physical health, psychological, social relationships, and environment). QOL was compared to country specific reference populations by using unpaired t-tests. Linear regression models explained the additional variance of the diagnosis on QOL.

Result(s): Three hundred one individuals with Turner Syndrome, 219 with Klinefelter Syndrome (including XYY), 226 with 46,XX CAH and 294 with rare DSD conditions (gonadal dysgenesis, androgen insensitivity syndrome, severe hypospadias, and androgen synthesis errors or other diagnosis) took part. Compared to healthy European populations, QOL was similar in

psychological, slightly worse in physical health, and slightly better in environment. In social relationships, QOL was significantly poorer compared to healthy and non-healthy reference populations. In linear regression models health status was the most important predictor of QOL; additional variance was explained by feelings about household's income in all domains, and the relationship status in social relationships. Diagnosis explained nearly no additional variance.

Conclusion(s): Except for social relationships, most people with DSD adapt well to their life circumstances and report a good QOL. Not diagnosis, but the individual's health status is much more important than previously thought. Therefore care for people with DSD should focus more on chronic physical or mental health problems both related and unrelated to the diagnosis itself.

Trial registration: German Clinical Trials Register DRKS00006072.

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Embase

Institution

(Rapp, Mueller-Godeffroy, Thyen) Universitat zu Lubeck, Klinik fur Kinder- und Jugendmedizin, Ratzeburger Allee 160, Lubeck 23538, Germany (Lee) Pediatrics, Penn State College of Medicine, Hershey, PA 17033, United States

(Roehle) Charite, Campus Virchow-Klinikum, Koordinierungszentrum fur Klinische Studien (KKS Charite), Berlin 13353, Germany

(Kreukels) VU Medisch Centrum, Medische Psychologie en Medisch Maatschappelijk Werk, PO Box 7057, Amsterdam, MB 1007, Netherlands

(Kohler) Charite, Campus Virchow-Klinikum, Klinik fur Padiatrie mit Schwerpunkt Endokrinologie und Diabetologie, Berlin 13353, Germany

(Nordenstrom) Karolinska University Hospital, Department of Women's and Children's Health, Karolinska Institutet, Stockholm 171 76, Sweden

(Bouvattier) Universite Paris-Sud, Endocrinologie pediatrique, Centre de reference des maladies rares du developpement sexuel, Hopital Bicetre, 78 rue du General Leclerc, Le Kremlin Bicetre 94270, France

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Year of Publication

2018

377.

The aberrant urethral meatus as a possible aetiological factor of recurrent post-coital urinary infections in young women.

Gyftopoulos K.

Embase

Medical Hypotheses. 113 (pp 6-8), 2018. Date of Publication: April 2018.

[Article]

AN: 620592278

Recurrent urinary tract infections (rUTI) in women is a common cause of morbidity worldwide. In young women recurrent cystitis associated with sexual intercourse is quite common; however not all sexually active women will suffer a post-coital UTI episode. A variety of possible predisposing factors has been described, including behavioral risk patterns, genetic factors and increased virulence of certain subtypes of uropathogens. It has long been regarded that anatomical anomalies are rare in this setting of rUTIs. The shorter distance of the urethra to the anus has been the only anatomical risk factor described that is marginally significant in predisposing women for rUTIs. We hereby present a hypothesis that focuses on the shorter distance of the urethral meatus to the vaginal opening as the mainstay of sexually-induced rUTIs. In this theory of "functional hypospadias" the low misplacement of the urethral opening allows for easier advancement of pathogen-laden mucous by penile thrust and increased friction during intercourse. Our hypothesis suggests that a shorter distance of the urethral meatus to the vaginal opening (and not the anus itself) is an anatomical risk factor for recurrent post-coital cystitis. Verification of this hypothesis might lead to a more patient-oriented approach by alerting clinicians in looking for an aberrant meatus and save the patient from unnecessary imaging and endoscopic examinations often used in these cases. Moreover it may also aid the patient in understanding her anatomy and modify behavioral risk practices.

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Status

Embase

Institution

(Gyftopoulos) Department of Anatomy University of Patras Medical School and Olympion Hospital, Patras, Greece

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Churchill Livingstone

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378.

Risk factors for failed urethrocutaneous fistula repair after transverse preputial island flap urethroplasty in pediatric hypospadias.

Han W., Zhang W., Sun N.

Embase

International Urology and Nephrology. 50 (2) (pp 191-195), 2018. Date of Publication: 01 Feb 2018.

[Article]

AN: 620057031

Objectives: We review our experience in urethrocutaneous fistula (UCF) repair after transverse preputial island flap urethroplasty (Duckett) in pediatric hypospadias to investigate the risk factors for unsuccessful outcome.

Method(s): Two hundred and nineteen UCFs repairs conducted in our center from January 2015 to December 2016 after Duckett technique were retrospectively reviewed and 182 had a follow-up for more than 6 months. The age of patients, size, localization and numbers of UCFs, interval time of two operations, length of neourethra, times of UCFs repairs, complications other than UCFs, hospital of primary surgery and postoperative infection were analyzed as potential risk factors. Stratified analysis and assessment of additive interaction were performed to have a better understanding of the relation between the risk factors. Logistic regression analysis was used for multivariate analysis.

Result(s): UCF repairs were succeeded in 121 (66.5%) and failed in 61 (33.5%) at first operation. In the univariate analysis, size and numbers of UCFs ($P = 0.01$ and $P = 0.035$, respectively), interval time of two operations ($P = 0.042$) and hospital of primary surgery ($P = 0.02$) were statistically related with the outcome. In the multivariate analysis, UCF ≥ 2 (OR 2.71, 95% CI 1.095-6.692, $P = 0.031$), size ≥ 2 mm (OR 2.45, 95% CI 1.267-4.757, $P = 0.08$) and primary surgery not at our hospital (OR 2.21, 95% CI 1.094-4.126, $P = 0.026$) were identified as independent risk factors for unsuccessful outcome. In the stratified analysis, multiplicative interaction between the 3 risk factors was not found.

Conclusion(s): Our study suggested that UCF repairs after transverse preputial island flap urethroplasty were easier to fail if the UCF was ≥ 2 mm, the numbers of UCFs was ≥ 2 , or the surgeon had less experience. The age of patients, site of UCFs, interval time of two operations, length of neourethra, times of UCFs repairs, complications other than UCF and postoperative infection were not significantly related to the success rate of UCF repair.

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Status

Embase

Institution

(Han, Zhang, Sun) Department of Urology, Beijing Children's Hospital Affiliated to Capital Medical University, No. 56 Nanlishi Road, Xicheng District, Beijing 100045, China

Publisher

Springer Netherlands (E-mail: rbk@louisiana.edu)

Year of Publication

2018

379.

Optimisation of sonourethrography: the clamp method.

Berna-Mestre J.D., Balmaceda T., Martinez D., Escudero J.F., Martinez G., Garcia J.A., Canteras M., Berna-Serna J.D.

Embase

European Radiology. 28 (5) (pp 1961-1968), 2018. Date of Publication: 01 May 2018.

[Article]

AN: 619763570

Purpose: To describe the clamp method for performing retrograde sonourethrography (RSUG) and contrast-enhanced voiding sonourethrography (CE-VSUG) via the transperineal approach in male adults.

Material(s) and Method(s): Prospective study of 113 males (14-86 years) with urethral strictures confirmed by urethrography who received sonourethrography via the clamp method between

2011 and 2015. The characteristic parameters of the quantitative variables were calculated and a comparative analysis of the qualitative variables was conducted using the McNemar test.

Result(s): RSUG was performed successfully in all the cases ($n = 113$) and detected 49 cases with anterior urethral strictures; the strictures in the proximal bulbar cone in five of them (10.2%) were not visualised on retrograde urethrography (RUG) ($p < 0.05$). CE-VSUG was performed successfully in 97 cases and observed posterior urethral strictures in 82; the bladder neck strictures in 6 of them (7.3%) were not observed on voiding cystourethrography (VCUG) ($p < 0.05$). Retrograde bladder filling was achieved in approximately 6 min.

Conclusion(s): The clamp method enables RSUG and CE-VSUG to be performed simply, effectively and painlessly by a single operator. It also allows the evaluation of cases with urethromeatal alterations (stricture, hypospadias and meatotomy). Key Points: * The clamp method enables RSUG to be performed simply and painlessly. * The clamp method requires only one operator and allows assessing urethromeatal alterations. * RSUG shows greater capacity for detecting anterior urethral strictures than RUG. * The clamp method achieves retrograde bladder filling in approximately 6 min. * CE-VSUG shows greater capacity for detecting strictures than VCUG.

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Status

Embase

Author NameID

Berna-Mestre, Juan de Dios; ORCID: <http://orcid.org/0000-0001-9990-2386>

Institution

(Berna-Mestre, Balmaceda, Martinez, Berna-Serna) Department of Radiology, Virgen de la Arrixaca University Hospital, Ctra. Madrid-Cartagena, s/n, El Palmar, Murcia 30120, Spain

(Escudero, Martinez, Garcia) Department of Urology, Virgen de la Arrixaca University Hospital, Ctra. Madrid-Cartagena, s/n, El Palmar, Murcia 30120, Spain

(Canteras) Department of Statistics, University of Murcia, Faculty of Medicine, Campus de Espinardo, Murcia, Spain

Publisher

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Year of Publication

2018

380.

Sir Denis Browne, the father of modern pediatric surgery.

Nakayama D.K.

Embase

Journal of Pediatric Surgery. 53 (3) (pp 576-579), 2018. Date of Publication: March 2018.

[Review]

AN: 619431444

Sir Denis John Wolko Browne (1892-1967), while not the first in the British Isles to devote his entire surgical practice to pediatric surgery, is accepted as "the father of pediatric surgery in the United Kingdom." He made contributions to operations as varied as tonsillectomy, pyloromyotomy, and hypospadias repair, and provided fundamental insights into the proper treatment of club foot, congenital dislocation of the hip, and cryptorchidism. He introduced the transverse laparotomy incision, primary repair of congenital intestinal obstruction, and the end-to-back anastomosis for intestinal atresia, techniques so commonly used that it is difficult to think of pediatric surgical operations done any differently. In addition, he invented the elegant Denis Browne retractor that remains in use today, one of the few eponymic instruments known by its originator's first and last name. He was among the founders of the British Association of Pediatric Surgeons, one of the first professional organizations in the field, and served as its first president. His legendary status was enhanced by an acerbic temperament that often surfaced in an outspoken and uncompromising advocacy on the behalf of the proper care of children. A larger-than-life figure in pediatric surgery, Browne's legacy is so wide-ranging and enduring that his unofficial title has been broadened to "the father of modern pediatric surgery."

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Status

Embase

Institution

(Nakayama) Department of Surgery, Florida International University, Sacred Heart Medical Group, Pensacola, FL 32504, United States

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W.B. Saunders

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2018

381.

Autophagy, Apoptosis, and Cell Proliferation in Exstrophy-Epispadias Complex.

Shabaninia M., Tourchi A., Di Carlo H., Gearhart J.P.

Embase

Urology. 111 (pp 157-161), 2018. Date of Publication: January 2018.

[Article]

AN: 619365066

Objective To investigate the state of autophagy and its interactions with apoptosis and cell proliferation in patients who underwent successful early closure or delayed closure of exstrophy. They compared those outcomes with cell culture samples from patients with vesicoureteral reflux as control. Patients and Methods Primary cultures of bladder smooth muscle cells (SMCs) were established from patients with successful neonatal bladder closure (group 1, N = 5), delayed closure because of small bladder template (group 2, N = 5), and vesicoureteral reflux as control (group 3, N = 5). The myogenicity of the cultures was determined using anti-Desmin antibody. Immunostainings for LC3 to assess autophagy and Ki67 to assess cell proliferation were applied. Apoptosis was assessed by the terminal deoxynucleotidyl transferase-mediated dUTP digoxigenin nick-end labeling assay. Results Autophagy marker (LC3) expression was significantly higher in the delayed closure group than in the other groups, whereas no significant difference was noted between the neonatal closure and the control groups. Apoptotic indices of the SMCs were remarkably higher in SMC cultures from the delayed closures than in the neonatal closure and the control groups. A significantly lower expression of proliferation marker (Ki67) in the delayed closure group compared with the control and the neonatal closure group was also of note. Conclusion Patients with small bladder template and delayed closure showed upregulated autophagic process and increased apoptotic indices while experiencing a dramatic decrease in the proliferation of their bladder SMCs. Finally, the concept of manipulating autophagy may lead to promising outcomes for patients with bladder exstrophy in the future.

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Status

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Author NameID

Gearhart, John P.; ORCID: <http://orcid.org/0000-0002-0177-6121>

Institution

(Shabaninia, Turchi, Di Carlo, Gearhart) Division of Pediatric Urology, James Buchanan Brady Urological Institute, The Johns Hopkins University School of Medicine, Baltimore, MD, United States

Publisher

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Year of Publication

2018

382.

Long-term Results of Ventral Penile Curvature Repair in Childhood.

Golomb D., Sivan B., Livne P.M., Nevo A., Ben-Meir D.

Embase

Urology. 112 (pp 161-163), 2018. Date of Publication: February 2018.

[Article]

AN: 619361367

Objective: To assess the postpubertal outcome of ventral penile curvature repaired in infancy in terms of recurrence and aesthetics.

Material(s) and Method(s): Postpubertal patients treated for hypospadias and ventral penile curvature in infancy at a tertiary medical center were invited to undergo assessment of the quality of the repair. Findings were compared between patients with a straight penis after skin release and patients who required dorsal plication.

Result(s): The cohort included 27 patients of mean age 16.5 years who were reported with straight penis after surgery. Postpubertal curvature was found in 6 of 14 patients (43%) successfully treated by skin release and 10 of 13 patients (77%) who underwent dorsal plication ($P = .087$). Significant curvature (≥ 30 degrees) was found in 1 of 14 patients in the skin-release group and 4 of 13 in the dorsal plication group ($P = .16$). Rates of redo urethroplasty were 2 of 14 (14%) and 5 of 10 (50%), respectively. Patient satisfaction with the appearance of the penis did not differ significantly.

Conclusion(s): Ventral penile curvature repaired in infancy often recurs after puberty. The need for dorsal plication has a trend-level association with recurrence of penile curvature in puberty. It might also be related to the degree of postpubertal penile curvature and the need for redo urethroplasty. Procedure type does not affect patient satisfaction with the postpubertal appearance of the penis.

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Status

Embase

Institution

(Golomb) Department of Urology, Rabin Medical Center-Beilinson Hospital, Petach Tikva, Israel

(Golomb) Sackler Faculty of Medicine, Tel Aviv University, Tel Aviv, Israel

(Sivan, Livne, Nevo, Ben-Meir) Unit of Pediatric Urology, Schneider Children's Medical Center of Israel, Petach Tikva, Israel

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Year of Publication

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383.

Utilization of Postpenile Surgery Illustrated Healing Atlas: A Comparative Study.

Chua M.E., Ming J.M., Sauders M.A., Fernandez N., Hannick J.H., Abu Awazayed I., Odeh R.I., Bagli D.J., Koyle M.A., Farhat W.A.

Embase

Urology. 112 (pp 151-154), 2018. Date of Publication: February 2018.

[Article]

AN: 619170393

Objective: To determine the effect of the postpenile surgery illustrated healing atlas on caregivers' anxiety levels pre- and postoperatively, the frequency of family's postoperative communication with the urology care team, and the number of unplanned emergency room (ER) return for wound checks.

Method(s): A prospective cohort enrolled children who underwent penile surgeries (distal hypospadias repair and phalloplasty) with no concomitant procedures from December 2016 to June 2017. A 6-item short-form Spielberger State-Trait Anxiety Inventory (STAI) was used to determine baseline and postoperative anxiety levels of the caregivers. Two groups were created: caregivers who did not view the illustrated atlas vs caregivers who were shown the atlas. Baseline characteristics and demographics were compared, along with caregiver's pre- to postoperative anxiety level difference, frequency of postoperative communication, and number of unplanned ER return for wound checks.

Result(s): Fifty-four patients were enrolled with 27 families in each group. Baseline characteristics and demographics were comparable with no significant differences. The assessment of the pre- to postoperative anxiety levels in both groups showed no significant differences (median difference -5 [interquartile range -8 to -5] vs -5 [interquartile range -8 to -4], $P = .94$). Although no differences were noted for ER-return rates between the groups (18% vs 11.1%, $P = .704$), significantly less postoperative calls and e-mails were noted among families who received or viewed the postpenile surgery illustrated healing atlas (51.9% vs 11.1%, $P = .003$).

Conclusion(s): The utilization of a postpenile surgery illustrated healing atlas as part of the postoperative support provided to families was able to decrease postoperative calls and e-mails.

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Status

Embase

Institution

(Chua, Ming, Sauders, Fernandez, Hannick, Abu Awazayed, Odeh, Bagli, Koyle, Farhat) Division of Urology, The Hospital for Sick Children, Toronto, ON, Canada

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2018

Expression of Xenobiotic Biomarkers CYP1 Family in Preputial Tissue of Patients with Hypospadias and Phimosis and Its Association with DNA Methylation Level of SRD5A2 Minimal Promoter.

Ohsako S., Aiba T., Miyado M., Fukami M., Ogata T., Hayashi Y., Mizuno K., Kojima Y.

Embase

Archives of Environmental Contamination and Toxicology. 74 (2) (pp 240-247), 2018. Date of Publication: 01 Feb 2018.

[Article]

AN: 619021410

Several epidemiological studies have suggested that the incidence of male reproductive organ malformations, including hypospadias or cryptorchidism, has increased due to fetal-stage exposure to environmental pollutants. However, the association of chemical exposure with the expression of target regulatory genes in the tissues of patients has not yet been reported. Because experimental approaches or clinical trials in human studies are limited, especially those using fetal and/or infants, it is difficult to obtain clear physiological evidence of mechanisms underlying male reproductive malformations. Thus, the lack of physiological evidence makes this issue controversial. We analyzed preputial tissues from patients with hypospadias (n = 23) and phimosis (n = 16). The atypical CYP1 family genes, CYP1A1 and CYP1B1, are potential biomarkers of environmental chemical exposure. We then compared the expression levels of CYP1A1 and CYP1B1 between hypospadias and phimosis samples by quantitative RT-PCR analysis. The mRNA expression levels of SRD5A2 and AR also were measured, because the androgen-related genes involved in the onset of disorders of male reproductive system. A significantly higher CYP1B1 expression level and a lower AR expression level were observed in the hypospadias groups than in the phimosis group. Positive correlations ($P < 0.001$) between the mRNA expression levels of the CYP1 family and SRD5A2 were found in patients with hypospadias but not in those with phimosis. Moreover, the methylation levels of the four genes were determined by bisulfite genomic sequencing. Although the SRD5A2 promoter region showed moderate methylation, no methylation was detected in CYP1A1, CYP1B1, or AR. There was no significant difference in SRD5A2 promoter methylation level between hypospadias and phimosis patients. Negative correlations were found between the methylation level of SRD5A2, especially at the - 221 Sp1 site, and the CYP1 family mRNA expression levels (CYP1A1, $p = 0.002$; CYP1B1, $p = 0.007$) in hypospadias patients, but not in phimosis patients. The significant positive association of mRNA expression level and the negative association of methylation level of the SRD5A2 gene with the mRNA expression levels of CYP1 family genes in the preputial tissue seem to indicate the chemical exposure of patients with hypospadias.

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Status

Embase

Institution

(Ohsako, Aiba) Laboratory of Environmental Health Sciences, Center for Disease Biology and Integrative Medicine, Graduate School of Medicine, The University of Tokyo, Bunkyo-ku, Tokyo 113-0033, Japan (Miyado, Fukami) Department of Molecular Endocrinology and Metabolism, National Research Institute for Child Health and Development, Setagaya-ku, Tokyo 157-8535, Japan

(Ogata) Department of Pediatrics, Hamamatsu University School of Medicine, Hamamatsu, Shizuoka 431-3192, Japan

(Hayashi, Mizuno) Department of Nephro-Urology, Nagoya City University Graduate School of Medical Sciences, Nagoya 467-8601, Japan

(Kojima) Department of Urology, Fukushima Medical University School of Medicine, Fukushima 960-1295, Japan

Publisher

Springer New York LLC (E-mail: barbara.b.bertram@gsk.com)

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385.

Hypospadias, Intrauterine Growth Restriction, and Abnormalities of the Placenta.

Toufaily M.H., Roberts D.J., Westgate M.-N., Hunt A.-T., Holmes L.B.

Embase

Birth Defects Research. 110 (2) (pp 122-127), 2018. Date of Publication: 30 Jan 2018.

[Article]

AN: 617594913

Background: Hypospadias is more common among male infants with growth restriction, defined as a birth weight less than the 10th centile, than in infants with a normal birth weight. Intrauterine growth retardation (IUGR) has been associated, also, with abnormalities of the placenta, such as maternal vascular malperfusion. In a consecutive sample of newborn infants, the association between hypospadias, IUGR and abnormalities of the placenta could be analyzed.

Method(s): Affected infants were identified among 289,365 liveborn and stillborn infants in the Active Malformations Surveillance Program between 1972 and 2012. The four anatomic locations of the ectopic urethral opening, based on the recorded physical examination findings, were: (1) glandular; (2) subcoronal; (3) penile; (4) penoscrotal. Affected infants with associated malformations, a chromosome abnormality, teratogenic exposure, maternal diabetes mellitus, or multiple gestations were excluded.

Result(s): Three hundred sixteen affected infants were identified: 52.2% glandular, 11.7% subcoronal, 27.8% penile, and 8.2% penoscrotal. The highest frequency of IUGR (34.6%) was in the infants with the most severe hypospadias (penoscrotal). The 39 reports of placenta findings showed a high frequency of abnormalities.

Conclusion(s): An increased rate of occurrence of hypospadias and abnormalities of the placenta were present in infants with intrauterine growth restriction. The postulated cause of this association is a deficiency in the function of the placenta during weeks 10 to 14 of gestation when normal masculinization occurs due to an increase in the level of placental human chorionic gonadotropin and fetal testosterone. The cause of the placental deficiency has not been established. Birth Defects Research 110:122-127, 2018.© 2017 Wiley Periodicals, Inc.

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Status

Embase

Author NameID

Toufaily, M. Hassan; ORCID: <http://orcid.org/0000-0003-4264-1730> Holmes, Lewis B.; ORCID: <http://orcid.org/0000-0002-0259-4848>

Institution

(Toufaily, Westgate, Holmes) Active Malformations Surveillance Program, Department of Pediatric Newborn Medicine, Brigham and Women's Hospital, Harvard Medical School, Boston, MA, United States (Toufaily, Westgate, Holmes) Medical Genetics Unit, MassGeneral Hospital for Children, Harvard Medical School, Boston, MA, United States

(Roberts) Department of Pathology, Massachusetts General Hospital, and Harvard Medical School, Boston, MA, United States

(Hunt) Hunt Consulting Associates, Chapel Hill, NC, United States

(Holmes) Department of Pediatrics, Harvard Medical School, Boston, MA, United States

Publisher

John Wiley and Sons Inc. (P.O.Box 18667, Newark NJ 07191-8667, United States)

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386.

Cystectomy in the Pediatric Exstrophy Population: Indications and Outcomes.

Ko J.S., Lue K., Friedlander D., Baumgartner T., Stuhldreher P., DiCarlo H.N., Gearhart J.P.

Embase

Urology. 116 (pp 168-171), 2018. Date of Publication: June 2018.

[Article]

AN: 2000718185

Objective: To explore the long-term outcomes and indications for cystectomy in patients with bladder exstrophy. Although rare, cystectomy is the final surgical alternative to bladder repair among these patients with a poor quality bladder template.

Method(s): A prospectively maintained database of 1298 patients with exstrophy-epispadias complex was reviewed for patients who underwent cystectomy between 1970 and 2015 at the authors' institution. Demographic data, indication for cystectomy, surgical history, postoperative outcomes, and continence status were collected.

Result(s): Eighteen (6 male; 12 female) patients with exstrophy (15 classic bladder exstrophy; 2 bladder exstrophy variants; 1 cloacal exstrophy) underwent cystectomy at a median age of 3.8 years. Six patients (33.3%) underwent primary cystectomy without attempted bladder closure. Eight patients (44.4%) had a history of failed primary closure with loss of capacity or inadequate growth after closure. Four patients (22.2%) had successful primary closure but underwent cystectomy secondary to poor bladder compliance with declining renal function or poor bladder growth or quality. Urinary diversion included 6 cutaneous ureterostomies, 4 bowel conduits (1 ileal; 3 colon), 6 continent urinary diversions with ileosigmoid reservoir, and 1 ureterosigmoidostomy. Of 8 patients who underwent a continence procedure, all were dry at a median of 25.3 months after cystectomy.

Conclusion(s): Cystectomy was most commonly indicated in intrinsically diseased bladder templates that remained too small despite permitting time for interval growth. These bladders often were of poor quality and compliance and did not reduce into the pelvis on examination. It was, however, possible to achieve urinary continence in these patients with cystectomy and urinary diversion.

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Status

Embase

Author NameID

Gearhart, John P.; ORCID: <http://orcid.org/0000-0002-0177-6121>

Institution

(Ko, Lue, Friedlander, Baumgartner, Stuhldreher, DiCarlo, Gearhart) Division of Pediatric Urology, James Buchanan Brady Urological Institute, Charlotte Bloomberg Children's Hospital, The Johns Hopkins University School of Medicine, Baltimore, MD, United States

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Year of Publication

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387.

Does grafted tubularized incised plate improve the outcome after repair of primary distal hypospadias: A prospective randomized study?.

Helmy T.E., Ghanem W., Orban H., Omar H., El-Kenawy M., Hafez A.T., Dawaba M.

Embase

Journal of Pediatric Surgery. 53 (8) (pp 1461-1463), 2018. Date of Publication: August 2018.

[Article]

AN: 2000671177

Objective: To detect whether grafting the incised plate during Snodgrass repair would improve outcome.

Material(s) and Method(s): Sixty patients with primary distal hypospadias were included. Patients were equally randomized using closed envelop method to either Snodgrass or grafted tubularized incised plate repair (GTIP). All operations were performed by a single surgeon. All intraoperative data were recorded. All patients were followed up for 1 year. Success was defined as slit shaped meatus at the tip of the glans with no stenosis, fistula or diverticulum.

Result(s): All 60 patients were evaluated at 1 year of follow-up. Mean age at surgery was 40 +/- 15 months. Both groups were comparable as regard to patients' age, meatus location, length and width and depth of urethral plate and glans width. Success was documented in 29/30 patients (96.7%) in the Snodgrass group. The only complication was meatal stenosis in one patient,

whereas success was documented in 28/30 patients (93.3%) in the GTIP group. The two failures were secondary to partial glans dehiscence. Success rate was not statistically different. Flow rate data at 1 year showed insignificant difference between both groups as regards Q-max and voiding time. The only statistically significant difference between both groups was a longer operative time 106 +/- 12 min in the GTIP group compared to only 77 +/- 9 for the Snodgrass group (p = 0.005).

Conclusion(s): Snodgrass and GTIP techniques for primary distal hypospadias repair have similar outcome. With a significantly shorter operative time, Snodgrass repair remains the first choice for primary distal hypospadias repair. Type of the study: Prospective randomized study.

Level of Evidence: Level I.

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Embase

Institution

(Helmy, Ghanem, Orban, Omar, El-Kenawy, Hafez, Dawaba) Urology and Nephrology Center,
Mansoura University, Egypt

Publisher

W.B. Saunders

Year of Publication

2018

388.

Complications after Hypospadias Correction: Prognostic Factors and Impact on Final Clinical Outcome.

Dokter E.M., Moues C.M., Rooij I.A.L.M.V., Biezen J.J.V.D.

Embase

European Journal of Pediatric Surgery. 28 (2) (pp 200-206), 2018. Date of Publication: 01 Apr 2018.

[Article]

AN: 614917136

Purpose The purpose of this study was to analyze the influence of patient and treatment characteristics on the occurrence of complications after hypospadias correction and the impact of complications on final clinical outcome. **Materials and Methods** The study cohort consisted of 205 hypospadias patients who had surgery in the Medical Centre Leeuwarden (1996-2011). Patient and treatment characteristics were hypospadias severity (preoperative meatal location and chordee), number of planned surgeries, reconstruction technique, operation year, and patient's age at the time of surgery. The final clinical outcome was measured with the Hypospadias Objective Scoring Evaluation (HOSE) (maximum score = 16) and compared between patients with and without complications. **Results** Sixty-four patients (31%) had complications, most of which were fistulas (n = 40). An increased complication risk was seen in patients with severe hypospadias (preoperative proximal meatus or chordee), multistage reconstruction, reconstruction techniques other than Mathieu, and surgeries performed before 2005. Uncomplicated treatment resulted only in a marginally higher HOSE (15.7) compared with complicated treatment (15.4). Fistulas and multiple complications reduced clinical outcome more (15.3 and 14.9, respectively), while urinary tract infections, wound dehiscence, or prepuce related complications did not (16.0, 16.0, and 15.8, respectively). **Conclusion** The complication risk after hypospadias correction is influenced by hypospadias severity and type and year of reconstruction. Certain, but not all complications diminish final clinical outcome.

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Status

Embase

Institution

(Dokter, Moues, Biezen) Department of Plastic Surgery and Hand Surgery, Medical Centre Leeuwarden, Leeuwarden, Netherlands (Dokter, Rooij) Department for Health Evidence, Radboud Institute for Health Sciences, Radboud University Medical Center, P.O. Box 9101, Nijmegen 6500 HB, Netherlands

Publisher

Georg Thieme Verlag (E-mail: iaorl@iaorl.org)

Year of Publication

2018

389.

Hypospadias reconstructive surgery modification using double layer urethral stent.

Mohammadipor A., Shojaeian R., Sharifabad P.S., Hiraifar M.

Embase

Nephro-Urology Monthly. 10 (5) (no pagination), 2018. Article Number: e80269. Date of Publication: September 2018.

[Article]

AN: 625077889

Background: Several surgical reconstructive procedures are introduced to repair a hypospadias while post-operative complications have still remained high. Poor tissue handling, anastomotic site ischemia, tension on the anastomotic line, and distal obstruction are suggested as the main risk factors contributing to post-operative complications. This article is the introduction of a simple technical modification to perform a uniform straight tension free urethroplasty that is supposed to decrease post hypospadias repair complications.

Method(s): We used a 6F silicon Foley catheter and covered it by a part of 10 F Nelaton catheter at the most distal part of the Foley. To facilitate the covering and uncovering process, we incised the Nelaton catheter in one side through the whole length. We performed a two-layer closure urethroplasty over our double layer stent and finally uncovered the Foley catheter, which remains in the bladder as a trans-urethral stent and also for further bladder drainage. Results and

Conclusion(s): In our innovative method of urethral stenting, we have suggested to replace the transurethral stent with a tension free lower size catheter easily and safely at the end of procedure, which helps decrease suture line tension and is supposed to improve the local circulation.

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Embase

Institution

(Mohammadipor, Shojaeian, Hiraifar) Department of Pediatric Surgery, Mashhad University of Medical Sciences, Mashhad, Iran, Islamic Republic of (Sharifabad) Department of Pediatric Urology, Mashhad University of Medical Sciences, Mashhad, Iran, Islamic Republic of

Publisher

Kowsar Corp (E-mail: supplort@kowsarpub.com)

Year of Publication

2018

390.

Fetal valproate syndrome: the Irish experience.

Mohd Yunus H., Green A.

Embase

Irish Journal of Medical Science. 187 (4) (pp 965-968), 2018. Date of Publication: 01 Nov 2018.

[Article]

AN: 621626855

Introduction: Fetal valproate syndrome was first described in 1984. Valproic acid crosses the placenta and can potentially lead to major congenital malformation, dysmorphism and neurodevelopmental disorder.

Method(s): A retrospective study of 29 cases of FVS diagnosed by geneticists from 1995 to 2016. The cases were diagnosed based on criteria of fetal anticonvulsant syndrome.

Result(s): A total of 29 cases reported in the last 21 years. Features commonly described are prominent metopic ridge, midface hypoplasia, epicanthic folds, micrognathia and broad and flat nasal bridge. Four (13.7%) had cleft palate, three (10%) had neural tube defect, four (13.7%) with cardiac malformation, 15 (52%) experienced developmental delay including six (40%) with speech delay, 11 (38%) with limb defects, four (13.7%) reported with neurodevelopmental disorder and two (7%) had hypospadias.

Conclusion(s): FVS is still seen in the Irish population even though the teratogenicity of the VPA has been known for over 32 years. It is very important to create public and professional awareness to prevent FVS whenever possible.

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Status

Embase

Author NameID

Mohd Yunus, Hamizah; ORCID: <http://orcid.org/0000-0003-4394-4215>

Institution

(Mohd Yunus, Green) Department of Clinical Genetics, Our Lady's Children's Hospital, Crumlin, Ireland

Publisher

Springer London

Year of Publication
2018

391.

Management of hypospadias in Saudi Arabia: A national survey.

Alsowayan O.S.

Embase

Urology Annals. 10 (4) (pp 391-394), 2018. Date of Publication: October-December 2018.

[Article]

AN: 624423701

Introduction: Hypospadias is one of the most common congenital anomalies of the penis.

Different methods of hypospadias management are described in the literature. We try in this study to evaluate the national trends and to compare them with international practices.

Material(s) and Method(s): A multiple choice survey was distributed among a sample of national practitioners using a weblink between September and December 2017. It included questions about participants demographics, number of cases operated on per year, perioperative care and preferences, long-Term follow-up, and complications. Data were analyzed and compared with international practices.

Result(s): Results of 47 practitioners were evaluated and analyzed in this study. The majority of the participants were pediatric urologists (48.9%) and from the central province (44.7%). Most of the participants prefer to operate on patients between the ages of 1 and 2 years (48.9%) and operate at ≥ 20 cases per year (76.6%). Tubularized incised plate (TIP) is the preferred technique for distal penile hypospadias repair whereas staged repair is preferred for proximal cases. All participants use a form of a second layer and a stent for their repairs. The majority reported an overall complication rate of $\leq 10\%$ for distal penile hypospadias (76.1%) and $>10\%$ for proximal penile cases (59.6%).

Conclusion(s): This study helped us identify national trends in hypospadias management, which were comparable to the international trends. TIP repair is the preferred technique for distal penile hypospadias repair whereas staged repair is preferred for more complex proximal variants.

Although data in this study come from reports of personal experience, it can serve as a backbone for the future prospective studies on this topic.

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Embase

Institution

(Alsowayan) Department of Urology, King Fahd Hospital, Imam Abdulrahman Bin Faisal University, PO Box 40086, Dammam 31952, Saudi Arabia

Publisher

Wolters Kluwer Medknow Publications (B9, Kanara Business Centre, off Link Road, Ghatkopar (E), Mumbai 400 075, India)

Year of Publication

2018

392.

Therapeutic effects of snodgrass urethroplasty in the treatment of distal hypospadias.

Xiong Z., Liang C., Deng X.

Embase

International Journal of Clinical and Experimental Medicine. 11 (9) (pp 10009-10014), 2018. Date of Publication: 2018.

[Article]

AN: 624250006

Objective: The aim of this study was to evaluate the efficacy and complications of Snodgrass urethroplasty in the treatment of hypospadias of the coronary sulcus type and middle penile shaft type, providing evidence for clinical treatment of distal hypospadias.

Method(s): From May 2015 to April 2017, 48 patients with distal hypospadias were divided into a coronary sulcus group (n=28) and middle penile shaft group (n=20). All patients were treated with Snodgrass urethroplasty and were followed up for more than 6 months. Surgical indicators and treatment effects of the patients in the two groups were compared. Operative methods are described and postoperative complications and their management methods are summarized.

Result(s): Patients in the coronary sulcus group had shorter operation times and urinary catheter retention times than those in the middle penis shaft group (both $P < 0.01$). There were no significant differences in postoperative complications (urethral stricture, meatal stenosis, urinary fistula, penile edema) between the two groups. Postoperative satisfaction rates of patients in the coronary sulcus group and middle penis shaft group were 92.86% and 95.00%, respectively, with

no significant differences. Follow up results showed that satisfaction rates for the appearance of the penis, glans penis, and urethral orifice in the coronary sulcus group were higher than those of the middle penis shaft group (all $P < 0.01$). There were no significant differences between the two groups in urinary range, urinary line, low urethral orifice, and penis rotation.

Conclusion(s): Snodgrass urethroplasty has good therapeutic effects in patients with hypospadias, specifically in the coronary sulcus and middle penile shaft. It has low complication rates and high patient satisfaction rates. It is worthy of clinical promotion.

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Status

Embase

Institution

(Xiong, Liang, Deng) Department of Urology Surgery, Zhangjiajie People's Hospital, Zhangjiajie City, Hunan Province, China

Publisher

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Year of Publication

2018

393.

Vascularization of vessel pedicle in hypospadias and its relationship to near-period complications.

Zhao Z., Sun N., Mao X.

Embase

Experimental and Therapeutic Medicine. 16 (3) (pp 2408-2412), 2018. Date of Publication: September 2018.

[Article]

AN: 623399782

The present study evaluated the relationship between the vascularization of vessel pedicle and the near-period complication by assessing the surgery effect and recognizing the blood supply of the material for urethra reconstruction and collected the follow-up of the hypospadias patients after the operation. We illuminated the prepuce by a lighting technique using cold light and then photographed, and recorded the distribution and the quantity of vessels. Then classified, analyzed and summarized the vascularization of vessel pedicle. Patients who were repaired by

Duckett technique were followed up for 1-5 months after the operation. We classified vessel pedicle vascularization in hypospadias cases into four patterns based on the predominant blood vessels. The number of predominant vessels had no exact relationship to the meatal location. The incidence of fistula and stricture had no exact relationship to the number of predominant vessels. In conclusion, there is no exact congruent relationship between the types of vascularization with the meatal location. Vascularization of vessel pedicle had no exact relationship with urethral fistula or stricture.

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Status

Embase

Institution

(Zhao, Mao) Department of Urology, Southern Medical University, Guangzhou, Guangdong 510515, China (Zhao) Department of Urology, Guangzhou Women and Children's Medical Center, Guangzhou, Guangdong 510623, China

(Sun) Department of Urology, Beijing Children's Hospital, Beijing 100045, China

(Mao) Department of Urology, Zhujiang Hospital, Guangzhou, Guangdong 510000, China

Publisher

Spandidos Publications (10 Vriaxidos Street, Athens 116 10, Greece. E-mail: subscriptions@spandidos-publications.com)

Year of Publication

2018

394.

Outcome after Hypospadias Repair: Evaluation Using the Hypospadias Objective Penile Evaluation Score.

Krull S., Rissmann A., Krause H., Mohnike K., Roehl F.-W., Koehn A., Hass H.-J.

Embase

European Journal of Pediatric Surgery. 28 (3) (pp 265-272), 2018. Date of Publication: 01 Jun 2018.

[Article]

AN: 616236146

Introduction The Hypospadias Objective Penile Evaluation Score (HOPE-Score) is a concise and reproducible way to describe hypospadias severity. We classified boys undergoing primary hypospadias repair to determine the correlation between the HOPE-Score and the severity of hypospadias first and the outcome after surgery second. **Materials and Methods** Patients who underwent primary hypospadias repair from 2005 to 2014 were identified. An independent physician assessed retrospectively the HOPE-Score, using photographs of the patients before, after primary surgery, and after all necessary surgeries. The correlation between the HOPE-Score and the severity of hypospadias, on the one hand, and the outcome after surgery, on the other hand, were analyzed. **Results** The HOPE-Score was assessed preoperatively for 79 boys, postoperatively for 66, and after all necessary surgeries for 21 patients. Mean HOPE-Score reached 30.2 ± 5.9 before surgery, 42.2 ± 6.1 after primary surgery, and 43.7 ± 3.4 after all necessary surgeries. A significant correlation between the HOPE-Score and the severity of hypospadias before surgery was observed. The boys with glanular hypospadias scored significantly higher (36.3 ± 5.4) than those with distal (29.6 ± 4.4) and proximal hypospadias (21.1 ± 3.5). Furthermore, a significant correlation between the HOPE-Score and the outcome after hypospadias repair was observed. Patients who needed no reintervention after primary hypospadias repair scored significantly higher postoperatively (45.1 ± 5.4) than those who needed a second (40.8 ± 4.2) or more than two surgeries (36.9 ± 7.4). **Conclusion** The HOPE-Score is a good system to assess the severity of hypospadias and the cosmetic outcome after hypospadias repair.

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Status

Embase

Institution

(Krull) Department of Pediatrics, Klinikum Magdeburg GGmbH, Magdeburg, Sachsen-Anhalt, Germany (Rissmann, Koehn) Malformation Monitoring Centre of Saxony-Anhalt, Medical Faculty Otto-von-Guericke, University Magdeburg, Leipziger Str. 44, Magdeburg 39120, Germany

(Krause, Hass) Division of Pediatric Surgery, Department of General, Abdominal and Vascular Surgery, University Hospital, Magdeburg, Sachsen-Anhalt, Germany

(Mohnike) Department of Pediatric Endocrinology, Universitätsklinikum Magdeburg Kinderklinik, Magdeburg, Sachsen-Anhalt, Germany

(Roehl) Department for Biometry and Medical Informatics, Universitätsklinikum Magdeburg, Sachsen-Anhalt, Germany

Publisher

Georg Thieme Verlag (E-mail: iaorl@iaorl.org)

Year of Publication

2018

395.

The possible impact of antenatal exposure to ubiquitous phthalates upon male reproductive function at 20 years of age.

Hart R.J., Frederiksen H., Doherty D.A., Keelan J.A., Skakkebaek N.E., Minaee N.S., McLachlan R., Newnham J.P., Dickinson J.E., Pennell C.E., Norman R.J., Main K.M.

Embase

Frontiers in Endocrinology. 9 (JUN) (no pagination), 2018. Article Number: 288. Date of Publication: 04 Jun 2018.

[Article]

AN: 622581985

Phthalates are ubiquitous environmental endocrine-disrupting chemicals suspected to interfere with developmental androgen action leading to adverse effects on male reproductive function. Prenatal exposure studies in rodents show cryptorchidism, hypospadias and reduced testicular volume (TV), testosterone and anogenital distance in males. It is postulated that there is a developmental window in utero when phthalate exposure has the most potent adverse effects. Some human studies show associations between prenatal phthalate exposure and reduced calculated "free" serum testosterone in infant boys and shorter anogenital distance. However, there are no data available yet which link antenatal exposure to long-term effects in men. We aimed to correlate antenatal phthalate exposure with adult TV, semen parameters and serum reproductive hormone concentrations. 913 men from the Western Australian (Raine) Pregnancy Cohort were contacted aged 20-22 years. 423 (56%) agreed to participate; 404 underwent testicular ultrasound examination; 365 provided semen samples, and reproductive hormones were measured in 384. Maternal antenatal serum phthalate metabolite measurements were available for 185 and 111 men, who provided serum and semen, respectively. Maternal serum collected at 18 and 34 weeks gestation, stored at -80degreeC, was pooled and analyzed for 32 phthalate metabolites by liquid chromatography-tandem mass spectrometry. TV was calculated, semen analysis performed by WHO approved methods, and serum concentrations of gonadotrophins, inhibin B, and testosterone measured. Eleven phthalate metabolites were detected. Primary and secondary metabolites of di-(2-ethyl-hexyl) phthalate (DEHP) and di-iso-

nonyl phthalate (DiNP) were positively correlated. After correction for adult height, BMI, presence of a varicocele and exposure to maternal smoking mono-iso-nonyl phthalate (MiNP) ($r = -0.22$) and sums of DEHP and DiNP metabolites ($r = -0.24$) and the sum of the metabolites of the high molecular weight phthalates ($r = -0.21$) were negatively correlated with TV (all $p < 0.05$). After adjustment for BMI adult serum total testosterone was positively associated with exposure to the following antenatal serum phthalate metabolites: mono-(2-ethylhexyl) phthalate ($r = 0.26$), MiNP ($r = 0.18$), the sum of metabolites for DEHP ($r = 0.21$) and DiNP ($r = 0.18$), and the sum of high molecular phthalates ($r = 0.20$) ($p = 0.0005$ to $p = 0.02$). Given sample size, storage duration and confounding through postnatal exposures, further studies are required.

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Status

Embase

Institution

(Hart, Doherty, Keelan, Newnham, Dickinson, Pennell) Division of Obstetrics and Gynaecology, University of Western Australia, Perth, WA, Australia (Hart) Fertility Specialists of Western Australia, Bethesda Hospital, Claremont, WA, Australia

(Frederiksen, Skakkebaek, Main) Department of Growth and Reproduction and EDMaRC, Rigshospitalet, University of Copenhagen, Copenhagen, Denmark

(Doherty, Keelan, Minaee, Newnham, Pennell) Women and Infants Research Foundation, King Edward Memorial Hospital, Perth, WA, Australia

(McLachlan) Hudson Institute of Medical Research, Monash Medical Centre, Melbourne, VIC, Australia

(Norman) Robinson Research Institute, University of Adelaide, Adelaide, SA, Australia

Publisher

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Year of Publication

2018

396.

Tubularized incised plate urethroplasty for distal and midpenile hypospadias reoperation: A single institute experience.

Salih E.M., Elemam A., Elfeky M., Abdrabuh A.M., Galal H.A.

Embase

Journal of Clinical Urology. 11 (3) (pp 178-183), 2018. Date of Publication: 01 May 2018.

[Article]

AN: 622163970

Objective: Our study aimed to evaluate our experience in tubularized incised plate urethroplasty in distal and midpenile hypospadias reoperation.

Patients and Methods: This was a retrospective study in which we reviewed the medical records of 56 patients who underwent tubularized incised plate urethroplasty in distal and midpenile hypospadias reoperation. Tubularized incised plate urethroplasty was performed using a surgical technique like the primary tubularized incised plate repair based on the Snodgrass's method. We recorded complications and functional outcomes of redo procedures.

Result(s): We reviewed 56 patients who ranged in age from 2-16 years (mean 7.02+/-3.8 years). The overall complication rate was 33.9% (n=19). Eight patients (14.3%) had fistula, six had meatal stenosis (10.7%), four had dehiscence (7.1%), and one had neourethral stenosis (1.8%). The complications occurred in 13 of mid-penile cases representing 50% of mid-penile cases but occurred in six cases of distal-penile cases (20%). The success rate of tubularized incised plate urethroplasty reoperation was 94.6% after repair of fistula and meatal stenosis by simple closure of fistula and meatoplasty.

Conclusion(s): Tubularized incised plate urethroplasty is a safe and effective alternative procedure for hypospadias reoperations. The outcome is favorable if the urethral plate has no scars and with distal meatal location. The complication rate increases if the third redo is done and with midpenile meatal location.

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Status

Embase

Institution

(Salih, Elemam, Elfeky, Abdrabuh, Galal) Urology Department, Al-Azhar Faculty of Medicine, Egypt

Publisher

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Year of Publication

2018

397.

Prospective study to evaluate the clinical outcome of intralesional interferon-2b in the management of Peyronie's disease.

Sokhal A., Jain N., Jhanwar A., Singh K., Saini D.

Embase

Urology Annals. 10 (2) (pp 154-158), 2018. Date of Publication: April-June 2018.

[Article]

AN: 621707200

Context: Interferon (IFN)-2b in Peyronie's disease (PD).

Aim(s): This study aims to evaluate clinical efficacy of the IFN-2b in both subjective and objective manner for the treatment of PD and compared with previously used intralesional verapamil in terms of cost-benefit analysis. Settings and Design: Prospective study.

Material(s) and Method(s): A prospective study conducted from January 2013 to July 2016 in the Department of Urology, Government Medical College, Kota, Rajasthan, India. We included patients with identifiable Peyronie's plaque with or without pain, curvature ranging between 30 and 90 degrees. We excluded patients with a calcified plaque and the ventral location of the plaque, any infective foci over the penis, erectile dysfunction due to other etiologies and patients who had received previous intralesional therapy. Patients were evaluated by clinical history, physical examination including plaque location, size, consistency, and penile curvature. Patients received intralesional IFN-2b in a dose of 3 x 106 IU. Patients completed the visual analogue pain (VAS) score for pain, and International Index of Erectile Function-5 (IIEF-5) questionnaire at first visit as well as at follow-up of 1 month and 3 months. Statistical Analysis Used: Comparisons were performed using the paired Student's t-test and Chi-square tests as appropriate. Patient's objective and subjective clinical characteristics were described as a means (standard deviation). Result(s): We included 86 patients in this study. Patients had a mean age of 48.6 years, mean plaque volume 256 mm³, and disease duration of 15.2 years. After 1 month of treatment, there was a significant change in plaque volume 256-60.8 mm³; $P < 0.01$) and penile curvature 34.8-24.6degree; $P < 0.01$). The patients reported significant improvement in pain score VAS and IIEF-5.

Conclusion(s): IFN-2b, as minimal invasive (intralesional) options for the treatment of PD, demonstrated significant improvement in plaque volume, penile curvature with minimal complications. Patients subjectively reported significant improvement in pain on erection and sexual activities. IFN-2b and verapamil had an almost similar clinical outcome, but verapamil at much lower cost.

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Status

Embase

Institution

(Sokhal, Jhanwar, Saini) Department of Urology, King George's Medical University, Lucknow, Uttar Pradesh, India (Jain, Singh) Department of Urology, Government Medical College, Kota, Rajasthan, India

Publisher

Medknow Publications (B9, Kanara Business Centre, off Link Road, Ghatkopar (E), Mumbai 400 075, India)

Year of Publication

2018

398.

Laparoscopic management of Mullerian duct remnants in the paediatric age: Evidence and outcome analysis.

Raicevic M., Saxena A.K.

Embase

Journal of Minimal Access Surgery. 14 (2) (pp 95-98), 2018. Date of Publication: April-June 2018.

[Article]

AN: 621284846

Background: This study performed a literature analysis to determine outcomes of laparoscopic management in Mullerian duct remnants (MDRs).

Patients and Methods: Literature was searched for terms 'Mullerian' 'duct' 'remnants' and 'laparoscopy'. Primary end points were age at surgery, laparoscopic technique, intraoperative complications and postoperative morbidity.

Result(s): The search revealed 10 articles (2003-2014) and included 23 patients with mean age of 1.5 years (0.5-18) at surgery. All patients were 46XY, n = 1 normal male karyotype with two cell lines. Explorative laparoscopy was performed in n = 2 and surgical management in n = 21. The 5-port technique was used in n = 10, 3-port in n = 9 and robot-assisted laparoscopic approach in n = 1 (n = 1 technique not described). Complete MDRs removal in n = 9, complete dissection and MDRs neck ligation with endoscopic loops in n = 11 and n = 1 uterus and cervix were split in the midline. After MDRs removal, there were n = 2 bilateral orchidopexy, n = 3

unilateral orchidopexy, n = 1 Fowler-Stephens stage-I and n = 1 orchiectomy. Mean operative time was 193 min (120-334), and there were no intraoperative complications. Mean follow-up was 20.5 months (3-54) and morbidity included 1 prostatic diverticula. There were 13 associations with hypospadias, of which 3 had mixed gonads and 3 bilateral cryptorchidism. Other associations were unilateral cryptorchidism and incarcerated inguinal hernia n = 1, right renal agenesis and left hydronephrosis n = 1 and n = 2 with transverse testicular ectopy.

Conclusion(s): This MDRs analysis suggests that the laparoscopic approach is an effective and safe method of treatment as no intraoperative complication has reported, and there is low morbidity in the long-term follow-up.

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Status

Embase

Institution

(Raicevic) Department of Pediatric Surgery and Orthopedics, Clinical Center Nis, Serbia

(Saxena) Department of Pediatric Surgery, Chelsea Children's Hospital, Chelsea and Westminster NHS Foundation Trust, Imperial College London, 369 Fulham Road, London SW109NH, United Kingdom

Publisher

Medknow Publications (B9, Kanara Business Centre, off Link Road, Ghatkopar (E), Mumbai 400 075, India)

Year of Publication

2018

399.

Hypospadias.

Buschel H., Carroll D.

Embase

Paediatrics and Child Health (United Kingdom). 28 (5) (pp 218-221), 2018. Date of Publication: May 2018.

[Review]

AN: 2000630934

Hypospadias is a relatively common abnormality of the external genitalia and the incidence seems to be increasing. Early recognition and appropriate referral to specialist centres allows for parents to be given timely and accurate information, although in general the surgical management of patients does not commence until one year of age. It is vital that assessment of children with hypospadias includes the position of the testes at the time of recognition. When both testes are adequately descended and palpable within a well-developed scrotum, no further investigation is required urgently. However, abnormalities of testicular descent or scrotal development can point to more serious problems and should be investigated urgently. For these children, hypospadias may be a part of a more complex abnormality in sexual differentiation. The operative management of hypospadias is undertaken by specialist surgeons. Results for distal hypospadias are excellent and few patients have long-term complications from surgery. Proximal hypospadias, particularly in association with other syndromes or disorders of sexual development have poorer long-term outcomes. There is no perfect operation for the correction of hypospadias, and long-term data outlining the advantages and disadvantages of different techniques is lacking in the literature.

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Status

Embase

Institution

(Buschel, Carroll) The Townsville Hospital, Townsville, Australia; Townsville, Australia

Publisher

Churchill Livingstone

Year of Publication

2018

400.

Role of uroflowmetry before and after hypospadias repair.

Piplani R., Aggarwal S.K., Ratan S.K.

Embase

Urology Annals. 10 (1) (pp 52-58), 2018. Date of Publication: January-March 2018.

[Article]

AN: 620799229

Aims: To study the role of uroflowmetry in the preoperative and early postoperative period in children undergoing hypospadias repair.

Material(s) and Method(s): Twenty-six cases undergoing hypospadias repair over 1 year (tubularized incised plate [TIP] Snodgrass [17 patients], TIP with Snodgraft [5 patients], Duckett's onlay flap [2 patients], and Duckett's tube [2 patients] urethroplasty) were prospectively evaluated with preoperative ultrasound and uroflowmetry and postoperative uroflowmetry at 3 months after the surgery and at 6 and 9 months interval if these dates fell within the study period on follow-up. The parameters studied were maximum flow rate (Qmax), average flow rate (Qav), total voided volume, voiding time, and type of curve. Preoperative and postoperative uroflow data were compared.

Result(s): Twenty-six cases comprised of anterior hypospadias (n = 8), mid penile (n = 11), and posterior hypospadias (n = 7). Fourteen patients had obstructed flow rates preoperatively. While 69% patients (18/26) had obstructed flow rates at 3 months postoperatively, it dropped to 43% at 9 months. Following TIP (Snodgrass) repair, 88% (15/17) had obstructed flow rates postoperatively. Best results were seen in patients undergoing circumferentially epithelialized urethral reconstruction (TIP with Snodgraft, Duckett's onlay flap, and Duckett's tube).

Conclusion(s): Abnormal uroflow is an inherent aspect of hypospadias in 50% of the cases. Both preoperative and postoperative uroflow evaluation is necessary for meaningful conclusion. Patients with preoperative normal flow rates but obstructed postoperative flow rates need clinical evaluation. Obstructive flow rates are more common after TIP (Snodgrass) repair. The urinary flow rates improve with time.

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Status

Embase

Institution

(Piplani, Aggarwal, Ratan) Department of Paediatric Surg., Maulana Azad Medical College and Assoc. Lok Nayak Hospital, New Delhi, India

Publisher

Medknow Publications (B9, Kanara Business Centre, off Link Road, Ghatkopar (E), Mumbai 400 075, India)

Year of Publication

2018

401.

Persistent pain following common outpatient surgeries in children: A multicenter study in Italy.
Mossetti V., Boretsky K., Astuto M., Locatelli B.G., Zurakowski D., Lio R., Nicoletti R., Sonzogni V., Maffioletti M., Vicchio N., Ivani G.

Embase

Paediatric anaesthesia. 28 (3) (pp 231-236), 2018. Date of Publication: 01 Mar 2018.

[Article]

AN: 625200025

INTRODUCTION: The prevalence of persistent postsurgical pain in children is over 20% after major surgeries; however, data are scarce on the prevalence, character, and risk factors among children undergoing common ambulatory surgeries. The primary aim of this study was to evaluate the prevalence of persistent pain following pediatric ambulatory surgery at 1, 3, and 6 months. Secondary aims were to identify risk factors and characterize the pain and consequences of persistent postsurgical pain.

METHOD(S): ASA I-II, ages 1 month to 16 years old, undergoing elective hypospadias repair, herniorrhaphy, orchiopexy, and orthopedic surgery were enrolled in a prospective, longitudinal, observational study at 3 pediatric centers in Italy. All patients received general plus regional anesthesia. Postoperative pain was evaluated using age appropriate pain scales at 1 and 3 hours. At 1, 3, and 6 months, pain scores were obtained and Parent's Postoperative Pain Measures (<8 yo) and Child Activity Limitations Interview (>8 yo) surveys were administered.

RESULT(S): About 350 patients completed the study. The prevalence of pain at 1, 3, and 6 months was 24% (84/350), 6.0% (21/350), and 4.0% (14/350), respectively. Inguinal herniorrhaphy patients experienced significantly higher pain at all 3-time points; 35.6%, 14.9%, and 9.2%. There was no significant association between mean pain scores >4 in PACU and persistent pain. Pain persisting at 6 months had neuropathic characteristics and frequently interfered with daily activities and sleep.

CONCLUSION(S): Our data support the presence of persistent pain in pediatric patients after common surgeries. Most patients who developed persistent pain at 6 months had pain at 1 month. We recommend questioning at follow-up visit about persistent pain and functional impairment with follow-up until resolution.

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PMID

29352738 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=29352738>]

Author NameID

Mossetti, Valeria; ORCID: <http://orcid.org/0000-0002-7862-4863>

Institution

(Mossetti, Lio, Vicchio, Ivani) Department of Anesthesia and Intensive Care, Regina Margherita Children's Hospital, Citta Della Salute e della Scienza, Torino, Italy (Boretsky, Zurakowski)

Department of Anesthesiology, Perioperative and Pain Medicine, Boston Children's Hospital, Boston, MA, United States

(Astuto, Nicoletti) Department of Anesthesia and Intensive Care, Vittorio Emanuele University Hospital, Catania, Italy

(Locatelli, Sonzogni, Maffioletti) Department of Anesthesia and Intensive Care, Papa Giovanni XXIII Hospital, Bergamo, Italy

Year of Publication

2018

402.

Is stent type used in snodgrass method a factor in fistula formation?.

Sarac M., Bakal U., Tartar T., Canpolat S., Kazez A.

Embase

Nigerian journal of clinical practice. 21 (9) (pp 1198-1202), 2018. Date of Publication: 01 Sep 2018.

[Article]

AN: 624726516

Context: Snodgrass method (tubularized incised plate urethroplasty [TIPU]) is a widely used technique for hypospadias repair. Aim: It was aimed to compare the outcome of hypospadias repair with stenting using feeding tube compare with those with Foley catheter.

Subjects and Methods: The demographic characteristics of the 123 patients who underwent hypospadias repair with Snodgrass method, the success of the applied method, and the factors affecting fistula complication were evaluated retrospectively. Patients were divided into two groups: those operated before January 2010 (Group A) and those who were operated after (Group B). In Group A patients, urethroplasty was performed using silicone Foley catheters, in which balloon of these catheters was filled by saline at appropriate size. In Group B, urethroplasty was performed using feeding catheter.

Results: Group A and Group B consisted of 32 and 91 patients, respectively. Fistula developed in 10 (31.3%) and 4 (4.39%) patients in Group A and Group B, respectively. There was a

statistically significant difference between the two groups in terms of the development of fistula complication ($P = 0.0002$).

Conclusion: The use of a feeding catheter in TIPU could be a more advantageous than using a Foley catheter.

PMID

30156207 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=30156207>]

Institution

(Sarac, Bakal, Tartar, Canpolat, Kazez) Department of Pediatric Surgery, Firat University School of Medicine, Elazig, Turkey

Year of Publication

2018

403.

Congenital anterior urethrocutaneous fistula: A systematic review.

Lin Y., Deng C., Peng Q.

Embase

African journal of paediatric surgery : AJPS. 15 (2) (pp 63-68), 2018. Date of Publication: 01 Apr 2018.

[Article]

AN: 628662546

Congenital anterior urethrocutaneous fistula (CAUF) is a rare anomaly characterized by fistulization of penile urethra to skin. It's usually seen as an isolated deformity or may accompany genitourinary or anorectal malformations. We aim to define the common properties of patients mentioned in literatures by systematic review. A comprehensive search of PubMed, Embase, Web of Science, and Cochrane Library was performed including cross-referencing independently by two assessors. Selections were restricted to human studies in English. Based on the systematic review, 63 patients in 34 articles were included in the study. Most common fistula site was subcoronal in 29 (46.0%) patients. Chordee was in 8 (14.5%) and associated genitourinary anomaly was detected in 19 (30.2%) of patients. Fistula recurrence ratio was 6/59 (11.3%) using different surgical techniques and 3/6 was closed spontaneously. CAUF is frequently located in subcoronal level and usually an intact urethra distal to it. Success rates are high with the principles of hypospadias surgery.

PMID

31290465 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=31290465>]

Institution

(Lin, Deng, Peng) Department of Pediatric Surgery, Chengdu Women's and Children's Central Hospital of Chongqing Medical University, Chengdu, China

Publisher

NLM (Medline)

Year of Publication

2018

404.

Gender Identity and Sexual Function in 46,XX Patients with Congenital Adrenal Hyperplasia Raised as Males.

Apostolos R.A.C., Cangucu-Campinho A.K., Lago R., Costa A.C.S., Oliveira L.M.B., Toralles M.B., Barroso U.

Embase

Archives of sexual behavior. 47 (8) (pp 2491-2496), 2018. Date of Publication: 01 Nov 2018.

[Article]

AN: 624991165

In individuals with congenital adrenal hyperplasia (CAH) and 46,XX karyotype, androgens produced by the adrenal glands during the intrauterine development promote virilization of the genitals, which may even result in the development of a well-formed penis. Some of these children with late diagnosis are registered as males after birth. After obtaining approval from the internal review board, we evaluated gender identity and sexual function in four 46,XX severely virilized patients with CAH, who were originally registered and raised as males, assisted in our Disorders of Sexual Development Clinic. The evaluation consisted of questionnaires to assess gender identity and sexual activity and interview with the multidisciplinary team that provides care for these patients. The patients underwent surgery to remove uterus, ovaries, and remaining vaginal structures, in addition to implantation of testicular prosthesis and correction of hypospadias, when necessary. All four patients have developed a clear male gender identity, and when evaluated for sexual activity, they have reported having erections, libido, orgasms, and sexual attraction to women only. Two of these 4 patients had satisfactory sexual intercourses

when assessed using the International Index of Erectile Function questionnaire. The other two patients who never had sexual intercourse reported not having a partner for sexual activity; one is 18 years old, and the other is 14 years old. This study showed that this group of 46,XX severely virilized patients with CAH, registered and raised as males, adapted well to the assigned male gender, with satisfactory sexual function in patients who had sexual intercourse.

PMID

30291599 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=30291599>]

Author NameID

Barroso, Ubirajara Jr; ORCID: <http://orcid.org/0000-0002-1067-2463>

Institution

(Apostolos, Cangucu-Campinho, Lago, Oliveira, Toralles, Barroso) Disorders of Sexual Development Clinic, Federal University of Bahia, Av Alphaville 1, Rua Pajucara, 335, Salvador, Bahia, 41701015, Brazil (Apostolos, Costa, Barroso) Bahiana School of Medicine and Public Health, Salvador, Bahia, Brazil

Publisher

NLM (Medline)

Year of Publication

2018

405.

Early detection of deep wound infection in bladder exstrophy and hypospadias using a novel intervention.

Sabetkish S., Sabetkish N., Kajbafzadeh A.-M.

Embase

Journal of wound care. 27 (10) (pp 686-691), 2018. Date of Publication: 02 Oct 2018.

[Article]

AN: 624496979

OBJECTIVE: To introduce a practical technique for the early detection and prompt management of a probable bladder dehiscence (BD) and glanular dehiscence (GD) in patients with bladder exstrophy epispadias complex (BEEC) and hypospadias.

METHOD(S): In this prospective study, paediatric patients with BEEC (group 1) and with proximal hypospadias (group 2) underwent body temperature measurement using a non-contact infrared

radiant digital temperature measurement device in four body regions, including the surgical wound, forehead, right hand, and right foot at eight hour intervals, postoperatively. This technique was performed to detect wound temperature rises before whole body temperature rise or visible local wound skin redness, cellulitis or any sign of inflammation or wound dehiscence (WD).

RESULT(S): A total of 24 paediatric patients were recruited. Temperature rise in the surgical wound area was discovered in two patients with BEEC. The temperature reached 39.2degreeC in the first case (12 days postoperative) and 39.4degreeC in the second case (16 days postoperative). Urinalysis, urine culture, and clean surgical wound sampling was performed and the presence of Gram-positive microorganisms was detected. Both patients were managed with intravenous imipenem and vancomycin. After changing the antibiotic regimen, wound temperature was gradually decreased to 37.2degreeC in the first patient by day 16, and to 36.9degreeC in the second patient by day 21, without rise in body temperature. Other patients in group 1 and all patients in group 2 had normal wound temperature fluctuations within the follow-up period.

CONCLUSION(S): Postoperative periodical temperature measurement by a non-contact infrared radiant digital temperature measurement device is a safe and feasible technique that has the ability to detect deep wound infection, and may prevent the occurrence of WD before any visible sign of inflammation.

PMID

30332360 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=30332360>]

Institution

(Sabetkish) Nurse; Pediatric Urology and Regenerative Medicine Research Center, Section of Tissue Engineering and Stem Cells Therapy, Children's Hospital Medical Center, Tehran University of Medical Sciences, Tehran, Iran (IRI) (Sabetkish) Pediatric Urology and Regenerative Medicine Research Center, Section of Tissue Engineering and Stem Cells Therapy, Children's Hospital Medical Center, Tehran University of Medical Sciences, Tehran, Iran, Islamic Republic of

(Kajbafzadeh) Professor of Pediatric Urology; Pediatric Urology and Regenerative Medicine Research Center, Section of Tissue Engineering and Stem Cells Therapy, Children's Hospital Medical Center, Tehran University of Medical Sciences, Tehran, Iran (IRI)

Publisher

NLM (Medline)

Year of Publication

2018

406.

A 1-year randomized controlled trial to compare the outcome of primary repair of hypospadias with vascular cover using tunica vaginalis flap with those using preputial dartos fascia.

Kurbet S.B., Koujalagi R.S., Geethika V., Nagathan V.

Embase

African journal of paediatric surgery : AJPS. 15 (1) (pp 42-47), 2018. Date of Publication: 01 Jan 2018.

[Article]

AN: 626638594

Background: : Tubularized incised plate (TIP) urethroplasty is the most common technique noted to correct hypospadias. However, urethrocuteaneous fistula (UCF) is still one of the most common complications of this technique. Several techniques of providing vascularized flaps to the neourethra have been recommended to decrease this complication rate. The aim of the study was to assess the outcome of primary repair of hypospadias using tunica vaginalis (TV) flap with those using preputial dartos (PD) fascia.

Patients and Methods: : Children diagnosed with hypospadias between the age group of 9 months to 18 years, who fulfilled the criteria were randomly divided into two groups by computerized randomization technique. Initially, TIP urethroplasty was done. Children with PD vascular cover were included in Group A and those with TV vascular cover were included in Group B. All the patients were followed up for a minimum of 6 months after surgery.

Result(s): : Two (10%) patients in Group A developed UCF and one (5%) patient developed stricture urethra in the follow-up period. None of the patients developed UCF in the Group B. One (5%) patient had stricture urethra in Group B. Two (10%) patient developed meatal stenosis in Group A. Two (10%) patient in Group B developed meatal stenosis.

Conclusion(s): TV flap could be an alternative to PD flap as a vascular cover of neourethra in patients undergoing primary hypospadias repair by TIP urethroplasty.

PMID

30829308 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=30829308>]

Institution

(Kurbet, Koujalagi, Geethika, Nagathan) Department of Paediatric Surgery, Jawaharlal Nehru Medical College, Belagavi, Karnataka, India

Publisher

NLM (Medline)

Year of Publication

407.

Pharmacologic and Environmental Endocrine Disruptors in the Pathogenesis of Hypospadias: a Review.

Raghavan R., Romano M.E., Karagas M.R., Penna F.J.

Embase

Current environmental health reports. 5 (4) (pp 499-511), 2018. Date of Publication: 01 Dec 2018.

[Review]

AN: 625705126

PURPOSE OF REVIEW: Endocrine disrupting chemicals (EDCs) potentially have a role in causing hypospadias malformation through modifiable in-utero exposure. Considering the emerging literature on the role of potential endocrine disrupting substances on the occurrence of hypospadias and the potential to inform public health efforts to prevent the occurrence of these malformations, we have summarized the current literature, identified areas of consensus, and highlighted areas that warrant further investigation. RECENT FINDINGS: Pharmaceuticals, such as diethylstilbestrol, progestin fertility treatments, corticosteroids, and valproic acid, have all been associated with hypospadias risk. Data on exposure to dichlorodiphenyltrichloroethane and hexachlorobenzene pesticides, as well as non-persistent pollutants, particularly phthalates, is less consistent but still compelling. Improving exposure assessment, standardizing sample timing to relevant developmental windows, using clear case identification and classification schemes, and elucidating dose-response relationships with EDCs will help to provide clearer evidence.

Promising directions for future research include identification of subgroups with genetic hypospadias risk factors, measurement of intermediate outcomes, and study of EDC mixtures that will more accurately represent the total fetal environment.

PMID

30578470 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=30578470>]

Institution

(Raghavan, Penna) Division of Pediatric Urology, Children's Hospital at Dartmouth, Dartmouth Geisel School of Medicine, 1 Medical Center Drive, NH 03756, United States

(Romano, Karagas) Department of Epidemiology, Dartmouth Geisel School of Medicine, NH, United States

Publisher
NLM (Medline)
Year of Publication
2018

408.

Hypospadias Repair in Ethiopia: A Five Year Review.

Mammo T.N., Negash S.A., Negussie T., Getachew H., Dejene B., Tadesse A., Derbew M.

Embase

Ethiopian journal of health sciences. 28 (6) (pp 735-740), 2018. Date of Publication: 01 Nov 2018.

[Article]

AN: 625783042

Background: Hypospadias repair is one of the problematic issues in pediatric surgery. As a result of the multiple complications following the procedure, a variety of techniques have been used and newer methods continue to emerge. There is still controversy regarding the best method of repair. We aimed to determine the outcome of surgery and factors contributing to unfavorable outcomes in children with hypospadias.

Material(s) and Method(s): This is a retrospective review undertaken from September 2009 to August 2014. The research was conducted at Tikur Anbessa Specialized Hospital, Addis Ababa, Ethiopia. All children who underwent hypospadias repair and had regular follow-up were included in the study.

Result(s): A total of 202 boys aged less than 13 years were assessed. Most surgeries (80.3%) were performed in children older than 18 months. Transverse incised plate urethroplasty (TIP) was frequently performed for distal hypospadias (71.2 %), while transverse ventral preputal flap (TVPF) was the most common procedure done for proximal hypospadias (62.8 %). Overall success rate for first surgery was 55.9 %. There was a high rate of major post-operative complications (44.1%) of which urethrocutaneous fistula (UCF) was the most common (31.2%) followed by meatal stenosis and glans breakdown (7.4 % each). These complications were found to be higher in those who were operated at a later age and those with proximal hypospadias ($p=0.03$ and $p=0.01$ respectively). There was also a significant difference among the type of procedures with TIP and TVPF having the least complications ($p<0.01$).

Conclusion(s): From our experience, we found TIP a relatively safe and reliable method of repair for distal hypospadias while TVPF single stage repair was superior in the proximal ones. The high rate of complications in our institution was associated with higher burden of severe hypospadias and older age at surgery.

PMID

30607090 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=30607090>]

Institution

(Mammo, Negash, Negussie, Getachew, Dejene, Tadesse, Derbew) Unit of pediatric of surgery, Department of surgery, Addis Ababa University, Ethiopia

Publisher

NLM (Medline)

Year of Publication

2018

409.

Patients with disorders of sex development and proximal hypospadias are at high risk for reoperation.

Saltzman A.F., Carrasco A., Colvin A., Campbell J.B., Vemulakonda V.M., Wilcox D.

Embase

World journal of urology. 36 (12) (pp 2051-2058), 2018. Date of Publication: 01 Dec 2018.

[Article]

AN: 625563902

PURPOSE: Surgical repair for proximal hypospadias has been associated with long-term success rates of 32-68%. In a prior study, outcomes for proximal hypospadias in patients with a diagnosis of disorders of sex development (DSD) were no different than those of patients without DSD. The objective of our study is to report our experience with proximal hypospadias repair in patients with and without DSD.

METHOD(S): We retrospectively reviewed patients who underwent repair of proximal hypospadias between 2005 and 2016. Data collected included patient and disease characteristics, operative details, complications, and follow-up. The primary outcome was unplanned reoperation.

RESULT(S): Sixty seven patients were identified; 30 (44.8%) with DSD and 37 (55.2%) without DSD. Median follow-up was 28.3 months (IQR 18.9-45.7). 41 patients (61.2%) underwent at least one unplanned reoperation, median time to unplanned reoperation 10.3 months. More patients with DSD needed an unplanned reoperation (80 vs. 45.9%, $p=0.024$). During the first 12 months after initial repair, there was no difference in unplanned reoperation rates (40 vs. 32.4%, $p=0.611$), but there was a difference in the first 24 months post-operatively (76.7 vs. 43.2%, $p=0.007$). On multivariate logistic regression, older age at initial repair (OR 1.144) and two stage repair (OR 7.644) were positively associated with unplanned reoperation in the first 2 years after repair.

CONCLUSION(S): Proximal hypospadias repair is associated with an overall 61.2% reoperation rate regardless of associated DSD diagnosis. Patients with DSD are more likely to undergo an unplanned reoperation in the first 2 years after repair.

PMID

29855697 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=29855697>]

Institution

(Saltzman, Carrasco, Colvin, Campbell, Vemulakonda, Wilcox) Division of Urology, Department of Surgery, University of Colorado and Children's Hospital Colorado, 13123 E 16th Ave. ,Box 463, Aurora, CO 80045, United States

Publisher

NLM (Medline)

Year of Publication

2018

410.

Single-stage Abdominoplasty Using Groin Flaps Without Osteotomies: Management of Exstrophy-epispadias Complex.

Giron A.M., Mello M.F., Berjeaut R.H., Machado M.G., Silva G.C.D.A., Cezarino B.N., Oliveira L.M., Lopes R.I., Denes F.T.

Embase

Urology. 120 (pp 266), 2018. Date of Publication: 01 Oct 2018.

[Article]

AN: 624915306

BACKGROUND AND OBJECTIVE: The optimal treatment for children born with exstrophy-epispadia complex is still a matter of debate.^{1,2,3} We demonstrate the Single-Stage Abdominoplasty using Groin Flap technique to close the abdominal wall of children with classic bladder exstrophy (CBE) without osteotomy neither radical soft tissue mobilization. Advantages over current techniques are less risk of penile tissue loss and avoidance of osteotomies.

MATERIAL AND METHODS: Abdominal wall repair consists in using the hypogastric skin, rectus, and obliquus externus abdominalis muscle fascial flaps. These groin flaps are rotated medially resulting in a very strong abdominal wall support. Groin flaps are made of rectus anterior fascia rotated medially, flipped over, and sutured with Prolene sutures to close the defect. By rotating the fascial flaps medially, complete reinforcement of the abdominal wall to the level of the pubic bone is achieved. This permits abdominal closure maintenance without tension.

RESULT(S): Groin flap was applied to 128 patients with CBE referenced from all over the country. Most of these patients returned to their home areas making difficult their follow up. However, we have 44 cases that have regular clinical visits. Mean follow-up was 10.3 +/- 4.5 years (2 years 8 months-16 years). Successful closure was achieved in 43 patients (97.7%) as a single procedure; one patient had a complete wound dehiscence and needed another reconstruction (2.2%). Four patients (9.1%) presented abdominal hernias that needed surgical management. When continence is evaluated, we present similar literature rates (60%).⁴

CONCLUSION(S): Abdominal reconstruction using Groin flaps has advantages over the traditional approaches to CBE. It reduces the surgical steps and facilitates the closure of the abdominal wall without the need of osteotomies and consequent immobilization during the postoperative period. It is feasible at any age and can be also very useful as a salvage technique even after previous failed procedures. Finally, it minimizes the number of surgeries.

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PMID

30031829 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=30031829>]

Institution

(Giron, Mello, Machado, Silva, Cezarino, Oliveira, Denes) Divisao de Urologia, Hospital das Clinicas HCFMUSP, Faculdade de Medicina, Universidade de Sao Paulo, Sao Paulo, Brazil

(Berjeaut) Divisao de Urologia, Hospital das Clinicas HCFMUSP, Faculdade de Medicina, Universidade de Sao Paulo, Sao Paulo, Brazil

(Lopes) Divisao de Urologia, Hospital das Clinicas HCFMUSP, Faculdade de Medicina, Universidade de Sao Paulo, Sao Paulo, Brazil; Division of Urology, The Hospital for Sick Children, Toronto, Ontario, Canada

Publisher

NLM (Medline)

Year of Publication

411.

The association of air pollution with congenital anomalies: An exploratory study in the northern Netherlands.

Salavati N., Strak M., Burgerhof J.G.M., de Walle H.E.K., Erwich J.J.H.M., Bakker M.K.

Embase

International journal of hygiene and environmental health. 221 (7) (pp 1061-1067), 2018. Date of Publication: 01 Aug 2018.

[Article]

AN: 626152403

BACKGROUND: There are a growing number of reports on the association between air pollution and the risk of congenital anomalies. However, the results are inconsistent and most studies have only focused on the association of air pollution with congenital heart defects and orofacial clefts.

OBJECTIVE(S): Using an exploratory study design, we aimed to identify congenital anomalies that may be sensitive to maternal exposure to specific air pollutants during the periconceptional period.

METHOD(S): We conducted a case-control study of 7426 subjects born in the 15 years between 1999 and 2014 and registered in the European Registration of Congenital Anomalies and Twins Northern Netherlands (EUROCAT NNL). Concentrations of various air pollutants (PM10, PM2.5, PM10-2.5, NO2, NOX, absorbance) were obtained using land use regression models from the European Study of Cohorts for Air Pollution Effects (ESCAPE). We linked these data to every subject in the EUROCAT NNL registry via their full postal code. Cases were classified as children or fetuses born in the 15-year period with a major congenital anomaly that was not associated with a known monogenic or chromosomal anomaly. Cases were divided into anomaly subgroups and compared with two different control groups: control group 1 comprised children or fetuses with a known monogenic or chromosomal anomaly, while control group 2 comprised all other non-monogenic and non-chromosomal registrations.

RESULT(S): Using control group 1 (n=1618) for analysis, we did not find any significant associations, but when we used control group 2 (ranges between n=4299 and n=5771) there were consistent positive associations between several air pollutants (NO2, PM2.5, PM10-2.5, absorbance) and the genital anomalies subgroup.

CONCLUSION(S): We examined various congenital anomalies and their possible associations with a number of air pollutants in order to generate hypotheses for future research. We found that air pollution exposure was positively associated with genital anomalies, mainly driven by hypospadias. These results broaden the evidence of associations between air pollution exposure during gestation and congenital anomalies in the child. They warrant further research, which should also focus on possible underlying mechanisms.

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30033390 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=30033390>]

Institution

(Salavati, Erwich) Department of Obstetrics and Gynecology, University Medical Centre of Groningen, University of Groningen, Groningen, Netherlands (Strak) Institute for Risk Assessment Sciences (IRAS), Utrecht University, Utrecht, Netherlands

(Burgerhof) Department of Epidemiology, University Medical Centre of Groningen, University of Groningen, Groningen, Netherlands

(de Walle) Department of Genetics, EUROCAT Registration Northern Netherlands, University Medical Centre of Groningen, University of Groningen, Groningen, Netherlands

(Bakker) Department of Obstetrics and Gynecology, University Medical Centre of Groningen, University of Groningen, Groningen, the Netherlands; Department of Genetics, EUROCAT Registration Northern Netherlands, University Medical Centre of Groningen, University of Groningen, Groningen, the Netherlands

Publisher

NLM (Medline)

Year of Publication

2018

412.

Shaping parents, shaping penises: How medical teams frame parents' decisions in response to hypospadias.

Roen K., Hegarty P.

Embase

British journal of health psychology. 23 (4) (pp 967-981), 2018. Date of Publication: 01 Nov 2018.

[Article]

AN: 624125559

INTRODUCTION: Psychological research provides insights into how parents approach medical decisions on behalf of children. The medical decision of concern here is the surgical alteration of a hypospadiac penis, whose urethral opening does not appear at the tip. Hypospadias surgery is routinely carried out in infancy, despite criticism by international organizations concerned about children's rights. The focus of this study is on the framing of hypospadias surgery.

OBJECTIVE(S): The objective was to examine how health professionals frame hypospadias and hypospadias surgery in medical and non-medical ways. DESIGN: This was a qualitative study designed to build on the experimental research of Streuli et al. who investigated how medical versus non-medical information affects decision-making about non-essential childhood genital surgery.

METHOD(S): Semi-structured interviews were undertaken with 32 health professionals.

Theoretically informed thematic analysis was used to examine how health professionals talk about hypospadias surgery and about supporting parents to make treatment decisions.

RESULT(S): The analysis suggests that medical professionals' engagement with parents underestimates the effect of framing in influencing parental decisions about hypospadias surgery. Some psychological specialists in this area are actively framing hypospadias in ways that enable some parents to choose a non-medical pathway. Psychologically informed ways of talking about a child's genital difference focus on psychological qualities, including affect, well-being, and unconditional positive regard.

CONCLUSION(S): The best interests of children with hypospadias may well be served when psychological pathways are highlighted, providing opportunities to support the flourishing of children whose genital appearance raises the question of medical intervention. Statement of Contribution What is already known on this subject? Framing significantly affects medical decision-making in ways that people typically fail to perceive. Parents frequently consent to non-essential hypospadias (penile) surgery for their sons, despite the risks and ethical concerns. What does this study add? Medical teams could do more to consider framing when counselling parents about their son's hypospadias. Psychological specialists can help parents to frame their son's penile difference in terms of well-being and love. The best interests of children with hypospadias may be served by highlighting psychological care pathways.

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PMID

30054962 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=30054962>]

Author NameID

Roen, Katrina; ORCID: <http://orcid.org/0000-0003-0421-2484>

Institution

(Roen) Department of Psychology, University of Oslo, Norway (Hegarty) School of Psychology,
University of Surrey, United Kingdom

Publisher

NLM (Medline)

Year of Publication

2018

413.

Urethral Meatus and Glanular Closure Line: Normal Biometrics and Clinical Significance.

Abbas T.O., Ali M.

Embase

Urology journal. 15 (5) (pp 277-279), 2018. Date of Publication: 26 Sep 2018.

[Article]

AN: 626980873

Purpose: The aim of this study is to explore the normal external urethral meatal and glans closure line in normal boys, and to investigate the correlation between these glans biometrics and the age of the participants. MATERIAL AND METHOD: 103 male children were asked to participate in the study during ritual circumcision. Par-ents of 94 of them (mean age 5.9 years, range 0.6-13) accepted while remaining 9 did not. Glans biometrics were measured using digital calipers.

RESULT(S): 100% of the study participants had a vertical slit-like meatal opening located at the tip of the glans. The length of the meatal opening was 5.3 (+/- 1) mm and of ventral glans closure was 4.8 (+/-1.1) mm. Significant cor-relation between both the external meatal opening and closure lines lengths and age was observed. Moreover, the meatal opening size was correlated to the glans closure line as well ($r = 0.36$, confidence interval 0.14-0.54, $P < .001$).

CONCLUSION(S): The site and size of the meatus opening in normal male children is consistent, and ventral glans clo-sure is equal to or slightly less than meatal length. These findings could aid in glanular reconstruction configuration during hypospadias surgery.

PMID

30221335 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=30221335>]

Institution

(Abbas, Ali) Department of Pediatric Surgery, Hamad General Hospital, Doha, Qatar

Year of Publication

414.

Further Evidence of the Association of the Diacylglycerol Kinase Kappa (DGKK) Gene With Hypospadias.

Hozyasz K.K., Mostowska A., Kowal A., Mydlak D., Tsibulski A., Jagodzinski P.P.

Embase

Urology journal. 15 (5) (pp 272-276), 2018. Date of Publication: 26 Sep 2018.

[Article]

AN: 626910241

PURPOSE: Hypospadias is a common developmental anomaly of the male external genitalia. In previous studies conducted on West European, Californian, and Han Chinese populations the relationship between polymorphic variants of the diacylglycerol kinase kappa (DGKK) gene and hypospadias have been reported. The aim was to study the possible associations between polymorphic variants of the DGKK gene and hypospadias using an independent sample of the Polish population. MATERIALS AND METHODS: Ten single nucleotide polymorphisms in DGKK, which were reported to have an impact on the risk of hypospadias in other populations, were genotyped using high-resolution melting curve analysis in a group of 166 boys with isolated anterior (66%) and middle (34%) forms of hypospadias and 285 properly matched controls without congenital anomalies.

RESULT(S): Two DGKK variants rs11091748 and rs12171755 were associated with increased risk of hypospadias in the Polish population. These results were statistically significant, even after applying the Bonferroni correction for multiple comparisons ($P < .005$). All the tested nucleotide variants were involved in haplotype combinations associated with hypospadias. The global p-values for haplotypes comprising of rs4143304-rs11091748, rs11091748-rs17328236, rs1934179-rs4554617, rs1934183-rs1934179-rs4554617 and rs12171755-rs1934183-rs1934179-rs4554617 were statistically significant, even after the permutation test correction.

CONCLUSION(S): Our study provides strong evidence of an association between DGKK nucleotide variants, haplotypes and hypospadias susceptibility.

PMID

29464676 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=29464676>]

Institution

(Hozyasz) Department of Paediatrics, Institute of Mother and Child, Warsaw, Poland
(Mostowska, Tsibulski, Jagodzinski) Department of Biochemistry and Molecular Biology, Poznan
University of Medical Sciences, Poznan, Poland
(Kowal, Mydlak) Department of Paediatric Surgery, Institute of Mother and Child, Warsaw, Poland
Year of Publication
2018

415.

Objective measurements of the penile angulation are significantly different than self-estimated magnitude among patients with penile curvature.

Liguori G., Salonia A., Garaffa G., Chiriaco G., Pavan N., Cavallini G., Trombetta C.

Embase

International braz j urol : official journal of the Brazilian Society of Urology. 44 (3) (pp 555-562),
2018. Date of Publication: 01 May 2018.

[Article]

AN: 623213495

INTRODUCTION: The study was aimed to assess the presence of actual differences between the objective and the perceived magnitude of a curvature between patients affected by Peyronie's disease (PD) and congenital penile curvature (CPC). MATERIALS AND METHODS: We analysed a cohort of 88 consecutive patients seeking medical help for either CPC or PD. All patients were invited to provide a self-made drawing of their penis in erection in order to obtain self-provided description of the deformity. An objective measurement of the deformity was also performed drawing two intersecting lines through the center of the distal and proximal straight section of the penile shaft.

RESULTS: Our findings showed significant differences between patient self-estimation and the objective measurements of the penile angulation performed by trained experts, with only 32% of patients correctly assessing their own curvature. Overall, patients tended to overestimate (56%) their degree of curvature, but the results are different in patients with PD than those with CPC. In the 60 men (68%) who did not accurately assess their curvature, PD patients generally overestimated their curvature versus CPC patients (67% vs 16%). On the contrary CPC patients underestimated their curvature compared to PD (42% vs. 4%).

CONCLUSION: In order to improve patients' satisfaction rates, the surgeon needs to take into consideration the patient's perception of the deformity when planning the type of surgical correction.

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PMID

29570261 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=29570261>]

Institution

(Liguori, Chiriaco, Pavan, Trombetta) Department of Urology, University of Trieste, Trieste, Italy

(Salonia) Department of Urology, Università Vita-Salute San Raffaele, Milan, Italy

(Garaffa) St Peter's Andrology and the Institute of Urology, University College London Hospitals, London, United Kingdom

(Cavallini) Andrological Unit, Gynepro Medical Team; Bologna, Italy

Year of Publication

2018

416.

Preoperative care of Polypoid exposed mucosal template in bladder exstrophy: the role of high-barrier plastic wraps in reducing inflammation and polyp size.

Sabetkish N., Sabetkish S., Kajbafzadeh A.-M.

Embase

International braz j urol : official journal of the Brazilian Society of Urology. 44 (3) (pp 591-599), 2018. Date of Publication: 01 May 2018.

[Article]

AN: 623212587

MATERIALS AND METHODS: Eight patients with bladder exstrophy-epispadias complex (BEEC) that had used a low density polyethylene (LDPE) wrap for coverage of the exposed polypoid bladder in preoperative care management were referred. The main complaint of their parents was increase in size and number of polyps. After a period of 2 months using the same wrap and observing the increasing pattern in size of polyps, these patients were recommended to use a high-barrier wrap which is made of polyvinylidene chloride (PVdC), until closure. Patients were monitored for the number and size of polyps before and after the change of barriers. The incidence of para-exstrophy skin infection/inflammation and skin allergy were assessed. Biopsies

were taken from the polyps to identify histopathological characteristics of the exposed polyps.

RESULTS: The high barrier wrap was applied for a mean $\pm$ SD duration of 12 $\pm$ 2.1 months.

Polyps' size and number decreased after 12 months. No allergic reaction was detected in patients after the usage of PVdC; three patients suffered from low-grade skin allergy when LDPE was applied. Also, pre-malignant changes were observed in none of the patients in histopathological examination after the application of PVdC.

CONCLUSION: Polyps' size and number and skin allergy may significantly decrease with the use of a high-barrier wrap. Certain PVdC wraps with more integrity and less evaporative permeability may be more "exstrophy-friendly".

OBJECTIVE: To assess the role of high-barrier plastic wrap in reducing the number and size of polyps, as well as decreasing the inflammation and allergic reactions in exstrophy cases, and to compare the results with the application of low-barrier wrap.

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Institution

(Sabetkish, Sabetkish, Kajbafzadeh) Pediatric Urology and Regenerative Medicine Research Center, Section of Tissue Engineering and Stem Cells Therapy, Children's Hospital Medical Center, Tehran University of Medical Sciences, Tehran, Iran, Islamic Republic of

Year of Publication

2018

417.

Sexual complications of penile fracture in men who have sex with men.

Barros R., Lacerda G., Schul A., Ornellas P., Koifman L., Favorito L.A.

Embase

International braz j urol : official journal of the Brazilian Society of Urology. 44 (3) (pp 550-554), 2018. Date of Publication: 01 May 2018.

[Article]

AN: 623211750

OBJECTIVES: Evaluate the demographic data, etiology, operative findings and results of surgical treatment of penile fracture (PF) in men who have sex with men(MSM) with emphasis on sexual

complications. MATERIALS AND METHODS: We studied 216 patients underwent surgical correction of PF at our hospital. Patients self-identified as MSM were followed for at least 6 months. Demographic data, presentation, operative findings, International Index of Erectile Function - 5 (IIEF-5) and the Premature Ejaculation Diagnostic Tool.

RESULTS: Of 216 PF cases, 4 (1.8%) were MSM. All cases resulted from sexual activity and all patients reported using the "doggy style" position during anal intercourse. Unilateral or bilateral injury of corpus cavernosum was found in 2 patients each. One (25%) patient had complete urethral injury associated with bilateral corpus cavernosum lesion. During the follow-up period, all patients developed some type of sexual complication. One patient reported penile pain during intercourse. Another patient experienced low sexual desire and premature ejaculation. This patient was also dissatisfied with the aesthetic result of the surgical scar and complained about decreased penis size after surgery. The third case developed delayed ejaculation. The fourth patient experienced mild to moderate erectile dysfunction. This same patient presented with penile curvature. Finally, palpable fibrotic nodules in the operative area were observed in all cases.

CONCLUSIONS: Sexual activity in the "doggy style" position was the commonest cause of PF in MSM. Sexual dysfunction is always present in gay man after surgery for PF. However, additional studies with larger samples should be conducted.

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Institution

(Barros, Lacerda, Schul, Ornellas, Koifman) Hospital Municipal Souza Aguiar, RJ, Rio de Janeiro, Brazil (Favorito) Universidade Estadual do Rio de Janeiro (UERJ), RJ, Brazil

(Favorito) Hospital Federal da Lagoa, RJ, Rio de Janeiro, Brazil

Year of Publication

2018

418.

Patient satisfaction with correction of congenital penile curvature. Satisfaccion en pacientes con correccion de pene curvo congenito. <Satisfaccion en pacientes con correccion de pene curvo congenito.>

Mayer M; Rey Valzacchi G; Silva Garreton A; Layus O; Gueglio G.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid

MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Actas Urologicas Espanolas. 2017 Dec 29.

[Journal Article]

UI: 29292041

BACKGROUND: Congenital penile curvature is a relatively rare disease, resulting from a deviation of the penis from the body's straight axis when erect. The prevalence is difficult to determine. Although it has been suggested that the condition could affect up to 10% of the male population, most of these deviations are minor, with no clinical or psychological importance, which leads to its underdiagnosis. Effective correction of the deviation can markedly improve the quality of life of adolescents with this condition.

OBJECTIVE: To assess the quality of the sex life of patients diagnosed with congenital penile curvature who underwent surgical correction.

MATERIALS AND METHODS: Design Retrospective, observational cohort study. Data was collected from the medical records of patients who underwent surgical correction of congenital penile curvature from June 2004 to August 2016. The patients completed the following self-administered questionnaires: Sexual Quality of Life Questionnaire-Male (SQOL-M), International Index of Erectile Function 5 (IIEF 5) and "How satisfied are you with the results of the surgery? From 0 to 10".

RESULTS: Twenty-two corporoplasties were performed to correct the patients' congenital penile curvature. The patients' average age was 23.4 years (range, 17-35). The mean deviation prior to surgery was 47.9degree (range, 20degree-90degree). The average score on the SQOL-M was 52 points (range, 6-66). The average score on the IIEF 5 was 22.4 points.

CONCLUSION: Congenital penile curvature profoundly decreases quality of life, and early surgery is fundamental for repairing the anatomical deformation and thereby significantly restores the patients' psychosocial and sexual wellbeing.

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1

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Authors Full Name

Mayer, M; Rey Valzacchi, G; Silva Garreton, A; Layus, O; Gueglio, G.

Institution

Mayer, M. Seccion de Andrologia, Servicio de Urologia, Hospital Italiano de Buenos Aires, Ciudad Autonoma de Buenos Aires, Argentina. Electronic address: mayermilena@hotmail.com.

Rey Valzacchi, G. Seccion de Andrologia, Servicio de Urologia, Hospital Italiano de Buenos Aires, Ciudad Autonoma de Buenos Aires, Argentina.

Silva Garreton, A. Seccion de Andrologia, Servicio de Urologia, Hospital Italiano de Buenos Aires, Ciudad Autonoma de Buenos Aires, Argentina.

Layus, O. Seccion de Andrologia, Servicio de Urologia, Hospital Italiano de Buenos Aires, Ciudad Autonoma de Buenos Aires, Argentina.

Gueglio, G. Seccion de Andrologia, Servicio de Urologia, Hospital Italiano de Buenos Aires, Ciudad Autonoma de Buenos Aires, Argentina.

Year of Publication

2017

419.

The hypospadias classification affected the surgical outcomes of staged oral mucosa graft urethroplasty in hypospadias reoperation: An observational study.

Zheng D; Fu S; Li W; Xie M; Guo J; Yao H; Wang Z.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present
Medicine. 96(47):e8238, 2017 Nov.

[Journal Article. Observational Study]

UI: 29381913

The staged graft urethroplasty is a recommended technique for repairing complex hypospadias. This retrospective study aimed to investigate the outcomes of this technique in hypospadias patients undergoing reoperation and to analyze the underlying contributing factors including age, meatus location, and graft and suture type. We retrospectively analyzed 40 hypospadias patients undergoing reoperation who received a staged oral graft urethroplasty, including 15 buccal mucosal grafts and 25 lingual mucosal grafts. Median age at presentation was 18.5 years, and median follow-up was 17.5 months (range 8-30 months). The patients were classified according to their original meatus location. Twenty-five complications developed in 12 of 40 (30%) cases, including 6 fistulas (15%), 7 infections (17.5%), 9 cases of glans dehiscence (22.5%), and 3 cases of stenosis (7.5%). There was no significant difference in the overall complication rates between prepuberty and postpuberty groups. In addition, no significant difference in complications was found between the 2 graft techniques. The complications were significantly

higher in the original perineal type compared with the original penoscrotal type (7/10 vs 5/30, $P = .0031$). Seven patients who originally had perineal hypospadias developed multiple complications. Based on this study, the staged graft urethroplasty is an effective technique in reoperative hypospadias repairs with reasonable complication risk. The hypospadias classification affects the surgical outcomes.

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Version ID

1

Status

MEDLINE

Authors Full Name

Zheng, Dachao; Fu, Shi; Li, Wenji; Xie, Minkai; Guo, Jianhua; Yao, Haijun; Wang, Zhong.

Institution

Zheng, Dachao. Department of Urology, Shanghai Ninth People's Hospital, Shanghai JiaoTong University School of Medicine, Shanghai, China.

PMID

<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5708912>

Year of Publication

2017

420.

Comparison of tubularized incised plate urethroplasty and onlay island flap urethroplasty techniques in the repair of primarily proximal hypospadias. Comparacion entre la uretroplastia tubularizada con incision de la placa uretral y las tecnicas de Uretroplastia con colgajo pediculado para la reparacion del hipospadias proximal primario. <Comparacion entre la uretroplastia tubularizada con incision de la placa uretral y las tecnicas de Uretroplastia con colgajo pediculado para la reparacion del hipospadias proximal primario.>

Aydogmus Y; Bagbanci S; Demirbas A; Hascicek AM.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid

MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Archivos Espanoles de Urologia. 70(8):740-745, 2017 Oct.

[Journal Article]

UI: 28976350

OBJECTIVES: To compare the success of the 'tubularized incised plate urethroplasty' (TIPU) and 'Onlay island flap urethroplasty' (OIFU) techniques for the repair of primary proximal hypospadias.

MATERIAL AND METHODS: A retrospective evaluation was made of the medical records of 68 patients with primary, non-complicated hypospadias, who were operated on in the Department of Urology, Ministry of Health Ankara Education and Research Hospital, between January 1997 and December 2012. Patients who underwent hypospadias surgery with TIPU were labelled as Group 1 and the OIFU technique as Group 2. Patient age, native meatus localization, operation time, diversion type, surgical success and complication rates were all assessed. Surgical success was evaluated with direct vision of voiding and uroflowmetry at the time of catheter removal, then at 2 weeks, 6 weeks, 6 months and 1 year.

RESULTS: Group 1 consisted of 43 patients and Group 2 of 25 patients. The mean age of patients was 6.4 ± 3.1 years in Group 1 and 8.0 ± 4.6 years in Group 2 ($p=0.09$). The mean operating time was significantly lower in Group 1. (Group 1- 104.8 ± 16.8 min, Group 2- 125.4 ± 24.7 min; $p<0.001$) The distribution of meatus localization was similar in both groups. At 1 year postoperatively, urethrocutaneous fistula was the only complication and the definitive success rates were 81.6% in Group 1 and 72% in Group 2.

CONCLUSION: Despite less frequent usage since the description and popularization of TIPU for proximal hypospadias repair, OIFU remains a valuable technique as a single-stage procedure in cases where the urethral plate is insufficient. Currently, both TIPU and OIFU are used successfully in the treatment of patients with proximal hypospadias.

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1

Status

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Authors Full Name

Aydogmus, Yasin; Bagbanci, Sahin; Demirbas, Arif; Hascicek, Ahmet Metin.

Institution

Aydogmus, Yasin. Department of Urology. Etimesgut Military Hospital. Ankara. Turkey.

Bagbanci, Sahin. Department of Urology. Ahi Evran University School of Medicine. Kirsehir. Turkey.

Demirbas, Arif. Department of Urology. Ankara Training and Research Hospital. Ankara. Turkey.

Hascicek, Ahmet Metin. Department of Urology. Ankara Training and Research Hospital. Ankara. Turkey.

Year of Publication

2017

421.

Correlation Between Primary Hypospadias Repair and Subsequent Urethral Strictures in a Series of 408 Adult Patients.

Barbagli G; Fossati N; Larcher A; Montorsi F; Sansalone S; Butnaru D; Lazzeri M.

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MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

European Urology Focus. 3(2-3):287-292, 2017 04.

[Journal Article. Observational Study]

UI: 28753858

BACKGROUND: The correlation between primary hypospadias repair and subsequent urethral strictures in adults has never been addressed.

OBJECTIVE: To evaluate the correlation between the site of primary hypospadias repair and the site of subsequent strictures and to investigate the predictive factors of failure after urethroplasty.

DESIGN, SETTING, AND PARTICIPANTS: An observational, retrospective, descriptive study of adult patients with urethral strictures following hypospadias surgery was carried out in a single centre.

INTERVENTION: Meatotomy, meatoplasty, end-to-end anastomosis, urethroplasty, perineostomy, urethrotomy, and fistula closure.

OUTCOME MEASUREMENTS AND STATISTICAL ANALYSIS: We performed correlations between the site of primary hypospadias and the site of subsequent strictures, treatment failure, and patient lack of motivation for definitive treatment. Cross-tables, Kaplan-Meier curves, and logistic or Cox regression were used.

RESULTS AND LIMITATIONS: A total of 408 patients, with median follow-up of 96 mo, were included. Concordance between the site of primary hypospadias repair and the site of subsequent strictures was observed. Multivariable analysis revealed that the number of previous operations needed for initial hypospadias repair was not associated with the risk of treatment failure (hazard ratio [HR] 0.96; 95% confidence Interval [CI] 0.88-1.04; $p=0.3$) or a lack of patient motivation (odds ratio 0.99, 95% CI 0.90-1.10; $p=0.9$). Length of stenosis (HR 1.38, 95% CI 1.11-1.71; $p=0.004$) and lichen sclerosus (HR 1.73, 95% CI 1.03-3.25; $p=0.035$) were associated with a higher risk of treatment failure. Our study is not representative of the entire population of patients with hypospadias repair.

CONCLUSIONS: The stricture site is usually consistent with the site of hypospadias. Stricture length, but not the number of previous operations needed for primary hypospadias repair, was associated with the risk of failure.

PATIENT SUMMARY: The number of operations needed for hypospadias repair was not associated with failure of subsequent urethroplasty.

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Version ID

1

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MEDLINE

Authors Full Name

Barbagli, Guido; Fossati, Nicola; Larcher, Alessandro; Montorsi, Francesco; Sansalone, Salvatore; Butnaru, Denis; Lazzeri, Massimo.

Institution

Barbagli, Guido. Centro Chirurgico Toscano, Arezzo, Italy. Fossati, Nicola. Division of Oncology, Unit of Urology, Urological Research Institute, IRCCS Ospedale San Raffaele, Milan, Italy.

Larcher, Alessandro. Division of Oncology, Unit of Urology, Urological Research Institute, IRCCS Ospedale San Raffaele, Milan, Italy.

Montorsi, Francesco. Division of Oncology, Unit of Urology, Urological Research Institute, IRCCS Ospedale San Raffaele, Milan, Italy.

Sansalone, Salvatore. Department of Experimental Medicine and Surgery, University of Tor Vergata, Rome, Italy.

Butnaru, Denis. Research Institute for Urology, Sechenov First Moscow State Medical University, Moscow, Russia.

Lazzeri, Massimo. Department of Urology, Istituto Clinico Humanitas IRCCS, Clinical and Research Hospital, Rozzano, Milan, Italy. Electronic address: massimo.lazzeri@humanitas.it.

Year of Publication

2017

Substitution urethroplasty for anterior urethral stricture repair: comparison between lingual mucosa graft and pedicled skin flap.

Fu Q; Zhang Y; Zhang J; Xie H; Sa YL; Jin S.

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MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Scandinavian Journal of Urology. 51(6):479-483, 2017 Dec.

[Comparative Study. Journal Article]

UI: 28738760

OBJECTIVES: The aim of this study was to compare the effectiveness and outcomes of lingual mucosa graft (LMG) urethroplasty versus pedicled skin flap (PSF) urethroplasty in the repair of anterior urethral strictures.

MATERIALS AND METHODS: A retrospective study in one urological center examined 293 male patients with anterior urethral strictures who received substitution urethroplasty from 2006 to 2015. Of these, 199 patients received PSF urethroplasty and 94 received LMG urethroplasty. The causes of stricture included catheter damage, transurethral instrumentation, trauma, infection, tumor and radiotherapy. Strictures caused by lichen sclerosus, hypospadias repair or failed urethroplasty were excluded. The success rate of PSF versus LMG in different urethra positions was analyzed.

RESULTS: Overall, PSF and LMG had similar success rates (83.4% vs 85.1%, $p = 0.713$). In distal penile urethra, penile skin flaps and LMG achieved similar success rates (87.7% vs 82.1%, $p = 0.297$), but in proximal penile urethra, scrotal flaps had relatively low success rates (69.0% vs LMG 83.3%, $p = 0.345$) and in bulbar urethra, perineal skin flaps had significantly lower success rates than LMG (66.7% vs 92.3%, $p = 0.036$).

CONCLUSION: Lingual mucosa is a good material for the repair of urethral defects and achieves results similar to or even better than those of PSF. Scrotal skin and perineal skin had lower success rates.

Version ID

1

Status

MEDLINE

Authors Full Name

Fu, Qiang; Zhang, Yumeng; Zhang, Jiong; Xie, Hong; Sa, Ying-Long; Jin, Sanbao.

Institution

Fu, Qiang. a Department of Urology, Shanghai 6th Hospital Affiliated to Shanghai Jiaotong University, Shanghai, PR China. Zhang, Yumeng. b Urology, The Second Hospital of Shandong University, Jinan, Shandong, PR China.

Zhang, Jiong. c Shanghai 6th Peoples Hospital Affiliated to Shanghai Jiaotong University School of Medicine, Shanghai, PR China.

Xie, Hong. c Shanghai 6th Peoples Hospital Affiliated to Shanghai Jiaotong University School of Medicine, Shanghai, PR China.

Sa, Ying-Long. d Urology, Shanghai 6th People's Hospital, Jinan, Shadong, PR China.

Jin, Sanbao. c Shanghai 6th Peoples Hospital Affiliated to Shanghai Jiaotong University School of Medicine, Shanghai, PR China.

Year of Publication

2017

423.

Steroid 5-alpha-reductase type 2 (SRD5A2) gene V89L polymorphism and hypospadias risk: A meta-analysis.

Zhang K; Li Y; Mao Y; Ma M.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid

MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Journal of pediatric urology. 13(6):630.e1-630.e9, 2017 Dec.

[Journal Article. Meta-Analysis]

UI: 28713005

BACKGROUND: Hypospadias is a common congenital malformation in males, in which the urethral orifice is found on the ventral side of the penis as a result of incomplete fusion of urethral folds. The etiology of hypospadias is poorly understood, and may be multifactorial, including genetic, endocrine and environmental factors. The steroid 5-alpha-reductase type 2 (SRD5A2) gene, which is mainly expressed in the ventral side of the urethra in the process of male genital development, plays an important role in urethral shaping.

OBJECTIVE: To investigate, with database searches of related published papers, whether SRD5A2 gene V89L polymorphism has an association with hypospadias risk.

METHODS: The following databases were searched for relevant papers, and all published case-control studies of hypospadias were used to perform a meta-analysis: PubMed, Embase, Springer Link, Cochrane Library, China National Knowledge Infrastructure (CNKI), Wanfang, and Weipu. A quality assessment was performed using the Newcastle-Ottawa scale of a case-control study. To assess the strength of the association under various genetic models, odds ratio (OR)

and its 95% confidence interval (CI) were calculated using fixed-effect or random-effects model according to the heterogeneity. Overall and stratified subgroup analyses, including ethnicity, source of controls, sample for DNA extraction, and hypospadias classification, were performed. All data were analyzed using Review Manager 5.3.

RESULTS: This analysis included six eligible case-control studies with 1130 cases and 1279 controls. Overall, there was a statistically significant association between hypospadias risk and V89L polymorphism for allele contrast (C vs G: OR 1.91, 95% CI 1.13-3.23), $P = 0.02$), codominant model (CC vs GG: OR 2.97, 95% CI 1.25-7.04, $P = 0.01$; GC vs GG: OR 2.36, 95% CI 1.35-4.13, $P = 0.003$), dominant model (GC + CC vs GG: OR 2.46, 95% CI 1.28-4.72, $P = 0.007$), and recessive model (CC vs GC + GG: OR 1.91, 95% CI 1.00-3.66, $P = 0.05$). Moreover, there was also a statistically significant association in some subgroups. The positive results are shown in the Summary Table.

CONCLUSION: This meta-analysis suggested that the V89L polymorphism definitely increases the risk of hypospadias, and the C allele is a genetic risk factor for hypospadias occurrence.

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Version ID

1

Status

MEDLINE

Authors Full Name

Zhang, K; Li, Y; Mao, Y; Ma, M.

Institution

Zhang, K. Department of Toxicology, School of Public Health, Shenyang Medical College, Shenyang, Liaoning Province 110034, People's Republic of China. Li, Y. Department of Toxicology, School of Public Health, Shenyang Medical College, Shenyang, Liaoning Province 110034, People's Republic of China.

Mao, Y. Department of Toxicology, School of Public Health, Shenyang Medical College, Shenyang, Liaoning Province 110034, People's Republic of China.

Ma, M. Department of Toxicology, School of Public Health, Shenyang Medical College, Shenyang, Liaoning Province 110034, People's Republic of China. Electronic address: mymacmu@163.com.

Year of Publication

2017

Assisted Reproductive Technology and Birth Defects: Effects of Subfertility and Multiple Births. Liberman RF; Getz KD; Heinke D; Luke B; Stern JE; Declercq ER; Chen X; Lin AE; Anderka M. OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present Birth Defects Research. 109(14):1144-1153, 2017 Aug 15.

[Journal Article]

UI: 28635008

BACKGROUND: Assisted reproductive technology (ART) has been associated with birth defects, but the contributions of multiple births and underlying subfertility remain unclear. We evaluated the effects of subfertility and mediation by multiple births on associations between ART and nonchromosomal birth defects.

METHODS: We identified a retrospective cohort of Massachusetts live births and stillbirths from 2004 to 2010 among ART-exposed, ART-unexposed subfertile, and fertile mothers using linked information from fertility clinics, vital records, hospital discharges, and birth defects surveillance. Log-binomial regression was used to estimate prevalence ratios and 95% confidence intervals (CIs). Mediation analyses were performed to deconstruct the ART-birth defects association into the direct effect of ART, the indirect effect of multiple births, and the effect of ART-multiples interaction.

RESULTS: Of 17,829 ART-exposed births, 355 had a birth defect, compared with 162 of 9431 births to subfertile mothers and 6183 of 445,080 births to fertile mothers. The adjusted prevalence ratio was 1.5 (95% CI, 1.3-1.6) for ART and 1.3 (95% CI, 1.1-1.5) in subfertile compared with fertile deliveries. We observed elevated rates of several birth defects with ART, including tetralogy of Fallot and hypospadias. Subfertility and multiple births affect these associations, with multiple births explaining 36% of the relative effect of ART on nonchromosomal birth defects.

CONCLUSION: Although the risk of birth defects with ART is small, a substantial portion of the relative effect is mediated through multiple births, with subfertility contributing an important role. Future research is needed to determine the impact of newer techniques, such as single embryo transfer, on these risks. Birth Defects Research 109:1144-1153, 2017. © 2017 Wiley Periodicals, Inc.

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Status

MEDLINE

Author Initials

Liberman, Rebecca F; ORCID: <http://orcid.org/0000-0002-7593-4145> Getz, Kelly D; ORCID: <http://orcid.org/0000-0003-2020-5153>

Authors Full Name

Liberman, Rebecca F; Getz, Kelly D; Heinke, Dominique; Luke, Barbara; Stern, Judy E; Declercq, Eugene R; Chen, Xiaoli; Lin, Angela E; Anderka, Marlene.

Institution

Liberman, Rebecca F. Center for Birth Defects Research and Prevention, Massachusetts Department of Public Health, Boston, Massachusetts. Getz, Kelly D. Division of Oncology and the Center for Pediatric Clinical Effectiveness, Children's Hospital of Philadelphia, Philadelphia, Pennsylvania.

Heinke, Dominique. Center for Birth Defects Research and Prevention, Massachusetts Department of Public Health, Boston, Massachusetts.

Luke, Barbara. Department of Obstetrics, Gynecology, and Reproductive Biology, Michigan State University, East Lansing, Michigan.

Stern, Judy E. Department of Obstetrics and Gynecology, Dartmouth-Hitchcock, Lebanon, New Hampshire.

Declercq, Eugene R. Department of Community Health Sciences, Boston University School of Public Health, Boston, Massachusetts.

Chen, Xiaoli. Center for Birth Defects Research and Prevention, Massachusetts Department of Public Health, Boston, Massachusetts.

Lin, Angela E. Medical Genetics Unit, MassGeneral Hospital for Children, Boston, Massachusetts.

Anderka, Marlene. Center for Birth Defects Research and Prevention, Massachusetts Department of Public Health, Boston, Massachusetts.

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2017

Clinical and molecular cytogenetic characterization of four unrelated patients carrying 2p14 microdeletions.

Mathieu ML; Demily C; Chantot-Bastaraud S; Afenjar A; Mignot C; Andrieux J; Gerard M; Catala-Mora J; Jouk PS; Labalme A; Edery P; Sanlaville D; Rossi M.

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American Journal of Medical Genetics. Part A. 173(8):2268-2274, 2017 Aug.

[Journal Article]

UI: 28599093

We report the clinical and molecular cytogenetic characterization of four unrelated patients from France and Spain, carrying 2p14 microdeletions and presenting with intellectual disability and dysmorphisms. 2p14 microdeletions are very rare. Seven patients have been reported so far harboring deletions including 2p14p15 and encompassing OTX1, whose haploinsufficiency is frequently associated with genitourinary defects. To date, only one patient has been reported carrying a more proximal 2p14 microdeletion which does not include OTX1. Here, we report three further patients carrying proximal 2p14 microdeletions not including OTX1 and one patient carrying a more distal 2p14p15 microdeletion including this gene, providing new insights into the associated phenotypic spectrum. First, our study and a review of the literature showed that 3/4 patients carrying proximal 2p14 microdeletions had sensorineural hearing loss, suggesting the presence of a previously unreported deafness-causing gene in this chromosomal region. Second, one patient developed a progressive cardiomyopathy, suggesting that a cardiac follow-up should be systematically warranted even in the absence of congenital heart disease. We speculate that ACTR2 and MEIS1 might respectively play a role in the pathogenesis of the observed deafness and cardiomyopathy. Third, we observed other previously unreported features such as glaucoma, retinopathy, and mild midline abnormalities including short corpus callosum, hypospadias and anteriorly placed anus. Finally, the patient carrying a 2p14p15 deletion including OTX1 had normal kidneys and genitalia, thus confirming that OTX1 haploinsufficiency is not invariably associated with genitourinary defects. In conclusion, our study contributes significantly to delineate the phenotypic spectrum of 2p14 microdeletions.

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Version ID

1

Status

MEDLINE

Author Initials

Rossi, Massimiliano; ORCID: <http://orcid.org/0000-0002-5797-8152>

Authors Full Name

Mathieu, Marie-Laure; Demily, Caroline; Chantot-Bastaraud, Sandra; Afenjar, Alexandra; Mignot, Cyril; Andrieux, Joris; Gerard, Marion; Catala-Mora, Jaume; Jouk, Pierre Simon; Labalme, Audrey; Edery, Patrick; Sanlaville, Damien; Rossi, Massimiliano.

Institution

Mathieu, Marie-Laure. Hospices Civils de Lyon, Service de Genetique, Centre de Reference Anomalies du Developpement, Bron, France. Demily, Caroline. Centre de depistage et de prises en charge des troubles psychiatriques d'origine genetique, CH le Vinatier et UMR 5229 (CNRS et Universite Lyon 1), Bron, France.

Chantot-Bastaraud, Sandra. Assistance Publique-Hopitaux de Paris, Groupe Hospitalier Universitaire de l'Est Parisien-Hopital Armand Trousseau, Departement de Genetique Medicale, Paris, France.

Afenjar, Alexandra. Assistance Publique-Hopitaux de Paris, Groupe Hospitalier Universitaire de l'Est Parisien-Hopital Armand Trousseau, Departement de Genetique Medicale, Paris, France.

Afenjar, Alexandra. Assistance Publique-Hopitaux de Paris, GHUEP, Hopital Armand Trousseau, Centre de Reference 'Malformations et maladies congenitales du cervelet', Paris, France.

Mignot, Cyril. Assistance Publique-Hopitaux de Paris, Groupe Hospitalier Universitaire de l'Est Parisien-Hopital Armand Trousseau, Departement de Genetique Medicale, Paris, France.

Mignot, Cyril. Assistance Publique-Hopitaux de Paris, Departement de Genetique and Centre de Reference Deficiences Intellectuelles de Causes Rares and Groupe de Recherche Clinique UPMC "Deficiences Intellectuelles et Autisme", Groupe Hospitalier Pitie-Salpetriere, Paris, France.

Andrieux, Joris. Institut de Genetique Medicale, Hopital Jeanne de Flandre, CHRU de Lille, Lille, France.

Gerard, Marion. Service de genetique, Hopital Clemenceau, CHU de Caen, Caen, France.

Catala-Mora, Jaume. Oftalmologia Infantil, Hospital el Pilar, Barcelona, Spain.

Jouk, Pierre Simon. Centre de reference des anomalies du developpement, Service de Genetique, Hopital Couple-Enfant, CHU de Grenoble, Grenoble, France.

Labalme, Audrey. Hospices Civils de Lyon, Service de Genetique, Centre de Reference Anomalies du Developpement, Bron, France.

Edery, Patrick. Hospices Civils de Lyon, Service de Genetique, Centre de Reference Anomalies du Developpement, Bron, France.

Edery, Patrick. INSERM U1028, CNRS UMR5292, UCB Lyon 1, Centre de Recherche en Neurosciences de Lyon, GENDEV Team, Lyon, France.

Sanlaville, Damien. Hospices Civils de Lyon, Service de Genetique, Centre de Reference Anomalies du Developpement, Bron, France.

Sanlaville, Damien. INSERM U1028, CNRS UMR5292, UCB Lyon 1, Centre de Recherche en Neurosciences de Lyon, GENDEV Team, Lyon, France.

Rossi, Massimiliano. Hospices Civils de Lyon, Service de Genetique, Centre de Reference Anomalies du Developpement, Bron, France.

Rossi, Massimiliano. INSERM U1028, CNRS UMR5292, UCB Lyon 1, Centre de Recherche en Neurosciences de Lyon, GENDEV Team, Lyon, France.

Year of Publication

2017

426.

Unexpected ethical dilemmas in sex assignment in 46,XY DSD due to 5-alpha reductase type 2 deficiency.

Byers HM; Mohnach LH; Fechner PY; Chen M; Thomas IH; Ramsdell LA; Shnorhavorian M; McCauley EA; Amies Oelschlager AE; Park JM; Sandberg DE; Adam MP; Keegan CE.

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American Journal of Medical Genetics. Part C, Seminars in Medical Genetics. 175(2):260-267, 2017 06.

[Journal Article]

UI: 28544750

Sex assignment at birth remains one of the most clinically challenging and controversial topics in 46,XY disorders of sexual development (DSD). This is particularly challenging in deficiency of 5-alpha reductase type 2 given that external genitalia are typically undervirilized at birth but typically virilize at puberty to a variable degree. Historically, most individuals with 5-alpha reductase deficiency were raised females. However, reports that over half of patients who underwent a virilizing puberty adopted an adult male gender identity have challenged this practice. Consensus guidelines on assignment of sex of rearing at birth are equivocal or favor male assignment in the most virilized cases. While a male sex of rearing assignment may avoid lifelong hormonal therapy and/or allow the potential for fertility, female sex assignment may be more consistent with external anatomy in the most severely undervirilized cases. Herein, we describe five patients with 46,XY DSD due 5-alpha-reductase type 2 deficiency, all with a severe phenotype. An interdisciplinary DSD medical team at one of two academic centers evaluated each patient. This case series illustrates the complicated decision-making process of assignment of sex of rearing at birth

in 5-alpha reductase type 2 deficiency and the challenges that arise when the interests of the child, parental wishes, recommendations of the medical team, and state law collide.

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Status

MEDLINE

Author Initials

Byers, Heather M; ORCID: <https://orcid.org/0000-0002-9971-1368> McCauley, Elizabeth A;

ORCID: <https://orcid.org/0000-0001-9763-9262>

Sandberg, David E; ORCID: <https://orcid.org/0000-0002-7389-8283>

Adam, Margaret P; ORCID: <https://orcid.org/0000-0001-9817-5121>

Keegan, Catherine E; ORCID: <https://orcid.org/0000-0002-7780-6542>

Authors Full Name

Byers, Heather M; Mohnach, Lauren H; Fechner, Patricia Y; Chen, Ming; Thomas, Inas H; Ramsdell, Linda A; Shnorhavorian, Margaret; McCauley, Elizabeth A; Amies Oelschlager, Anne-Marie E; Park, John M; Sandberg, David E; Adam, Margaret P; Keegan, Catherine E.

Institution

Byers, Heather M. Division of Medical Genetics, University of Washington, Seattle, Washington.

Mohnach, Lauren H. DSD Program, Department of Pediatrics, University of Michigan, Ann Arbor, Michigan.

Fechner, Patricia Y. Division of Endocrinology, Seattle Children's Hospital, Seattle, Washington.

Fechner, Patricia Y. Division of Pediatric Psychology, Seattle Children's Hospital, Seattle, Washington.

Chen, Ming. Division of Endocrinology, Department of Pediatrics, University of Michigan, Ann Arbor, Michigan.

Thomas, Inas H. Division of Endocrinology, Department of Pediatrics, University of Michigan, Ann Arbor, Michigan.

Ramsdell, Linda A. Division of Genetic Medicine, Seattle Children's Hospital, Seattle, Washington.

Shnorhavorian, Margaret. Division of Urology, Seattle Children's Hospital, Seattle, Washington.

McCauley, Elizabeth A. Department of Psychiatry and Behavior Medicine, University of Washington, Seattle, Washington.

Amies Oelschlager, Anne-Marie E. Division of Adolescent Gynecology, Seattle Children's Hospital, Seattle, Washington.

Park, John M. Department of Urology, University of Michigan, Ann Arbor, Michigan.

Sandberg, David E. Division of Pediatric Psychology, Department of Pediatrics, University of Michigan, Ann Arbor, Michigan.

Adam, Margaret P. Division of Medical Genetics, University of Washington, Seattle, Washington.

Adam, Margaret P. Division of Genetic Medicine, Seattle Children's Hospital, Seattle, Washington.

Keegan, Catherine E. Division of Genetics, Department of Pediatrics, University of Michigan, Ann Arbor, Michigan.

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427.

Use of Hormones, Tissue Factors and Bioengineering in the Management of Hypospadias.

[Review]

Mitra A; Sarin YK.

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MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Indian Journal of Pediatrics. 84(7):564-569, 2017 Jul.

[Journal Article. Review]

UI: 28429283

Hypospadiology is a rapidly evolving field. Progress in the understanding of how hormonal therapy affects the growth of the phallus has allowed surgeons to optimize the tissues for surgery. But conflicting data from a number of studies and a lack of consensus on drugs, their dosing, mode of delivery and timing of use means that the creation of protocols is unlikely to happen in the near future. Nonetheless, there is a hope and the standardization of scientific reporting will make it easier to compare data at the global level. There are reports of the increasing incidence of hypospadias and the etiology is thought to be multifactorial. Although complex interactions between genetic polymorphisms and the environment make it difficult to identify the exact factors responsible for hypospadias, the advent of massively parallel gene sequencing, large scale

epigenetic screens and CRISPR technology will definitely ease the process. The knowledge of culprit genes will not only broaden our understanding of embryology and growth but will also enable us to predict and/or modify tissue healing. Advances in tissue engineering are also expected to provide a plethora of biomaterials for urethral reconstruction. The development of this field is directly linked with the elucidation of the processes of proliferation and vascularization coupled with the cataloguing of the growth factors involved. One can safely conclude that the exciting new advances in the field will have far reaching consequences on patient care and counselling.

Version ID

1

Status

MEDLINE

Authors Full Name

Mitra, Aparajita; Sarin, Yogesh Kumar.

Institution

Mitra, Aparajita. Department of Pediatric Surgery, Maulana Azad Medical College and Associated Lok Nayak Hospital, New Delhi, 110002, India. Sarin, Yogesh Kumar. Department of Pediatric Surgery, Maulana Azad Medical College and Associated Lok Nayak Hospital, New Delhi, 110002, India. yksarin@hotmail.com.

Year of Publication

2017

428.

The incidence of isolated penile torsion in North India: A study of 5,018 male neonates.

Bhat A; Bhat M; Kumar V; Goyal S; Bhat A; Patni M.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid

MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Journal of pediatric urology. 13(5):491.e1-491.e6, 2017 Oct.

[Journal Article]

UI: 28344020

OBJECTIVE: Congenital penile torsion is a three-dimensional deformity with helical rotation of the distal corporal bodies with the penile crurae remaining fixed to the pubic rami. The first case of

congenital penile torsion (hypospadias) was described in 1857. Isolated penile torsion is an under-reported anomaly. The reported incidence of isolated penile torsion is 1.7-27% and severe torsion is 0.7%. There are no studies available from Indian subcontinent on the incidence of isolated penile torque. The objective of this study was to determine the overall incidence of isolated penile torque in a north Indian population.

MATERIALS AND METHODS: A prospective study of deliveries of male children was conducted at our institute between April 2014 and June 2015. Penile torsion was measured using a small protractor either by the deviation of the median raphae or the direction of the meatus. Data were collected on the incidence of congenital isolated penile torsion, including the degree and direction (left or right) of torsion. Torsion was classified as mild (<450), moderate (450-900), and severe (>900). Statistical analysis was done using the chi-square test with variables of age and parity of the mother and weight of the child.

RESULTS: There were 99 cases of isolated penile torque among 5018 male neonates assessed for penile torque. The incidence of isolated penile torque was 19.7 per 1000 births. The degree of torsion varied from 30 to 110degree (average 51.46degree). Seventy-nine percent (79%) of them had left side and 21% had right side torque (4:1). The degree of torsion was mild in 30%, with 20% having left side torque and 10% having right side torque (2:1). A moderate degree of torsion was seen in 69%: 84% of them had left torque and only 16% had right sided torque (5:1). Only one patient had severe left torque. The incidence of isolated congenital penile torsion was highest in the maternal age group of >30 years followed by the 26-30-year age group, and was lowest in 21-25 year age group. In multiparous women, the incidence of isolated congenital penile torsion was highest (2.54%), and it was lowest in primiparous women (1.36%).

CONCLUSION: The incidence of isolated penile torsion was 1.97% and the left-to-right ratio was 3:1, but for moderate torque it was 5:1. There was a strong association between incidence of penile torque with the age of the mother ($p = 0.012$) and parity ($p = 0.008$) but not with the weight of the baby ($p = 0.415$).

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Status

MEDLINE

Authors Full Name

Bhat, Amilal; Bhat, Mahakshit; Kumar, Vinay; Goyal, Suresh; Bhat, Akshita; Patni, Madhu.

Institution

Bhat, Amilal. Department of Urology, Dr S.N. Medical College Jodhpur, Rajasthan, India.
Electronic address: bhatamilal@gmail.com. Bhat, Mahakshit. Department of Surgery, M.G.
Medical College, Jaipur, Rajasthan India.

Kumar, Vinay. Department of Urology, S.P. Medical College, Bikaner, Rajasthan, India.

Goyal, Suresh. Department of Urology, Dr S.N. Medical College Jodhpur, Rajasthan, India.

Bhat, Akshita. Department of Urology, S.P. Medical College, Bikaner, Rajasthan, India;
Department of Obstetric and Gynecology, S.P. Medical College Bikaner, Rajasthan, India.

Patni, Madhu. Department of Obstetric and Gynecology, S.P. Medical College Bikaner,
Rajasthan, India.

Year of Publication

2017

429.

Voiding patterns of adult patients who underwent hypospadias repair in childhood.

Jaber J; Kocherov S; Chertin L; Farkas A; Chertin B.

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Journal of pediatric urology. 13(1):78.e1-78.e5, 2017 Feb.

[Comparative Study. Evaluation Study. Journal Article]

UI: 27887915

OBJECTIVE: This study aimed at evaluating the voiding patterns of adult patients who underwent hypospadias repair in childhood.

METHOD: Following IRB approval 103 (22.7%) of 449 adult patients who underwent hypospadias repair between 1978 and 1993 responded to the following questionnaires: International Prostate Symptom Score (I-PSS) and Short Form 12 questionnaire (SF-12). Uroflowmetry (UF) was performed for all patients. The patients were divided into three groups according to the primary meatus localization. Group I had 63 patients (61.5%) treated for glanular hypospadias, group II had 19 patients (18.4%) treated for distal hypospadias, and group III comprised the remaining 21 patients (20.4%) treated for proximal hypospadias.

RESULTS: The mean +/- SD I-PSS score for all patients who responded to the questionnaire was 2.3 +/- 2.4, and UF was 21.1 +/- 4.3 mL/s. The patients from groups I and III had fewer urinary symptoms compared with those of the group II: 1.3 +/- 1.5, 5.5 +/- 2.4, and 1.6 +/- 1.4,

respectively ($p < 0.0001$). With regards to UF, the patients from the groups I and III did better compared with those from the group II: 22.1 $\pm$ 4.1 mL/s, 18.91 $\pm$ 4.2 mL/s, and 20.11 $\pm$ 3.42 mL/s, respectively ($p = 0.021$) (Figure). The UF was better in patients with normal vs. abnormal IPSS ($p = 0.0064$). The physical component summary was 49.8 $\pm$ 10.3, 51.1 $\pm$ 3.6, and 46.4 $\pm$ 0.3 in groups I, II, and III, respectively. The mental summary component was 42.64 $\pm$ 4.1, 42.2 $\pm$ 2.4, and 39.89 $\pm$ 2.9 in groups I, II, and III, respectively.

CONCLUSIONS: Most of the adult patients who underwent hypospadias repair in childhood had normal or mild voiding disturbance, with no effects on their physical or mental status.

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Version ID

1

Status

MEDLINE

Authors Full Name

Jaber, Jawdat; Kocherov, Stanislav; Chertin, Leonid; Farkas, Amicur; Chertin, Boris.

Institution

Jaber, Jawdat. Department of Pediatric Urology, Shaare Zedek Medical Center, Faculty of Medical Science, Hebrew University, Jerusalem, Israel. Kocherov, Stanislav. Department of Pediatric Urology, Shaare Zedek Medical Center, Faculty of Medical Science, Hebrew University, Jerusalem, Israel.

Chertin, Leonid. Department of Pediatric Urology, Shaare Zedek Medical Center, Faculty of Medical Science, Hebrew University, Jerusalem, Israel.

Farkas, Amicur. Department of Pediatric Urology, Shaare Zedek Medical Center, Faculty of Medical Science, Hebrew University, Jerusalem, Israel.

Chertin, Boris. Department of Pediatric Urology, Shaare Zedek Medical Center, Faculty of Medical Science, Hebrew University, Jerusalem, Israel. Electronic address:

boris.chertin@gmail.com.

Year of Publication

2017

Hypospadias repair with the glanular-frenular collar (GFC) technique.

Ozbey H; Etker S.

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Journal of pediatric urology. 13(1):34.e1-34.e6, 2017 Feb.

[Journal Article]

UI: 27847256

BACKGROUND: In the normal human penis, the glans wings merge in the midline ventrally, but are separated by the 'septum glandis' in conjunction with the frenulum. The frenulum is also included in the formation of the distal (glanular and subcoronal) urethra, which has a special part known as the 'fossa navicularis'. This has inspired a hypospadias repair technique that simulates the development of the glanular and subcoronal urethra, which can be incorporated into the repair of all cases of hypospadias.

MATERIAL AND METHOD: A total of 121 patients with varying degrees of hypospadias underwent surgery with the described technique: a Y-V plasty was used to dissect the inner foreskin, in a fashion that allowed for its ventral mobilization as a frenular mucosal collar. After tubularization of the proximal urethra, a partial spongioplasty was performed that extended up to the subcoronal level. The glans wings were approximated only at their outermost convexities, with a couple of subepithelial sutures, leaving a slit for the meatus. The cleft-like area between the split wings of the glans penis was filled with the terminal ends of the spongiosum and the dartos of the mucosal collar, which converged to form a septum and a neo-frenulum (glanular-frenular collar, GFC). The midline skin closure of the ventral collar and the circumferential foreskin closure was completed as usual.

RESULTS: At a mean follow-up of 10 months, two patients developed urethral fistula (2%), six had meatal stenosis (5%), and two had glans dehiscence (2%) that resulted in meatal retraction. Overall, patients had a cosmetically satisfying appearance (Figure). Forty-one received secondary circumcision; the parents of 80 (66%) patients were satisfied with the final foreskin appearance obtained with this method.

DISCUSSION: The split wings of the glans penis or so-called ventral cleft between the glans wings that accommodate the frenulum is part of normal anatomy. Hence, in hypospadias surgery, the approximated glans wings should allow for ventral support of the glanular and subcoronal urethra through a reconstructed neo-frenulum. Neither glanular surface enhancement nor extensive dissection of the glans wings and their full-length approximation are necessary, and may in fact be counter-productive.

CONCLUSIONS: The employment of a GFC provided: 1) an anatomical restoration of the distal (glanular and subcoronal) urethra, supported by a frenulum; 2) a protective (undissected) dartos

layer over the distal part of the tubularized neourethra; and 3) a space for the re-formation of the fossa navicularis.

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Version ID

1

Status

MEDLINE

Authors Full Name

Ozbey, Huseyin; Etker, Seref.

Institution

Ozbey, Huseyin. Department of Pediatric Surgery and Division of Pediatric Urology, Istanbul Medical Faculty, Istanbul University, Istanbul, Turkey; Society for Sexual Development and Hypospadias (DSDturk), Istanbul, Turkey. Electronic address: hozbey@istanbul.edu.tr. Etker, Seref. Society for Sexual Development and Hypospadias (DSDturk), Istanbul, Turkey.

Comments

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Year of Publication

2017

431.

To Finish the Cut or Not.

Zamilpa I; Patel A; Booth J; Canon S.

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Clinical Pediatrics. 56(2):157-161, 2017 Feb.

[Journal Article]

UI: 27162177

We retrospectively evaluated the management of patients with unrecognized glanular hypospadias and a completed (group 1) or aborted (group 2) neonatal circumcision. The rate and type of subsequent surgeries performed were analyzed. Penile curvature, urinary stream deviation, and their impact on management were evaluated. Surgery was done in 55% of

patients-40% of group 1 and 86% of group 2. Completion of the circumcision was done in 63% of group 2. Hypospadias repair was performed in 56% of group 1 and in 34% of group 2. Penile curvature rate did not affect the rate or type of surgery performed. Urinary stream deviation did not affect the rate of repair, but was a significant factor leading to hypospadias repair. We concluded that providers performing neonatal circumcisions do not have to abort the procedure when a glanular hypospadias is noticed. Most patients will require circumcision completion only.

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1

Status

MEDLINE

Authors Full Name

Zamilpa, Ismael; Patel, Ashay; Booth, Joshua; Canon, Stephen.

Institution

Zamilpa, Ismael. 1 Arkansas Children's Hospital, Little Rock, AR, USA. Patel, Ashay. 1

Arkansas Children's Hospital, Little Rock, AR, USA.

Booth, Joshua. 2 Arkansas Children's Hospital, Fayetteville, AR, USA.

Canon, Stephen. 3 University of Arkansas for Medical Sciences, Little Rock, AR, USA.

Year of Publication

2017

432.

EXPERIENCE WITH MANAGING CHILDHOOD URETHROCUTANEOUS FISTULA AT IBADAN.

Takure AO; Adebayo SA; Sotunbi PT; Olapade-Olaopa EO; Okeke LI; Shittu OB.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid

MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Journal of the West African Colleges of Surgeons. 7(3):44-58, 2017 Jul-Sep.

[Journal Article]

UI: 30525002

Background: Urethrocuteaneous fistula could be a distressing condition to the child and parents alike. Its management could be challenging and requires adequate expertise.

Aim: To review the characteristics and aetiology of urethrocuteaneous fistula managed in our division over a ten-year period.

Methodology: All children with urethrocuteaneous fistulae from July 2006 to June 2015 were subject of this review. The demography, aetiology, type of fistula, operation performed and the outcome were retrieved from the division operation book and case notes of the patients. The data was analyzed using SPSS Inc. version 20 and odd ratio.

Results: Thirty-five children were managed over a period of ten years. The age ranged from 6 months to 13 years with a mean of 4+/-1.9years. Seventy one percent of urethrocuteaneous fistulae resulted from complication of childhood male circumcision procedures performed in private hospitals and by nurses. One child (3%) had isolated perineal urethrocuteaneous fistula while 26% complicated hypospadias repair at these locations: glandular in 1% case, subcoronal in 3% cases, penile in 2% cases, and penoscrotal in 3% cases. Ten (29%) children with abnormal haemoglobin AC was noted in 3 (9%) patients and haemoglobin AS in 7 (20%) patients. The odd ratio between abnormal haemoglobin and normal haemoglobin was 3.8. The surgical repair of post-circumcision urethrocuteaneous fistulae and post-hypospadias had a recurrent fistulae in 4 (16%) and 3 (33%) respectively. Majority of the fistulae were repaired by simple closure in 80% post-circumcision and in 44% post-hypospadias repair. in the more difficult cases, penile degloving with urethral mobilization was done in 16% post-circumcision fistula and 22% post-hypospadias fistula with no recurrence.

Conclusion: in this study, post-circumcision urethrocuteaneous fistula was the commonest cause of childhood urethrocuteaneous fistula, the severe ones could require penile degloving to achieve repair without tension; recurrence was a major complication.

Version ID

1

Status

PubMed-not-MEDLINE

Authors Full Name

Takure, A O; Adebayo, S A; Sotunbi, P T; Olapade-Olaopa, E O; Okeke, L I; Shittu, O B.

Institution

Takure, A O. Department of Surgery, University College Hospital, Ibadan, Nigeria. Adebayo, S

A. Department of Surgery, University College Hospital, Ibadan, Nigeria.

Sotunbi, P T. Department of Surgery, University College Hospital, Ibadan, Nigeria.

Olapade-Olaopa, E O. Department of Surgery, University College Hospital, Ibadan, Nigeria.

Okeke, L I. Department of Surgery, University College Hospital, Ibadan, Nigeria.

Shittu, O B. Department of Surgery, University College Hospital, Ibadan, Nigeria.

PMID

<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC6237404>

Year of Publication

2017

433.

Current Status of Tissue Engineering in the Management of Severe Hypospadias. [Review]

Abbas TO; Mahdi E; Hasan A; AlAnsari A; Pennisi CP.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid

MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Frontiers in Pediatrics. 5:283, 2017.

[Journal Article. Review]

UI: 29404308

Hypospadias, characterized by misplacement of the urinary meatus in the lower side of the penis, is a frequent birth defect in male children. Because of the huge variation in the anatomic presentation of hypospadias, no single urethroplasty procedure is suitable for all situations.

Hence, many surgical techniques have emerged to address the shortage of tissues required to bridge the gap in the urethra particularly in the severe forms of hypospadias. However, the rate of postoperative complications of currently available surgical procedures reaches up to one-fourth of the patients having severe hypospadias. Moreover, these urethroplasty techniques are technically demanding and require considerable surgical experience. These limitations have fueled the development of novel tissue engineering techniques that aim to simplify the surgical procedures and to reduce the rate of complications. Several types of biomaterials have been considered for urethral repair, including synthetic and natural polymers, which in some cases have been seeded with cells prior to implantation. These methods have been tested in preclinical and clinical studies, with variable degrees of success. This review describes the different urethral tissue engineering methodologies, with focus on the approaches used for the treatment of hypospadias. At present, despite many significant advances, the search for a suitable tissue engineering approach for use in routine clinical applications continues.

Version ID

1

Status

PubMed-not-MEDLINE

Authors Full Name

Abbas, Tariq O; Mahdi, Elsadig; Hasan, Anwarul; AlAnsari, Abdulla; Pennisi, Cristian Pablo.

Institution

Abbas, Tariq O. Department of Health Science and Technology, Faculty of Medicine, Aalborg University, Aalborg, Denmark. Abbas, Tariq O. Department of Pediatric Surgery and Urology, Hamad General Hospital, Doha, Qatar.

Abbas, Tariq O. College of Medicine, Qatar University, Doha, Qatar.

Mahdi, Elsadig. Department of Mechanical and Industrial Engineering, Qatar University, Doha, Qatar.

Hasan, Anwarul. Department of Mechanical and Industrial Engineering, Qatar University, Doha, Qatar.

AlAnsari, Abdulla. Department of Urology, Hamad Medical Corporation, Doha, Qatar.

Pennisi, Cristian Pablo. Department of Health Science and Technology, Faculty of Medicine, Aalborg University, Aalborg, Denmark.

PMID

<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5786532>

Year of Publication

2017

434.

A Boy with a Congenital Sagittal Fissure of the Glans Penis Representing an Abortive Isolated Urethral Duplication.

Sfoungaris D; Valioulis I; Mitroudi M; Patoulas I; Panteli C.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid

MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Journal of Clinical and Diagnostic Research JCDR. 11(9):PJ01-PJ02, 2017 Sep.

[Journal Article]

UI: 29207781

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Status

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Authors Full Name

Sfoungaris, Dimitrios; Valioulis, Ioannis; Mitroudi, Magdalini; Patoulas, Ioannis; Panteli, Christina.

Institution

Sfoungaris, Dimitrios. Assistant Professor, Department of Paediatric Surgery, Aristotle University of Thessaloniki, Thessaloniki, Greece. Valioulis, Ioannis. Assistant Professor, Department of Paediatric Surgery, Aristotle University of Thessaloniki, Thessaloniki, Greece.

Mitroudi, Magdalini. Staff Surgeon, Department of Paediatric Surgery, Aristotle University of Thessaloniki, Thessaloniki, Greece.

Patoulas, Ioannis. Staff Surgeon, Department of Paediatric Surgery, Aristotle University of Thessaloniki, Thessaloniki, Greece.

Panteli, Christina. Staff Surgeon, Department of Paediatric Surgery, Aristotle University of Thessaloniki, Thessaloniki, Greece.

PMID

<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5713803>

Year of Publication

2017

435.

Pseudoverrucous Papules and Nodules in an Infant with Penoscrotal Hypospadias.

Kamat D; Malathi M; Prabhakaran N; Thappa DM.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Indian Dermatology Online Journal. 8(6):503-505, 2017 Nov-Dec.

[Journal Article]

UI: 29204405

Version ID

1

Status

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Authors Full Name

Kamat, Divya; Malathi, Munisamy; Prabhakaran, Nagendran; Thappa, Devinder M.

Institution

Kamat, Divya. Department of Dermatology, Jawaharlal Institute of Post Graduate Medical Education and Research, Puducherry, India. Malathi, Munisamy. Department of Dermatology, Jawaharlal Institute of Post Graduate Medical Education and Research, Puducherry, India.

Prabhakaran, Nagendran. Department of Dermatology, Jawaharlal Institute of Post Graduate Medical Education and Research, Puducherry, India.

Thappa, Devinder M. Department of Dermatology, Jawaharlal Institute of Post Graduate Medical Education and Research, Puducherry, India.

PMID

<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5707854>

Year of Publication

2017

436.

Management of High-Grade Penile Curvature Associated With Hypospadias in Children. [Review]
Moscardi PRM; Gosalbez R; Castellan MA.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid
MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present
Frontiers in Pediatrics. 5:189, 2017.

[Journal Article. Review]

UI: 28929092

Penile curvature is a frequent feature associated with hypospadias with also a great variability of severity among each patient. While the low-grade curvature (<30degree) can be relatively easily corrected by simple techniques like penile degloving and dorsal plication, severe cases often demand more complex maneuvers to manage it. A great number of surgical techniques have been developed to adequately correct curvatures greater than 30degree; however, each one of them should be individualized to different patients and local conditions encountered. In this article, we will review the evaluation of the pediatric patient with penile curvature associated with hypospadias with a special attention to high-grade cases, their management, indications for surgical treatment, and several surgical options for their definitive treatment.

Version ID

1

Status

PubMed-not-MEDLINE

Authors Full Name

Moscardi, Paulo R M; Gosalbez, Rafael; Castellan, Miguel Alfredo.

Institution

Moscardi, Paulo R M. Pediatric Urology, Children's Urology Associates, Miami, FL, United States.

Gosalbez, Rafael. Pediatric Urology, Children's Urology Associates, Miami, FL, United States.

Castellan, Miguel Alfredo. Pediatric Urology, Children's Urology Associates, Miami, FL, United States.

Comments

Comment in (CIN)

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<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5591333>

Year of Publication

2017

437.

A rare case of complete penoscrotal transposition with hypospadias in a newborn.

Beyazit F; Pek E; Aylanc H.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid

MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Turkish Journal of Obstetrics & Gynecology. 14(1):74-75, 2017 Mar.

[Journal Article]

UI: 28913140

Version ID

1

Status

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Authors Full Name

Beyazit, Fatma; Pek, Eren; Aylanc, Hakan.

Institution

Beyazit, Fatma. Canakkale 18 Mart University Faculty of Medicine, Department of Obstetrics and Gynecology, Canakkale, Turkey. Pek, Eren. Canakkale 18 Mart University Faculty of Medicine, Department of Obstetrics and Gynecology, Canakkale, Turkey.

Aylanc, Hakan. Canakkale 18 Mart University Faculty of Medicine, Department of Pediatrics, Canakkale, Turkey.

PMID

<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5558323>

Year of Publication

2017

438.

Two Stages Repair of Proximal Hypospadias: Review of 700 Cases.

Shukla AK; Singh AP; Sharma P; Shukla J.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Journal of Indian Association of Pediatric Surgeons. 22(3):158-162, 2017 Jul-Sep.

[Journal Article]

UI: 28694573

OBJECTIVE: Surgical repair of hypospadias is challenging and problematic even for the most experienced specialists, and this is especially true when severe and complicated case is confronted. Many operations had been described for the management of this deformity.

MATERIALS AND METHODS: During the period from May 2004 to December 2015, we performed 700 cases with proximal hypospadias, at our institute in the Department of Pediatric Surgery by a single surgeon. Data were collected retrospectively and included patient's age at operation, degree of the hypospadias, degree of associated chordee, complications, and cosmetic outcome. All patients underwent 2 two-stage procedures with 9-12 months interval in between.

RESULTS: Seven hundred patients with proximal hypospadias were operated upon in a period of 11 years. Byars's 2 two-stage operation was used in all 700 cases. Neither complete disruption nor urethral diverticula occurred 2 two-stage procedures in the 700 patients. 677 (96.7%) patients had no complication. Fistula was present in 21 (3%) patients. There was no case of meatal stenosis in our study.

CONCLUSION: Two stages procedure using the principles of Byar's technique is a versatile operation that can be used for 2 two-stage procedures the proximal hypospadias. It decreases the rate of fistula formation, disruption, and stenosis and gives a satisfactory cosmetic appearance.

Version ID

1

Status

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Authors Full Name

Shukla, Arvind Kumar; Singh, Aditya Pratap; Sharma, Pramila; Shukla, Jyotsna.

Institution

Shukla, Arvind Kumar. Department of Pediatric Surgery, SMS Medical College, Jaipur, Rajasthan, India. Singh, Aditya Pratap. Department of Pediatric Surgery, SMS Medical College, Jaipur, Rajasthan, India.

Sharma, Pramila. Department of Pediatric Surgery, SMS Medical College, Jaipur, Rajasthan, India.

Shukla, Jyotsna. Department of Pediatric Surgery, SMS Medical College, Jaipur, Rajasthan, India.

PMID

<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5473302>

Year of Publication

2017

439.

Congenital urogenital abnormalities in children with congenital hypothyroidism.

Yousefi Chaijan P; Dorreh F; Sharafkhah M; Amiri M; Ebrahimimonfared M; Rafeie M; Safi F.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid

MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Medical Journal of the Islamic Republic of Iran. 31:7, 2017.

[Journal Article]

UI: 28638814

Background: Congenital hypothyroidism (CH), as one of the most common congenital endocrine disorders, may be significantly associated with congenital malformations. This study investigates urogenital abnormalities in children with primary CH (PCH). Methods: This case-control study was conducted on 200 children aged three months to 1 year, referred to Amir-Kabir Hospital, Arak, Iran. One hundred children with PCH, as the case group, and 100 healthy children, as the control group, were selected using convenient sampling. For all children, demographic data checklists

were filled, and physical examination, abdomen and pelvic ultrasound and other diagnostic measures (if necessary) were performed to evaluate the congenital urogenital abnormalities including anomalies of the penis and urethra, and disorders and anomalies of the scrotal contents.

Results: Among 92 (100%) urogenital anomalies diagnosed, highest frequencies with 37 (40.2%), 26(28.2%) and 9 (9.7%) cases including hypospadias, Cryptorchidism, and hydrocele, respectively. The frequency of urogenital abnormalities among 32 children with PCH, with 52 cases (56.5%) was significantly higher than the frequency of abnormalities among the 21 children in the control group, with 40 cases (43.4%). (OR=2.04; 95%CI: 1.1-3.6; p=0.014). Conclusion: Our study demonstrated that PCH is significantly associated with the congenital urogenital abnormalities. However, due to the lack of evidence in this area, further studies are recommended to determine the necessity of conducting screening programs for abnormalities of the urogenital system in children with CH at birth.

Version ID

1

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Authors Full Name

Yousefi Chaijan, Parsa; Dorreh, Fatemeh; Sharafkhah, Mojtaba; Amiri, Mohammad; Ebrahimimonfared, Mohsen; Rafeie, Mohammad; Safi, Fatemeh.

Institution

Yousefi Chaijan, Parsa. Department of Pediatrics Nephrology, AmirKabir Hospital, School of Medicine, Arak University of Medical Sciences, Arak, Iran. Dorreh, Fatemeh. Department of Pediatrics Nephrology, AmirKabir Hospital, School of Medicine, Arak University of Medical Sciences, Arak, Iran.

Sharafkhah, Mojtaba. Students Research Committee, School of Medicine, Arak University of Medical Sciences, Arak, Iran.

Amiri, Mohammad. Department of Emergency Medicine, Valiasr Hospital, School of Medicine, Arak University of Medical Sciences, Arak, Iran.

Ebrahimimonfared, Mohsen. Department of Neurology, Valiasr Hospital, School of Medicine, Arak University of Medical Sciences, Arak, Iran.

Rafeie, Mohammad. Department of Biostatistics and Epidemiology, School of Medicine, Arak University of Medical Sciences, Arak, Iran. rafeie@yahoo.com.

Safi, Fatemeh. Department of Radiology, Valiasr Hospital, School of Medicine, Arak University of Medical Sciences, Arak, Iran.

PMID

<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5473016>

Year of Publication

2017

440.

Hypospadiac Duplication of Anterior Urethra-a Rare Congenital Anomaly.

Goyal B; Gupta S; Goyal P.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid

MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Indian Journal of Surgery. 79(1):62-63, 2017 Feb.

[Journal Article]

UI: 28331269

Duplication of the urethra is a complex and rarely seen congenital anomaly with three anatomic variants: epispadiac (dorsal), hypospadiac (ventral), and Y-type. We report here a case of hypospadiac duplication of anterior urethra with dorsal blind ending urethra in a 9-year-old boy who presented with complaint of passing urine from the ventral aspect of penis.

Version ID

1

Status

PubMed-not-MEDLINE

Author Initials

Goyal, Bhawana; ISNI: 0000 0004 1768 2079 Goyal, Bhawana; GRID: 0000 0004 1768 2079

Gupta, Suresh; ISNI: 0000 0004 1768 2079

Gupta, Suresh; GRID: 0000 0004 1768 2079

Goyal, Parag; ISNI: 0000 0004 1768 2079

Goyal, Parag; GRID: 0000 0004 1768 2079

Authors Full Name

Goyal, Bhawana; Gupta, Suresh; Goyal, Parag.

Institution

Goyal, Bhawana. Department of Pediatric Surgery, IGM, Shimla, HP India. Gupta, Suresh.

Department of Pediatric Surgery, IGM, Shimla, HP India.

Goyal, Parag. Department of Radio-Diagnosis & Imaging, IGM, Shimla, HP India.

PMID

<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5346086>

Year of Publication

2017

441.

Pediatric Penile and Glans Anthropometry Nomograms: An Aid in Hypospadias Management.

Puri A; Sikdar S; Prakash R.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid

MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Journal of Indian Association of Pediatric Surgeons. 22(1):9-12, 2017 Jan-Mar.

[Journal Article]

UI: 28082769

OBJECTIVE: To establish pediatric penile and glans anthropometry nomograms. This may be used as a reference model for penile assessment while managing hypospadias.

PATIENTS AND METHODS: Between October 2012 and September 2013, 263 boys of varying ages (0-16 years) were included in the study. Those with genetic, endocrine disorders, having genital anomaly, undescended testis, neonates, and infants with a nonretractile prepuce, with multiple congenital anomalies and refusal to take part in the study were excluded. Evaluated outcome variables were stretched penile length, glans circumference (GC) at coronal sulcus, glans diameter at coronal sulcus (Gdcl), mid glans diameter, and ventral glans length. Glans ratios were generated by dividing Gdcl by GC. Data were expressed as mean, median, and standard deviation. Correlation between age and variables was evaluated using nonparametric Spearman's rank correlation coefficient.

RESULTS: The patients were divided in six age groups, namely 0-1 (n = 61), 1-3 (n = 37), 3-5 (n = 36), 5-7 (n = 36), 7-12 (n = 45), and >12 years (n = 48). Gdcl was the maximum transverse glans diameter and based on it small glans size varied widely from 8.9 to 35.04 mm for various age groups. Although glans anthropometry showed age-related changes, glans ratio remained relatively constant between 0.49 and 0.53 (mean: 0.5 +/- 0.051, r = 0.29). All the variables except glans ratio showed a significant positive correlation with age (r = 0.954-0.98, P < 0.01).

CONCLUSION: Penile anthropometry nomograms provide a reference model for hypospadias.

This may aid in (a) objective preoperative assessment of glans size (b) patient selection for preoperative hormonal stimulation (c) provides a yardstick for postoperative cosmesis.

Version ID

1

Status

PubMed-not-MEDLINE

Authors Full Name

Puri, Archana; Sikdar, Satyajit; Prakash, Raghu.

Institution

Puri, Archana. Department of Pediatric Surgery, Lady Hardinge Medical College, New Delhi, India. Sikdar, Satyajit. Department of Pediatric Surgery, Lady Hardinge Medical College, New Delhi, India.

Prakash, Raghu. Department of Pediatric Surgery, Lady Hardinge Medical College, New Delhi, India.

PMID

<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5217151>

Year of Publication

2017

442.

Identification of the underlying androgen receptor defect in the Dallas Reifenstein family.

Ahmad Z., Xing C., Panach K., Kittler R., McPhaul M.J., Wilson J.D.

Embase

Journal of the Endocrine Society. 1 (7) (pp 836-842), 2017. Date of Publication: 2017.

[Article]

AN: 624189065

Context: The Dallas Reifenstein family - first described in 1965 - includes 14 known members with partial androgen insensitivity syndrome (PAIS). However, the underlying molecular defect was never identified.

Objective(s): To identify the underlying genetic defect for PAIS in the Dallas Reifenstein family.

Design(s): DNA was purified from scrotal skin fibroblasts, and whole exome sequencing was then performed in four affected men in the family. Additional family members - both affected and unaffected - were subsequently recruited to confirm segregation of the candidate mutations with the PAIS phenotype.

Patient(s): The affected men have PAIS with infertility associated with azoospermia, hypospadias, and gynecomastia.

Result(s): All four men harbored an intronic variant NC_000023.10:g.66788676A > C between exon 1 and exon 2 of the androgen receptor (AR) canonical transcript NM_000044 (complementary DNA position NM_000044: c.1616+22072A > C) predicted to cause an alternatively spliced AR transcript. Reverse transcription (RT) polymerase chain (PCR) experiments detected the predicted PCR product of the alternatively spliced AR transcript, and the mutation segregated with the PAIS phenotype in this family. The transcript includes the insertion of 185 nucleotides with a premature stop codon at chrX:66863131- 66863133, likely resulting in a reduction in AR protein expression due to nonsense-mediated decay.

Conclusion(s): An intronic AR mutation was identified in the Dallas Reifenstein family. The findings suggest that in cases of PAIS without identifiable AR mutations in coding regions, intronic AR mutations should be considered.

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Status

Embase

Institution

(Ahmad) Division of Nutrition and Metabolic Diseases, Center for Human Nutrition, UT Southwestern Medical Center, Dallas, TX 75390, United States (Panach, McPhaul, Wilson)

Division of Endocrinology and Department of Internal Medicine, UT Southwestern Medical Center, Dallas, TX 75390, United States

(Xing, Kittler) McDermott Center for Human Growth and Development, UT Southwestern Medical Center, Dallas, TX 75390, United States

(McPhaul) Quest Diagnostics Nichols Institute, San Juan Capistrano, CA 92665, United States

Publisher

Oxford University Press (E-mail: Chris.Payne@oup.com)

Year of Publication

2017

443.

Duckett versus modified Bracka technique for proximal hypospadias repair a 10-year experience.

Sadeghi A., Mirshemirani A., Tabari A.K., Sadeghian N., Rozroukh M., Ghoroubi J.,
Mohajerzadeh L., Sarafi M.

Embase

Iranian Journal of Pediatrics. 27 (6) (no pagination), 2017. Article Number: e7752. Date of
Publication: December 2017.

[Article]

AN: 623888354

Background: There are various techniques for treatment of proximal hypospadias
disease. Surgical correction is often associated with complications. Proximal hypospadias can be
repaired by Duckett or Bracka two-stage repair procedure. This study was to evaluate the
outcomes, complications and long term follow-up of these two techniques in our referral hospital.

Method(s): From January 2006 to January 2015 totally 1550 cases of hypospadias were
diagnosed in our hospital, of which 164 patients with high type hypospadias underwent Duckett
(78 cases) and modified Bracka (86 cases) surgical repair procedures. Sufficient data were
analyzed for age at operation, type of pathology, chordee type, number of operations,
complications, outcomes and follow-up.

Result(s): One hundred sixty four cases with a mean age of 2.70 ± 2.6 (range 0.5-13) years
underwent proximal hypospadias repair. Follow up ran in average to 5 (range 1-10)
years. Chordee was seen in 19 (11.58%) cases which 15 (78.9%) cases released and 4 (21.1%)
corrected by dorsal Nesbit plication. Fifteen of 164 (9.1%) cases had meatal stenosis: 5 in Bracka
and 10 in Duckett group, all of which were repaired by dilatation. Six patients in Duckett group
and only one in Bracka series had urethrocutaneous fistula. One case in Bracka group and five
cases in Duckett series underwent re-operation.

Conclusion(s): Our study shows that proximal hypospadias repair with modified Bracka procedure
has significantly a lower complication rate, also a better and more cosmetic outcome than Duckett
technique.

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Status

Embase

Institution

(Sadeghi) Department of Pediatric Anesthesiology, Shahid Beheshti University of Medical
Sciences, Tehran, Iran, Islamic Republic of (Mirshemirani, Tabari, Sadeghian, Rozroukh,
Ghoroubi, Mohajerzadeh, Sarafi) Pediatric Surgery Research Center, Research Institute for
Children Health, Shahid Beheshti University of Medical Sciences, Tehran, Iran, Islamic Republic
of

Publisher

Tehran University of Medical Sciences (TUMS) (E-mail: editor@anesthpain.com)

Year of Publication
2017

444.

Male hypogonadism resulting from mutations in the genes for the gonadotropin subunits and their receptors.

Huhtaniemi I.T.

Embase

Contemporary Endocrinology. (pp 127-152), 2017. Date of Publication: 2017.

[Chapter]

AN: 627405078

Mutations in the genes of gonadotropin subunits (CGA, LHB, FSHB, and CGB) and receptors (LHCGR and FSHR) are extremely rare causes of male hypogonadism. No germ line mutations of CGA have been reported, apparently because of the incompatibility of pregnancy maintenance in the absence of hGG. Five inactivating LHB mutations have been described in men with normal prenatal masculinization but arrested pubertal development. The three men so far described with FSHB mutation were azoospermic. Constitutively activating mutations of the LHCGR gene give rise to very early onset familial male-limited precocious puberty (FMPP) also termed testotoxicosis. Inactivating LHCGR mutation results in an array of male phenotypes ranging from micropenis and hypospadias to complete sex reversal (XY, disorder of sexual development), depending on the completeness of receptor inactivation. Inactivating FSHR mutations in men cause a decrease in testicular size and suppressed quality and quantity of spermatogenesis but not azoospermia, and some affected men may be fertile. Only two cases of activating FSHR mutations have been detected, and they suggest that the mutation does not have phenotype in men with otherwise normal endocrine function. The discrepancy between the phenotypes of men with inactivating FSHB (azoospermia) and FSHR (no azoospermia) mutations must be clarified with additional subjects. Information about the genotypic effects of common polymorphisms in gonadotropin and gonadotropin receptor genes is gradually mounting. A common polymorphism in LHB affects bioactivity of the hormone and has multiple mild phenotypic effects, including slow tempo of puberty in boys and is enriched in post-term boys with cryptorchidism. Some FSHB and FSHR polymorphisms have been shown to affect spermatogenesis and the response of

oligozoospermic men to FSH therapy. Such polymorphisms may represent important targets for the pharmacogenetic evaluation of gonadotropin treatment in infertility.

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Status

Embase

Institution

(Huhtaniemi) Institute of Reproductive and Developmental Biology, Department of Surgery and Cancer, Imperial College, London W12 0NN, United Kingdom

Publisher

Humana Press Inc. (E-mail: humana@humanapr.com)

Year of Publication

2017

445.

The Surgical Management of Bladder Polyps in the Setting of Exstrophy Epispadias Complex.

Jayman J., Tourchi A., Shabaninia M., Maruf M., DiCarlo H., Gearhart J.P.

Embase

Urology. 109 (pp 171-174), 2017. Date of Publication: November 2017.

[Article]

AN: 617757118

Objective To investigate the surgical course for treating bladder polyps in patients with exstrophy epispadias complex (EEC). EEC bladder polyps are unique pathologic entities, with a distinct difference between polyps discovered at birth and polyps developed after failed exstrophy closure. Methods A prospectively maintained database of 1300 patients with EEC was reviewed for bladder exstrophy patients with bladder polyps. The following data were obtained: patient demographics, polyp type, past medical and surgical history, and continence outcomes. Polyps were categorized as (1) primary, bladder polyps at birth or during neonatal period; and (2) secondary, bladder polyps following a failed exstrophy closure. Results Of 1300 EEC patients, 43 patients with polyps met the inclusion criteria. All closures for primary polyp patients were successful, and continence outcomes remained similar in early and delayed closures ($P = .689$). Secondary polyp patients were more likely to require augmentation to increase bladder capacity ($P = .033$). Bladder neck reconstruction ($n = 8$) was successful in 83.0% of patients with primary

polyps, and none were successful for those with secondary polyps ($P = .035$). Conclusion
Delaying primary bladder closure for patients with small polypoid bladder templates did not affect
closure or continence outcomes. However, primary and secondary polyps may require different
surgical interventions. Bladder neck reconstruction was markedly less successful in secondary
polyp patients, and secondary polyp patients were more likely to need augmentation to increase
bladder capacity. This study emphasizes the importance of a successful primary closure and
suggests that secondary polyps are an indicator of decreasing potential for bladder growth.

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Author NameID

Gearhart, John P.; ORCID: <http://orcid.org/0000-0002-0177-6121>

Institution

(Jayman, Tourchi, Shabaninia, Maruf, DiCarlo, Gearhart) James Buchanan Brady Urological
Institute, Division of Pediatric Urology, Charlotte Bloomberg Children's Hospital, The Johns
Hopkins University School of Medicine, Baltimore, MD, United States

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446.

A Critical Analysis of Stented and Unstented Tubularized Incised Plate Urethroplasty Through a
Prospective Randomized Study and Assessment of Factors Influencing the Functional and
Cosmetic Outcomes.

El-Karamany T.M., Al-Adl A.M., Omar R.G., Abdel Aal A.M., Eldakhakhny A.S., Abdelbaki S.A.

Embase

Urology. 107 (pp 202-208), 2017. Date of Publication: September 2017.

[Article]

AN: 617402824

Objective To compare, functionally and aesthetically, stented and unstented tubularized incised plate urethroplasty (TIPU). In addition, predictors of repair success were investigated. **Materials and Methods** One hundred ten children with distal hypospadias were included in this study. The length and width of the urethral plate (UP) and transverse glans diameter (TGD) were measured. Patients were randomized into two groups: stented and unstented TIPU. Postoperative assessment of pain was done using Face, Legs, Activity, Cry, Consolability (FLACC) scale, and cosmetic outcome was evaluated using hypospadias objective scoring evaluation. Uroflow was assessed at 6 months in toilet-trained boys. Complications were graded by the Clavien classification system. **Results** In 93 cases, Face, Legs, Activity, Cry, Consolability score, hospital stay, and dressing time were in favor of the unstented group, whereas hypospadias objective scoring evaluation score and uroflow parameters were comparable. TGD and UP width varied significantly between successful and failed cases. Cutoff values of 13.5 mm and 8.5 mm for TGD and UP width, respectively, are required for successful outcome. Acute urinary retention was 12.8% after unstented repair. Detrusor spasm was 47.8% after stented and 8.5% after unstented repair. In both groups, urethrocutaneous fistula of 5.4%, glanular dehiscence of 1.1%, and meatal stenosis of 5.4% were found in cases with small TGD and narrow UP. **Conclusion** Unstented TIPU evades the associated drawbacks of the stent along with lower postoperative pain, hospital stay, and dressing time. Even without a postoperative stent, the complication rate in infants with distal hypospadias is low, and short-term functional and aesthetic outcomes were not compromised.

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Embase

Author NameID

Al-Adl, Ahmed M.; ORCID: <http://orcid.org/0000-0001-7655-0437>

Institution

(El-Karamany, Al-Adl, Omar, Abdel Aal, Eldakhakhny, Abdelbaki) Urology Department, Benha University, Benha, Egypt

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447.

Use of Bipedicled Dorsal Penile Flap With Z Release Incision: A New Option in Redo Hypospadias Surgery.

Elmoghazy H.

Embase

Urology. 106 (pp 188-192), 2017. Date of Publication: August 2017.

[Article]

AN: 616895875

Objective To solve the challenge in redo hypospadias surgery, we tried to use a bipedicled dorsal penile flap with a Z release incision in failed hypospadias cases and reported the outcome.

Materials and Methods Thirty male children with 3 or 4 previous unsuccessful hypospadias surgeries were included in our study. Our technique was done after at least 6 months from the last surgery. A flap of the dorsal penile skin was preserved and the skin lateral to the flap was dissected on each side. A small opening was done in the dartos proximal to flap. The glans was withdrawn through this opening with a ventral transposition of the flap. Z-plasty was used to compensate for the deficient dorsal skin; the Z-plasty had 3 limbs and all were made of equal length. **Results** The mean age of the patients was 5.4 +/- 1.8 years and the mean follow-up was 2.1 +/- 0.7 years. The technique was successful in 80%. Reoperation was required in 3 cases; all cases were managed using a 2-stage buccal mucosal graft. A small fistula at the coronal level developed in 2 cases but closed spontaneously within 1 month. All patients were voiding well and had a vertically oriented meatus at the tip of the glans and satisfactory cosmetic results.

Conclusion Repair of failed hypospadias using a bipedicled dorsal penile skin flap with Z release incision is a safe and simple procedure offering high success rates.

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Institution

(Elmoghazy) Department of Urology, Sohag University, Sohag, Egypt

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448.

Ointment Fistulography: Introducing a Novel Technique for Single or Multiple Urethrocutaneous Fistula Diagnosis After Hypospadias Surgery.

Mozafarpour S., Kajbafzadeh A.-M., Abbasioun R., Habibi A.A., Nabavizadeh B.

Embase

Urology. 106 (pp 231-232), 2017. Date of Publication: August 2017.

[Article]

AN: 616583530

Introduction and Objective Hypospadias is a common congenital malformation of the male genital tract. The most frequent complication after hypospadias repair is urethrocutaneous fistula.¹ Its incidence has been reported up to 35% worldwide.² The diagnosis of these fistulas is sometimes challenging particularly with tiny and multiple fistulas. Usually, parents complain of urinary spraying, sprinkling, or passing a single stream of urine from the undersurface of the penis after the surgery. Urethrocutaneous fistulas are not always visible on physical examination. Locating the fistula and status of the surrounding skin is very important in order to choose the surgical repair technique.³ The patency and anatomy of the distal urethra should also be determined before the repair surgery.⁴ That is why urologists usually use retrograde urethrography or cystoscopy to find the location of fistula. However, these modalities have their own risk for children and are not always helpful. Sometimes the contrast media inserted can clog some fistulas especially near the glans and preclude the diagnosis. We present an easy technique to screen for urethrocutaneous fistulas after hypospadias surgery. Technique In this technique (Video 1), an antibiotic ointment is inserted through the meatus while the base of the penis is held; the ointment is then gently pushed through the meatus. As shown in the video, the ointment will protrude through the fistulas anywhere along the shaft. This technique can also be performed reversely as the ointment inserted from the fistulas will protrude from the meatus. For younger children under the age of 3, we exclusively perform this technique under anesthesia on the repair surgery day, whereas in toddlers we perform this technique in the office and once again under anesthesia right before the repair surgery. Result This technique shows even tiny and multiple fistulas not detectable on physical examinations. We have used this technique in our clinic and have not missed any fistulas. The diameter of the protruded ointment in reverse fistulography is an indicator of the distal urethral diameter. Therefore, in case of narrow ointment diameter, distal

urethral strictures should be suspected as correction of distal obstruction is an important determinant in the success rate of fistula repair surgery.⁵ Conclusion We suggest this diagnostic technique as a safe, inexpensive, easy, office-based, feasible, and reproducible method. Negative urine culture is not required for this examination. Also, there is no need to insert contrast media as in retrograde urethrography or induce anesthesia as in cystoscopy. We believe this simple technique help urologists around the world to diagnose this common complication of hypospadias surgery without the need for special equipment.

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Author NameID

Kajbafzadeh, Abdol-Mohammad; ORCID: <http://orcid.org/0000-0001-7804-6285>

Institution

(Mozafarpour) Department of Urology, Massachusetts General Hospital, Harvard Medical School, Boston, MA, United States (Kajbafzadeh, Abbasioun, Habibi, Nabavizadeh) Pediatric Urology and Regenerative Medicine Research Center, Children's Hospital Medical Center, Tehran University of Medical Sciences, Tehran, Iran, Islamic Republic of

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449.

Emerging role of preputial vascular pattern and postoperative microvessel density in cases of proximal hypospadias: A pilot study.

Narang V., Sen A., Bhardwaj M.

Embase

Indian Journal of Pathology and Microbiology. 60 (4) (pp 521-523), 2017. Date of Publication: October-December 2017.

[Article]

AN: 620505228

Introduction: Understanding of anatomical vascular patterns and anatomy of prepuce is critical for a good outcome in hypospadias surgery. A well-vascularized neourethral and preputial flaps used for repair are exceptionally important for a successful outcome, especially in cases of proximal hypospadias undergoing one-stage procedures.

Objective(s): To evaluate the vascular anatomy of prepuce in cases of proximal hypospadias and to evaluate microvessel density (MVD) by immunohistochemistry and its correlation with postoperative complications.

Material(s) and Method(s): This was a prospective observational study done between November 2013 and March 2015; 33 cases of proximal hypospadias undergoing surgery were evaluated for vascular pattern by intraoperative cold light method and postoperatively by MVD.

Result(s): Twenty-six cases with a predominant vessel pattern were identified (18 of type 1, 7 of type 2, and 1 of type 3), while seven cases had a reticular pattern (type 4) on cold light transillumination. The mean MVD in cases with predominant vascular pattern (Type 1-3) was 64.83. In cases of Type 4 pattern, mean MVD was found to be low 55.57 ($P = 0.37$). Patients who underwent single-stage surgery and developed postoperative complications had a low MVD score (mean 45.88, $P = 0.040$).

Conclusion(s): Cold light transillumination is an effective perioperative test, reliable in the assessment of preputial vascularity. There is no statistically significant difference between the MVD of predominant vascular pattern and reticular pattern signifying that MVD may or may not be good in a given vascular pattern. MVD can be a helpful marker in assessing prognosis of repair in proximal hypospadias.

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Status

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Institution

(Narang, Sen) Department of Pediatric Surgery, MMIMSR, New Delhi, Mullana, Punjab Ambala - 133 207, India (Bhardwaj) Department of Pathology, Dr. RML Hospital and PGIMER, New Delhi, India

Publisher

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450.

Modified tubularized incised plate urethroplasty reduces the risk of fistula and meatal stenosis for proximal hypospadias: a report of 63 cases.

Arshadi H., Sabetkish S., Kajbafzadeh A.-M.

Embase

International Urology and Nephrology. 49 (12) (pp 2099-2104), 2017. Date of Publication: 01 Dec 2017.

[Article]

AN: 618923053

Purpose: To report the feasibility of modified tubularized incised plate (TIP) urethroplasty technique for proximal hypospadias in 63 cases.

Method(s): From January 2004 to March 2010, 63 patients underwent one-stage TIP urethroplasty (modified Snodgrass technique repair) using 2-3 of three covering layers (corpus spongiosum, dartos, and tunica vaginalis). The primary meatus was proximal penile, penoscrotal, scrotal, and perineal in 38, 13, 10, and 2 patients, respectively. All patients had chordee that was corrected with dorsal plication. Glanuloplasty was performed in all cases. Complications and cosmetic results were documented after 6-72 months of follow-up.

Result(s): A total of 63 boys with proximal hypospadias underwent Snodgrass hypospadias repair at a mean age of 8.5 months (range 6-54). Mean operative time was 210 +/- 35 min. Patients were followed up with 6-month intervals for up to 6 years postoperatively. After 6 years of follow-up, nine urethrocutaneous fistulae, four bleeding, four meatal stenoses, and one urethral stricture were reported. Cosmetic result was satisfactory according to parent's opinion and another surgeon. No residual chordee was observed in any cases (without artificial correction).

Conclusion(s): In conclusion, this preliminary report can be estimated as an alternative technique with acceptable complication and cosmetic results for proximal hypospadias correction.

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Embase

Institution

(Arshadi, Sabetkish, Kajbafzadeh) Pediatric Urology and Regenerative Medicine Research Center, Section of Tissue Engineering and Stem Cells Therapy, Children's Hospital Medical Center, Tehran University of Medical Sciences, No. 62, Dr. Qarib's Street, Keshavarz Boulevard, Tehran 1419433151, Iran, Islamic Republic of

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Springer Netherlands (E-mail: rbk@louisiana.edu)

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451.

A prospective audit of pain profiles following general and urological surgery in children.

Wilson C.A., Sommerfield D., Drake-Brockman T.F.E., Lagrange C., Ramgolam A., von Ungern-Sternberg B.S.

Embase

Paediatric Anaesthesia. 27 (11) (pp 1155-1164), 2017. Date of Publication: November 2017.

[Article]

AN: 618759112

Background: Postoperative pain is frequently undertreated in children both in hospital and at home following discharge. Pain has both short- and long-term consequences for children, their families, and the healthcare system. A greater understanding of procedure-specific postoperative pain trajectories is required to improve pain management.

Aim(s): To determine the duration and severity of acute postoperative pain experienced by children undergoing 8 different general and urological procedures (primary outcomes). Behavioral disturbance rates, nausea and vomiting scores, and parental satisfaction were also examined during the follow-up period (secondary outcomes).

Method(s): Families of children (0-18 years) undergoing common general and urological procedures were invited to enroll in the study. Children's pain scores, measured using a parental proxy 0-10 numerical rating scale, were collected by telephone interview until pain was resolved. Analgesia prescribed and given, behavioral disturbance, nausea and vomiting scores, the method of medication education communication, and parental satisfaction were also measured.

Result(s): Of 360 patients recruited, 326 complete datasets were available. Patients underwent laparoscopic appendicectomy (57), open appendicectomy (19), circumcision (50), cystoscopy

(52), hypospadias repair (22), inguinal hernia repair (51), orchidopexy (51), or umbilical hernia repair (24). Postoperative pain peaked on the day of or the day after surgery in all groups, and decreased over time. Pain lasted a median duration of 5 postoperative days following open appendicectomy, and 0-2 postoperative days for other procedures. Behavioral disturbance rates closely followed pain scores. Analgesia administration at home varied widely between and within groups.

Conclusion(s): Pain management was inadequate in most of the groups studied, particularly after appendicectomy or umbilical hernia repair, with most children experiencing at least moderate pain on the day of and day after surgery. There was a need for a standardized management, with increased dual analgesia prescribing, to ensure that children receive adequate postoperative analgesia in hospital and at home.

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Author NameID

Drake-Brockman, Thomas F. E.; ORCID: <http://orcid.org/0000-0002-7649-3951> von Ungern-Sternberg, Britta S.; ORCID: <http://orcid.org/0000-0002-8043-8541>

Institution

(Wilson) Department of Paediatric Anaesthesia, Bristol Royal Hospital for Children, Bristol, United Kingdom (Sommerfield, Drake-Brockman, Lagrange, von Ungern-Sternberg) Department of Anaesthesia and Pain Management, Princess Margaret Hospital for Children, Perth, WA, Australia

(Drake-Brockman, von Ungern-Sternberg) School of Medicine and Pharmacology, The University of Western Australia, Perth, WA, Australia

(Ramgolam) Telethon Kids Institute, Perth, WA, Australia

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2017

A new modification of the Duckett technique for one-stage repairing urethral plate transected hypospadias: Another option for severe hypospadias?.

Huang L.-Q., Guo Y.-F., Ge Z., Lu R.-G., Deng Y.-J., Ma G., Chen F.

Embase

International Urology and Nephrology. 49 (12) (pp 2091-2097), 2017. Date of Publication: 01 Dec 2017.

[Article]

AN: 618320411

Objectives: The study aimed to evaluate the new modification of the Duckett technique in decreasing the incidence of urethral strictures for urethral plate transected hypospadias and also explored its clinical application.

Method(s): Thirty-three patients (aged 7 months to 12 years, mean age 2.8 years) who underwent repair of primary hypospadias using the new one-stage urethroplasty were enrolled. Clinical data, including cosmetic and its complications, and uroflowmetry data were documented. Uroflowmetry data of 19 patients who underwent Duckett urethroplasty were used as a comparison.

Result(s): The length of the urethral defect ranged from 2.5 to 5.0 cm. The postoperative follow-up was 14-30 months. Ten patients (30.3%) had fistulas; no patients had strictures or diverticula. All ten fistulas were small (<0.5 cm) and repaired with fistula repairing operation. The appearance of the penis remained satisfactory, and the meatus was located in the normal anatomic position. Among 17 patients who underwent uroflowmetry, all patients were bell-shaped or platforms, and Qmax was 7.37 +/- 2.45 ml/s. Compared with 14 of 19 patients who underwent Duckett urethroplasty, the urethral function achieved with new one-stage urethroplasty was significantly better ($p < 0.05$).

Conclusion(s): The incidence of strictures was dramatically lowered in patients with proximal hypospadias. Small fistulas are common complications and can be repaired easily. Based on the uroflow pattern results, the quality of neourethra and function of it were better than Duckett urethroplasty. These preliminary results suggested that the modified procedure seems to be reliable and can be a suitable option for proximal hypospadias.

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Institution

(Huang, Guo, Ge, Lu, Deng, Ma) Department of Urology, Children's Hospital of Nanjing Medical University, 72 Guangzhou Road, Nanjing, Jiangsu 210008, China (Chen) Department of Urology, Shanghai Children's Hospital, Shanghai Jiaotong University, Shanghai, China

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453.

Clinical and genetic features of 64 young male paediatric patients with congenital hypogonadotropic hypogonadism.

Wang Y., Gong C., Qin M., Liu Y., Tian Y.

Embase

Clinical Endocrinology. 87 (6) (pp 757-766), 2017. Date of Publication: December 2017.

[Article]

AN: 618284675

Context: The diagnosis of congenital hypogonadotropic hypogonadism (CHH) in prepuberty has always been challenging. Here, we aimed at studying the clinical and genetic features of paediatric CHH, especially the phenotype of hypospadias and dual defects (patients showing hypothalamic and/or pituitary defects and testicular hypoplasia), so as to have a better understanding of CHH.

Design(s): The clinical and genetic features of patients with CHH were analysed, and the relationships between hypospadias, dual defects and genetics were investigated.

Patient(s): Patients who visited Beijing Children's Hospital and were positively diagnosed with CHH. Measurements: The collected data included sex hormones, MRI of the olfactory bulb, human chorionic gonadotrophin (hCG) test and genetic testing. We analysed clinical features and genetic results, especially hypospadias and dual defects, and compared the stimulated testosterone (T) levels in patients with and without cryptorchidism.

Result(s): Sixty-four patients were positively diagnosed, and forty-seven (73.4%) had Kallmann syndrome (KS). Four patients (6.3%) had hypospadias, including 2 KS. Micropenis combined with cryptorchidism was the most common phenotype (39%). Approximately two-third of patients showed a poor response to hCG; 15 cases were diagnosed with dual defects, and there were no

significant differences between those with and without cryptorchidism. Twenty-six cases (51%) of 51 patients were identified as having classical HH mutations, affecting 10 different genes, with oligogenic mutations in 5 cases (9.8%). The most common mutations were in PROKR2 (17.6%), FGFR1 (13.7%) and CHD7 (7.8%). The frequency of PROKR2 mutations was higher in dual HH when compared to other HH cases (6/15 vs 3/36, $P = .021$).

Conclusion(s): Micropenis and/or cryptorchidism can serve as important signs for the diagnosis of HH in paediatrics, and the coexistence of hypospadias does not exclude the diagnosis of CHH, including KS or normosmic isolated HH (nHH). Testicular function may be impaired earlier than expected, and PROKR2 mutations need to be evaluated to identify presumed dual defects.

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Status

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Author NameID

Gong, Chunxiu; ORCID: <http://orcid.org/0000-0002-1262-7383>

Institution

(Wang, Gong, Qin, Liu, Tian) National Center for Children's Health, Capital Medical University, Beijing, China (Wang, Gong, Qin, Tian) Department of Endocrinology, Genetics, Metabolism and Adolescent Medicine, Beijing Children's Hospital, Capital Medical University, Beijing, China (Wang, Gong, Qin, Tian) Beijing Key Laboratory for Genetics of Birth Defects, Beijing Children's Hospital, Capital Medical University, Beijing, China

(Liu) Department of Pharmacy, Beijing Children's Hospital, Capital Medical University, Beijing, China

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Year of Publication

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454.

Is Concealed Epispadias a Rare Variant?.

Shahat A., Safwat A.S., Elderwy A., Abdelkawi I.F., Abonnoor A.E.I., Abdelhafez M.F., Moeen A., Hammouda H.

Embase

Urology. 109 (pp 165-170), 2017. Date of Publication: November 2017.

[Article]

AN: 618000269

Objective To present our experience with concealed epispadias and to estimate its actual share in the isolated male epispadias cases and its effect on the surgical outcome. **Materials and Methods** Consecutive patients with isolated male epispadias treated in our center between 2008 and 2015 were classified into concealed and classic epispadias. The 2 groups were compared regarding age at presentation, meatal location, incontinence, dorsal curvature, success rate, and complications. **Results** Out of 51 patients with isolated male epispadias, 11 (21.6%) were concealed: 7 balanic and 4 penile shaft epispadias. Concealed epispadias cases were found to have significantly delayed age at presentation, more distal meatal location, and less incontinence rate than classic epispadias cases. None of the surgical outcome parameters showed significant difference between the 2 groups. **Conclusion** Concealed epispadias represents about one-fifth of isolated male epispadias cases. Impediment and delay of diagnosis are its main clinical impacts, with insignificant effect on the surgical outcome.

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Status

Embase

Author NameID

Safwat, Ahmed S.; ORCID: <http://orcid.org/0000-0003-0096-5323>

Institution

(Shahat, Safwat, Elderwy, Abdelkawi, Abonnoor, Abdelhafez, Moeen, Hammouda) Assiut Urology and Nephrology Hospital, Assiut University, Assiut, Egypt

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455.

Current challenges with proximal hypospadias: We have a long way to go.

Gong E.M., Cheng E.Y.

Embase

Journal of Pediatric Urology. 13 (5) (pp 457-467), 2017. Date of Publication: October 2017.

[Review]

AN: 616379114

Background and purpose Proximal hypospadias repair has continued to challenge the pediatric urologist despite great efforts to develop better techniques for repair. **Methods** In this article, we review the literature regarding modern proximal hypospadias repair approaches and outcomes. We also present our current approach to proximal hypospadias repair surgery and outcome tracking. Finally, we review the literature on surgical outcome tools that may help us standardize research. **Results** Proximal hypospadias repair has significant variability in technique. Overall modern data would suggest that we require great improvement in our surgical management of this challenging problem. Though tools exist to begin to standardize the reporting of hypospadias outcomes, collaborative efforts are still required for future research. **Conclusion** Future research in proximal hypospadias repair needs to utilize new reporting tools as well as engage in collaborative efforts between institutions.

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Status

Embase

Author NameID

Gong, Edward M.; ORCID: <http://orcid.org/0000-0002-1240-7380>

Institution

(Gong, Cheng) Ann & Robert H. Lurie Children's Hospital of Chicago, Northwestern University
Feinberg School of Medicine, Chicago, IL, United States

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456.

Update on the surgical approach for reconstruction of the male genitalia.

Romao R.L.P., Pippi Salle J.L.

Embase

Seminars in Perinatology. 41 (4) (pp 218-226), 2017. Date of Publication: June 2017.

[Article]

AN: 615923217

The majority of patients with DSD will be found to carry an XY karyotype and be assigned male gender. From a phenotypical standpoint, most will present with proximal hypospadias +/- cryptorchidism. In this review article, the authors present the current status of reconstruction of the male genitalia in this setting. The report addresses the following topics: surgical input in the evaluation of the newborn with an undervirilized external genitalia, including gender assignment considerations; controversies surrounding timing and indication for hypospadias surgery in proximal cases as well as use of testosterone; surgical techniques and decision-making process for one- vs. two-stage repairs; complications of hypospadias surgery based on technique used for repair; and long-term follow-up. The high complication rates observed in the treatment of proximal hypospadias attest to its challenging nature. Concentration of experience, tracking carefully identified patient-centered outcomes and long-term follow-up of this patient population are recommended.

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Status

Embase

Institution

(Romao) IWK Health Centre, Dalhousie University, Halifax, Nova Scotia, Canada (Pippi Salle)
Division of Urology, Department of Surgery, Sidra Medical and Research Center, PO Box 26999,
Doha, Qatar

Publisher

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457.

Association between a maternal history of miscarriages and birth defects.

Campana H., Rittler M., Gili J.A., Poletta F.A., Pawluk M.S., Gimenez L.G., Cosentino V.R.,
Castilla E.E., Lopez Camelo J.S.

Embase

Birth Defects Research. 109 (4) (pp 254-261), 2017. Date of Publication: 2017.

[Article]

AN: 619490781

BACKGROUND: Some studies, mainly in the older literature, observed a significant association between miscarriages and birth defects (BDs) occurring in the same sibship. However, few studies examined the BD/ miscarriage relationship in depth. In addition nothing has been added to the underlying mechanisms possibly linking both events. The purpose of this work was to identify specific BDs associated with maternal miscarriages. In particular, it examined whether the risk depended on the number of losses, and to suggest the existence of specific factors for each BD/miscarriage association observed.

METHOD(S): The study relied on the Latin American Collaborative Study on Congenital Malformations (ECLAMC) database registries including 26,906 live and stillborn infants with one of 19 selected isolated BDs and 93,853 normal controls. Infants born to primigravid mothers were excluded from the present study. Demographic and reproductive variables were compared between control mothers With and Without previous miscarriages. The number, frequency, and distribution of miscarriages were observed for each BD and controls. A conditional logistic regression was applied to evaluate the miscarriage risk for each BD.

RESULT(S): Control mothers with previous miscarriages were older, had had more pregnancies, and were less educated. Three risk patterns of miscarriages were observed: a very high risk of miscarriages associated with gastroschisis, omphalocele, and talipes; only one miscarriage associated with spina bifida, and two or more miscarriages associated with hypospadias.

CONCLUSION(S): These three patterns suggest that different factors underly each BD/miscarriage association: infertility for hypospadias, vascular disruption for gastroschisis and talipes, while for spina bifida, the much debated trophoblastic cell residue theory could not be discarded.

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Institution

(Campana, Gili, Poletta, Pawluk, Gimenez, Cosentino, Castilla, Lopez Camelo) Latin American Collaborative Study of Congenital Malformations (ECLAMC), Center for Medical Education and Clinical Research (CEMIC), Buenos Aires, Argentina (Rittler) ECLAMC, Hospital Materno Infantil Ramon Sarda, University of Buenos Aires, Argentina

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458.

Glanular hypospadias management with modified circumcision and firilit procedure.

Iqbal N., Hussain I., Cheema M.A., Hijazi M.T., Khan M.A., Lodhi O.H., Akhter S.

Embase

Journal of the College of Physicians and Surgeons Pakistan. 27 (11) (pp 707-710), 2017. Date of Publication: 01 Nov 2017.

[Article]

AN: 619267428

Objective: To document the results of using simple technique of Firilit procedure with modified circumcision for treatment of glanular hypospadias.

Study Design: Descriptive case series. Place and Duration of Study: Department of Urology, Shifa International Hospital, Islamabad, from January 2011 till May 2017. Methodology: The subjects included pediatric patients who underwent repair of glanular hypospadias by using modified circumcision and Firilit procedure. Patients were analyzed for operative time, age, hospital stay, and peroperative and postoperative complications. Chart review was done for collection of data on specified proforma.

Result(s): There were 73 patients with mean age of 1.81 +/-0.92 years. Children were discharged on the same day. None of them needed catheterization and dressing. Temporary dressing was used for mild bleeding immediate post op in 13 (17.80%) cases only. Postoperative complication such as urinary retention, fever, severe pain, or urinary tract infection were not seen in children.

Meatal stenosis was not seen on a 2-24 months' follow-up. Satisfactory urine stream and good cosmesis was noted by the parents.

Conclusion(s): The Firlit technique for the repair of glanular hypospadias was simple and satisfactory in terms of urine stream and cosmesis with minimal complications.

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Status

Embase

Institution

(Iqbal, Hussain, Cheema, Akhter) Department of Urology and Kidney Transplant, Pakistan Kidney Institute, Shifa International Hospital, Islamabad, Pakistan (Hijazi, Khan, Lodhi) Medical Student, Shifa College of Medicine, Islamabad, Pakistan

Publisher

College of Physicians and Surgeons Pakistan (7th Central Street, Karachi 755000, Pakistan)

Year of Publication

2017

459.

Complications of bladder closure in cloacal exstrophy: Do osteotomy and reoperative closure factor in?.

Friedlander D.A., Di Carlo H.N., Sponseller P.D., Gearhart J.P.

Embase

Journal of Pediatric Surgery. 52 (11) (pp 1836-1841), 2017. Date of Publication: November 2017.

[Article]

AN: 614007195

Background/purpose The aims of surgical management in cloacal exstrophy (CE) have shifted to optimizing outcomes and quality of life while minimizing morbidity. This report reviews the single-institution experience of complications of bladder closure in CE. Methods Patients with CE were identified from a prospectively-maintained bladder exstrophy-epispadias complex database.

Operative and follow-up data were analyzed to compare complications and failure rates of bladder closure between closures performed with and without osteotomy and primary versus reoperative closures. Results Of 134 patients followed with CE, 112 met inclusion criteria. Median

follow-up time was 3.05 years. The failure rate among 112 primary closures (mean age 8.4 months) was 31.3% versus 51.9% in reoperative closures (mean age 19.7 months) ($p = 0.044$). Complication rate among primary and reoperative closures was 17.9% and 33.3%, respectively ($p = 0.076$). For closures with pelvic osteotomy, failure rate was 24.0% versus 45.9% without osteotomy ($p = 0.018$). Among primary closures with osteotomy, the complication rate was 21.3% versus 10.8% without osteotomy ($p = 0.171$). Conclusions Complications of bladder closure are common in CE. Pelvic osteotomy reduces failure rates without a significant rise in complications, which are often minor. There was no statistically significant difference in complication rates between reoperative and primary closures. However, reoperative closures were more likely to fail, emphasizing the importance of a successful primary closure. Level of evidence II: retrospective study.

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Status

Embase

Author NameID

Gearhart, John P.; ORCID: <http://orcid.org/0000-0002-0177-6121>

Institution

(Friedlander, Di Carlo, Gearhart) Jeffs Division of Pediatric Urology, The James Buchanan Brady Urological Institute, The Johns Hopkins University School of Medicine, Baltimore, MD, United States (Sponseller) Division of Pediatric Orthopaedics, Department of Orthopaedics, The Johns Hopkins University School of Medicine, Baltimore, MD, United States

Publisher

W.B. Saunders

Year of Publication

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460.

Associations of TGFBR1 and TGFBR2 gene polymorphisms with the risk of hypospadias: A case-control study in a Chinese population.

Han X.-R., Wen X., Wang S., Hong X.-W., Fan S.-H., Zhuang J., Wang Y.-J., Zhang Z.-F., Li M.-Q., Hu B., Shan Q., Sun C.-H., Bao Y.-X., Lin M., He T., Wu D.-M., Lu J., Zheng Y.-L.

Embase

Bioscience Reports. 37 (5) (no pagination), 2017. Article Number: BSR20170713. Date of Publication: 31 Oct 2017.

[Article]

AN: 619228375

This case-control study investigated the association of transforming growth factor-beta (TGF-beta) receptor type I and II (TGFB1 and TGFB2) gene polymorphisms with the risk of hypospadias in a Chinese population. One hundred and sixty two patients suffering from hypospadias were enrolled as case group and 165 children who underwent circumcision were recruited as control group. Single nucleotide polymorphisms (SNPs) in TGFB1 and TGFB2 genes were selected on the basis of genetic data obtained from HapMap. PCR-restriction fragment length polymorphism (PCR-RFLP) was performed to identify TGFB1 and TGFB2 gene polymorphisms and analyze genotype distribution and allele frequency. Logistic regression analysis was conducted to estimate the risk factors for hypospadias. No significant difference was found concerning the genotype and allele frequencies of TGFB1 rs4743325 polymorphism between the case and control groups. However, genotype and allele frequencies of TGFB2 rs6785358 in the case group were significantly different in contrast with those in the control group. Patients carrying the G allele of TGFB2 rs6785358 polymorphism exhibited a higher risk of hypospadias compared with the patients carrying the A allele ($P < 0.05$). The TGFB2 rs6785358 genotype was found to be significantly related to abnormal pregnancy and preterm birth (both $P < 0.05$). The frequency of TGFB2 rs6785358 GG genotype exhibited significant differences amongst patients suffering from four different pathological types of hypospadias. Logistic regression analysis revealed that preterm birth, abnormal pregnancy, and TGFB2 rs6785358 were the independent risk factors for hypospadias. Our study provides evidence that TGFB2 rs6785358 polymorphism might be associated with the risk of hypospadias.

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Institution

(Han, Wen, Wang, Fan, Zhuang, Wang, Zhang, Li, Hu, Shan, Sun, Wu, Lu, Zheng) Key Laboratory for Biotechnology on Medicinal Plants of Jiangsu Province, School of Life Science, Jiangsu Normal University, Xuzhou 221116, China (Hong) Department of Immunology, School of Basic Medical Sciences, Fudan University, Shanghai 200032, China

(Zhuang) Jiangsu Key Laboratory for Eco-Agricultural Biotechnology around Hongze Lake,
School of Life Sciences, Huaiyin Normal University, Huaian 223300, China
(Bao) Department of Orthopedics, Affiliated Municipal Hospital, Xuzhou Medical University,
Xuzhou 221009, China
(Lin, He) Department of Urology Surgery, Peking Union Medical College Hospital, Beijing 100730,
China
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461.

Pre-incision urethral plate width does not impact short-term Tubularized Incised Plate
urethroplasty outcomes.

Bush N.C., Snodgrass W.

Embase

Journal of Pediatric Urology. 13 (6) (pp 625.e1-625.e6), 2017. Date of Publication: December
2017.

[Article]

AN: 619224325

Introduction Two reports have found that urethral plate (UP) widths <8 mm before tubularized incised plate (TIP) incision increased urethroplasty complications. The present study measured pre-incision UP width in consecutive boys undergoing TIP to determine if it affected outcomes. **Methods** The present study followed the method previously used by Holland and Smith, and Sarhan et al. to measure UP width before creating glans wings or performing midline plate incision in consecutive patients with primary hypospadias and ventral curvature <30degree who all underwent TIP repair (Summary Fig.). Glans width at its widest point was also measured. Multiple logistic regression assessed urethroplasty complications (fistula, glans dehiscence, meatal stenosis/urethral stricture, diverticulum) based on pre-incision UP width, glans width, patient age, and meatal location. **Results** The UP widths were determined in 224 consecutive primary TIP repairs during 2012-2015: 200 distal, 11 midshaft, and 13 proximal. The UP width was <8 mm in 192/224 (86%) patients. Mean pre-incision width was 6.1 mm (SD 1.5, range 2-

11), without difference in UP widths according to meatal location ($P = 0.06$). Mean post-incision UP width was 12 mm (SD 2.2, range 10-16). Mean change in width after incision (delta/original UP width) was 116% (SD 63, range 20-250). There was follow-up in 186 patients for a mean of 6 months. Urethroplasty complications (five fistulas, six glans dehiscence) were diagnosed in 11 (6%): 9/165 distal, 1/9 midshaft, and 1/12 proximal repairs. There was no difference in those <8 vs ≥ 8 mm (11/160 vs 0/26, $P = 0.17$). Similarly, UP width was not different between patients with and without urethroplasty complications. Multiple logistic regression in these 186 patients - including meatal location, UP width, glans width, and age - found only glans width <14 mm was associated with increased odds of urethroplasty complications (OR 19.2, 95% CI 3.5-106, AUC = 0.799). Discussion The data show that pre-incision UP width is not an independent risk factor for urethroplasty complications. However, it is possible that technical factors, such as how deeply the dorsal incision is made or size of the urethral stent, might contribute to this finding by other authors. After watching the TIP repair, Smith stated that the plate incision was deeper than he made. Sarhan et al. reported a mean change of 57% in UP width after incision, whereas the present one was double at 116% (i.e. from 6 mm pre-incision to 12 mm post incision), and they used an 8-Fr catheter. While they stated that they incised the plate deeply, the lower percentage increase in width suggests that it was not as deep as was recommended. Conclusions The UP width before incision did not increase urethroplasty complications. Surgeons do not need to measure or categorize the UP to determine suitability for TIP repair, as long as the plate incision is made deeply to the corpora.[Figure presented]

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Status

Embase

Author NameID

Bush N.C.; ORCID: <http://orcid.org/0000-0002-9887-3680>

Institution

(Bush, Snodgrass) PARC Urology, 5680 Frisco Square Blvd., Suite 2300, Frisco, TX 75033,
United States

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462.

Buck's fascia repair with glanuloplasty in hypospadias surgery: A simple approach with excellent outcome.

Baba A.A., Wani S.A., Bhat N.A., Mufti G.N., Lone T.N., Nazir S.

Embase

Journal of Pediatric Urology. 13 (6) (pp 633.e1-633.e5), 2017. Date of Publication: December 2017.

[Article]

AN: 617693540

Background Hypospadias is a common problem encountered in surgical practice and its repair is challenging because of various complications. Urethro-cutaneous fistula is the most common postoperative complication (incidence of 0-33%). Different types of intermediate protective layers have been used in attempts to decrease UCF formation; however, no single surgical technique is ideal. **Objective** The aim of this study was to compare use of dartos fascia and Buck's fascia as intermediate layers in prevention of the formation of UCF. **Study design** This was a prospective, comparative study conducted over a period of 2 years from January 2014 to December 2015. Patients with primary hypospadias without or with mild ventral penile curvature were included in the study. Patients were categorised into two groups, A and B, with alternate patients assigned to each group. Patients in group A underwent Snodgrass repair with urethroplasty by two-layer subepithelial closure and dartos tissue as an intermediate layer. Patients in group B underwent a urethral repair followed by Buck's fascia repair as intermediate layer and glanuloplasty after excision of a triangular skin strip on either side of the urethral plate. Patients were followed at regular intervals for a minimum of 6 months and complications were noted. **Results** Over a period of 2 years, 160 patients were included in the study: 80 patients in group A and 80 patients in group B. The age of patients ranged from 1 year to 4.6 years, with a mean age of 1.8 years. Postoperative complications are listed in the summary table. **Discussion** A protective intermediate layer between the neourethra and the skin can be used to reduce fistula formation. We describe a technique of urethroplasty using Buck's fascia as intermediate layer and glanuloplasty, with excellent results. Buck's fascia over the corpora spongiosum which is deficient ventrally in hypospadias is not completely absent, and can be easily used to cover the neourethra, needs minimal dissection and hence vascularity of tissues is preserved (summary Fig.). We used this Buck's fascia as a second protective layer over the neourethra in an attempt to decrease UCF formation, and compared it with use of dartos fascia. In our study, UCF occurred in 2.5% of patients in the Buck's fascia group and 12.5% of patients in the dartos group, a statistically

significant difference. We believe that the native Buck's fascia lateral to spongiosum is a more appropriate, natural, and strong layer to cover the neourethra. Conclusion Use of Buck's fascia as an intermediate layer along with glanuloplasty is simple and very effective in preventing UCF formation and glanular dehiscence. We recommend the use of Buck's fascia as an intermediate layer to cover the neourethra to reduce incidence of postoperative complications and improve results. [Table presented] [Figure presented]

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Status

Embase

Institution

(Baba, Wani, Bhat, Mufti) Department of Paediatric Surgery, SKIMS, Soura, Srinagar, Jammu and Kashmir, India (Lone) Department of Paediatrics Health Services, Srinagar, Jammu and Kashmir, India

(Nazir) Health Services, Srinagar, Jammu and Kashmir, India

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463.

Urinary tract infection after retrograde urethrogram in children: A multicenter study.

Malhotra N.R., Green J.R., Rigsby C.K., Holl J.L., Cheng E.Y., Johnson E.K.

Embase

Journal of Pediatric Urology. 13 (6) (pp 623.e1-623.e5), 2017. Date of Publication: December 2017.

[Article]

AN: 617201834

Introduction The risk of post-procedural urinary tract infection (ppUTI) after retrograde urethrogram (RUG) has not been well quantified. Prophylactic antibiotics may reduce the risk, but must be weighed against the risks of antibiotic resistance. Prior research has shown that this risk

is variable after voiding cystourethrogram (0-42% reported ppUTI rate) and appears to be low after urodynamics (0-4.8%) but this risk has not been well documented for patients undergoing RUG. Objective We aimed to (1) describe the rate of ppUTI after RUG, and (2) examine factors associated with use of antibiotics at the time of RUG. Study design We conducted a retrospective cohort study of children undergoing RUG at two hospitals January 1, 2004, to December 31, 2014. ppUTI within 7 days was measured. Antibiotic prophylaxis was determined. Relationships between clinical characteristics and receipt of pre-procedure antibiotics were evaluated using univariate statistics. Results Forty-two patients (100% male, median age 11.7 years) underwent 47 RUGs. Indications included trauma (27%), hypospadias (17%), and non-hypospadias (27%) stricture. Study indication and antibiotic administration practices are illustrated in the Figure. Three patients (6.4%) had a history of posterior urethral valves (PUV); one had neurogenic bladder (NGB) (2.4%). Two (4%) studies were performed within 30 days of a clinical UTI and 11% had a positive urinalysis or culture within 30 days pre-RUG. UTI in the 30 days before RUG was not associated with antibiotic prescription ($p > 0.99$). One child (2.1%; 95% CI 0-6.3%) had a ppUTI: a 7-year-old, uncircumcised male with a history of PUV and voiding dysfunction who was asymptomatic at the time of RUG. Discussion Although the risk of ppUTI after RUG appears to be low, the rare occurrence of an iatrogenic UTI can lead to high cost and patient morbidity, particularly in cases similar to our patients who required inpatient admission for intravenous antibiotics. To help balance the risk of UTI with the associated healthcare costs and morbidity with the concerns about antibiotic overuse and potential exposure to drug reaction, data from this study may inform future development of evidence-based guidelines targeting only patients at highest risk for ppUTI with antibiotic prophylaxis. Conclusions The risk of ppUTI after RUG is low. Antibiotic prophylaxis was unrelated pre-RUG UTI in our population. These results indicate the need for pre-procedural antibiotic prescribing guidelines, and suggest that routine antibiotics prior to RUG are not indicated unless the patient has another indication for antibiotics.[Figure presented]

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Status

Embase

Institution

(Malhotra) Department of Urology, University of Illinois at Chicago, Chicago, IL, United States

(Green, Rigsby) Department of Medical Imaging, Ann & Robert H. Lurie Children's Hospital of Chicago, Chicago, IL, United States

(Green, Rigsby) Department of Radiology, Northwestern University Feinberg School of Medicine, Chicago, IL, United States

(Rigsby) Department of Pediatrics, Northwestern University Feinberg School of Medicine,
Chicago, IL, United States

(Holl, Johnson) Center for Healthcare Studies, Northwestern University Feinberg School of
Medicine, Chicago, IL, United States

(Cheng, Johnson) Division of Urology, Ann & Robert H. Lurie Children's Hospital of Chicago,
Chicago, IL, United States

(Cheng, Johnson) Department of Urology, Northwestern University Feinberg School of Medicine,
Chicago, IL, United States

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464.

Two-stage flap repair of severe hypospadias: Usefulness of the tubularized incised plate
urethroplasty.

Tijani K.H., Idiodi-Thomas H.O., Elebute O.A., Alakaloko F.M., Ojewola R.W., Ademuyiwa A.O.

Embase

Journal of Pediatric Urology. 13 (5) (pp 483.e1-483.e5), 2017. Date of Publication: October 2017.

[Article]

AN: 616588069

Purpose The use of flaps in the two-stage repair of posterior hypospadias associated with severe chordee has been well established. Despite the almost certain guarantee of flap take, complications such as diverticuli are still relatively high. While different applications of the tubularized incised plate have been described, experience with the application of the technique to the two-stage flap repair is very limited. A previous local review of 15 cases performed at the present institution during the period 1998-2003, using the technique as described by Rekit, revealed fistulae and diverticulum rates of 20% and 27%, respectively. With the primary objective of improving surgical outcome, the midline incision was incorporated into the two-stage flap repair. **Materials and methods** Between 2004 and 2015, 35 boys (aged 2-15 years) with severe hypospadias that required excision of the urethral plate were operated using the two-stage flap technique. The first stage involved mobilization of preputal or dorsal penile skin (if circumcised)

to the ventral surface, as described by Retik. After a minimum interval of 6 months, the second-stage operation was performed in a way similar to the technique of tubularized incised plate urethroplasty, as popularized by Snodgrass, and involved a preliminary midline incision on the neo-urethral plate followed by tubularization and multilayered closure. Results All but one flap took successfully. The outcome was satisfactory in 80% of patients, and there was a fistula rate of 14% (Summary Table). One patient had a complete breakdown of the flap and was successfully treated about 12 months later by repeating the second stage of the operation. No case of diverticulum or stricture was recorded. Discussion Even though there was a marginal improvement in the fistula rate, the most striking observation was the complete absence of diverticulum or stricture. With a reported incidence rate of 20-63%, different authors have reported diverticulum formation (despite the absence of distal obstruction) to be a major problem of the two-stage flap technique. Attempts by these authors at reducing the risk of diverticulum by reducing flap size have tended to increase the risk of strictures. This has been the main reason given by some authors for abandoning the technique. The main limitations of the present study included the wide age range of the patients and the small sample size. Conclusion The inclusion of a midline incision in a two-stage flap urethroplasty for proximal hypospadias appears to prevent the development of diverticulum. [Table presented]

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Status

Embase

Author NameID

Tijani K.H.; ORCID: <http://orcid.org/0000-0001-7429-7183>

Institution

(Tijani, Ojewola) Section of Urology, Department of Surgery, College of Medicine, University of Lagos (CMUL)/Lagos University Teaching Hospital (LUTH), Lagos, Nigeria (Idiodi-Thomas, Elebute, Alakaloko, Ademuyiwa) Section of Pediatric Surgery, Department of Surgery, College of Medicine, University of Lagos (CMUL)/Lagos University Teaching Hospital (LUTH), Lagos, Nigeria

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465.

Clinical heterogeneity in children with gonadal dysgenesis associated with non-mosaic 46,XY karyotype.

Wong Y.S., Tam Y.H., Pang K.K.Y., To K.F., Chan S.S.C., Chan K.W., Lee K.H.

Embase

Journal of Pediatric Urology. 13 (5) (pp 508.e1-508.e6), 2017. Date of Publication: October 2017.

[Article]

AN: 615612198

Introduction Gonadal dysgenesis is unique in disorders of sex development (DSD), in that it can be associated with 46,XX, 46,XY or mosaic 45,X/46,XY karyotypes. Gonadal dysgenesis can be partial or complete. Gonadal dysgenesis associated with the Y-chromosome has increased risk of gonadal germ cell neoplasms. Most of the literature focus on 45,X/46,XY gonadal dysgenesis, while there are scanty data on the condition when the karyotype is non-mosaic 46,XY. **Objective** To investigate the diversity of clinical pictures of children presenting with 46,XY DSD due to gonadal dysgenesis. **Methods** A retrospective study on consecutive patients diagnosed with 46,XY gonadal dysgenesis at age ≤ 18 years in a tertiary center from 1985 to 2015. The clinical presentations, phenotypes, gonadal features and associated anomalies were investigated. **Results** Twenty-eight patients with Y-chromosome gonadal dysgenesis were identified during the study period and six (21.4%) had non-mosaic 46,XY karyotype. Three had complete gonadal dysgenesis (CGD) with normal female phenotype, while the other three had partial gonadal dysgenesis (PGD). Of the three patients with CGD, two presented with the classical Swyer syndrome at adolescence, while the third presented at birth with multiple congenital anomalies. The three PGD patients presented with ambiguous genitalia at birth ($n = 2$), and isolated hypospadias ($n = 1$), which was associated with Frasier syndrome. Three patients had germ cell neoplasms: bilateral gonadoblastoma ($n = 1$), bilateral intratubular germ cell neoplasia unclassified ($n = 1$), and dysgerminoma + gonadoblastoma ($n = 1$). Two patients had global developmental delay with other congenital anomalies, and another patient had learning difficulties with borderline intelligence (Table). **Discussion** The findings suggest that 46,XY gonadal dysgenesis is much rarer than 45,X/46,XY gonadal dysgenesis. Patients differed in their clinical presentations and well-established syndromes happened in half of them. Overall, the risk of germ cell neoplasms and the association with other somatic anomalies appeared to be high. The study was limited by: its small number, single-center experience, and the possibility of missing the diagnosis in some male patients with mild undervirilization. **Conclusion** Heterogeneity was noted

in the clinical, phenotypic and gonadal features among pediatric patients with 46,XY gonadal dysgenesis. [Table presented]

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Status

Embase

Institution

(Wong, Tam, Pang, Chan, Lee) Division of Paediatric Surgery and Paediatric Urology,
Department of Surgery, Prince of Wales Hospital, The Chinese University of Hong Kong, China
(To) Department of Anatomical and Cellular Pathology, Prince of Wales Hospital, The Chinese
University of Hong Kong, Hong Kong

(Chan) Department of Obstetrics and Gynaecology, Prince of Wales Hospital, The Chinese
University of Hong Kong, Hong Kong

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Year of Publication

2017

466.

Hypospadias surgery in England: Higher volume centres have lower complication rates.

Wilkinson D.J., Green P.A., Beglinger S., Myers J., Hudson R., Edgar D., Kenny S.E.

Embase

Journal of Pediatric Urology. 13 (5) (pp 481.e1-481.e6), 2017. Date of Publication: October 2017.

[Article]

AN: 615610933

Introduction Hypospadias surgery has progressed steadily over recent years. There remains considerable variation in the operative management of boys with hypospadias in the UK, and it is therefore difficult to identify acceptable standards with regards to reoperation rates. Objective To determine the frequency of reoperations and complications from all centres performing hypospadias surgery in England and to identify variables that influence outcome. Methods All children undergoing NHS hypospadias surgery in England between 1999 and 2009 were

identified using the Hospital Episode Statistics database. Patient demographics, centre type, and associated diagnostic (ICD-10) and treatment codes (OPCS4.6) were collected for both primary repairs and postoperative complications. Centres were classed as high volume if they performed an average of 20 or more operations a year. Operative complications were split into revisions (repeat repairs), repairs of urethral fistulae, repairs of meatal stenosis, or urethral stricture repairs. Statistical analysis included logistic regression, Spearman's correlation, and Mann-Whitney U for non-parametric data, with $p < 0.05$ taken as significant. Data are presented as median (interquartile range) unless otherwise stated. Results children underwent a total of 23,962 operations at 75 centres in England during the study period. The median age at primary repair was 21 (15-38) months. The overall complication rate was 18.1%. The median complication rate for individual centres was 20.0% (13.9-27.4%) overall; 10.8% (4.7-15.9%) for revision procedures, 8.1% (5.5-11.7%) for urethral fistulae, 2.3% (1.1-3.7%) for meatal stenosis repairs, and 1.8% (0-2.8%) for urethral strictures. High volume centres had significantly lower complication rates than low volume centres (17.5% vs. 25%, $p = 0.01$) (Figure), and this was proven to be an independent predictor of outcomes ($p = 0.01$). Staged repairs were associated with more complications ($p < 0.001$); however, patient age and centre type were not. Median time to repair of complication was 13 (8-22) months. Discussion This national population-based study used hospital episode statistics data. While accuracy is high and it has been validated for use in research, it has intrinsic limitations which affect our study. We are unable to fully account for the severity of hypospadias or the number of operating surgeons within institutions. Conclusions This study has found a clear relationship between caseload volume and complications following hypospadias surgery. Furthermore, there is significant variability between centres in terms of their surgical outcomes. Taken together these results suggest that surgeons, particularly those in centres with small caseloads should assess their results against such benchmarks when evaluating the service they provide.[Figure presented]

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Status

Embase

Author NameID

Green, Patrick A.; ORCID: <http://orcid.org/0000-0002-4805-7298>

Institution

(Wilkinson, Green, Beglinger, Myers, Hudson, Edgar, Kenny) University of Liverpool, Liverpool,

United Kingdom (Wilkinson, Kenny) Department of Paediatric Surgery, Alder Hey Children's

Hospital, NHS Foundation Trust, Liverpool, United Kingdom

(Green) Royal Liverpool Hospital, Prescott Street, Liverpool, United Kingdom

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467.

Ultrasound diagnosis of fetal hypospadias: Accuracy and outcomes.

Epelboym Y., Estrada C., Estroff J.

Embase

Journal of Pediatric Urology. 13 (5) (pp 484.e1-484.e4), 2017. Date of Publication: October 2017.

[Article]

AN: 615330062

Objectives The purpose of this study was to assess the positive predictive value of a prenatal ultrasound diagnosis of hypospadias when compared with postnatal diagnosis based on physical exam. **Methods** We retrospectively identified all pregnant women between 2004 and 2014 who were either referred to our fetal care center carrying a fetus with an ultrasound diagnosis of possible hypospadias or who had a new diagnosis of hypospadias after imaging in our center. **Results** A total of 32 cases of possible hypospadias were identified, with our fetal center ultrasound suggesting hypospadias in 25 of the 32 cases (78%). Of the 25 cases, 18 infants were confirmed to have hypospadias on postnatal physical exam (Table), reflecting a positive predictive value of 72%. Twenty-one of twenty-five cases with suggested hypospadias on ultrasound were found to have either hypospadias or another penile anomaly on postnatal physical exam, reflecting a positive predictive value for any genital anomaly of 84%. Infants with confirmed hypospadias often had several associated GU anomalies on postnatal clinical exam. **Conclusions** Our single center experience with the fetal ultrasound diagnosis of hypospadias demonstrates a high positive predictive value for a penile anomaly (21/25, 84%), and a moderately high positive predictive value for the specific diagnosis of hypospadias (18/25, 72%) when compared with the postnatal diagnosis. [Table presented]

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Status

Embase

Institution

(Epelboym) Brigham & Women's Hospital, Boston, MA, United States (Estrada, Estroff) Boston Children's Hospital, Boston, MA, United States

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Year of Publication

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468.

Long-term follow-up after traditional versus modified perineal approach in the management of female epispadias.

Alyami F., Fernandez N., Lee L., Metcalfe P., Lorenzo A., Pippi Salle J.

Embase

Journal of Pediatric Urology. 13 (5) (pp 497.e1-497.e5), 2017. Date of Publication: October 2017.

[Article]

AN: 615246835

Objective Isolated female epispadias (IFE) is a rare congenital anomaly. The defect extends to the bladder neck, which is usually incompetent. The traditional surgical approach includes urethral and genital reconstruction in the first year, followed by bladder neck reconstruction (Young-Dees-Leadbetter cervicoplasty (YDL)) at the age of social continence. An alternative single-stage technique includes urethral, bladder neck and clitoris repair by a perineal approach. The aim of the present study was to describe long-term follow-up of patients who underwent the traditional vs alternative approach. Materials and methods A retrospective review was performed of all female epispadias cases managed between 2000 and 2013. The YDL procedure (Group 1) vs single-stage perineal approach (Group 2) cases were followed and compared. Collected variables included: patients' demographics, age at diagnosis and surgery, presence of associated anomalies, clinical presentation, presence of vesicoureteral reflux (VUR), and pre-operative and postoperative continence. Results A total of 12 cases of female epispadias were managed and followed between 2000 and 2013. No major complications occurred in either group. Urinary continence evaluated in seven children showed that none (0/3) and 4/7 (57%) were continent following the initial procedure in Group 1 and Group 2, respectively. All patients in Group 1 failed

to achieve continence and required re-intervention. Conclusions Female epispadias could be successfully repaired using a single-stage modified perineal approach that achieved good continence with volitional voiding, good cosmetic results and compared favorably with the ones repaired with the YDL technique. The additional step of performing bladder neck tailoring to achieve a funneling configuration seemed to be useful in improving continence. [Table presented]

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Author NameID

Pippi Salle J.; ORCID: <http://orcid.org/0000-0002-0200-3884> Alyami F.; ORCID: <http://orcid.org/0000-0002-4519-4510>

Institution

(Alyami, Fernandez, Lee, Lorenzo, Pippi Salle) Division of Urology, The Hospital for Sick Children and University of Toronto, Toronto, Ontario, Canada (Pippi Salle) Department of Surgery, Division of Urology, Sidra Medical and Research Center, Doha, Qatar

(Alyami) Urology Division, Department of Surgery, King Saud University, King Saud University Medical City and College of Medicine, Riyadh, Saudi Arabia

(Metcalf) Division of Pediatric Urology, Department of Pediatric Surgery, University of Alberta, Mackenzie Health Sciences Centre, Edmonton, Canada

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469.

The prostatic utricle: An under-recognized condition resulting in significant morbidity in boys with both hypospadias and normal external genitalia.

Hester A.G., Kogan S.J.

Embase

Journal of Pediatric Urology. 13 (5) (pp 492.e1-492.e5), 2017. Date of Publication: October 2017.

[Article]

AN: 614885522

Introduction Pediatric presentations of a prostatic utricle have received only scant attention. Though recognized with increased frequency in boys with hypospadias, little is described about their incidence and potential for morbidity in boys with normal external genitalia. **Methods** We initially reviewed a cohort of 64 patients with hypospadias seen over a 3-year period to determine the frequency of investigative lower urinary tract studies and utricle identification. Children with disorders of sexual differentiation were excluded from this review. A subsequent group of 70 boys with hypospadias and 23 boys with normal external genitalia presenting with lower urinary tract symptoms (LUTS) who were found to have an unsuspected utricle were reviewed. This comparative group was investigated since symptomatology was the indication for evaluation, contrasting with those in the hypospadias group who were investigated because of hypospadias presence alone. **Results** In our initial review of 64 patients only 24 (37.5%) underwent an investigative study and six (9.4%) had a utricle. Three (50%) required surgical excision, allowing their hypospadias repair to proceed. Results in the subsequent group with hypospadias confirmed these findings with increased rates of investigation and identification. The boys with normal external male genitalia all required surgery since symptoms were the result of the utricle alone. Penile pain with voiding, hematuria, epididymitis, and urinary infection were the most common causes for interventions. **Conclusions** The prostatic utricle should be considered as a cause of morbidity in boys with both normal external genitalia and those with hypospadias. Endoscopic or radiological evaluation (see Figure) should be undertaken in all boys with proximal hypospadias, boys with hypospadias and associated cryptorchidism, and those with hypospadias with associated urinary symptoms. Boys with normal external genitalia with lower urinary tract symptoms not explained with imaging should undergo cystoscopy, as an unidentified unsuspected utricle may be the underlying cause.[Figure presented]

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Status

Embase

Author NameID

Hester, Austin G.; ORCID: <http://orcid.org/0000-0003-1495-6487>

Institution

(Hester, Kogan) Department of Urology, Wake Forest University, Winston-Salem, NC, United States

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470.

Clinical and molecular characteristics in 15 patients with androgen receptor gene mutations from South China.

Su L., Cheng J., Yin X., Liu G., Lu Z., Sheng H., Cai Y., Shi Q., Liu L.

Embase

Andrologia. 49 (10) (no pagination), 2017. Article Number: e12763. Date of Publication: December 2017.

[Article]

AN: 614697884

A variety of mutations in the androgen receptor (AR) gene are linked to androgen insensitivity syndrome (AIS) or sexual development disorder. Here, we studied 15 patients with various degrees of disorders of genital hypoplasia from South China. Clinical data including basal hormone level, phenotype, karyotyping and SRY gene identification were documented. Exons with flanking intronic region of the AR gene were sequenced and analysed for mutations, and a total of eight mutations were identified in the AR gene. Of eight mutations, two novel mutations c.2518G>T (p.Asp840Tyr) and c.1186G>C (p.Gly396Arg) were predicted to be damaging by SIFT and Polyphen2 online software. Previously reported mutations: c.528C>A (p.Ser176Arg), c.1789G>A (p.Ala597Thr), c.2612C>T (p.Ala871Val), c.1752C>A (p.Phe584Leu), c.171_172insCTG (p.57_58insLeu) and c.2659A>G (p.Met887Val) were also detected in our subjects. Most of them are involved in hypospadias, penis dysplasia or other disorders of sexual development. A complete AIS case (p.Phe584Leu) with female phenotype and high serum concentrations of dihydrotestosterone (DHT) was also found. This study presented a wide range of spectrum of AIS (from partial AIS to complete AIS) caused by AR mutations in South China population. It suggests that further study with larger data set need to be performed to elucidate the differences of the phenotypes in our study.

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Embase

Institution

(Su, Cheng, Yin, Lu, Sheng, Cai, Liu) Department of Genetics and Endocrinology, Guangzhou Women and Children's Medical Center, Guangzhou Medical University, Guangzhou, Guangdong, China (Liu) Department of Pediatric Urology, Guangzhou Women and Children's Medical Center, Guangzhou Medical University, Guangzhou, Guangdong, China (Shi) Department of Pathology, Guangzhou Women and Children's Medical Center, Guangzhou Medical University, Guangzhou, Guangdong, China

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471.

Comparison of the peak inspiratory pressure and lung dynamic compliance between a classic laryngeal mask airway and an endotracheal tube in children under mechanical ventilation.

Mahdavi A., Razavi S.S., Malekianzadeh B., Sadeghi A.

Embase

Tanaffos. 16 (4) (pp 289-294), 2017. Date of Publication: 2017.

[Article]

AN: 621870462

Background: The present study was performed with the aim of comparing the peak inspiratory pressure and lung dynamic compliance between a classic laryngeal mask airway (LMA) and an endotracheal tube in children under mechanical ventilation.

Material(s) and Method(s): In this study, 30 children aged 1 to 7 years with a physical condition of ASA I-II who were admitted for operations to repair inguinal hernias, hydroceles, or hypospadias were randomly enrolled. After induction of anesthesia, the appropriate laryngeal mask was used for each patient and they were placed under pressure-controlled mechanical ventilation. The peak inspiratory pressure was adjusted and recorded to obtain an appropriate tidal volume, then the laryngeal mask was removed and the appropriate size uncuffed endotracheal tube was inserted and the patient was placed again under controlled mechanical ventilation. The required settings were adjusted and peak inspiratory pressure and tidal volume were measured and recorded by

the ventilator. Dynamic compliance was also calculated in both cases using the appropriate formula.

Result(s): The results showed that peak inspiratory pressure (PIP) with the use of LMA in children under mechanical ventilation was less than the PIP with the use of an endotracheal tube ($p<0.05$). Also, the pulmonary dynamic compliance with a laryngeal mask was greater than the use of an endotracheal tube ($p<0.05$).

Conclusion(s): A laryngeal mask airway due to its low airway resistance and high dynamic compliance is an acceptable alternative to a tracheal tube during mechanical ventilation and it can be a good alternative to the endotracheal tube, especially during mechanical ventilation of children, in whom avoidance of barotrauma is desirable.

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Status

Embase

Institution

(Mahdavi, Razavi, Malekianzadeh, Sadeghi) Department of Pediatric Anesthesia, Shahid Beheshti University of Medical Science, Tehran, Iran, Islamic Republic of

Publisher

Shaheed Beheshti University of Medical Sciences and Health Services (E-mail: info@tanaffosjournal.ir)

Year of Publication

2017

472.

Surgery for distal hypospadias: What about the catheter?.

Scarpa M.-G., Perin G., Di Grazia M., Codrich D., Pederiva F., Guida E., Lembo M.A., Giannotta A., Schleef J.

Embase

Pediatrica Medica e Chirurgica. 39 (3) (pp 75-78), 2017. Article Number: 145. Date of Publication: 2017.

[Article]

AN: 619244639

No agreed recommendations exist for timing of urethral stent removal, after distal hypospadias surgery. We compared our preliminary case series with outcomes from literature: 18/44 patients were treated with catheter and 26/44 without it. The surgical outcome was comparable in the two groups. After hypospadias surgery, the main advantage of the immediate postoperative catheter removal was the shorter hospital stay without negatively affecting the care and home management.

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Status

Embase

Institution

(Scarpa, Perin, Codrich, Pederiva, Guida, Lembo, Giannotta, Schleef) Pediatric Surgery, Institute for Maternal and Child Health, IRCCS Burlo Garofolo, Trieste, Italy (Di Grazia) Institute for Maternal and Child Health, IRCCS Burlo Garofolo, Trieste, Italy

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2017

473.

Risk comparison for prenatal use of analgesics and selected birth defects, National Birth Defects Prevention Study 1997-2011.

Interrante J.D., Ailes E.C., Lind J.N., Anderka M., Feldkamp M.L., Werler M.M., Taylor L.G., Trinidad J., Gilboa S.M., Broussard C.S.

Embase

Annals of Epidemiology. 27 (10) (pp 645-653.e2), 2017. Date of Publication: October 2017.

[Article]

AN: 618695498

Purpose To compare the use of nonsteroidal anti-inflammatory drugs (NSAIDs) and/or opioids to the use of acetaminophen without NSAIDs or opioids with respect to associations with birth defects. Methods We used data from the National Birth Defects Prevention Study (1997-2011).

Exposure was self-reported maternal analgesic use from the month before through the third month of pregnancy (periconceptional). Adjusted odds ratios (aORs) were calculated to examine associations with 16 birth defects. Results Compared to acetaminophen, mothers reporting NSAIDs were significantly more likely to have offspring with gastroschisis, hypospadias, cleft palate, cleft lip with cleft palate, cleft lip without cleft palate, anencephaly, spina bifida, hypoplastic left heart syndrome, pulmonary valve stenosis, and tetralogy of Fallot (aOR range, 1.2-1.6). Opioids were associated with tetralogy of Fallot, perimembranous ventricular septal defect, and ventricular septal defect with atrial septal defect (aOR range, 1.8-2.3), whereas use of both opioids and NSAIDs was associated with gastroschisis, cleft palate, spina bifida, hypoplastic left heart syndrome, and pulmonary valve stenosis (aOR range, 2.0-2.9). Conclusions Compared to periconceptional use of acetaminophen, selected birth defects occurred more frequently among infants of women using NSAIDs and/or opioids. However, we could not definitely determine whether these risks relate to the drugs or to indications for treatment.

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Status

Embase

Author NameID

Trinidad, James; ORCID: <http://orcid.org/0000-0002-8226-3925>

Institution

(Interrante, Ailes, Lind, Gilboa, Broussard) Division of Congenital and Developmental Disorders, National Center on Birth Defects and Developmental Disabilities, Centers for Disease Control and Prevention, Atlanta, GA, United States (Interrante) Oak Ridge Institute for Science and Education, Oak Ridge, TN, United States

(Lind) United States Public Health Service, Atlanta, GA, United States

(Anderka) Birth Defects Monitoring Program, Center for Birth Defects Research and Prevention, Massachusetts Department of Public Health, Boston, MA, United States

(Feldkamp) Division of Medical Genetics, Department of Pediatrics, University of Utah School of Medicine, Salt Lake City, United States

(Werler) Department of Epidemiology, School of Public Health, Boston University, Boston, MA, United States

(Taylor, Trinidad) Division of Epidemiology, Office of Surveillance and Epidemiology, Center for Drug Evaluation and Research, Food and Drug Administration, Silver Spring, MD, United States

Publisher

Elsevier Inc. (E-mail: usjcs@elsevier.com)

Year of Publication

474.

Prevalence of untreated surgical conditions in Rural Rwanda: A population-based cross-sectional study in burera district.

Maine R.G., Linden A.F., Riviello R., Kamanzi E., Mody G.N., Ntakiyiruta G., Kansayisa G., Ntaganda E., Niyonkuru F., Mubiligi J.M., Mpunga T., Meara J.G., Hedt-Gauthier B.L.

Embase

JAMA Surgery. 152 (12) (no pagination), 2017. Article Number: e174013. Date of Publication: December 2017.

[Article]

AN: 620071761

IMPORTANCE In low- and middle-income countries, community-level surgical epidemiology is largely undefined. Accurate community-level surgical epidemiology is necessary for surgical health systems planning. **OBJECTIVE** To determine the prevalence of surgical conditions in Burera District, Northern Province, Rwanda. **DESIGN, SETTING, AND PARTICIPANTS** A cross-sectional study with a 2-stage cluster sample design (at village and household level) was carried out in Burera District in March and May 2012. A team of surgeons randomly sampled 30 villages with probability proportionate to village population size, then sampled 23 households within each village. All available household members were examined. **MAIN OUTCOMES AND MEASURES** The presence of 10 index surgical conditions (injuries/wounds, hernias/hydroceles, breast masses, neck masses, obstetric fistulas, undescended testes, hypospadias, hydrocephalus, cleft lip/palate, and clubfoot) was determined by physical examination. Prevalence was estimated overall and for each condition. Multivariable logistic regression was performed to identify factors associated with surgical conditions, accounting for the complex survey design. **RESULTS** Of the 2165 examined individuals, 1215 (56.2%) were female. The prevalence of any surgical condition among all examined individuals was 12%(95%CI, 9.2-14.9%). Half of conditions were hernias/hydroceles (49.6%), and 44%were injuries/wounds. In multivariable analysis, children 5 years or younger had twice the odds of having a surgical condition compared with married individuals 21 to 35 years of age (reference group) (odds ratio [OR], 2.2; 95%CI, 1.26-4.04; P = .01). The oldest group, people older than 50 years, also had twice the odds of having a surgical condition compared with the reference group (married, aged >50 years: OR, 2.3; 95%CI, 1.28-

4.23; $P = .01$; unmarried, aged >50 years: OR, 2.38; 95%CI, 1.02-5.52; $P = .06$). Unmarried individuals 21 to 35 years of age and unmarried individuals aged 36 to 50 years had higher odds of a surgical condition compared with the reference group (aged 21-35 years: OR, 1.68; 95%CI, 0.74-3.82; $P = .22$; aged 36-50 years: OR, 3.35; 95%CI, 1.29-9.11; $P = .02$). There was no statistical difference in odds by sex, wealth, education, or travel time to the nearest hospital.

CONCLUSIONS AND RELEVANCE The prevalence of surgically treatable conditions in northern Rwanda was considerably higher than previously estimated modeling and surveys in comparable low- and middle-income countries. This surgical backlog must be addressed in health system plans to increase surgical infrastructure and workforce in rural Africa.

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Status

Embase

Institution

(Maine) Department of Surgery, Harborview Medical Center, Seattle, WA, United States (Maine)

Now with Department of Surgery, University of North Carolina, School of Medicine, 4008 Burnett-Womack Building, Campus Box 7228, Chapel Hill, NC 27599-7228, United States

(Linden, Riviello, Mody, Meara) Program in Global Surgery and Social Change, Harvard Medical School, Boston, MA, United States

(Linden) Section of Pediatric Surgery, General Surgery, University of Chicago Comer's Children Hospital, Chicago, IL, United States

(Riviello, Mody) Center for Surgery and Public Health, Brigham And Women's Hospital, Boston, MA, United States

(Kamanzi) University of Global Health Equity (UGHE), Kigali, Rwanda

(Ntakiyiruta) Ejo Heza Surgical Centre, Kigali, Rwanda

(Kansayisa) Rwanda Military Hospital, Kigali, Rwanda

(Ntaganda, Niyonkuru) University Teaching Hospital-Kigali (CHUK), Kigali, Rwanda

(Mubiligi, Hedt-Gauthier) Partners in Health/Inshuti Mu Buzima, Kigali, Rwanda

(Mpunga) Rwanda Ministry of Health, Butaro District Hospital, Burera, Rwanda

(Meara) Department of Plastic and Oral Surgery, Boston Children's Hospital, Boston, MA, United States

(Hedt-Gauthier) Department of Global Health and Social Medicine, Harvard Medical School, Boston, MA, United States

Publisher

American Medical Association (E-mail: smcleod@itsa.ucsf.edu)

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475.

The hypospadias classification affected the surgical outcomes of staged oral mucosa graft urethroplasty in hypospadias reoperation.

Zheng D., Fu S., Li W., Xie M., Guo J., Yao H., Wang Z.

Embase

Medicine (United States). 96 (47) (no pagination), 2017. Article Number: e8238. Date of Publication: 01 Nov 2017.

[Article]

AN: 619527796

The staged graft urethroplasty is a recommended technique for repairing complex hypospadias. This retrospective study aimed to investigate the outcomes of this technique in hypospadias patients undergoing reoperation and to analyze the underlying contributing factors including age, meatus location, and graft and suture type. We retrospectively analyzed 40 hypospadias patients undergoing reoperation who received a staged oral graft urethroplasty, including 15 buccal mucosal grafts and 25 lingual mucosal grafts. Median age at presentation was 18.5 years, and median follow-up was 17.5 months (range 8-30 months). The patients were classified according to their original meatus location. Twenty-five complications developed in 12 of 40 (30%) cases, including 6 fistulas (15%), 7 infections (17.5%), 9 cases of glans dehiscence (22.5%), and 3 cases of stenosis (7.5%). There was no significant difference in the overall complication rates between prepuberty and postpuberty groups. In addition, no significant difference in complications was found between the 2 graft techniques. The complications were significantly higher in the original perineal type compared with the original penoscrotal type (7/10 vs 5/30, $P = .0031$). Seven patients who originally had perineal hypospadias developed multiple complications. Based on this study, the staged graft urethroplasty is an effective technique in reoperative hypospadias repairs with reasonable complication risk. The hypospadias classification affects the surgical outcomes.

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Status

Embase

Institution

(Zheng, Fu, Li, Xie, Guo, Yao, Wang) Department of Urology, Shanghai Ninth People's Hospital, Shanghai JiaoTong University, School of Medicine, Shanghai, China

Publisher

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Year of Publication

2017

476.

Fertility outcome and information on fertility issues in individuals with different forms of disorders of sex development: findings from the dsd-LIFE study.

Hirschberg A.L., Cohen Kettenis P., Roehle R., Kohler B., Cohen-Kettenis P., de Vries A., Arlt W., Wiesemann C., Slowikowska-Hilczer J., de la Perriere A.B., Sultan C., Paris F., Bouvattier C., Thyen U., Reisch N., Richter-Unruh A., Claahsen-van der Grinten H., Nordenstrom A., Pienkowski C., Szarras-Czapnik M.

Embase

Fertility and Sterility. 108 (5) (pp 822-831), 2017. Date of Publication: November 2017.

[Article]

AN: 618325046

Objective To investigate fertility outcome in individuals with different forms of disorders of sex development (DSD), if assisted reproductive technology (ART) was used, and the patients' satisfaction with the information they had received. Design A cross-sectional multicenter study, dsd-LIFE. Setting Not applicable. Patient(s) A total of 1,040 patients aged ≥ 16 years with different DSD diagnoses participated. Intervention(s) A web-based questionnaire was filled out by all participants. The participants could chose to take part in somatic investigations including ultrasonography. Main Outcome Measure(s) Information on partner, number of children, ART, adoption and step-children, general health, presence of gonads and uterus, current education and economic situation, received information on fertility issues, and satisfaction with the information, was collected. Result(s) In the total cohort, mean age 32 years, 33% lived with a partner, but only 14% reported having at least one child including 7% with ART, 4% adopted. Only 3.5% of the total cohort had been able to reproduce without ART, most frequently women

with congenital adrenal hyperplasia, and only 0.7% of participants with other diagnoses. Of the participants, 72% had received information on fertility, but 17% were not satisfied with the information. Conclusion(s) Fertility outcome is significantly reduced in all types of DSD; however, fertility potential should be assessed individually. The satisfaction with how fertility problems have been discussed can be improved. The care of patients with DSD is complex, should be individualized, and new treatment possibilities incorporated. A close collaboration in multidisciplinary teams is therefore essential to improve the situation for individuals with DSD.

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Status

Embase

Institution

(Slowikowska-Hilczer) Department of Andrology and Reproductive Endocrinology, Medical University of Lodz, Lodz, Poland (Hirschberg) Department of Gynecology, Karolinska University Hospital, Stockholm, Sweden

(Hirschberg, Nordenstrom) Department of Women's and Children's Health, Karolinska Institutet, Karolinska University Hospital, Stockholm, Sweden

(Nordenstrom) Department of Paediatric Endocrinology, Astrid Lindgren Children's Hospital, Karolinska University Hospital, Stockholm, Sweden

(Claahsen-van der Grinten) Department of Pediatric Endocrinology, Radboud University Nijmegen Medical Centre, Nijmegen, Netherlands

(Cohen Kettenis) Department of Medical Psychology, VU Medisch Centrum, Amsterdam, Netherlands

(Reisch) Medizinische Klinik and Poliklinik IV, Department of Endocrinology, University Hospital Munich, Munich, Germany

(Thyen) Klinik für Kinder-und Jugendmedizin, Universität zu Lubeck, Lubeck, Germany

(Roehle) Charite, Campus Virchow-Klinikum, Koordinierungszentrum für Klinische Studien (KKS Charite), Berlin, Germany

(Kohler) Klinik für Pädiatrische Endokrinologie und Diabetologie, Charite-Universitätsmedizin Berlin, corporate member of Freie Universität Berlin, Humboldt-Universität zu Berlin, Berlin Institute of Health, Berlin, Germany

(Bouvattier) Service d'Endocrinologie, Groupement Hospitalier Est, Centre Hospitalo-Universitaire de Lyon, Bron Cedex, France

(Bouvattier) Fac Med Paris-Sud, Université Paris-Sud, Université Paris Saclay, Le Kremlin Bicetre, France

(Bouvattier) Departement d'Endocrinologie Pediatrique, Hopital de Bicetre, Hopitaux
Universitaires Paris Sud, Assistance Publique-Hopitaux de Paris, Le Kremlin Bicetre, France
(Bouvattier) Centre de reference des maladies rares du developpement sexuel, Le Kremlin
Bicetre, France

Publisher

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477.

Transdisciplinary management of patients with disorders of sexual development in Colombia.

Limiting factors for appropriate management. Manejo transdisciplinario de pacientes con
desordenes del desarrollo sexual en Colombia. Limitantes para un manejo oportuno e integral
<Manejo transdisciplinario de pacientes con desordenes del desarrollo sexual en Colombia.

Limitantes para un manejo oportuno e integral.>

Fernandez N., Moreno O., Rojas A., Cespedes C., Forero C., Mora L., Suarez F., Auli J., Perez J.

Embase

Urologia Colombiana. 26 (3) (pp 164-168), 2017. Date of Publication: September 2017.

[Article]

AN: 613456778

Introduction Disorders of sex development (DSD) are present in 0.76 per 4,500 births. The management is complex and requires the ability of a multi-disciplinary team, which can have a positive impact on the prognosis. The aim of this article is to describe a population managed by a cross-disciplinary DSD group in our environment, and presenting the limitations faced by our patients in the care of their conditions. Methodology Description of the follow-up of patients with DSD by a cross-disciplinary group since 2007. The DSD clinical approach and genetic and diagnostic processes are described. Also a description of cases taken to surgery and their postoperative course was also evaluated from a medical and administrative point of view. Result The study included 55 patients with DSD, with the most common disease being congenital adrenal hyperplasia. The mean follow-up was 17.2 months. Surgery was performed in 36% of cases due to their condition, with a mean age of 5 years at the time of surgery. An initial extra-institutional initial assessment was never made by a multidisciplinary group, and only 72% were

studied by karyotype analysis by analysing 25 metaphases. None of the cases were diagnosed as having mosaicism when 25 metaphases were analysed, but were detected after extending the analysis to 100 metaphases. Eighteen percent had gender reassigned. Conclusions DSD are very complex diseases that require comprehensive treatment by multiple specialties. This is demonstrated by clear reduction in complications, costs and most importantly, an improvement in prognosis and reduction in disability. In our environment, limitations by the health system provides is alarming. A comprehensive treatment by a cross-disciplinary team to these patients is far from optimal. It is important that health care providers are aware of cross-disciplinary groups in order to promote proper treatment.

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Status

Embase

Institution

(Fernandez, Forero) Urologo, PhD Ciencias Basicas-Genetica, Hospital Universitario San Ignacio, Pontificia Universidad Javeriana, Bogota, Colombia (Moreno) Biol, MSc Biologia, cPhD Ciencias, Instituto de Genetica Humana, Facultad de Medicina, Pontificia Universidad Javeriana, Bogota, Colombia

(Rojas) Lic. en Biol y Quim, MSc Ciencias Biologicas, PhD Ciencias, Instituto de Genetica Humana-Facultad de Medicina, Pontificia Universidad Javeriana, Bogota, Colombia

(Cespedes) Endocrinologa pediatria Hospital Universitario San Ignacio, Pontificia Universidad Javeriana, Bogota, Colombia

(Mora) Genetista Hospital Universitario San Ignacio, Pontificia Universidad Javeriana, Bogota, Colombia

(Suarez) Genetista Instituto de Genetica Humana, Hospital Universitario San Ignacio, Pontificia Universidad Javeriana, Bogota, Colombia

(Auli) Psiquiatra Infantil, Hospital Universitario San Ignacio Pontificia Universidad Javeriana, Bogota, Colombia

(Perez) Urologo, Hospital Universitario San Ignacio, Profesor Asistente Facultad de Medicina Pontificia Universidad Javeriana, Bogota, Colombia

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478.

Long-Term Voiding Outcomes After Adult Urethral Reconstruction for Stricture Disease.

McAbee K.E., Rasper A.M., Terlecki R.P.

Embase

Current Bladder Dysfunction Reports. 12 (4) (pp 246-251), 2017. Date of Publication: 01 Dec 2017.

[Review]

AN: 619486267

Purpose of Review: While urethral reconstruction may restore luminal patency, voiding symptoms may persist or develop de novo. This review investigates contemporary literature regarding long-term voiding dysfunction after adult urethroplasty. Recent Findings: Following anterior repairs, voiding dysfunction occurs in 12.5% after graft-based urethroplasty, compared to 14% after excision and primary anastomosis. After posterior repair, 76% of patients may anticipate normal voiding and maintain continence. Voiding dysfunction in males with prior hypospadias repair(s) has been demonstrated to improve post-puberty. Success following female urethroplasty is reported as 94%.

Summary: Long-term success following urethral reconstruction is generally excellent, but elements of voiding dysfunction may be unmasked by relief of obstruction, or may stem from the repair itself. Continued emphasis on collection and validation of patient-reported outcome measures will better identify and characterize voiding dysfunction, and potentially guide preoperative counseling and surgical therapy.

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Status

Embase

Institution

(McAbee, Rasper, Terlecki) Department of Urology, Wake Forest Baptist Medical Center, 1 Medical Center Blvd, Winston-Salem, NC 27157, United States

Publisher

Current Medicine Group LLC 1 (E-mail: info@phl.cursci.com)

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2017

479.

Accuracy of the hypospadias diagnoses and surgical treatment registrations in the danish national patient register.

Arendt L.H., Ernst A., Lindhard M.S., Jonsson A.A., Henriksen T.B., Olsen J., Thorup J., Olsen L.H., Ramlau-Hansen C.H.

Embase

Clinical Epidemiology. 9 (pp 483-489), 2017. Date of Publication: 03 Oct 2017.

[Article]

AN: 619342737

Purpose: The Danish National Health registers provide a valuable data source that offers unique opportunities for observational research, including studies on the congenital anomaly hypospadias. The accuracy of the diagnosis and surgical treatment registration of hypospadias in the Danish National Patient Register (DNPR) remains unknown.

Patients and Methods: We randomly sampled 500 patients diagnosed with hypospadias in the DNPR from January 1, 1995 to December 31, 2012. Among these, 384 patients were also registered with surgical treatment for hypospadias. Medical records were collected and reviewed independently by two investigators. Any classification disagreements were resolved by consensus. Using the medical records as the gold standard, we estimated positive predictive values (PPVs) with 95% confidence intervals (CIs) for the hypospadias diagnoses and surgical treatment registrations overall, as well as for the clinical subtypes.

Result(s): We were able to retrieve medical records for 463 (92.6%) patients with hypospadias diagnoses and for 329 (85.7%) patients registered with surgical treatment. Presence of hypospadias was confirmed in 450 of 463 patients, yielding an overall PPV (95% CI) of 97.6% (95.8%-98.7%). For subtypes of hypospadias, the PPVs ranged between 37.5% and 72.7%. For surgical treatment of hypospadias, the overall PPV was 99.7% (97.9%-99.9%).

Conclusion(s): The validity of the registration of hypospadias diagnoses as well as surgical treatment for hypospadias in the DNPR is overall very high. For the specific subtypes of hypospadias diagnoses codes and the specific surgical treatment codes, the PPVs are lower and cautious use is warranted. However, the DNPR remains a valuable tool for future observational research on hypospadias.

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Institution

(Arendt, Ernst, Jonsson, Ramlau-Hansen) Department of Public Health, Section for Epidemiology, Aarhus University, Aarhus, Denmark (Arendt, Lindhard, Henriksen) Department of Paediatrics and Adolescent Medicine, Aarhus University Hospital, Aarhus, Denmark (Ernst, Olsen) Department of Urology, Section for Paediatric Urology, Aarhus University Hospital, Aarhus, Denmark (Olsen) Department of Clinical Epidemiology, Aarhus University Hospital, Aarhus, Denmark (Thorup) Department of Paediatric Surgery, Rigshospitalet and Institute of Clinical Medicine, University of Copenhagen, Copenhagen, Denmark
Publisher
Dove Medical Press Ltd (PO Box 300-008, Albany, 44 Corinthian Drive, Albany, Auckland 0752, New Zealand. E-mail: angela@dovepress.com)
Year of Publication
2017

480.

Comparison of interrupted- and continuous-suture urethroplasty in tubularised incised-plate hypospadias repair: A prospective study.

Gupta R., Srivastav P., Gupta A.

Embase

Arab Journal of Urology. 15 (4) (pp 312-318), 2017. Date of Publication: December 2017.

[Article]

AN: 619331398

Objective To compare the effect of interrupted- and continuous-suture urethroplasty on complication rates in Snodgrass tubularised incised-plate (TIP) hypospadias repair. **Patients and methods** This was a prospective randomised study comprising 100 boys (age range 1-5 years) with primary subcoronal, distal- and mid-penile hypospadias who underwent repair from October 2010 to March 2015 in a tertiary care hospital. Boys with glanular, recurrent, and proximal hypospadias were excluded from the study. The boys were prospectively randomised into two groups: Group A, comprised 50 boys who underwent interrupted subcuticular suture Snodgrass TIP urethroplasty; and Group B, comprised 50 boys who underwent continuous subcuticular suture Snodgrass TIP urethroplasty. Outcomes were assessed in terms of complication rates and aesthetic appearance during follow-up. **Results** There was no significant difference in the

occurrence of complications between the groups. There were 21 complications, with 10 occurring in Group A and 11 in Group-B. Urethrocutaneous fistula was the most common complication in both groups (six in Group A and seven in Group B), the fistulae were <2 mm in nine patients and 3-5 mm in the remaining four. Partial glans dehiscence occurred in one patient in each group. One patient from each group also had superficial wound infection, meatal stenosis and urethral stricture respectively, all of which were managed conservatively. The resultant urinary stream was single and good in all patients of both groups. Conclusions The type of suture technique had no significant effect on complication rates after Snodgrass hypospadias repair and thus the choice of technique depends on surgeon preference.

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Embase

Institution

(Gupta) Department of Pediatric Surgery, King George's Medical University, Lucknow, India

(Gupta, Srivastav) Unit of Pediatric Surgery, Department of Surgery, SN Medical College, Agra, India

(Gupta) Unit of Urology, Department of Surgery, SN Medical College, Agra, India

Publisher

Arab Association of Urology (E-mail: araburo@yahoo.com)

Year of Publication

2017

481.

Advances in paediatric urology.

Diamond D.A., Chan I.H.Y., Holland A.J.A., Kurtz M.P., Nelson C., Estrada C.R., Bauer S., Tam P.K.H.

Embase

The Lancet. 390 (10099) (pp 1061-1071), 2017. Date of Publication: 9 - 15 September 2017.

[Review]

AN: 619146569

Paediatric urological surgery is often required for managing congenital and acquired disorders of the genitourinary system. In this Series paper, we highlight advances in the surgical management

of six paediatric urological disorders. The management of vesicoureteral reflux is evolving, with advocacy ranging from a less interventional assessment and antimicrobial prophylaxis to surgery including endoscopic injection of a bulking agent and minimally invasive ureteric reimplantation. Evidence supports early orchidopexy to improve fertility and reduce malignancy in boys with undescended testes. A variety of surgical techniques have been developed for hypospadias, with excellent outcomes for distal but not proximal hypospadias. Pelvi-ureteric junction obstruction is mostly detected prenatally; indications for surgery have been refined with evidence, and minimally invasive pyeloplasty is now standard. The outlook for patients with neurogenic bladder has been transformed by a combination of clean intermittent catheterisation, algorithms of diagnostic investigations, and innovative medical and surgical therapies. Posterior urethral valves are associated with considerable mortality; fetal diagnosis allows stratification of candidates for intervention, but ongoing bladder dysfunction in patients after valve ablation remains a cause of long-term morbidity.

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Institution

(Diamond, Kurtz, Nelson, Estrada, Bauer) Department of Urology, Boston Children's Hospital, Harvard Medical School, Boston, MA, United States (Chan, Tam) Department of Surgery, The University of Hong Kong, Queen Mary Hospital, Hong Kong

(Holland) Department of Paediatric Surgery, The Children's Hospital at Westmead, Westmead, NSW, Australia

(Holland) Sydney Medical School, University of Sydney, Sydney, NSW, Australia

Publisher

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Year of Publication

2017

482.

Molecular diagnosis in hypospadias. Diagnostico molecular en hipospadias <Diagnostico molecular en hipospadias.>

Abello A., Ayala P., Ortiz A.M., Fernandez N.

Embase

Urologia Colombiana. 26 (3) (pp 180-185), 2017. Date of Publication: September 2017.

[Article]

AN: 619097303

Objective The aim of this study is to perform a systematic review of the principal genetic and molecular diagnostic methods for hypospadias and their usefulness. **Methods** A Pubmed and EMBASE search was carried out using the following MESH terms: "molecular diagnosis", "genes", "hypospadias", "karyotype", "wgs", "fish", "chg", "sanger", "microarray", "mps", "wes", and "gwas". Meta-analyses, systematic reviews, Cochrane reviews, clinical trials, narrative reviews and case series were included, between 2001 and 2016, in both Spanish and English. A total of 33 items were selected for review after reviewing titles, abstracts, and cross references. **Results** Hypospadias are the birth defect of the ventral aspect of the penis, accompanied by an ectopic location of the urethral meatus. 30% of all birth malformations in the newborns are urological malformations make up 30% of all birth malformations in newborns. Within the genetic and molecular test available for diagnosis, many are of varying usefulness. These include, among others, karyotyping, FISH, and Sanger sequencing. **Conclusions** Due to advances in technology, there are multiple molecular diagnosis methods that can widen the knowledge of the aetiology of hypospadias. They also allow them to be used in the everyday practice for a complete study of patients.

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Institution

(Abello) Medico Cirujano, Pontificia Universidad Javeriana, Bogota, Colombia (Ayala) Instituto de Genetica, Pontificia Universidad Javeriana, Bogota, Colombia

(Ortiz) Medico Interno, Pontificia Universidad Javeriana, Bogota, Colombia

(Fernandez) Medico Urologo, Unidad de Urologia, Hospital Universitario San Ignacio, Pontificia Universidad Javeriana, Bogota, Colombia

Publisher

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Year of Publication

2017

483.

Hypospadias repair during adulthood: Case series.

Al Taweel W.M., Seyam R.M.

Embase

Urology Annals. 9 (4) (pp 366-371), 2017. Date of Publication: October-December 2017.

[Article]

AN: 619006295

Purpose: We report our complications and success rate in adult hypospadias repair.

Patients and Methods: This was a retrospective study of patients aged ≥ 17 years who underwent hypospadias repair during 2006-2014. We analyzed two groups, one with primary repair and the other that had secondary repair after failed childhood hypospadias surgery.

Outcome was compared between the two groups and among different surgical procedures.

Descriptive statistics and Fisher's exact test were used and significance level was set at $P < 0.05$.

Result(s): Forty patients were included, of which 26 presented for a secondary repair and 14 for primary repair. The meatus was distal in 15 patients, mid-penile in 16, and penoscrotal in 9. The median age of patients at the time of surgery was 21 years (standard deviation [SD] =4, range 17-30). The median follow-up period was 37 months (SD = 8, range 5-75). In the primary repair group, the success rate was 71% (10/14), whereas in the secondary repair group, the success rate was 55% (14/26). The overall complication rate was 60%. Following a subsequent repair, the overall success rate reached 95% (38/40). There was no significant difference in success or complications between patients who presented with primary or secondary hypospadias or between methods of repair.

Conclusion(s): Delayed hypospadias repair in adults is associated with a high success rate of 95% with no difference between primary and secondary repair. Secondary repair however may require more than one procedure most of the time.

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Institution

(Al Taweel, Seyam) Department of Urology, King Faisal Specialist Hospital and Research Center, P.O. Box: 3354, Riyadh 11211, Saudi Arabia

Publisher

Medknow Publications (B9, Kanara Business Centre, off Link Road, Ghatkopar (E), Mumbai 400 075, India)

Year of Publication

2017

484.

Choosing an ideal vascular cover for Snodgrass repair.

Basavaraju M., Balaji D.K.

Embase

Urology Annals. 9 (4) (pp 348-352), 2017. Date of Publication: October-December 2017.

[Article]

AN: 619006258

Aim: The aim of this study is to compare tunica vaginalis (TV), dorsal dartos, and ventral dartos flap as a second layer vascular cover during Snodgrass repair.

Material(s) and Method(s): Data of 83 patients who underwent primary hypospadias repair with Snodgrass technique (age range: 1.6-12 years) were retrospectively collected and compared. They were divided into three groups. Group A (26 patients) included cases using TV flap, Group B (36 patients) included those where dorsal dartos from prepuce was used as second cover, and Group C (21 patients) included those with ventral dartos as cover.

Result(s): In Group A, no complications recorded. Mild scrotal edema was present in 5 patients which was conservatively managed. In Group B, there were 8 fistulas, 2 glans breakdown, and 1 meatal stenosis. In Group C, there were 3 fistulas and 1 glans breakdown.

Conclusion(s): TV flap is better than dorsal dartos and ventral dartos as vascular cover for primary hypospadias repair with Snodgrass technique.

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Institution

(Basavaraju, Balaji) Departments of Pediatric Surgery, Vydehi Institute of Medical Science, 178/2, Shivakrupa Venkatapura, 5th Cross, Koramangala, Bengaluru, Karnataka 560 034, India

Publisher

Medknow Publications (B9, Kanara Business Centre, off Link Road, Ghatkopar (E), Mumbai 400 075, India)

Year of Publication

2017

485.

Amniotic therapeutic biomaterials in urology: Current and future applications.

Oottamasathien S., Hotaling J.M., Craig J.R., Myers J.B., Brant W.O.

Embase

Translational Andrology and Urology. 6 (5) (pp 943-950), 2017. Date of Publication: 01 Oct 2017.

[Review]

AN: 618986266

To examine the rationale and applications of amniotic tissue augmentation in urological surgery.

Published literature in English-language was reviewed for basic science and clinical use of amniotic or amnion-chorionic tissue in genitourinary tissues. Basic science and animal studies support the likely benefit of clinical applications of amnion-derived tissues in a variety of urologic interventions. The broad number of properties found in amniotic membrane, coupled with its immunologically privileged status presents a number of future applications in the urological surgical realm. These applications are in their clinical infancy and suggest that further studies are warranted to investigate the use of these products in a systematic fashion.

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Institution

(Oottamasathien, Hotaling, Craig, Myers, Brant) Department of Surgery, Section of Pediatric Urology, University of Utah, Salt Lake City, UT, United States (Oottamasathien) Primary Children's Hospital, University of Utah, Salt Lake City, UT, United States

(Hotaling, Craig, Myers, Brant) Department of Surgery, Division of Urology Section of Men's Health, University of Utah, Salt Lake City, UT, United States

Publisher

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Year of Publication

2017

486.

Molecular diagnosis in hypospadias. Diagnostico molecular en hipospadias <Diagnostico molecular en hipospadias.>

Abello A., Ayala P., Ortiz A.M., Fernandez N.

Embase

Urologia Colombiana. 26 (3) (pp 192-197), 2017. Date of Publication: September 2017.

[Article]

AN: 618781425

Objective The aim of this study is to perform a systematic review of the principal genetic and molecular diagnostic methods for hypospadias and their usefulness. **Methods** A Pubmed and EMBASE search was carried out using the following MESH terms: "molecular diagnosis", "genes", "hypospadias", "karyotype", "wgs", "fish", "chg", "sanger", "microarray", "mps", "wes", and "gwas". Meta-analyses, systematic reviews, Cochrane reviews, clinical trials, narrative reviews and case series were included, between 2001 and 2016, in both Spanish and English. A total of 33 items were selected for review after reviewing titles, abstracts, and cross references. **Results** Hypospadias are the birth defect of the ventral aspect of the penis, accompanied by an ectopic location of the urethral meatus. 30% of all birth malformations in the newborns are urological malformations make up 30% of all birth malformations in newborns. Within the genetic and molecular test available for diagnosis, many are of varying usefulness. These include, among others, karyotyping, FISH, and Sanger sequencing. **Conclusions** Due to advances in technology, there are multiple molecular diagnosis methods that can widen the knowledge of the etiology of hypospadias. They also allow them to be used in the everyday practice for a complete study of patients.

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Institution

(Abello) Medico Cirujano, Pontificia Universidad Javeriana, Bogota, Colombia (Ayala) Instituto de Genetica, Pontificia Universidad Javeriana, Bogota, Colombia

(Ortiz) Medico Interno, Pontificia Universidad Javeriana, Bogota, Colombia

(Fernandez) Medico Urologo, Unidad de Urologia, Hospital Universitario San Ignacio, Pontificia Universidad Javeriana, Bogota, Colombia

Publisher

487.

Role of urethral plate and fossa navicularis biopsies in the detection of balanitis xerotica obliterans in boys undergoing redo hypospadias repair.

Sultan M., El-Shazly M., Elsherif E., Younes S., Selim M.

Embase

Arab Journal of Urology. 15 (4) (pp 326-330), 2017. Date of Publication: December 2017.

[Article]

AN: 618314717

Objectives To evaluate the presence of balanitis xerotica obliterans (BXO), clinically and pathologically, in the urethra of boys with failed previous hypospadias repair and where surgical management was planned. **Patients and methods** Between February 2010 and March 2015, boys with failed distal penile hypospadias repair who were planned for surgical management were evaluated for the presence of clinical and pathological evidence of BXO. Samples were obtained from the urethral plate and fossa navicularis, after obtaining informed consent and ethical approval. The samples were fixed, sectioned, and haematoxylin and eosin stained for light microscopic examination. **Results** In all, 157 boys were enrolled in our study, with a mean (SD) age of 6.4 (2.8) years. All the boys had a history of failed hypospadias repair surgeries (once or more). The presentation was fistula in 34 boys (21.7%), meatal stenosis in 45 (28.7%), urethral stricture in 28 (17.8%), and total dehiscence in 50 (31.8%). BXO was detected clinically in 46 boys (29.3%). The total number of biopsies taken was 314, of which 124 (39.5%) were pathologically BXO-positive samples. Of the 157 boys, BXO-positive cases were clinically associated with fistula in seven boys (4.5%), meatal stenosis in 18 (10.8%), urethral stricture in seven (4.5%), and total dehiscence in 15 (9.6%). Of the 314 pathological samples, pathologically BXO-positive samples were associated with fistula in 20 samples (6.4%), meatal stenosis in 40 (12.7%), urethral stricture in 22 (7%), and total dehiscence in 42 (13.4%). **Conclusions** In failed hypospadias cases BXO should be considered, especially for cases with multiple failures, meatal stenosis, and total dehiscence. Urethral plate and fossa navicularis biopsies are important in planning a proper approach for subsequent repair.

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Institution

(Sultan, El-Shazly, Elsherif, Selim) Department of Urology, Faculty of Medicine, Menoufia University, Menoufia Governorate, Egypt (Younes) Department of Pathology, Faculty of Medicine, Menoufia University, Menoufia Governorate, Egypt

Publisher

Arab Association of Urology (E-mail: araburo@yahoo.com)

Year of Publication

2017

488.

Staged Tubularized Autograft Repair for Primary Proximal Hypospadias with 30-Degree or Greater Ventral Curvature.

Snodgrass W., Bush N.

Embase

Journal of Urology. 198 (3) (pp 680-686), 2017. Date of Publication: September 2017.

[Article]

AN: 617478500

Purpose We report outcomes in consecutive patients with primary proximal hypospadias and ventral curvature 30 degrees or greater after degloving, all repaired with 2-stage tubularized autografts, a variation of the Nicolle-Bracka procedure. **Materials and Methods** Consecutive boys with proximal hypospadias and ventral curvature 30 degrees or greater after degloving underwent transection of the urethral plate. In addition, those with persistent ventral curvature 30 degrees or greater underwent 3 ventral corporotomies without corporal grafting for straightening. Staged graft urethroplasty was performed using prepuce, or labial mucosa, when the family requested preputioplasty. The goal in all cases was a straight penis with a normally positioned neomeatus. **Results** A total of 43 boys completed both stages with postoperative followup. Mean ventral curvature was 70 degrees, which necessitated corporotomy in 65%. There was no recurrent ventral curvature. Preputial grafts were used in 88%. A skin graft or scrotal flap was needed for ventral shaft coverage at the second stage in 4 boys (9%). During followup of an average of 22

months urethroplasty complications were diagnosed in 10 (23%), mostly glans dehiscence and fistulas. Of these patients 9 underwent 12 reoperations. Success was achieved in 42 (98%) patients. Conclusions Staged tubularized autograft resulted in a straight penis with a normally positioned neomeatus in boys with the most severe proximal hypospadias in 77%, increasing to 98% after 1 or 2 distal reoperations for complications. There was no recurrent ventral curvature after 3 corporotomies without corporal grafting. Urethroplasty graft take was reliable.

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Embase

Institution

(Snodgrass, Bush) PARC Urology, Frisco, Texas, United States

Publisher

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Year of Publication

2017

489.

Failed Primary Bladder Exstrophy Closure with Osteotomy: Multivariable Analysis of a 25-Year Experience.

Sirisreetreerux P., Lue K.M., Ingviya T., Friedlander D.A., Di Carlo H.N., Sponseller P.D., Gearhart J.P.

Embase

Journal of Urology. 197 (4) (pp 1138-1143), 2017. Date of Publication: 01 Apr 2017.

[Article]

AN: 614663936

Purpose Successful primary bladder exstrophy closure provides the best opportunity for patients to achieve a functional closure and urinary continence regardless of the method of repair. Use of osteotomy during initial closure has significantly improved success rates. However, failures can still occur. We identify factors that contribute to a failed primary exstrophy closure with osteotomy. Materials and Methods We reviewed a prospectively maintained institutional database for classic

bladder exstrophy cases primarily closed with osteotomy at our institution or referred after primary closure between 1990 and 2015. Data were collected regarding patient gender, closure, osteotomy, immobilization, orthopedics and perioperative pain control. Univariate and multivariable analyses were performed to determine predictors of failure. Results A total of 156 patients met inclusion criteria. Overall failure rate was 30% (13% from our institution and 87% from referrals). On multivariable analysis use of Buck traction (OR 0.11, 95% CI 0.02-0.60, $p = 0.011$) and immobilization time greater than 4 weeks (OR 0.19, 95% CI 0.04-0.86, $p = 0.031$) had significantly lower odds of failure. Osteotomy performed by general orthopedic surgeons had significantly higher odds of failure (OR 23.47, 95% CI 1.45-379.19, $p = 0.027$). Type of osteotomy and use of epidural anesthesia did not significantly impact failure rates. Conclusions Proper immobilization with modified Buck traction and external fixation, immobilization time greater than 4 weeks and undergoing osteotomy performed by a pediatric orthopedic surgeon are crucial factors for successful primary closure with osteotomy.

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27720781 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=27720781>]

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Institution

(Sirisreetreerux, Lue, Friedlander, Di Carlo, Gearhart) Jeffs Division of Pediatric Urology, James Buchanan Brady Urological Institute, The Johns Hopkins University School of Medicine, Baltimore, Maryland, United States (Ingviya) Department of Environmental Health Sciences, Bloomberg School of Public Health, The Johns Hopkins University School of Medicine, Baltimore, Maryland, United States

(Sponseller) Division of Pediatric Orthopaedics, The Johns Hopkins University School of Medicine, Baltimore, Maryland, United States

Publisher

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Year of Publication

2017

Cause and Effect versus Confounding-Is There a True Association between Caudal Blocks and Tubularized Incised Plate Repair Complications?.

Braga L.H., Jegatheeswaran K., McGrath M., Easterbrook B., Rickard M., DeMaria J., Lorenzo A.J.

Embase

Journal of Urology. Part 2. 197 (3) (pp 845-851), 2017. Date of Publication: 01 Mar 2017.

[Article]

AN: 614208638

Purpose We studied the impact of caudal block vs dorsal penile block on the rate of urethrocuteaneous fistula and glans dehiscence in children who underwent hypospadias repair. **Materials and Methods** We retrospectively reviewed the records of 849 consecutive patients who underwent tubularized incised plate repair between 2004 and 2015. A total of 331 cases with incomplete medical records, other techniques and redo repair were excluded. The preference for caudal block was based on anesthesiologist discretion. Age at surgery, meatal location, preoperative testosterone stimulation, type of regional anesthesia (caudal block vs dorsal penile block), degree of ventral curvature, surgeon expertise and complications (urethrocuteaneous fistula/glans dehiscence) were captured. Univariate and multivariable analyses were done of risk factors for complications. **Results** Median age at surgery was 18 months and median followup was 6 months. Of 518 patients 405 (78%) had distal and 113 (22%) had mid shaft/proximal defects. Complications developed in 37 cases (7%), including urethrocuteaneous fistula in 21 (19 with a caudal block and 2 with a dorsal penile block) and glans dehiscence in 16 (13 with a caudal block and 3 with a dorsal penile block). On univariate analysis preoperative testosterone stimulation vs no preoperative testosterone stimulation (13.0% vs 6.2% of cases, $p = 0.04$), mid shaft/proximal vs distal defects (15.9% vs 4.7%, $p < 0.01$) and caudal block (8.7% vs 3.3%, $p = 0.03$) were significantly associated with more complications. However, on multivariable analysis the associations of preoperative testosterone stimulation (OR 1.2, 95% CI 0.4-3.7) and caudal block (OR 2.4, 95% CI 0.9-6.4) with complications did not hold. Only the combination of meatal location/ventral curvature remained as an independent risk factor for urethrocuteaneous fistula/glans dehiscence (OR 2.4, 95% CI 1.1-5.7, $p = 0.04$). **Conclusions** Our data indicate that hypospadias severity and not the type of regional anesthesia was the only risk factor significantly associated with postoperative complications. To confirm these findings and provide strong and definitive evidence on this topic a well powered, randomized, controlled trial is clearly required.

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Embase

Institution

(DeMaria) Division of Urology, Department of Surgery, McMaster University, Hamilton, Ontario, Canada (Jegatheeswaran, McGrath, Easterbrook) McMaster Pediatric Surgery Research Collaborative, McMaster University, Hamilton, Ontario, Canada

(Braga, Jegatheeswaran, McGrath, Easterbrook, Rickard, DeMaria) Clinical Urology Research Enterprise Program, McMaster Children's Hospital, Hamilton, Ontario, Canada

(Lorenzo) Division of Urology, Hospital for Sick Children, Toronto, Ontario, Canada

Publisher

Elsevier Inc. (E-mail: usjcs@elsevier.com)

Year of Publication

2017

491.

Resurfacing the Penis of Complex Hypospadias Repair ("Hypospadias Cripples").

Fam M.M., Hanna M.K.

Embase

Journal of Urology. Part 2. 197 (3) (pp 859-864), 2017. Date of Publication: 01 Mar 2017.

[Article]

AN: 614202379

Purpose After the creation of a neourethra in a "hypospadias cripple," resurfacing the penis with healthy skin is a significant challenge because local tissue is often scarred and unusable. We reviewed our experience with various strategies to resurface the penis of hypospadias cripples. **Materials and Methods** We retrospectively reviewed the records of 215 patients referred after multiple unsuccessful hypospadias repairs from 1981 to 2014. In 130 of 215 patients we performed resurfacing using local penile flaps using various techniques, including Byars flaps, Z-plasty or double Z-plasty, or a dorsal relaxing incision. Of the 215 patients 85 did not have adequate healthy local penile skin to resurface the penis after urethroplasty. Scrotal skin was used to resurface the penis in 54 patients, 6 underwent tissue expansion of the dorsal penile skin during a 12 to 16-week period prior to penile resurfacing, 23 underwent full-thickness skin grafting and another 4 received a split-thickness skin graft. **Results** Of the 56 patients who underwent fasciomyocutaneous rotational flaps, tissue expansion or a combination of both approaches 54 (96.4%) finally had a successful outcome. All 6 patients who underwent tissue expansion had a

successful outcome without complications and were reported on previously. All 23 full-thickness skin grafts took with excellent results. All 4 patients who underwent fenestrated split-thickness skin grafting had 100% graft take but secondary contraction and ulceration were associated with sexual activity. Conclusions In our experience scrotal skin flaps, tissue expansion of the dorsal penile skin and full-thickness skin grafts serve as reliable approaches in resurfacing the penis in almost any hypospadias cripple lacking healthy local skin.

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Status

Embase

Institution

(Fam, Hanna) Division of Urology, Rutgers New Jersey Medical School, Newark, New Jersey, United States (Fam, Hanna) Department of Urology, New York-Presbyterian Weill Cornell Medical Center, New York, New York, United States

Publisher

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Year of Publication

2017

492.

Intermediate-Term Followup of Proximal Hypospadias Repair Reveals High Complication Rate. Long C.J., Chu D.I., Tenney R.W., Morris A.R., Weiss D.A., Shukla A.R., Srinivasan A.K., Zderic S.A., Kolon T.F., Canning D.A.

Embase

Journal of Urology. Part 2. 197 (3) (pp 852-858), 2017. Date of Publication: 01 Mar 2017.

[Article]

AN: 614202306

Purpose Results following distal hypospadias repair are favorable. Grouping proximal and distal hypospadias repair artificially increases the perceived success rate of proximal hypospadias. We identified our complication rate of proximal hypospadias repair and hypothesized a higher complication rate for 1-stage repair. Materials and Methods We retrospectively reviewed the

records of consecutive boys who underwent proximal hypospadias from 2007 to 2014. Proximal hypospadias was defined as a urethral meatus location at or more proximal than the penoscrotal junction after penile degloving. We further stratified boys into those with planned 1-stage vs 2-stage repair. Univariate and Cox regression analyses were performed to assess associations with covariates and compare time to the first complication, respectively. Results A total of 167 boys met study inclusion criteria. Median followup was 31.7 months for 1-stage repair in 86 patients and staged repair in 81. The overall complication rate was 56%. Complications developed in 53 of 86 1-stage (62%) vs 40 of 81 staged (49%) repairs ($p = 0.11$). The number of unplanned procedures per patient was higher in the 1-stage than in the staged group (0.99 vs 0.69, $p = 0.06$), as was the number of patients who had at least 2 complications (29 of 86 or 33% vs 13 of 81 or 16%, $p = 0.03$). Cox regression showed no difference in time to the first complication for staged compared to 1-stage repair (HR 0.77, 95% CI 0.43-1.39). Conclusions Our 56% complication rate of proximal hypospadias warrants further long-term patient followup. More patients in the 1-stage group experienced at least 2 complications. However, when complications developed, they developed no differently in the 2 groups.

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Institution

(Long, Chu, Tenney, Morris, Weiss, Shukla, Srinivasan, Zderic, Kolon, Canning) Division of Urology, Children's Hospital of Philadelphia, Perelman School of Medicine at University of Pennsylvania, Philadelphia, Pennsylvania, United States

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493.

Assessing quality of life of patients with hypospadias: A systematic review of validated patient-reported outcome instruments.

Sullivan K.J., Hunter Z., Andrioli V., Guerra L., Leonard M., Klassen A., Keays M.A.

Embase

Journal of Pediatric Urology. 13 (1) (pp 19-27), 2017. Date of Publication: 01 Feb 2017.

[Review]

AN: 614105869

Background Patient-reported outcomes have the potential to provide invaluable information for evaluation of hypospadias patients, aid in decision-making, performance assessment, and improvement in quality of care. To appropriately measure patient-relevant outcomes, well-developed and validated patient-reported outcome (PRO) instruments are essential. **Objective** To identify and evaluate existing PRO instruments designed to measure quality of life and/or satisfaction of individuals with hypospadias that have been developed and validated in a hypospadias population. **Methods** A systematic search of MEDLINE, EMBASE, PsycINFO, CINAHL and Health and Psychosocial Instruments was conducted in April 2016. Two reviewers independently assessed studies and identified PRO instruments for inclusion. Data were extracted on study characteristics, instrument development and validation, and content domains. **Results** A total of 32 studies were included that used or described five PRO instruments: Hypospadias Objective Scoring Evaluation (HOSE), Pediatric Penile Perception Score (PPPS), Penile Perception Score (PPS), Genital Perception Scale (GPS) for adults, and GPS for children/adolescents. Instrument development and validation was limited. The majority of identified instruments focused on postoperative cosmetic satisfaction, with only one instrument considering urinary function, and no instruments evaluating sexual function and psychosocial sequelae. **Conclusions** While many hypospadias studies have acknowledged the necessity of a patient-reported element, few have used validated PRO instruments developed in a hypospadias population. Existing instruments to measure patient-reported outcomes in hypospadias require improvement in both the breadth of content and in their development and validation methodology.

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Embase

Author NameID

Hunter Z.; ORCID: <http://orcid.org/0000-0002-0991-3071> Keays M.A.; ORCID: <http://orcid.org/0000-0002-8523-8610>

Institution

(Sullivan, Andrioli, Guerra, Leonard, Keays) Department of Pediatric Surgery, Division of Urology, Children's Hospital of Eastern Ontario, 401 Smyth Road, Ottawa, ON, Canada (Hunter, Guerra, Leonard, Keays) Faculty of Medicine, University of Ottawa, 451 Smyth Rd, Ottawa, ON, Canada

(Klassen) Department of Pediatrics, McMaster University, 3N27, 1280 Main Street West,
Hamilton, ON, Canada

(Klassen) Department of Clinical Epidemiology & Biostatistics, McMaster University, 3N27, 1280
Main Street West, Hamilton, ON, Canada

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494.

A simple technique for small-diameter urethrocuteaneous fistula repair: Ligation.

Karakus S.C., User I.R., Akcaer V., Ozokutan B.H., Ceylan H.

Embase

Journal of Pediatric Urology. 13 (1) (pp 88-90), 2017. Date of Publication: 01 Feb 2017.

[Article]

AN: 614063244

Objective To describe a simple and effective technique for repairing a small-diameter urethrocuteaneous fistula. Methods and technique A total of 13 patients with a solitary and small-diameter (≤ 2 mm) urethrocuteaneous fistula underwent repair with a ligation technique. Results None of the patients had voiding difficulties. One recurrent urethrocuteaneous fistula occurred and it was successfully repaired with the same technique. Conclusion This is a simple, quick and useful technique, particularly for small-diameter (≤ 2 mm) urethrocuteaneous fistulas.

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Institution

(Karakus, User, Akcaer, Ozokutan, Ceylan) Department of Pediatric Surgery, Faculty of Medicine,
University of Gaziantep, Gaziantep, Turkey

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495.

Two-stage hypospadias repair with a free graft for severe primary and revision hypospadias: A single surgeon's experience with long-term follow-up.

Pfistermuller K.L.M., Manoharan S., Desai D., Cuckow P.M.

Embase

Journal of Pediatric Urology. 13 (1) (pp 35.e1-35.e7), 2017. Date of Publication: 01 Feb 2017.

[Article]

AN: 614020440

Introduction Repair of severe primary and revision hypospadias is a demanding procedure.

Debate continues as to whether a two-stage approach or single-stage technique is superior. The two-stage procedure with a free graft involves penile straightening followed by application of a graft for the neourethral plate at stage one; with tubularization at stage two after graft maturation.

Objective To report the outcomes of a single surgeon's experience with the two-stage repair using a free graft for both severe primary and revision hypospadias with long-term follow-up.

Materials and methods Between July 1998 and January 2010, 301 boys underwent a two-stage reconstruction. The surgical technique is described in the manuscript. Primary repairs (n = 208): indications for a two-stage approach with a free graft included meatal position, presence of corporal chordee, and poor glans development. Median follow-up from completion of the second stage was 75 months. Revision repairs (n = 93): indications included urethral fistula, excessive scarring/meatal stenosis, balanitis xerotica obliterans (BXO), and residual or untreated chordee. Median follow-up from completion of the second stage was 85 months. Results For the primary repairs (n = 208), the graft took well in all but one case. Second-stage complications included fistula (7), meatal stenosis (3), partial glans dehiscence (3), and all were re-operated (13). For the revision repairs (n = 93), the graft took well in all but four cases. Second-stage complications included fistula (5), meatal stenosis (3), breakdown (1) and reoperation (8). Discussion In a systematic review of 20 years of publications on the repair of primary severe hypospadias, the two-stage procedure with a free graft demonstrated an overall complication rate of 22%; this was a distinct overall benefit when compared with the single-stage procedures in terms of lower complication rates (Castagnetti and El-Ghoneimi, 2010). Our results for the severe primary

repairs revealed significantly lower complication rates than those in the literature, with an overall re-operation rate of 6.3%, a fistula rate of 3.4%, and meatal stenosis and partial glans dehiscence at 1.4% each. Several papers have documented outcomes following the single-stage tubularized incised plate urethroplasty for re-operative hypospadias, giving overall complication rates ranging from 15.4 to 30%. Our data show a re-operative rate of 8.6%, a fistula rate of 5.3%, breakdown in 1.1%, and meatal stenosis in 3.2%. Conclusion The two-stage repair with a free graft for correction of both severe primary and failed primary hypospadias is a safe, viable, and durable procedure offering low morbidity and excellent cosmetic results. The authors advocate the two-stage repair with a free graft as the technique of choice for treatment of both of these challenging groups of the deformity.[Table presented]

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Status

Embase

Institution

(Pfistermuller, Manoharan, Desai, Cuckow) Department of Paediatric Urology, Great Ormond Street Hospital, Great Ormond Street, London WC1N 3JN, United Kingdom

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496.

The SIGHT questionnaire: A novel assessment tool for Satisfaction In Genital Hypospadias Treatment.

Ardelt P.U., Cederqvist M., Barth M., Frankenschmidt A.

Embase

Journal of Pediatric Urology. 13 (1) (pp 33.e1-33.e8), 2017. Date of Publication: 01 Feb 2017.

[Article]

AN: 613872883

Purpose Psychosexual development is currently underrepresented in hypospadias outcome research. The aim of this study was to develop and validate a questionnaire addressing psychosexual long-term satisfaction, specifically of adolescent patients, after hypospadias repair.

Materials and methods In a multistep participative design we identified key interests of adolescent patients with hypospadias. Next, a questionnaire addressing specifically the psychosexual satisfaction of adolescents after hypospadias repair was established. A population of 109 former patients with hypospadias was then assessed using this questionnaire. Furthermore, functional and cosmetic aspects, behavioural anomalies, and sexual activity were investigated. Age-matched patients undergoing circumcision served as control patients. Possible influence factors on patient satisfaction were investigated. Clinical trial registry site: German Registry of Clinical Trials DRKS, Freiburg, Germany (Reference: DRKS00003432).

Results Key interests of adolescent patients were "normal appearance of the penis", "normal function of the penis regarding voiding and sexual activity", "no limitations regarding cosmetic appearance to others", "no limitations to sexual activity", and an "unimpaired masculine identity". The "Satisfaction In Genital Hypospadias Treatment" (SIGHT) questionnaire was developed using these items and using previously published evaluation systems. Nine questions address psychosexual aspects and two additional questions address current sexual activity. Internal consistency was high and retest reliability acceptable. The patient population showed a normal strength and difficulties score (SDQ). Overall satisfaction was high and similar to that of the control group. In a Spearman correlation a high SDQ value, erectile problems, and complications correlated negatively with satisfaction.

Conclusion To date, few studies have examined patients' satisfaction and psychosexuality. To our knowledge, the SIGHT questionnaire is the first to be developed participatively and in a stepwise fashion in collaboration with a paediatric psychologist and an open approach to determine items specifically important for adolescents. The SIGHT questionnaire can thus offer a relevant assessment of patients' psychosexual satisfaction. It is most suitable to supplement current strategies that so far mostly neglect the impact on psychosexual wellbeing.

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Status

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Institution

(Ardelt) Department of Urology, University Hospital Basel, Switzerland (Cederqvist, Frankenschmidt) Department of Urology and Paediatric Urology, Medical School, Albert-Ludwigs-University of Freiburg, Germany

(Barth) Department of Paediatric Psychology, Medical School, Albert-Ludwigs-University of Freiburg, Germany

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497.

Modified tubularized incised plate urethroplasty in distal hypospadias repair: Stepwise technique with validated functional and cosmetic outcome.

Spinoit A.-F., Radford A., Ashraf J., Gopal M., Subramaniam R.

Embase

Journal of Pediatric Urology. 13 (1) (pp 86-87), 2017. Date of Publication: 01 Feb 2017.

[Article]

AN: 613862136

Objective To describe modification of the tubularized incised plate urethroplasty (TIP) for distal hypospadias, and assess its efficacy, and functional and cosmetic outcomes. **Methods** A prospective evaluation of a consecutive series of patients operated for primary distal hypospadias was conducted at a tertiary reference center. A standardized modification of the TIP (mTIP) procedure was performed on a 10 French catheter. Clinical data were collected in a dedicated database. Intraoperative variables, postoperative complications and outcomes, by means of uroflowmetries and a validated (HOPE) questionnaire, were assessed. Efficacy was evaluated with the reported complications: functional outcome was evaluated with uroflowmetries and cosmetic assessment by a validated questionnaire (HOPE). A descriptive statistical analysis was performed. **Results** Of the 112 boys operated between 30/09/2011 and 1/04/2014, 50 completed long-term follow-up with functional and esthetic evaluation, as required for inclusion. Median age at surgery was 25 months (range 14-156); median follow-up time was 21.5 months (range 6-48). Complications requiring re-intervention occurred in 2/50 boys. Uroflowmetry presented a bell-shaped curve in 47/50 boys, and the median HOPE score was 9.5 (range 7.6-10.0). **Conclusion** The mTIP procedure provided satisfactory long-term functional and cosmetic outcomes, as validated by uroflowmetries and standardized questionnaire.

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Status

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Author NameID

Spinoit A.-F.; ORCID: <http://orcid.org/0000-0002-9045-9127>

Institution

(Radford, Ashraf, Gopal, Subramaniam) Department of Paediatric Surgery and Urology, Leeds Teaching Hospital, Leeds, United Kingdom (Spinoit) Department of Urology, Ghent University Hospital, Ghent, Belgium

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498.

Erratum to: Hypospadias, all there is to know (European Journal of Pediatrics, (2017), 176, 4, (435-441), 10.1007/s00431-017-2864-5).

van der Horst H.J.R., de Wall L.L.

Embase

European Journal of Pediatrics. 176 (10) (pp 1443), 2017. Date of Publication: 01 Oct 2017.

[Erratum]

AN: 618003764

The original version of this article, unfortunately, contained an error. In the original article, the statement found on page 438, left column, third paragraph, the bibliographic information of reference citation [12] should be the below: Jones BC, O'Brien M, Chase J, Southwell BR, Hutson JM (2009) Early hypospadias surgery may lead to a better longterm psychosexual outcome. J Urol 182 (4 suppl): 1744-1749.

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Embase

Institution

(van der Horst) Department of Urology, VUmc, De Boelelaan 1117, P.O. Box 7057, Amsterdam 1007 MB, Netherlands (de Wall) Department of Urology, Radboudumc, Geert Grooteplein 10, P.O. Box 9101, Nijmegen 6500 HB, Netherlands

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Year of Publication

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499.

Spinal anesthesia for pediatric urological surgery: Reducing the theoretic neurotoxic effects of general anesthesia.

Whitaker E.E., Wiemann B.Z., DaJusta D.G., Alpert S.A., Ching C.B., McLeod D.J., Tobias J.D., Jayanthi V.R.

Embase

Journal of Pediatric Urology. 13 (4) (pp 396-400), 2017. Date of Publication: August 2017.

[Article]

AN: 618497614

Background Spinal anesthesia (SA) is an effective technique that has been used in children for years. With growing concern with regard to the risks of general anesthesia (GA), we developed a SA program to provide an alternative option. We present our initial experience with this program.

Objective To implement a SA program at a large tertiary care pediatric center and assess the safety and efficacy of the technique as an alternative to GA for urologic surgery. Study design/methods We prospectively collected data on all children undergoing SA at our institution. We recorded demographics, procedure, time required for placement of the SA, length of surgery, success of lumbar puncture, success of attaining adequate surgical anesthesia, need for supplemental systemic sedation, conversion to GA, and perioperative complications. Results SA was attempted in 105 consecutive children (104 boys, 1 girl) with a mean age of 7.4 +/- 4.3 months (range 19 days-24 months) and mean weight of 8.3 +/- 1.7 kg (range 3.5-13.7).

Placement of the SA was successful in 93/105 children (89%). Inability to achieve lumbar puncture (cerebrospinal fluid was not obtained) meant that SA was abandoned in seven (7%)

patients and GA was administered. In five patients in whom SA was successful and surgery was begun, 5/93 (5%) required conversion to GA: two because of evisceration of intestine through large hernia defects related to coughing and abdominal irritation, two because of lack of motor blockade despite an adequate sensory block, and one because of an inability to place an intravenous catheter in the lower extremities (required per SA protocol). If necessary, an intravenous catheter can be placed in the upper extremity, but this must be weighed against the fact that the block has already been placed and is of limited duration. Overall, SA was successful (SA was placed and surgery was completed without conversion to GA) in 88/105 children (84%). No additional sedation and no systemic anesthetic agents were required in 75/88 children (85%). The average time required to place the SA was 3.8 +/- 2.7 min (range 1-12). The average time for the surgical procedure was 38.3 +/- 23.1 min (range 10-122). No patient required conversion to GA because of recession of block. There were no surgical complications. Discussion/conclusions SA is a safe and efficacious technique for routine pediatric urological procedures. SA should be considered for cases such as neonatal torsion or patients with significant cardiac or pulmonary comorbidities when the risks of GA are often weighed against the risks of non-intervention.

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Status

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Author NameID

Whitaker, Emmett E.; ORCID: <http://orcid.org/0000-0002-1362-1653> Alpert, Seth A.; ORCID: <http://orcid.org/0000-0001-7034-603X>

Institution

(Whitaker, Tobias) Department of Anesthesiology, The Ohio State University College of Medicine, Columbus, OH 43210, United States (Whitaker, Tobias) Department of Anesthesiology and Pain Medicine, Nationwide Children's Hospital, Columbus, OH 43205, United States

(DaJusta, Alpert, Ching, McLeod, Jayanthi) Division of Pediatric Urology, Nationwide Children's Hospital, Columbus, OH 43205, United States

(DaJusta, Alpert, Ching, McLeod, Jayanthi) Department of Urology, The Ohio State University College of Medicine, Columbus, OH 43212, United States

(Wiemann) The Ohio State University College of Medicine, Columbus, OH 43210, United States

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500.

Foreskin reconstruction vs circumcision in distal hypospadias.

Rampersad R., Nyo Y.L., Hutson J., O'Brien M., Heloury Y.

Embase

Pediatric Surgery International. 33 (10) (pp 1131-1137), 2017. Date of Publication: 01 Oct 2017.

[Article]

AN: 618098276

Purpose: The purpose of the study was to determine if there were differences in the complication rates between foreskin reconstruction (FR) and circumcision (CIRC) in distal hypospadias repairs. The primary outcomes were urethrocutaneous fistula (UF) and dehiscence.

Method(s): The data of distal hypospadias operated between 2005 and 2013 were retrospectively reviewed. The inclusion criteria were any distal hypospadias repair that required an urethroplasty. The exclusion criteria were follow-up <1 year, redo procedures, chordee greater than 20degree, and incomplete data. Univariate and multivariate analysis was performed on the results.

Result(s): 213 patients were included (95 FR and 118 CIRC). The 2 groups were comparable for age at surgery 19.32 months in FR and 14.25 months in CIRC. Mathieu repair was more common in FR (47/95-49.47%) than in CIRC (45/118-38.14%). The total subsequent procedures required were 23 in FR and 57 in CIRC. The incidence of UF was 6.3% (6/95) in FR and 27.1% (32/118) in CIRC ($p < 0.001$, OR 5.52, 95% CI 2.2-13.9). Complete dehiscence rates were 3.16% (3/95) FR vs 11.02% (13/118) for CIRC ($p = 0.037$, OR 3.8, 95% CI 1.05-13.74). The incidence of patients requiring reoperation was 18.9% (18/95) in FR versus 45.8% (54/118) in CIRC ($p < 0.001$, OR 3.61, 95% CI 1.93-6.76).

Conclusion(s): Foreskin Reconstruction conferred a significantly lower rate of complications, particularly the UF rate, dehiscence rate, and number of patients that required reoperation. Our rate of complications in the CIRC group is much higher than other published data.

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Author NameID

Rampersad, Rajay; ORCID: <http://orcid.org/0000-0003-1758-3487>

Institution

(Rampersad, Nyo, Hutson, O'Brien, Heloury) Department of Paediatric Urology, The Royal Children's Hospital Melbourne, 50 Flemington Road, Parkville, VIC 3052, Australia (Hutson)

Department of Paediatrics, University of Melbourne, Melbourne, VIC, Australia

(Heloury) Murdoch Children's Research Institute, Parkville, VIC, Australia

Publisher

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Year of Publication

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501.

Exstrophy Bladder - Reconstruction or Diversion for the Underprivileged.

Sarin Y.K., Sekhon V.

Embase

Indian Journal of Pediatrics. 84 (9) (pp 715-720), 2017. Date of Publication: 01 Sep 2017.

[Review]

AN: 617411815

The surgical techniques for management of bladder exstrophy epispadias complex have evolved from staged reconstruction, complete primary repair to radical mobilization. Post-operative complications add to the multiplicity of surgical procedures at each step. The end results are variable with many achieving continence rates of 85-89% only after bladder augmentation and clean intermittent catheterization. The situation is further complicated in resource-poor settings, where illiteracy and poverty are the driving factors for choosing a single operative procedure for creation of low pressure reservoir aiming at upper tract preservation and good primary continence. Thus, primary urinary diversion should be offered as a surgical option to patients with limited access to health care facilities. Yogesh's cystorectostomy is a modification of Heitz-Boyer-Hovelacque procedure, wherein the bladder plate is directly anastomosed to the recto-sigmoid pouch, without mobilizing the ureters from their original location. The short-term follow-ups are encouraging with all achieving total urinary continence over the ensuing months. The upper tract functions are well preserved, along with huge parental and patient satisfaction and overall improvement in the quality of life.

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Status

Embase

Institution

(Sarin) Department of Pediatric Surgery, Maulana Azad Medical College, New Delhi 110002, India (Sekhon) Division of Urology, Renal Transplant and Robotics, Medanta - The Medicity, Gurgaon, Haryana, India

Publisher

Springer

Year of Publication

2017

502.

Early childhood development of boys with genital anomalies.

Schneuer F.J., Bentley J.P., Holland A.J.A., Lain S.J., Jamieson S.E., Badawi N., Nassar N.

Embase

Birth Defects Research. 109 (8) (pp 535-542), 2017. Date of Publication: 2017.

[Article]

AN: 617403395

Background: Male genital anomalies often require surgery in early life to address functional and cosmetic consequences. However, there has been little assessment of developmental outcomes of affected boys.

Method(s): We conducted a population-based cohort study of all boys born in New South Wales, Australia, and undergoing school-entry developmental assessment in 2009 or 2012. Health and developmental information was obtained by means of record-linkage of birth, hospital and Australian Early Development Census data. Boys with hypospadias or undescended testis (UDT) were compared with those without. Developmental outcomes were assessed in five domains (physical health, emotional maturity, communication, cognitive skills, and social competence), and boys were categorized as vulnerable (<10th centile of national scores), developmentally high risk (DHR; vulnerable in 21 domains), and special needs.

Result(s): We included 420 boys with hypospadias, 873 with UDT, and 77,176 unaffected boys. There was no difference in the proportion of boys developmentally vulnerable in any domain or DHR between boys with hypospadias (DHR: n 5 49; 13.1%; p 5 0.9), UDT (n 5 116; 15.2%; p 5 0.06), and unaffected boys (n 5 9278; 12.9%). Compared with unaffected boys (n 5 4826; 6.3%), boys with hypospadias (n 5 43; 10.2%; p < 0.001) or UDT (n 5 105; 12.0%; p < 0.001) were more likely to have special needs. Stratified analyses revealed that only boys with UDT and coexisting anomalies had increased risk of being DHR (odds ratio: 2.65; 95% confidence interval, 1.61-4.36) or special needs (odds ratio: 2.91; 95% confidence interval, 2.00-4.22).

Conclusion(s): We found no increased risk of poor development among boys with hypospadias or UDT. However, boys with UDT and coexisting anomalies were more likely to have poorer development and special needs.

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Status

Embase

Institution

(Schneuer, Bentley, Lain, Nassar) Menzies Centre for Health Policy, School of Public Health, University of Sydney, Sydney, Australia (Holland) Discipline of Paediatrics and Child Health, The Children's Hospital at Westmead, Sydney Medical School, The University of Sydney, NSW, Australia

(Jamieson) Telethon Kids Institute, The University of Western Australia, Crawley, WA, Australia (Badawi) Grace Centre for Newborn Care, The Children's Hospital at Westmead, Sydney, Australia

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503.

Maternal use of mild analgesics during pregnancy associated with reduced anogenital distance in sons: A cohort study of 1027 mother-child pairs.

Lind D.V., Main K.M., Kyhl H.B., Kristensen D.M., Toppari J., Andersen H.R., Andersen M.S., Skakkebaek N.E., Jensen T.K.

Embase

Human Reproduction. 32 (1) (pp 223-231), 2017. Date of Publication: 01 Jan 2017.

[Article]

AN: 616972953

STUDY QUESTION Is maternal use of mild analgesics in pregnancy associated with anogenital distance (AGD) - the distance from the anus to the genitals - in the offspring? **SUMMARY**

ANSWER Maternal use of mild analgesics [especially simultaneous use of paracetamol and nonsteroidal anti-inflammatory drugs (NSAIDs)] during pregnancy was associated with a shorter AGD in boys whereas no effect was found in girls. **WHAT IS KNOWN ALREADY** Mild analgesics including paracetamol (acetaminophen) and NSAIDs (e.g. ibuprofen and acetyl salicylic acid) have endocrine disrupting properties and in utero exposure reduces AGD in male rats. In humans, maternal exposure has been associated with cryptorchidism and hypospadias in male offspring but no studies have examined AGD. **STUDY DESIGN, SIZE, DURATION** A prospective birth cohort study. Between 2010 and 2012, 2500 pregnant women were recruited from the Odense Child Cohort. Children were examined 3 months after the expected date of birth.

PARTICIPANTS/MATERIALS, SETTING, METHODS Pregnant women were asked about use of medication including mild analgesics (paracetamol and NSAID) during pregnancy at recruitment (gestational age (GA) week 10-27) and at GA week 28. AGD and penile width were measured 3 months after expected date of birth by trained personnel. A total of 1027 women answered both questionnaires and their children were examined. Associations between prenatal exposure to mild analgesics and AGD and penile width were estimated using multivariable linear regression adjusting for age and weight-for-age SD score. **MAIN RESULTS AND THE ROLE OF CHANCE** A total of 40% of the women reported use of paracetamol and/or NSAIDs (4.4%) during the first 28 weeks of pregnancy. Exposure to analgesics during pregnancy was associated with a reduced AGD in boys, although statistically significant only for NSAIDs. The association was significant among 20 boys exposed to both paracetamol and NSAIDs (AGD -4.1 mm; CI 95%: -6.4; -1.7). Maternal intake of analgesics did not show any clear association with AGD in female offspring. No effect on penile width was found. **LIMITATIONS REASONS FOR CAUTION** Only 27 boys and 18 girls were exposed to NSAIDs and most of them were also exposed to paracetamol. This makes it impossible to distinguish between exposures to NSAIDs alone and a potential mixture effect. Moreover, use of mild analgesics was self-reported up to 2 months after intake, which could have caused misclassification of exposure but is probably not associated with AGD as this was unknown to the women at time of reply to the questionnaire thereby underestimating the association. Confounding by indication may also explain our findings, as the condition for which the analgesic was taken may be associated with a reduction in AGD, rather than the use of the

analgesic medication. This is the first study to report such an association in humans and further studies are needed to confirm our findings. WIDER IMPLICATIONS OF THE FINDINGS A negative association was observed between exposure to analgesics during pregnancy and AGD in boys, suggesting disruption of androgen action. The health implications of a shorter AGD are still uncertain, but in cross-sectional studies among adult men a shorter AGD is associated with poorer semen quality and lower testosterone. As 41% of the women used these painkillers the finding are of public health importance and pregnant women should be advised about the potentially harmful effects of painkiller use.

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Status

Embase

Institution

(Lind, Andersen, Jensen) Department of Environmental Medicine, Institute of Public Health, University of Southern Denmark, Odense, Denmark (Main, Skakkebaek) Department of Growth and Reproduction, Rigshospitalet, University of Copenhagen, Blegdamsvej 9, Copenhagen 2100, Denmark

(Kyhl, Jensen) Odense University Hospital, Hans Christian Andersen Children's Hospital, Sdr. Boulevard 29, Odense C 5000, Denmark

(Kyhl) Odense Patient data Exploratory Network (OPEN), Sdr. Boulevard 29, Odense C 5000, Denmark

(Kristensen) Laboratory of Genomic and Molecular Biomedicine, Department of Biology, University of Copenhagen, Universitetsparken 13, Copenhagen DK-2100, Denmark

(Toppari) Department of Physiology, Institute of Biomedicine, University of Turku, Kiinamylynkatu 10, Turku FI-20520, Finland

(Toppari) Department of Pediatrics, Turku University Hospital, Kiinamylynkatu 10, Turku FI-20520, Finland

(Andersen) Odense University Hospital, Endocrinological Department, Odense C 5000, Denmark

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Oxford University Press

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504.

The Fisher Technique for Correction of Penile Torsion in Children: Who Are the Candidates?.

Marret J.-B., Ravasse P., Raffoul L., Rod J.

Embase

Urology. 104 (pp 179-182), 2017. Date of Publication: June 2017.

[Article]

AN: 616484071

Objective To report our experience in the treatment of penile torsion with a special reference to the Fisher technique. **Patients and Methods** We reviewed 30 cases of congenital penile torsion treated from 2009 to 2015 in a single center. Sixteen patients presented a moderate rotation of 45degree-90degree and 14 had a severe rotation with an angle greater than 90degree. Chordee was present in 17 cases and distal glanular hypospadias in 15 cases. Median age at surgery was 10 months. In 14 cases, degloving along with skin realignment, alone, allowed correction of the torsion. In 16 cases, the Dartos flap (Fisher technique) was used. **Results** Out of the 30 patients, 29 had a good result, with a complete correction of the torsion. Only 1 patient had a residual torsion of 30degree. Complications were minor and consisted of skin excess in 11 patients; 1 single case needed to be reoperated for that skin complication. The need for the Dartos flap procedure was significantly correlated to the degree of torsion ($P = .001$): it was used in 4 out of 16 patients (25%) with a torsion of less than 90degree, and in 12 out of 14 patients (86%) with a torsion of 90degree or more. **Conclusion** Most of penile rotations less than 90degree can be corrected by skin degloving and realignment. The Fisher technique is a simple and safe procedure for correction of persistent rotation after skin degloving; such cases have usually a penile rotation greater than 90degree.

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Status

Embase

Institution

(Marret, Ravasse, Rod) Department of Pediatric Surgery, University of Caen Hospital, France

(Ravasse, Rod) Caen University Hospital, Avenue de la Cote de Nacre, Caen, France

(Raffoul) Hotel-Dieu de France Hospital, Beirut, Lebanon

Publisher

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Year of Publication
2017

505.

Prophylactic antibiotic use in pediatric patients undergoing urinary tract catheterization: A survey of members of the Society for Pediatric Urology.

Glaser A.P., Rosoklija I., Johnson E.K., Yerkes E.B.

Embase

BMC Urology. 17 (1) (no pagination), 2017. Article Number: 76. Date of Publication: 06 Sep 2017.

[Review]

AN: 618128012

Background: Current organizational guidelines regarding use of antibiotics during urinary tract catheterization are based on limited evidence and are not directly applicable to the pediatric urology population. We seek to improve understanding of this population by first evaluating current practices. This study aims to investigate practice patterns and attitudes of pediatric urologists regarding the use of antibiotics in the setting of urinary tract catheterization.

Method(s): An online survey was sent to members of the Society for Pediatric Urology.

Questionnaire sections included demographics, general questions about antibiotic use with catheterization, and specific clinical scenarios. Descriptive statistics were used, and chi-square analysis was performed to examine associations between demographics and specific responses.

Result(s): Of 448 pediatric urologists surveyed, 154 (34%) responded to the survey. A majority of surveyed urologists (78%) prescribe daily prophylactic antibiotics with a hypospadias stent in place, but extensive variation in use of antibiotics was reported with other catheters and tubes. Extensive variation in practice patterns was also reported for three case scenarios regarding antibiotic prophylaxis with catheterization. Urologists > 50 years of age and fellowship-trained urologists were more likely to prescribe antibiotics for hypospadias stents ($p = 0.02$, $p = 0.03$), but no other significant associations between demographic characteristics and antibiotic use were found.

Conclusion(s): There is substantial variation in practice patterns among surveyed pediatric urologists regarding prophylactic antibiotic use with urinary catheterization. This variation, combined with a lack of objective data and increasing pressure to decrease infectious

complications and combat antibiotic resistance, highlights the need for development of management guidelines for this unique population.

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Embase

Institution

(Glaser, Rosoklija, Johnson, Yerkes) Department of Surgery, Division of Urology, Ann and Robert H. Lurie Children's Hospital of Chicago, 225 E. Chicago Ave, Chicago, IL 60611, United States

Publisher

BioMed Central Ltd. (E-mail: info@biomedcentral.com)

Year of Publication

2017

506.

Maternal endometriosis and genital malformations in boys: a Danish register-based study.

Arendt L.H., Lindhard M.S., Henriksen T.B., Forman A., Olsen J., Ramlau-Hansen C.H.

Embase

Fertility and Sterility. 108 (4) (pp 687-693), 2017. Date of Publication: October 2017.

[Article]

AN: 618057208

Objective To investigate the association between maternal endometriosis and occurrence of the genital anomalies cryptorchidism and hypospadias in sons. Design Population-based cohort study. Setting Not applicable. Patient(s) All live-born singleton boys born from 1978 to 2012. Intervention(s) None. Main Outcome Measure(s) Cryptorchidism and hypospadias in boys based on information from the Danish National Patient Register. Result(s) The study included 1,073,026 live-born singleton boys. A total of 6,443 boys were sons of women diagnosed with endometriosis before pregnancy. Altogether, 27,342 boys were diagnosed with cryptorchidism, of whom 16,446 had corrective surgery. Hypospadias was diagnosed in 4,853 boys. As compared with unexposed boys, a tendency towards a slightly higher occurrence of cryptorchidism was observed among boys of women with endometriosis (adjusted hazard ratio [aHR] 1.18; 95% confidence interval

[CI], 0.97, 1.44). When stratified by medically assisted reproduction (MAR) technologies, the association was slightly stronger among boys born to women with endometriosis who had conceived via MAR, yet it remained moderate and statistically insignificant (aHR 1.27; 95% CI, 0.97, 1.70). When women who conceived with MAR were excluded, the association between endometriosis and cryptorchidism disappeared. For hypospadias, we observed no association, either in the main analysis or the stratified analysis. Conclusion(s) The findings from this register-based study do not provide strong evidence for a higher occurrence of the studied genital anomalies among boys of women with endometriosis.

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Author NameID

Arendt, Linn Hakonsen; ORCID: <http://orcid.org/0000-0001-9692-3501> Olsen, Jorn; ORCID: <http://orcid.org/0000-0001-7462-2740>

Institution

(Arendt, Ramlau-Hansen) Department of Public Health, Section for Epidemiology, Aarhus University, Aarhus, Denmark (Arendt, Lindhard, Henriksen) Perinatal Epidemiology Research Unit, Department of Child and Adolescent Health, Aarhus University Hospital, Aarhus, Denmark (Forman) Department of Gynaecology and Obstetrics, Aarhus University Hospital, Aarhus, Denmark

(Olsen) Department of Clinical Epidemiology, Aarhus University Hospital, Aarhus, Denmark

Publisher

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Year of Publication

2017

507.

Genotype-phenotype correlation and identification of two novel SRD5A2 mutations in 33 Chinese patients with hypospadias.

Yuan S., Meng L., Zhang Y., Tu C., Du J., Li W., Liang P., Lu G., Tan Y.-Q.

Embase

Steroids. 125 (pp 61-66), 2017. Date of Publication: 2017.

[Article]

AN: 617351997

Hypospadias, a common congenital malformation of male external genitalia, is characterized mainly by an aberrant opening of the urethra on the ventral side of the penis. Depending on the severity of the disease, it can be classified into three types: anterior, middle, and posterior. In our study, 33 patients with hypospadias were recruited, including eight with anterior hypospadias and 25 with posterior hypospadias. We performed mutation analysis of the SRD5A2, AR and HSD17B3 genes in these patients. Eight different SRD5A2 mutations were detected in 15 patients with posterior hypospadias (60%, 15/25), including six previously described mutations (p.Q6X, Q71X, p.L20P, p.G203S, p.R227Q, and p.R246Q) and two novel mutations (p.G196R and p.L73Pfs*17). One AR gene mutation (p.A597T) was found in a patient and no HSD17B3 mutations were detected. Additionally, we carried out routine semen analyses in all adult patients. Combining mutation analysis with semen examination results, showed that whole five adult patients who carried SRD5A2 mutations had abnormal semen quality. In summary, all the detected mutations were responsible for the clinical features observed in these 16 patients. Our data suggest that mutations of the SRD5A2 gene are the main causes of posterior hypospadias and seem to affect the semen quality of adult patients, whereas mutations in AR and HSD17B3 gene were rare in these hypospadias patients. Our study expanded the SRD5A2 mutation spectrum in the Han Chinese population and provided useful information for genetic and reproductive counselling for patients with hypospadias.

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Institution

(Yuan, Meng, Tu, Du, Li, Lu, Tan) Institute of Reproduction and Stem Cell Engineering, Central South University, Changsha, Hunan 410078, China (Yuan, Meng, Du, Li, Lu, Tan) Reproductive and Genetic Hospital of Citic-Xiangya, Changsha, Hunan 410078, China

(Zhang) Maternal and Child Health Hospital of Hunan Province, Changsha, Hunan 410078, China

(Liang) Brock University Faculty of Mathematics and Science, St. Catharines, Ontario L2S 3A1, Canada

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Year of Publication

508.

Maternal Overweight and Obesity and Genital Anomalies in Male Offspring: A Population-Based Swedish Cohort Study.

Arendt L.H., Ramlau-Hansen C.H., Lindhard M.S., Henriksen T.B., Olsen J., Yu Y., Cnattingius S.
Embase

Paediatric and Perinatal Epidemiology. 31 (4) (pp 317-327), 2017. Date of Publication: July 2017.
[Article]

AN: 617012803

Background: Overweight and obese pregnant women face higher risk of several critical birth outcomes, including an overall increased risk of congenital abnormalities. Only few studies have focused on associations between maternal overweight and the genital anomalies in boys, cryptorchidism and hypospadias, and results are inconclusive.

Method(s): We performed a population-based cohort study and assessed the associations between maternal body mass index (BMI) in early pregnancy and occurrence of cryptorchidism and hypospadias. All live-born singleton boys born in Sweden from 1992 to 2012 were included. From the Swedish Patient Register, information on cryptorchidism and hypospadias was available. Data were analysed using Cox proportional hazards regression adjusted for potential confounders. Mediation analyses were performed to estimate how much of the association between BMI and genital anomalies were mediated through obesity-related diseases.

Result(s): Of the 1 055 705 live-born singleton boys born from 1992 to 2012, 6807 (6.4 per 1000) were diagnosed with hypospadias and 16 469 (15.6 per 1000) were diagnosed with cryptorchidism, of which 9768 (9.3 per 1000) underwent corrective surgery for cryptorchidism. We observed dose-response associations between maternal BMI and hypospadias and cryptorchidism. Boys of mothers with BMI ≥ 40.0 kg/m² had the highest adjusted hazard ratios for hypospadias (HR 1.35, 95% confidence interval [CI] 1.04, 1.76) and cryptorchidism (HR 1.25, 95% CI 1.00, 1.58). A substantial proportion of the associations between BMI and the genital anomalies were mediated through preeclampsia.

Conclusion(s): This large register-based study adds to the current literature and indicates that the occurrence of hypospadias and cryptorchidism increase with maternal overweight and obesity severity.

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Status

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Author NameID

Arendt, Linn Hakonsen; ORCID: <http://orcid.org/0000-0001-9692-3501> Cnattingius, Sven;

ORCID: <http://orcid.org/0000-0002-0805-8093>

Institution

(Arendt, Ramlau-Hansen, Olsen) Department of Public Health, Section for Epidemiology, Aarhus University, Aarhus, Denmark (Arendt, Lindhard, Henriksen) Perinatal Epidemiology Research Unit, Department of Child and Adolescent Health, Aarhus University Hospital, Aarhus, Denmark (Olsen, Yu) Department of Clinical Epidemiology, Aarhus University Hospital, Aarhus, Denmark (Cnattingius) Clinical Epidemiology Unit, Department of Medicine, Karolinska University Hospital, Stockholm, Sweden

Publisher

Blackwell Publishing Ltd

Year of Publication

2017

509.

Short-term outcomes of the multi-institutional bladder exstrophy consortium: Successes and complications in the first two years of collaboration.

Borer J.G., Vasquez E., Canning D.A., Kryger J.V., Bellows A., Weiss D., Groth T., Shukla A., Kurtz M.P., Mitchell M.E.

Embase

Journal of Pediatric Urology. 13 (3) (pp 275.e1-275.e6), 2017. Date of Publication: June 2017. [Article]

AN: 616489961

Introduction/background Bladder exstrophy is a rare diagnosis that presents major reconstructive challenges. To increase experience and proficiency in the care of bladder exstrophy (BE), the Multi-Institutional BE Consortium (MIBEC) was formed, with a focus on refining technical aspects

of complete primary repair of bladder exstrophy (CPRE) and subsequent care. Objective Outcome measures included successful CPRE (absence of dehiscence), complications, and integrated points of technique and care over the short-term. Study design Boston Children's Hospital, Children's Hospital of Philadelphia and Children's Hospital of Wisconsin alternately served as the host, with observation, commentary and critique by visiting collaborating surgeons. CPRE with bilateral iliac osteotomy was performed at 1-3 months of age. High-definition video capture of the surgery allowed local and distant broadcast to facilitate real-time observation and teaching, and recording of all procedures. Results From February 2013 to February 2015, MIBEC participating surgeons performed CPRE on 27 consecutive patients (22 classic BE, five epispadias). There were no dehiscences in 27 patients (0%, 95% CI 0-12.5%). Thirteen girls and 14 boys underwent CPRE at a median age of 2.3 months (range 0.1-51.6). One boy had a hypospadiac urethral meatus at CPRE completion. Hydronephrosis of mild or moderate grade was present postoperatively in eight girls and two boys. Additional results, per gender, are presented in the Summary table below. Discussion Absence of dehiscence in this cohort was comparable or compared favorably with the literature. However, several girls had significant obstructive complications following CPRE. The rate of bladder outlet obstruction (BOO) in girls was increased compared with published reports. A low complication rate was noted in the boys following CPRE, which was comparable to reports in the literature, and early signs of continence and spontaneous voiding were noted in some boys and girls. Limitations included variation in patient age at presentation, thereby introducing a wide age range at CPRE. Outcome data were limited by short follow-up regarding voiding with continence. Conclusion This collaborative effort proved beneficial regarding significantly increased surgeon exposure to CPRE, refinement of CPRE technique, surgeon learning and expertise. Technical refinement of CPRE is ongoing.

[Table presented]

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Institution

(Borer, Vasquez, Bellows, Kurtz) Department of Urology, Boston Children's Hospital, Boston, MA, United States (Canning, Weiss, Shukla) Department of Urology, Children's Hospital of Philadelphia, Philadelphia, PA, United States

(Kryger, Groth, Mitchell) Department of Urology, Children's Hospital of Wisconsin, Milwaukee, WI, United States

Publisher

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2017

510.

Association of reproductive disorders and male congenital anomalies with environmental exposure to endocrine active pesticides.

Gonzalez J.G., Miranda M.I.V., Mullor M.R., Jerez A.F.H., Carreno T.P., Rodriguez R.A.

Embase

Reproductive Toxicology. 71 (pp 95-100), 2017. Date of Publication: 01 Aug 2017.

[Article]

AN: 616107670

There is growing evidence that environmental exposure to pesticides may increase the risk of developing reproductive and developmental disorders. This study determined the prevalence and risk of developing gestational disorders and male congenital genitourinary malformations in areas with distinct exposure to pesticides, many of them with potential endocrine disrupting properties. A population-based case-control study was carried out on pregnant women and male children living in ten health districts of Andalusia classified as areas of high and low environmental exposure to pesticides according to agronomic criteria. The study population included 45,050 cases and 950,620 controls matched for age and health district. Data were collected from computerized hospital records between 1998 and 2005. Prevalence rates and risk of miscarriage, low birth weight, hypospadias, cryptorchidism and micropenis were significantly greater in areas with higher use of pesticides in relation to those with lower use, thus supporting and extending previous information.

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Status

Embase

Institution

(Gonzalez) Rafael Mendez Hospital Lorca, Murcia (Spain) and University of Lorca, Spain

(Miranda, Mullor, Carreno, Rodriguez) University of Almeria, Department of Neurosciences and Health Sciences, Almeria, Spain

(Jerez) University of Granada School of Medicine, Granada, Spain
(Carreno) Andalusian Council of Health at Almeria Province, Almeria, Spain
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Year of Publication
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511.

Better Defining the Spectrum of Adult Hypospadias: Examining the Effect of Childhood Surgery on Adult Presentation.

Hoy N.Y., Rourke K.F.

Embase

Urology. 99 (pp 281-286), 2017. Date of Publication: 01 Jan 2017.

[Article]

AN: 613841634

Objective To describe the spectrum of adult presentations with hypospadias-related complications and examine the effect of childhood surgical repair on these adult presentations. **Methods** A retrospective chart review over a 10-year period, from August 2004 to December 2014, demonstrated 93 adult patients who presented to a reconstructive urologist with complications related to hypospadias. Patients were divided into 2 groups: those with no prior hypospadias surgery (Group 1, N=19) and those who underwent surgical correction as a child (Group 2, N=74). Charts were reviewed for age at presentation, initial complaints, history of repair, and surgical intervention required. **Results** The mean age at presentation was 34.6+/-0.6 years. Overall, lower urinary tract symptoms (LUTS) (49%) was the most common presenting complaint, followed by spraying (24%), urethrocutaneous fistula (18%), recurrent urinary tract infections (UTIs) (15%), and chordee (14%). Comparison demonstrated that Group 2 patients were more likely to present with LUTS (55% vs 26%; P=.038) and recurrent UTIs (19% vs 0%; P=.050). There was a trend toward Group 1 patients presenting more commonly with cosmetic dissatisfaction (16% vs 4%; P=.06). Urethral stricture was demonstrated more commonly in Group 2 (47% vs 11%; P=.0043). Of these, strictures were significantly longer in the previous surgery group (5.5+/-0.6cm vs 3.0+/-0.6cm, P=.019). **Conclusion** Correction of hypospadias as a child likely increases the future risk of urethral stricture, recurrent UTIs, and subsequent LUTS,

with a trend toward improving patient satisfaction with cosmesis compared to nonsurgical management. Follow-up of hypospadias repair patients should extend into adulthood, as a significant portion of adult presentations ultimately require surgical intervention.

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Status

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Institution

(Hoy, Rourke) Division of Urology, University of Alberta, Edmonton, AB, Canada

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Year of Publication

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512.

Results of Epispadias Repair Using the Modified Cantwell-Ransley Technique.

Bar-Yosef Y., Sofer M., Ekstein M.P., Binyamini Y., Ben-Chaim J.

Embase

Urology. 99 (pp 221-224), 2017. Date of Publication: 01 Jan 2017.

[Article]

AN: 613254907

Objective To evaluate our results of epispadias repair with a modified Cantwell-Ransley (MCR) technique. Methods A retrospective Institutional Review Board-approved chart review of all patients who underwent an MCR epispadias repair was conducted between 1998 and 2015. Procedures were performed at birth or after the age of 8 months as part of the modern staged repair of exstrophy-epispadias complex (EEC) in patients with bladder exstrophy treated since birth, at presentation for older patients, and after the age of 6 months in isolated epispadias patients. Twenty-two children underwent MCR epispadias repair in our institute during the study period. Sixteen of them had EEC and 6 had isolated epispadias. Four patients underwent exstrophy and epispadias repair at the same session. Twelve children underwent epispadias repair as a second stage of modern staged repair at a mean age of 21 months (range 8-60). The

procedures involved dissection of the corporeal bodies and urethral plate from the penile base to the tip of the penile glans. Results After a mean follow-up of 6.9 years (range 0.5-18), there were no complications in the isolated epispadias group and 4 complications in the bladder exstrophy group: urethrocutaneous fistula (n=1), residual dorsal curvature (n=1), and excess of penile skin (n=2). The meatal location was orthotopic in all cases. All of the complications were successfully addressed in a single subsequent surgical session. Conclusion MCR technique continues to be a reliable, reproducible option for epispadias repair in EEC patients and in cases of isolated epispadias.

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Status

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Author NameID

Bar-Yosef, Yuval; ORCID: <http://orcid.org/0000-0003-3769-6606>

Institution

(Bar-Yosef, Sofer, Binyamini, Ben-Chaim) Pediatric Urology, Dana-Dwek Children's Hospital, Tel-Aviv Medical Center, Sackler Faculty of Medicine, Tel-Aviv University, Tel-Aviv, Israel (Ekstein) Pediatric Anesthesia, Dana-Dwek Children's Hospital, Tel-Aviv Medical Center, Sackler Faculty of Medicine, Tel-Aviv University, Tel-Aviv, Israel

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Year of Publication

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513.

Early vs. late-presenting urethroplasty complications after hypospadias repair: A retrospective analysis of patient follow-up.

Faasse M.A., Liu D.B.

Embase

Journal of Pediatric Urology. 13 (4) (pp 354.e1-354.e5), 2017. Date of Publication: August 2017.

[Article]

AN: 617037003

Introduction Complications after primary hypospadias repair often present late, that is, more than 1 year postoperatively. This has important implications for clinical practice and prospective research protocols. Follow-up regimen-or lack thereof-may contribute to delayed diagnosis of complications. **Objective** To characterize and compare the follow-up regimens of patients with early and late-presenting urethroplasty complications after primary hypospadias repair, specifically noting the length of time between encounters during which complications emerged. **Patients and methods** Boys who underwent repair of urethroplasty complications after hypospadias surgery were identified, and retrospective chart reviews were performed. Late complications were defined by presentation more than 1 year after primary repair; all others were designated as early. We recorded the encounter at which each patient was first noted to have a complication, as well as the last encounter prior to this. Follow-up intervals during which complications emerged were determined. Comparisons of the type of primary repairs, complications, and follow-up regimens (prescribed as well as actual) were performed between patients with early and late complications. **Results** A total of 51 patients underwent repair of 57 hypospadias complications. Eighteen patients (35%) had complications that presented late. Complications after a midshaft/distal hypospadias repair were more likely to present late than complications following a one- or two-stage proximal repair (59% vs. 31% vs. 6%, respectively; $p = 0.003$). The median interval between encounters during which late complications emerged was 24 months (IQR 16-43), compared with 1.2 (0.7-2.2) months for early-presenting complications ($p < 0.001$). Eleven of the 18 patients with late complications (61%) had not had an encounter beyond 3 months postoperatively prior to presentation of their complications; only four patients (8%) had a late complication that was unrecognized at a follow-up visit more than 6 months postoperatively (Figure). Patients with late complications had a greater discrepancy between prescribed and actual follow-up intervals, averaging 11 months ($p = 0.001$). **Conclusions** Late presentation of urethroplasty complications after hypospadias repair is relatively common. There is typically a lengthy follow-up interval during which late complications emerge. Many patients who presented with late complications had not previously been examined beyond the early postoperative period. Adherence to a more structured follow-up regimen that includes a visit outside of the early postoperative period (e.g. routine encounters at 6 months postoperatively) may facilitate earlier detection and reduce late presentation of complications, especially among patients with midshaft or distal primary repairs. Earlier diagnosis would allow secondary procedures to be completed sooner.[Figure presented]

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Status

Embase

Institution

(Faasse, Liu) Division of Urology, Ann & Robert H. Lurie Children's Hospital of Chicago,
Northwestern University Feinberg School of Medicine, Chicago, IL, United States (Faasse)
Advocate Children's Hospital, Chicago, IL, United States

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514.

The modified Ulaanbaatar procedure: Reduced complications and enhanced cosmetic outcome for the most severe cases of hypospadias.

Jayanthi V.R., Ching C.B., DaJusta D.G., McLeod D.J., Alpert S.A.

Embase

Journal of Pediatric Urology. 13 (4) (pp 353.e1-353.e7), 2017. Date of Publication: August 2017.

[Article]

AN: 616844342

Introduction/objective Proximal hypospadias is one of the most challenging conditions that pediatric urologists have to deal with. Many procedures have been devised over the years, but nothing has been proven to be the best option. Although there have been some attempts at correcting severe hypospadias in one procedure, most have advocated a staged approach. The classic approach - laying penile skin or a graft within a split glans followed by glanuloplasty at the second stage - by definition requires two operations on the glans. In the Ulaanbaatar procedure the distal glanular urethra is constructed at the first stage, allowing for a single glans procedure and thus potentially better cosmetic outcomes. The present study discusses experience with the Ulaanbaatar procedure for severe hypospadias. Study design The study retrospectively reviewed every child who underwent both stages of this procedure at the present institution. It reviewed age, associated diagnoses, surgical technique and outcomes. Surgical technique The first stage was analogous to a classic first-stage procedure with regard to division of the urethral plate and correction of penile curvature. However, an island flap of preputial skin was mobilized and tubularized to create the glanular urethra. No attempt was made to bridge the native meatus and

this reconstructed urethra, and the remaining penile skin was placed between the two. The second stage was performed 6 months later by tubularizing the penile skin between the two meatuses. Results The series consisted of 34 boys. Mean age at surgery was 18.3 months (range 6-118). Nineteen underwent evaluation for genital ambiguity at birth (56%). Thirty (88%) received pre-operative testosterone or human chorionic gonadotropin (HCG). After urethral plate transection, persistent curvature was addressed during the first stage, with dorsal plication in 12 (35%), urethral plate transection alone in six (18%) or ventral grafting with small intestinal submucosa in 16 (47%). Twenty-three boys (67%) had the neourethra tunneled through the glans, and 11 (33%) had the glans split followed by glanuloplasty. Average time between the two stages was 7 months (range 4.0-13.9). Four patients (12%) developed urethral diverticula that required repair. One developed recurrent epididymitis related to an abnormal ejaculatory duct (no stricture) and underwent vasectomy. No patient developed a fistula. Mean length of follow-up was 15.2 months (range 0.3-55.5). Discussion This modification of the classic staged hypospadias repair may allow for better cosmetic outcome, since the majority of boys required no formal glanuloplasty. There were reduced complications, perhaps because the urethral defect acted like a controlled fistula, allowing for better tissue healing prior to final urethral reconstruction.[Figure presented]

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Author NameID

Jayanthi V.R.; ORCID: <http://orcid.org/0000-0002-6865-9026> Alpert S.A.; ORCID:
<http://orcid.org/0000-0001-7034-603X>

Institution

(Jayanthi, Ching, DaJusta, McLeod, Alpert) Division of Urology, Nationwide Children's Hospital,
The Ohio State University, Columbus, OH, United States

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Echocardiographic evaluation of left ventricular mass index in children with hypospadias after hormonal stimulation with topical testosterone: A randomized controlled trial.

de Andrade E.C., de Castro Paiva K.C., da Silva Guedes S., Souza M.L.C., Pereira M.N., Miana L.P., de Figueiredo A.A., de Bessa J., Netto J.M.B.

Embase

Journal of Pediatric Urology. 13 (4) (pp 352.e1-352.e7), 2017. Date of Publication: August 2017.

[Article]

AN: 615611157

Introduction Testosterone is often used in the preoperative period of hypospadias surgery. Previous studies have demonstrated the presence of androgen receptors in cardiac myocytes that can modulate the phenotype. The use of supraphysiological doses of androgens can lead to toxicity on the heart muscle and, in some cases, to left ventricular hypertrophy. This randomized double blind controlled clinical trial aims to evaluate the effect of topical testosterone on left ventricular mass index in boys with hypospadias. Materials and methods Boys with hypospadias aged 6 months to 9 years were included. Children were divided into two groups: G1 - boys who received testosterone propionate 1% ointment twice a day for 30 days, and G2 - boys receiving placebo ointment in the same regimen. All children were submitted to bi-dimensional echocardiographic evaluation to compare the left ventricular mass index, blood pressure, and body mass index before and after treatment (30 and 90 days). Levels of serum testosterone, LH, and FSH were measured. Results Thirty-five children were analyzed: 17 in G1 and 18 in G2. No differences were found in left ventricular mass index (left ventricular mass indexed by body surface area) prior to treatment. Left ventricular mass index was 59.21 ± 11.91 g/m² in G1 and 55.12 ± 8.29 g/m² in G2 ($p = 0.244$) after 30 days of treatment, and 61.13 ± 11.69 g/m² in G1 and 62.84 ± 35.99 g/m² in G2 ($p = 0.852$) after 90 days. Serum testosterone levels were 12 (7-80) ng/dL in G1 and 5 (5-7) ng/dL in G2 ($p = 0.018$) after 30 days of treatment, and 10 (5-11) ng/dL in G1 and 5 (4-5) ng/dL in G2 ($p = 0.155$), after 90 days (Figure). There was a small increase in systolic blood pressure (SBP) after 30 days (83.82 ± 7.18 mmHg) in the group who receive testosterone (G1) compared with controls (77.5 ± 6.69 mmHg) ($p = 0.010$). After 90 days, SBP levels returned to basal levels in G1 (82.35 ± 5.62 mmHg) and in G2 (81.38 ± 4.79 mmHg) ($p = 0.588$). Conclusion Topical testosterone can be considered safe in the preoperative period of children with hypospadias with no risk of left ventricular hypertrophy. An increase in systolic blood pressure occurs while using testosterone but it is transitory, returning to normal levels after 90 days.[Figure presented]

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Status

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Author NameID

Netto, Jose Murillo B.; ORCID: <http://orcid.org/0000-0002-9959-6160>

Institution

(de Andrade, de Castro Paiva, da Silva Guedes, Souza, Pereira, Miana) Department of Pediatrics, Federal University of Juiz de Fora, UFJF, Brazil (de Figueiredo, Netto) Department of Surgery, Division of Urology, Federal University of Juiz de Fora, UFJF, Brazil

(de Bessa) Department of Surgery, Division of Urology, State University of Feira de Santana, UEFS, Brazil

(Netto) Department of Surgery, Division of Urology, Hospital e Maternidade Therezinha de Jesus da Faculdade de Ciencias Medicas e da Saude de Juiz de Fora, Suprema, Brazil

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516.

Tubularized proximally-incised plate in distal/midshaft hypospadias repair.

Marte A., Pintozzi L.

Embase

Pediatrica Medica e Chirurgica. 39 (2) (pp 39-42), 2017. Article Number: 151. Date of Publication: 2017.

[Article]

AN: 616978509

The aim of this study was to verify the validity, feasibility, and the functional results, by uroflowmetry, of Tubularized proximallyincised plate technique in selected case of distal/midshaft hypospadias. Out of 120 patients scheduled to undergo TIP (or Snodgrass) procedure, 23 were selected between January 2013 and January 2016 (19.1%). This case series comprised 16 patients with distal and 7 with midshaft hypospadias. Mean age at surgery was 2.9 years. The inclusion criteria were a deep and wide glandular groove and a proximal narrow urethral plate.

The procedure was carried out as described by Snodgrass but the incision of the urethral plate, including the mucosal and submucosal tissue, was made only proximally, between the original meatus and the glandular groove in no case extending to the entire length of the plate.

Postoperatively a foley catheter was left in place from 4 to 7 days. Uroflowmetry was performed when the patients age ranged from 2.5 to 5.7 years (mean age 3.11 years and mean follow-up 1.8 years, body surface <1.1 m²). Patients were included if they were old enough to void volitionally and fistula-free. The results of flow pattern were expressed as percentiles and compared with those reported by Toguri. At the time of uroflowmetry their ages ranged from 2.5 to 5.7 years (mean age 3.11 years and mean follow-up 1.8 years, body surface <1.1 m²). No patient presented fistulas nor perioperative complications. At uroflowmetry, eighteen patients presented values above the 25th percentile and 5 showed a borderline flow. All patients in this group remained stable without urinary symptoms. In selected cases, the tubularized proximally-incised plate yields satisfactory cosmetic and functional results for the treatment of midshaft proximal hypospadias. A long-term follow-up study is needed for further evaluation. Patient selection is crucial for the success of this technique.

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Institution

(Marte, Pintozzi) Department of Pediatric Surgery, University of Campania Luigi Vanvitelli, Naples, Italy

Publisher

Page Press Publications (E-mail: info@pagepress.org)

Year of Publication

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517.

The association between caudal anesthesia and increased risk of postoperative surgical complications in boys undergoing hypospadias repair.

Taicher B.M., Routh J.C., Eck J.B., Ross S.S., Wiener J.S., Ross A.K.

Embase

Paediatric Anaesthesia. 27 (7) (pp 688-694), 2017. Date of Publication: July 2017.

[Article]

AN: 615331448

Introduction: Recent reports have suggested that caudal anesthesia may be associated with an increased risk of postoperative surgical complications. We examined our experience with caudal anesthesia in hypospadias repair to evaluate for increased risk of urethrocutaneous fistula or glanular dehiscence.

Method(s): All hypospadias repairs performed by a single surgeon in 2001-2014 were reviewed. Staged or revision surgeries were excluded. Patient age, weight, hypospadias severity, surgery duration, month and year of surgery, caudal anesthesia use, and postoperative complications were recorded. Bivariate and multivariate statistical analyses were performed.

Result(s): We identified 395 single-stage primary hypospadias repairs. Mean age was 15.6 months; 326 patients had distal (83%) and 69 had proximal (17%) hypospadias. Caudal anesthetics were used in 230 (58%) cases; 165 patients (42%) underwent local penile block at the discretion of the surgeon and/or anesthesiologist. Complications of urethrocutaneous fistula or glanular dehiscence occurred in 22 patients (5.6%) and were associated with caudal anesthetic use (OR 16.5, 95% CI 2.2-123.8, $P = 0.007$), proximal hypospadias (OR 8.2, 95% CI 3.3-20.0, $P < 0.001$), increased surgical duration (OR 1.01, 95% CI 1.01-1.02, $P < 0.001$), and earlier year of practice (OR 3.0, 95% CI 1.2-7.9, $P = 0.03$ for trend). After adjusting for confounding variables via multivariable logistic regression, both caudal anesthetic use (OR 13.4, 95% CI 1.8-101.8, $P = 0.01$) and proximal hypospadias (OR 6.8, 95% CI 2.7-16.9, $P < 0.001$) remained highly associated with postoperative complications.

Conclusion(s): In our experience, caudal anesthesia was associated with an over 13-fold increase in the odds of developing postoperative surgical complications in boys undergoing hypospadias repair even after adjusting for urethral meatus location. Until further investigation occurs, clinicians should carefully consider the use of caudal anesthesia for children undergoing hypospadias repair.

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Status

Embase

Author NameID

Taicher, Brad M.; ORCID: <http://orcid.org/0000-0001-9985-8637>

Institution

(Taicher, Eck, Ross) Division of Pediatric Anesthesia, Department of Anesthesiology, Duke University Medical Center, Durham, NC, United States (Routh, Wiener) Division of Urology, Department of Surgery, Duke University Medical Center, Durham, NC, United States (Ross) Department of Urology, University of North Carolina at Chapel Hill, Chapel Hill, NC, United States
Publisher
Blackwell Publishing Ltd (E-mail: customerservices@oxonblackwellpublishing.com)
Year of Publication
2017

518.

Urethral Mobilization as an Alternative Procedure for Distal Hypospadias Repair.

El Darawany H.M., Al Damhogy M.E.

Embase

Urology. 104 (pp 183-186), 2017. Date of Publication: June 2017.

[Article]

AN: 615248094

Objective To evaluate the urethral mobilization procedure as the main technique in management of glanular, coronal, subcoronal, and distal penile hypospadias. **Patients and Methods** Sixty patients with distal hypospadias were included in this prospective study between January 2013 and January 2015. All of them had urethral mobilization. All patients were followed up at 2 weeks, and at 1, 3, 6, and 12 months postoperatively. Duration of the operation, hospital stay, position of meatus and its shape before and after the operation, and the postoperative complications were recorded. The force and direction of the postoperative voiding stream were observed and assessed subjectively. **Results** The mean operative time and hospital stay were 38 minutes and 2.2 days, respectively. Postoperatively, 2 patients developed wound infection, 1 of them developed glanular suture dehiscence managed by simple glanular suturing 3 months later. None of our patients developed urethrocutaneous fistula. Three months postoperatively, 2 patients developed meatal stenosis, 1 of them managed by meatal dilation whereas the other patient required ventral meatotomy. **Conclusion** Hypospadias surgery aim to construct a functionally and cosmetically normal penis with minimal complications. Recently, urethral mobilization started to regain its position in distal hypospadias repair. Its results are comparable with other popular

procedures or even better. Urethral mobilization is an easy and simple procedure and it has an excellent cosmetic and functional outcome with minimal complications.

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Status

Embase

Institution

(El Darawany) Department of Urology, King Fahd Hospital of the University, Imam Abdulrahman Bin Faisal University, Khobar, Saudi Arabia (Al Damhogy) Tanta Insurance Hospital, Egypt

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Year of Publication

2017

519.

One-stage repair of proximal hypospadias with severe chordee by in situ tubularization of the transverse preputial island flap.

Huang Y., Xie H., Lv Y., Yu L., Sun L., Li X., Chen Y., Sun H., Chen F.

Embase

Journal of Pediatric Urology. 13 (3) (pp 296-299), 2017. Date of Publication: June 2017.

[Article]

AN: 615175405

Introduction To investigate a modified transverse preputial island flap repair which is performed in an attempt to reduce the complications following one-stage repair of proximal hypospadias with chordee. **Methods** Briefly, the two ends of the flap were trimmed into V shape and anastomosed with the spatulated urethra proximally and urethral plate distally before tubularization. Then the in situ tubularization of the flap was performed. The procedure was performed in our hospital on 32 patients (mean age = 11 months). They were followed for 12-38 months. **Results** The length of the urethral defect ranged from 4.0-6.0 cm after chordee correction. Urethrocutaneous fistulae occurred in 6 (18.7%) cases. No urethral strictures or meatal stenoses were observed. 29/32

families were satisfied with the cosmetic results. Conclusion This procedure seems straightforward and reliable, leading to good result after a short-term follow-up.

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Embase

Institution

(Huang, Xie, Lv, Yu, Sun, Li, Chen, Sun, Chen) Department of Urology, Shanghai Children's Hospital, Shanghai Jiaotong University, China

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2017

520.

A population-based study of prevalence trends and geospatial analysis of hypospadias and cryptorchidism compared with non-endocrine mediated congenital anomalies.

Lane C., Boxall J., MacLellan D., Anderson P.A., Dodds L., Romao R.L.P.

Embase

Journal of Pediatric Urology. 13 (3) (pp 284.e1-284.e7), 2017. Date of Publication: June 2017.

[Article]

AN: 615003876

Introduction Several reports have suggested an increase in the prevalence of hypospadias and cryptorchidism over the last few decades. Endocrine disruption caused by exposure to environmental chemicals has been postulated as a possible cause. Objectives The objectives of our study were: 1) to determine whether the prevalence of hypospadias and cryptorchidism is increasing compared with other congenital anomalies not known to be mediated by endocrine factors; and 2) to perform a geospatial analysis of these congenital malformations looking for clustering that could offer insight into environmental risk factors. Material and methods Data were obtained from the Nova Scotia ATLEE Perinatal Database containing the perinatal records of all live births in Nova Scotia, Canada since 1988. Records from 1988 to 2013 defined the study

cohort. Overall prevalence rates and prevalence trends by year were calculated for hypospadias, cryptorchidism, gastroschisis, and clubfoot. County of residence was collected and spatial autocorrelation testing for clustering was performed for each of the congenital anomalies. Results There were 258,147 live births during the study period. Overall prevalence rates for the four malformations over the study period were: hypospadias 78 per 10,000 male births, cryptorchidism 75 per 10,000 male births, clubfoot 24 per 10,000 total births, and gastroschisis 4 per 10,000 total births. Incidence rate ratios per year for hypospadias, cryptorchidism, clubfoot, and gastroschisis were 1.00 (0.99-1.01), 0.99 (0.98-1.00), 0.98 (0.97-0.99), and 1.04 (1.04-1.07), respectively. During the study period, the prevalence rates in the region were unchanged for hypospadias, slightly reduced for cryptorchidism and clubfoot, and rising for gastroschisis (Figure). Spatial autocorrelation testing revealed statistically significant clustering for hypospadias ($p = 0.03$) and cryptorchidism ($p = 0.03$), while no spatial autocorrelation was observed for the other malformations. Discussion Contrary to previous studies we show that hypospadias and cryptorchidism prevalence rates are not increasing over time in our region. Nonetheless, rates for these conditions in our area are high compared with other regions of the world. Local clustering of these congenital anomalies without clustering of the control, non-endocrine mediated congenital malformations supports a possible unique spatial distribution associated with environmental exposure. The hotspots identified for hypospadias and cryptorchidism are associated with intense agricultural activity. Conclusions Our study found no increase in hypospadias and cryptorchidism prevalence over a 26-year period compared with other congenital anomalies not known to be associated with endocrine factors. Geospatial analysis supports high clustering for hypospadias and cryptorchidism in areas of intense agricultural activity.[Figure presented]

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Status

Embase

Institution

(Lane, MacLellan, Anderson, Romao) Division of Urology, Department of Surgery, IWK Health Centre, Department of Urology, Faculty of Medicine, Dalhousie University, Halifax, NS, Canada

(Boxall) Department of Earth Sciences, Dalhousie University, Halifax, NS, Canada

(Dodds) Perinatal Research Epidemiology Unit, IWK Health Centre, Department of Pediatrics and Obstetrics/Gynecology, Dalhousie University, Halifax, NS, Canada

Publisher

Elsevier Ltd

Year of Publication

2017

521.

In pursuit of the perfect penis: Hypospadias repair outcomes.

Winship B.B., Rushton H.G., Pohl H.G.

Embase

Journal of Pediatric Urology. 13 (3) (pp 285-288), 2017. Date of Publication: June 2017.

[Review]

AN: 615002698

Hypospadias is commonly assessed and repaired by pediatric urologists. Mild, distal hypospadias is generally more a cosmetic problem than a functional one and is more frequently encountered than severe, proximal hypospadias. Outcomes following repair, especially of mild phenotypes, are important to understand, but range widely in timing and measurability. Surgical complications, postoperative satisfaction of parents, patients, surgeons and even lay observers, urinary function, sexual function, and quality of life all may be considered as relevant outcomes of hypospadias repair. Existing studies examining these outcomes are diverse in their conclusions, but are important to understand when counseling parents and patients prior to any surgical intervention.

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Status

Embase

Institution

(Winship) George Washington University School of Medicine, Washington, DC, United States

(Rushton, Pohl) Children's National Medical Center, Washington, DC, United States

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522.

Proximal hypospadias repair with bladder mucosal graft: Our 10 years experience.

Lanciotti M., Betti M., Elia A., Landi L., Taverna M., Cini C., Masieri L.

Embase

Journal of Pediatric Urology. 13 (3) (pp 294.e1-294.e6), 2017. Date of Publication: June 2017.

[Article]

AN: 614976418

Introduction and objective A great variety of different surgical techniques has been described for proximal hypospadias repair and an ideal tissue has not been determined yet. We present our 10 years of surgical experience using a bladder mucosal graft for urethroplasty. **Study design** Bladder mucosal graft urethroplasty was performed between 2005 and 2015 in 50 patients with severe proximal hypospadias. The mean age of patients was 45.1 months (range 24-164 months). Hypospadias were perineal in 18 patients, scrotal in 22, and penoscrotal in 10. In all cases a chordee correction was performed and median time between the first and the second stages was 12 months (mean 17 months, range 4-68 months). Both foley catheter and a suprapubic cystostomy were positioned and maintained for 2-4 weeks. Follow-up was performed at 1-3 and 6-12 months after surgery, and afterwards annually with clinical examination and flowmetry test. **Results** Mean follow-up was 5.3 years (median 5, range 1-10 years). Mean graft length was 57.4 mm (median 55 mm, range 35-85 mm). Among all the only early complication registered was a postoperative infection in one patient (2%) at the site of anastomosis. The long-term complications observed were urethrocutaneous fistula in nine patients (18%), urethral stricture in 15 patients (30%), meatal stricture in four patients (8%), and prolapse of meatus in seven patients (14%). The mean time of complication occurrence was 15 months (median 15.5 months, range 1-96 months). The functional and cosmetic appearance after surgery was satisfactory in 42/50 patients (84%) during the follow-up period. **Discussion** There is still an open debate regarding the optimal surgical approach for management of severe proximal hypospadias. Compared with other approaches, our technique showed acceptable results even though encumbered by slightly higher complication rates. **Conclusion** Our results show that bladder mucosal graft for primary severe proximal hypospadias in selected patients is a possible alternative to other commonly used techniques, with the aim of restoring recovery of the normal continuity of the distal urinary tract see figure below.[Figure presented]

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Author NameID

Lanciotti, Michele; ORCID: <http://orcid.org/0000-0002-3368-8652>

Institution

(Lanciotti, Betti, Elia, Landi, Taverna, Cini, Masieri) Urology Unit, Interdisciplinary Specialist
Department, Meyer University Hospital, Viale Pieraccini 24, Firenze 50139, Italy

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2017

523.

The Staged Urethroplasty with Vascularised Scrotal Flap and Buccal Mucosa Graft after Failed
Hypospadias Surgery: A Reliable Technique with a Novel Tool.

Pandey A., Barta-Kelemen A.-M., Borisenkov M., Keller H.

Embase

Urologia Internationalis. 99 (1) (pp 36-42), 2017. Date of Publication: 01 Jul 2017.

[Article]

AN: 614917525

Background/Aims/Objectives: To report the outcome of staged urethroplasty with buccal mucosa
and vascularised scrotal flap after failed hypospadias surgery.

Method(s): n = 35. Inclusion criteria: destroyed urethral plate, deficient local skin, fistula and penile
deviation. Five out of 35 patients also had a secondary proximal stricture. Stage 1: excision of
scarred tissue and fistulae, correction of the penile deviation through dorsal plication and the
ventral placement and quilting of buccal graft. Stage 2: tubularisation of the neourethral plate; in
20 patients with deficient penile skin a vascularised scrotal flap was developed and transferred on
the tubularised urethra. In 5 patients, the proximal stricture was repaired during a separate
operation by using buccal graft, the distal urethra was marsupialised. The repair of the distal
urethra was performed later as described above.

Result(s): Thirty-three patients are recurrence-free without further interventions; successful
reoperation was done in 2 cases. Complications: 1 graft necrosis; 1 coronary fistula; 1 scrotal flap
necrosis and 1 case of hematoma.

Conclusion(s): Complicated strictures after multiple failed hypospadias repair are well managed by using buccal graft. The vascularised scrotal flap is a very useful tool in case of deficient and scarred penile skin and could explain the low rate of fistula formation in our series.

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Institution

(Pandey, Barta-Kelemen, Borisenkov, Keller) Department of Urology, Urologic Oncology and Paediatric Urology, Sana Klinikum Hof, Eppenreuther Strasse 9, Hof DE-95032, Germany

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524.

Psychosexual development management of bladder exstrophy epispadias in complex patients.

Di Grazia M., Pellizzoni S., Tonegatti L.G., Rigamonti W.

Embase

Journal of Pediatric Urology. 13 (2) (pp 199.e1-199.e5), 2017. Date of Publication: April 2017.

[Article]

AN: 614488900

Introduction Bladder-exstrophy-epispadias complex (BEEC) represents a spectrum of urogenital step-wise malformations: epispadias, complete exstrophy, and cloacal exstrophy. Psychosexual development in adolescent patients with BEEC may become especially problematic. At present, there are few contributions in the literature investigating the validity of psychosexual treatment in order to tackle this particularly emotional and personal development phase. Objective The study aimed at verifying the efficacy of an intervention methodology for psychosexual support of a group of adolescents with BEEC. The main goal of the intervention program was to educate the adolescents and re-frame how they see themselves or feel about themselves, especially in relation to BEEC. In particular it was predicted that the program could: (1) improve the perception

of pleasure concerning the body, particularly regarding the genital area, giving proper and specific information on pleasure, masturbation and medical history of BEEC; and (2) elicit a more relational-affective perspective on sexuality. Study design 13 adolescent patients took part in the 1-year program. The effects of the intervention program were verified through a test-retest methodology using Sexuality Evaluation Schedule Assessment Monitoring (SESAMO). Results The results showed that participants changed their attitude in several psychosexual areas, more specifically: psycho-environmental situations, body experience, areas of pleasure, medical and sexual history, and motivation and conflict areas (Summary Table). Discussion This study demonstrated, for the first time, that a targeted program may significantly improve the psychosexual condition of adolescents with BEEC. In particular, this research showed that adolescents need to be able to discuss and tackle topics of a psychological and sexual nature, as well as receive understandable answers that can be put into practice in their everyday lives. The study had several methodological limitations, especially owing to the limited number of participants, the absence of a follow-up period of a few months after the intervention, and the overall exploratory nature of the program. Conclusion This intervention methodology may be considered a first attempt at improving the self-esteem of adolescents with BEEC, by contrasting forms of psychological difficulties in order to improve the quality of life of these young people. [Table presented]

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Status

Embase

Author NameID

Di Grazia M.; ORCID: <http://orcid.org/0000-0002-1302-7785>

Institution

(Di Grazia, Rigamonti) Department of Medical, Surgical and Health Science, University of Trieste, Trieste, Italy (Pellizzoni) Department of Life Science, University of Trieste, Via Weiss 21, Building W, Trieste, Italy

(Tonegatti, Rigamonti) Institute for Maternal and Child Health, IRCCS 'Burlo Garofolo', Trieste, Italy

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525.

Variation in the clinical and genetic evaluation of undervirilized boys with bifid scrotum and hypospadias.

Swartz J.M., Ciarlo R., Denhoff E., Abrha A., Diamond D.A., Hirschhorn J.N., Chan Y.-M.

Embase

Journal of Pediatric Urology. 13 (3) (pp 293.e1-293.e6), 2017. Date of Publication: June 2017.

[Article]

AN: 614461837

Background Bifid scrotum and hypospadias can be signs of undervirilization, yet boys presenting with these findings often do not undergo genetic evaluation. In some cases, identifying an underlying genetic diagnosis can help to optimize clinical care and improve guidance given to patients and families. **Objectives** The aim of this study was to characterize current practice for genetic evaluation of patients with bifid scrotum, and to identify approaches with a good diagnostic yield. **Methods** A retrospective study of the Boston Children's Hospital electronic medical records (1993-2015) was conducted using the search term "bifid scrotum" and clinical data were extracted. Data were abstracted into a REDCap database for analysis. Statistical analysis was performed using SPSS, SAS, and Excel software. **Results** The search identified 110 subjects evaluated in the Urology and/or Endocrinology clinics for bifid scrotum. Genetic testing (including karyotype, microarray, or targeted testing) was performed on 64% of the subjects with bifid scrotum; of those tested, 23% (15% of the total cohort of 110 subjects) received a confirmed genetic diagnosis. Karyotype analysis, when performed, led to a diagnosis in 17% of patients. Of the ten instances when androgen receptor gene sequencing was performed, a pathogenic mutation was identified 20% of the time. **Conclusion** This study demonstrated that the majority of individuals with moderate undervirilization resulting in bifid scrotum do not receive a genetic diagnosis. Over a third of the analyzed subjects did not have any genetic testing, even though karyotype analysis and androgen receptor (AR) sequencing were both relatively high yield for identifying a genetic etiology. Increased utilization of traditional genetic approaches could significantly improve the ability to find a genetic diagnosis.[Figure presented]

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Embase

Author NameID

Swartz J.M.; ORCID: <http://orcid.org/0000-0003-1017-9318>

Institution

(Swartz, Ciarlo, Abrha, Hirschhorn, Chan) Department of Endocrinology, Boston Children's Hospital, Boston, MA, United States (Denhoff) Clinical Research Center, Boston Children's Hospital, Boston, MA, United States

(Diamond) Department of Urology, Boston Children's Hospital, Boston, MA, United States

(Hirschhorn) Program in Medical and Population Genetics, Cambridge, MA, United States

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526.

Neo-yoke repair for severe hypospadias: A simple modification for better outcome.

Seleim H.M., Morsi H., Elbarbary M.M.

Embase

Journal of Pediatric Urology. 13 (3) (pp 290.e1-290.e7), 2017. Date of Publication: June 2017.

[Article]

AN: 614278977

Background Although staged repair for reconstructing severe hypospadias is more popular, various one-stage repairs have been attempted. Koyanagi repair (parameatal-based and fully extended circumferential foreskin flap urethroplasty) has enabled correction of severe hypospadias in one stage. However, its un-acceptably high incidence of complications has initiated a series of technical modifications, including the "yoke" repair. Objectives To retrospectively analyze the outcome of a proposed modification of the originally described yoke repair, for patients with severe hypospadias. This modification was developed to reduce complications. Study design Over 4 years (between Jan 2011 and Jan 2015), all cases of severe hypospadias were included in this study; except those with prior attempts at repair, circumcised cases, and cases with severe hypogonadism - because of partial androgen insensitivity - not responding to hormonal manipulations. The make-up of the neo-urethra in this modification is the urethral plate with its spongiosal tissue proximally, a circum-coronal preputial pedicled flap in the

middle, and an incorporated part of the augmented preputial flap and the preserved V-shaped glanular urethra, distally. Close postoperative follow-up was conducted to investigate the outcome. Results Thirty-one children with a median age of 32.48 months had repair of severe hypospadias using the neo-yoke technique. After a median follow-up of 26.7 months, the overall complication rate was 16.1%. Four children developed urethrocutaneous fistula (12.9%). Meatal drop-back occurred in one case (3.2%). No meatal stenosis or urethral sacculation was detected during follow-up of the studied group. Almost all cases had cosmetically appealing outlook. Single-staged repair of severe hypospadias using parametatal foreskin-based urethroplasty has passed through different modifications, all aimed at optimizing the outcome (Table). Conclusion Neo-yoke repair for severe hypospadias is a natural development of established one-stage techniques, which resulted in better mid-term outcomes. However, an extended study is needed to declare the long-term results. [Table presented]

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Status

Embase

Institution

(Seleim) Pediatric Surgery Unit, Tanta University Hospital, Tanta, Egypt (Morsi) Pediatric

Urology Unit, Cairo University Specialized Children Hospital, Cairo, Egypt

(Elbarbary) Pediatric Surgery Unit, Cairo University Specialized Children Hospital, Cairo, Egypt

Publisher

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Year of Publication

2017

527.

Durham Smith Vest-Over-Pant Technique: Simple Procedure for a Complex Problem (Post-Hypospadias Repair Fistula).

Gite V.A., Patil S.R., Bote S.M., Siddiqui M.A.K.N., Nikose J.V., Kandi A.J.

Embase

Urologia Internationalis. 99 (1) (pp 29-35), 2017. Date of Publication: 01 Jul 2017.

[Article]

AN: 614243487

Objective: Urethrocuteaneous fistula, which occurs after hypospadias surgery, is often a baffling problem and its treatment is challenging. The study aimed to evaluate the results of the simple procedure (Durham Smith vest-over-pant technique) for this complex problem (post-hypospadias repair fistula).

Method(s): During the period from 2011 to 2015, 20 patients with post-hypospadias repair fistulas underwent Durham Smith repair. Common age group was between 5 and 12 years. Site wise distribution of fistula was coronal 2 (10%), distal penile 7 (35%), mid-penile 7 (35%), and proximal-penile 4 (20%). Out of 20 patients, 15 had fistula of size <5 mm (75%) and 5 patients had fistula of size >5 mm (25%). All cases were repaired with Durham Smith vest-over-pant technique by a single surgeon. In case of multiple fistulas adjacent to each other, all fistulas were joined to form single fistula and repaired.

Result(s): We have successfully repaired all post-hypospadias surgery urethrocuteaneous fistulas using the technique described by Durham Smith with 100% success rate.

Conclusion(s): Durham Smith vest-over-pant technique is a simple solution for a complex problem (post hypospadias surgery penile fistulas) in properly selected patients.

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Embase

Institution

(Gite, Patil, Bote, Siddiqui, Nikose) Department of Urology, Grant Government Medical College, Sir J.J. Group of Hospitals, Saikrupa, H-21, TirupatiSupreme Enclave, Jalan Nagar, Mumbai, Maharashtra 431005, India (Kandi) Department of General Surgery, Government Medical College, Aurangabad, India

Publisher

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2017

Outcome of urethroplasty after parenteral testosterone in children with distal hypospadias.

Menon P., Rao K.L.N., Handu A., Balan L., Kakkar N.

Embase

Journal of Pediatric Urology. 13 (3) (pp 292.e1-292.e7), 2017. Date of Publication: June 2017.

[Article]

AN: 614137714

Introduction Pre-operative testosterone use in hypospadias surgery is known to increase penile dimensions and vascularity, which should facilitate tension-less formation of the urethral tube and tissue healing. However, androgens can have a negative effect on wound healing. There are very few randomized studies on postoperative results after androgen use, and this study attempted to understand the utility of pre-operative testosterone in distal hypospadias. **Objective** To study the effect of parenteral testosterone in children undergoing single stage urethroplasty for distal hypospadias, especially the occurrence of urethrocuteaneous fistula and wound dehiscence.

Design Patients were prospectively enrolled and randomized into two groups: Group 1 (control group) and Group 2 (receiving three injections of pre-operative intramuscular testosterone enanthate (2 mg/kg) at 1 monthly intervals; they were further subdivided into those operated 1 month (Group 2A) or 3 months (Group 2B) later. Patients with micropenis, previous testosterone use or any surgical intervention were excluded. Preputial skin was studied with hematoxylin and eosin (H&E) staining and CD31 immunohistochemistry. Patients were followed up for at least 18 months. **Results** Ninety four patients underwent urethroplasty over a 3.5-year period. Penile dimensions increased significantly after testosterone use (Summary table). On H&E staining, proliferating blood vessels and increased lymphocytic infiltrates were significantly increased in Group 2B. Group 2 patients tended to have more postoperative edema and inflammation.

Although urethrocuteaneous fistula rates were similar in Group 1 (n = 7) and Group 2 (n = 5) (P = 0.438), wound dehiscence occurred only in Group 2 (P = 0.01). **Discussion** The total number of patients in this study was small and this was a drawback. Although, several factors played a role in wound healing, the overall higher complication rate, especially wound dehiscence in Group 2, pointed to a higher incidence of inflammatory reaction and healing complication rates with testosterone use. **Conclusion** Testosterone should be used judiciously in distal hypospadias.

While tissue availability significantly increased, there was an increase in inflammatory reaction and edema, which increased the risk of wound dehiscence in cases of precocious surgery. [Table presented]

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Status

Embase

Institution

(Menon, Rao, Handu) Department of Pediatric Surgery, Postgraduate Institute of Medical Education and Research, Chandigarh 160012, India (Balan, Kakkar) Department of Histopathology, Postgraduate Institute of Medical Education and Research, Chandigarh 160012, India

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2017

529.

Bleeding after circumcision is more likely in children with lichen sclerosus (balanitis xerotica obliterans).

Somov P., Chan B.K.Y., Wilde C., Corbett H.

Embase

Journal of Pediatric Urology. 13 (2) (pp 208.e1-208.e4), 2017. Date of Publication: April 2017.

[Article]

AN: 614101103

Introduction Over 27,000 circumcisions were performed in England in 2012-13. The complication rate is generally perceived to be low, although published figures vary widely. Balanitis xerotica obliterans, more correctly termed Lichen Sclerosus et atrophicus (LS), is one of the commonest indications for medical circumcision. To test the hypothesis that children undergoing circumcision for LS have a higher rate of postoperative bleeding than those undergoing the procedure for other reasons, we retrospectively reviewed records for patients undergoing circumcision. Methods The disease and procedure coding system was used to identify patients who underwent circumcision (ICD10 code N303) between 2000-2010. Cases with a diagnosis unrelated to circumcision and children circumcised during hypospadias repair were excluded. Bleeding which required return to theatre for surgical arrest was considered significant. Cases were identified by review of medical records if there was: a second procedure during the same admission, or readmission coded for circumcision within 2 weeks. Only cases with histologically confirmed LS were included in the LS cohort. GraphPad online calculator was used for statistical analysis (two tailed Fisher's exact test.

Results 2385 boys with a median age of 4 years (range 0-16) were included in the study. Indication for circumcision included religious (1305, 54.7%), phimosis or redundant prepuce (512, 21.5%), suspected LS (366, 15.4%) and balanoposthitis (202, 8.5%). LS was histologically confirmed in 262 (10.9%) boys. Fourteen (0.6%) patients returned to theatre for surgical arrest of bleeding following circumcision; 6 had LS and 8 did not (Table 1). The bleeding rate was higher in those with LS (2.3%) than in those without (0.3%), $P = 0.0003$ with a relative risk of 6.08. Conclusion Post-operative complications are distressing, especially if further surgery is required. Published figures for complications following circumcision vary widely making counseling regarding risk difficult. Since LS includes an inflammatory element and circumcision in widespread LS can be challenging, the observation of more post-operative bleeding in patients with histologically confirmed LS during a previous audit prompted the hypothesis that this may be a significant finding. Thus we reviewed all patients requiring return to theatre within 2 weeks of circumcision, finding that whilst the overall bleeding rate was low, circumcision for LS significantly increased the risk. Although factors such as the severity of LS and surgical technique were not assessed, this is still a notable finding which should be reflected during pre-operative counseling.

[Table presented]

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Status

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Author NameID

Somov P.; ORCID: <http://orcid.org/0000-0001-8161-2815> Chan B.K.Y.; ORCID:

<http://orcid.org/0000-0001-9500-2380>

Institution

(Somov, Chan, Wilde, Corbett) Alder Hey Children's NHS Foundation Trust, Department of Paediatric Surgery, E Prescott Rd, Liverpool L14 5AB, United Kingdom

Publisher

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Re-operative urethroplasty after failed hypospadias repair: how prior surgery impacts risk for additional complications.

Snodgrass W., Bush N.C.

Embase

Journal of Pediatric Urology. 13 (3) (pp 289.e1-289.e6), 2017. Date of Publication: June 2017.

[Article]

AN: 614076123

Purpose The primary aim of this report was to compare urethroplasty complications for primary distal and proximal repairs with those after 1, 2, 3, and 4 or more re-operations. **Methods** Prospectively collected data on consecutive hypospadias repairs (tubularized incised plate (TIP), inlay, two-stage graft) from 2000 to 2015 were reviewed. Isolated fistula closures were excluded. Extracted information included patient age, meatal location, repair type, primary vs. re-operative surgery, number of prior operations, any testosterone use, glans width, and urethroplasty complications. Pre-operative testosterone stimulation was used during the study period until 2012. Initially, it was given for a subjectively small-appearing glans, but from 2008 to 2012 use was determined by glans width <14 mm. Patients initially managed elsewhere were queried for any testosterone treatment. The number of prior operations was determined by patient history and confirmed by review of records. Calibrations, dilations, cystoscopies, and/or isolated skin revisions were not considered as prior urethroplasty operations. Multiple logistic regression was performed for all patients, and for the subset of patients undergoing re-operation, using stepwise regression for the following potential risk factors: meatal location (distal vs. midshaft/proximal), number of prior surgeries (0, 1, 2, 3, >=4), pre-operative testosterone use (yes/no), small glans (<14 vs. >=14), surgery type (TIP, inlay and two-stage graft), and age (continuous in months), with P-values <0.05 considered statistically significant. **Results** In contrast to the 135/1085 (12%) complication rate in patients undergoing primary distal and proximal TIP repair, re-operative urethroplasty complications occurred in 61/191 (32%) TIP, 16/46 (35%) inlay, and 49/124 (40%) two-stage repairs, $P < 0.0001$. Data regarding testosterone use was available for 1490 (96%) patients. A total of 139 received therapy, of which 65 (46%) had urethroplasty complications vs. 229 of 1351 (16%) without treatment, $P = 0.0001$. Logistic regression in 1536 patients demonstrated that each prior surgery increased the odds of subsequent urethroplasty complications 1.5-fold (OR 1.51, 95% CI 1.25-1.83), along with small glans <14 mm (OR 2.40, 95% CI 1.48-3.87), mid/proximal meatal location (OR 2.54, 95% CI 1.65-3.92), and use of pre-operative testosterone (OR 2.57, 95% CI 1.53-4.31); age and surgery type did not increase odds (AUC = 0.739). **Discussion** Urethroplasty complications doubled in people undergoing a second hypospadias urethroplasty compared with those undergoing primary repair. This risk increased to 40% with three or more re-operations. Logistic regression demonstrates that each surgery

increases the odds for additional complications 1.5-fold. Mid/proximal meatal location, small glans <14 mm, and use of pre-operative testosterone also significantly increase odds for complications. These observations support the theory that previously operated tissues have less robust vascularity than assumed in a primary repair, and suggest additional adjunctive therapies are needed to improve wound healing in re-operations. The finding that even a single re-operative urethroplasty has twice the risk for additional complications vs. a primary repair emphasizes the need for hypospadias surgeons to 'get it right the first time'. The fact that 40% of the re-operative urethroplasties in this series followed distal repairs emphasizes that there is no 'minor' hypospadias. Conclusions A single re-operative hypospadias urethroplasty has twice the risk for additional complications vs. the primary repair, which increases to 40% with three or more re-operations. These results support a theory that vascularity of penile tissues decreases with successive operations, and suggest the need for treatments to improve vascularity. The higher risk for complications during re-operative urethroplasties also emphasizes the need to get the initial repair correct.

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Status

Embase

Author NameID

Snodgrass W.; ORCID: <http://orcid.org/0000-0001-8715-9919>

Institution

(Snodgrass, Bush) PARC Urology, 5680 Frisco Square Blvd, Suite 2300, Frisco, TX 75034,
United States

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531.

Recurrence after management of meatal balanitis xerotica obliterans.

Snodgrass W., Blanquel J.S., Bush N.C.

Embase

Journal of Pediatric Urology. 13 (2) (pp 204.e1-204.e6), 2017. Date of Publication: April 2017.

[Article]

AN: 614013785

Introduction We review outcomes after management of meatal balanitis xerotica obliterans (BXO). The primary outcome was recurrent meatal BXO. **Methods** A database comprising mostly hypospadias patients was queried for meatal BXO. The disease was confirmed histologically in all cases. Management included topical steroids and/or immunosuppressants, and/or surgical excision of BXO with two-stage oral mucosa graft circumferential replacement urethroplasty.

Results A total of 12 patients had meatal BXO (8 boys and 4 adults). Of these, 10 had hypospadias, two presenting without prior surgery and eight returning 5-30 years after one or multiple (n = 2) repairs. Another two boys did not have hypospadias: one developing BXO 10 years after newborn circumcision and the other having persistent meatal BXO following therapeutic circumcision. Topical and intraluminal steroids (1% betamethasone or clobetasol) and tacrolimus were used for ≥ 12 weeks each as primary therapy or for meatal recurrence in a total of six cases. Complete response with resolution of white discoloration and relief of stranguria only occurred in two of the three receiving clobetasol, with follow up ≤ 12 weeks. BXO excision and urethroplasty was done in 11 patients, 10 using oral mucosa grafts; one with a focal lesion and a negative frozen section had reoperative TIP. Of the 10 undergoing excision with two-stage replacement urethroplasty, six remain disease free at a mean follow-up of 23 months (8-48 months), and four had recurrent stranguria and visible meatal BXO at a median of 26 months (22-105 months). Three of the four with recurrences had additional treatment and one was lost to follow-up. All initially had topical steroids, and two also used tacrolimus, without clinical resolution. These three then underwent a second BXO excision and two-stage oral graft replacement urethroplasty. In two recurrences, BXO was found invading from the meatus proximally within oral mucosa (Figure). Of these three with secondary urethroplasties, two are free of disease at 6 and 18 months, and the third had another meatal recurrence 6 months after the second stage.

Discussion We found topical steroids and immunosuppressants to have limited efficacy, with two clinical complete responses achieved only with clobetasol in patients with short follow-up. Forty percent of patients recurred at 2-9 years after visually complete BXO excision and two-stage oral mucosa graft replacement urethroplasty, and in two cases disease invaded into oral mucosa, the first well-documented cases of this occurrence.[Figure presented]

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Status

Embase

Author NameID

Snodgrass, Warren; ORCID: <http://orcid.org/0000-0001-8715-9919> Blanquel, Juan Soto;

ORCID: <http://orcid.org/0000-0001-7862-2131>

Institution

(Snodgrass, Bush) PARC Urology, Frisco, TX, United States (Blanquel) Hospital del Nino y el Adolescente Morelense, Emiliano Zapata, Mexico

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532.

High GMS score hypospadias: Outcomes after one- and two-stage operations.

Huang J., Rayfield L., Broecker B., Cerwinka W., Kirsch A., Scherz H., Smith E., Elmore J.

Embase

Journal of Pediatric Urology. 13 (3) (pp 291.e1-291.e4), 2017. Date of Publication: June 2017.

[Article]

AN: 614007338

Introduction Established criteria to assist surgeons in deciding between a one- or two-stage operation for severe hypospadias are lacking. While anatomical features may preclude some surgical options, the decision to approach severe hypospadias in a one- or two-stage fashion is generally based on individual surgeon preference. This decision has been described as a dilemma as outcomes range widely and there is lack of evidence supporting the superiority of one approach over the other. Objectives The aim of this study is to determine whether the GMS hypospadias score may provide some guidance in choosing the surgical approach used for correction of severe hypospadias. Study design GMS scores were preoperatively assigned to patients having primary surgery for hypospadias. Those patients having surgery for the most severe hypospadias were selected and formed the study cohort. The records of these patients were reviewed and pertinent data collected. Complications requiring further surgery were assessed and correlated with the GMS score and the surgical technique used for repair (one-stage vs. two-stage). Results Eighty-seven boys were identified with a GMS score (range 3-12) of 10 or higher. At a mean follow-up of 22 months the overall complication rate for the cohort after

final planned surgery was 39%. For intended one-stage procedures (n = 48) an acceptable result was achieved with one surgery for 28 patients (58%), with two surgeries for 14 (29%), and with three to five surgeries for six (13%). For intended two-stage procedures (n = 39) an acceptable result was achieved with two surgeries for 26 patients (67%), three surgeries for eight (21%), and four surgeries for three (8%). Two other patients having two-stage surgery required seven surgeries to achieve an acceptable result. Complication rates are summarized in the Table. The complication rates for GMS 10 patients were similar (27% and 33%, $p = 0.28$) for one- and two-stage repairs, respectively. GMS 11 patients having a one-stage repair had a significantly higher complication rate (69%) than those having a two-stage repair (29%) ($p = 0.04$). GMS 12 patients had the highest complication rate with a one-stage repair (80%) compared with a complication rate of 37% when a two-stage repair was used ($p = 0.12$). Conclusions Guidelines to help standardize the surgical approach to severe hypospadias are needed. Staged surgery for GMS 11 and 12 patients may result in a lower complication rate but may not reduce the number of surgeries required for an acceptable result. Although further study is needed, the GMS score may be helpful for establishing such criteria. [Table presented]

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Embase

Author NameID

Elmore, James; ORCID: <http://orcid.org/0000-0002-1289-860X>

Institution

(Huang) Department of Urology, Emory University School of Medicine and Children's Healthcare of Atlanta, Atlanta, GA, United States (Rayfield) Department of Biostatistics, Rollins School of Public Health, Emory University, Atlanta, GA, United States

(Broecker, Cerwinka, Kirsch, Scherz, Smith, Elmore) Department of Pediatric Urology, Emory University School of Medicine and Children's Healthcare of Atlanta, Atlanta, GA, United States

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2017

Outcomes of seromuscular bladder augmentation versus standard ileocystoplasty: A single institution experience over 14 years.

Odeh R.I., Farhat W.A., Penna F.J., Koyle M.A., Lee L.C., Butt H., Alyami F.A.

Embase

Journal of Pediatric Urology. 13 (2) (pp 200.e1-200.e5), 2017. Date of Publication: April 2017.

[Article]

AN: 613194138

Introduction Ileocystoplasty is the standard technique used for bladder augmentation, and has been used widely for decades. However, it is known to be associated with complications such as stone formation, mucus production, metabolic acidosis, urinary tract infections, intestinal obstruction, and a long-term risk of bladder cancer. Seromuscular bladder augmentation (SMBA) is an alternative to the standard ileocystoplasty, and has been associated with a lower incidence of bladder stones. Few reports have been published on intermediate outcomes of SMBA. Herein, we report long-term outcomes of SMBA from a single institution compared with standard ileocystoplasty. **Methods** After Institutional Review Board approval, a retrospective chart review of all patients who underwent bladder augmentation at our institution over a 14-year period was performed. The status of patients after SMBA (10 patients) was compared according to age, sex, and diagnosis with patients who underwent traditional ileocystoplasty (30 patients). Parameters such as demographic information, pre- and postoperative bladder capacity as assessed by urodynamic studies, urinary tract infections (UTIs), bladder calculi, incontinence, need for secondary surgical procedures, and spontaneous bladder perforation were compared in the two groups. All the patients were on a clean intermittent catheterization (CIC) regimen. **Results** Over the study period, 10 patients underwent SMBA and 30 patients (according to age, sex, and diagnosis) underwent standard ileocystoplasty; the average age at surgery was 10.3 and 10 years respectively, with a mean follow up of 6.7 years in the SMBA group and 6 years in the ileocystoplasty group. There were no statistically significant differences in the rate of UTIs, urinary incontinence, subsequent surgery, or spontaneous bladder perforation. The mean bladder capacity increased significantly for both groups as assessed by pre- and postoperative urodynamic studies, although the difference in the rate of bladder calculi between the two groups (0 [0%] vs. 8 [27%], $p = 0.06$) did not reach statistical significance (Table). **Conclusions** SMBA is safe and efficacious and may result in a lower rate of stone formation than standard ileocystoscopy. SMBA should be considered as a viable alternative to standard ileocystoplasty [Table presented].

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Author NameID

Penna, Frank J.; ORCID: <http://orcid.org/0000-0003-0962-2220> Alyami, Fahad A.; ORCID: <http://orcid.org/0000-0002-4519-4510>

Institution

(Odeh, Farhat, Penna, Koyle, Lee, Butt, Alyami) Division of Paediatric Urology, The Hospital for Sick Children and University of Toronto, Toronto, Ontario, Canada (Alyami) Urology Division, Department of Surgery, King Saud University, King Khalid University Hospital, College of Medicine, Riyadh, Saudi Arabia

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2017

534.

Low-dose Human Chorionic Gonadotropin Stimulation Test as a Prognostic Incontinent Indicator in Boys With Bladder Exstrophy-epispadias Complex.

Sabetkish N., Eftekharzadeh S., Elmi A., Talab S.S., Kajbafzadeh A.-M.

Embase

Urology. 101 (pp 133-138), 2017. Date of Publication: 01 Mar 2017.

[Article]

AN: 614274642

Objective To investigate the effect of low-dose human chorionic gonadotropin (HCG) administration on structural changes in the lower urinary tract in boys with urinary incontinence in the setting of bladder exstrophy-epispadias complex (BEEC). Patients and Methods We prospectively evaluated 30 patients (median age 7.5 years) with BEEC and randomly divided them into 2 groups. Patients in the HCG group were administered 250IU HCG intramuscularly 3 times per week during a 4-week period. The other 15 patients served as the control group. The patients were followed up for a mean duration of 4 years. Incontinence rate, hormonal changes, penile length, prostate size, and bladder capacity were evaluated using 3D sonography or pelvic

magnetic resonance imaging and uroflowmetry studies before and after HCG administration. Results The incontinence score improvement was significantly higher in the HCG group ($P=.01$). A significant increase was detected in the health-related quality of life score of both patients and parents at the final follow-up ($P<.001$). The total prostate size ($P<.0001$) and bladder capacity ($P<.0001$) increased significantly in all patients of the HCG group. Basal serum testosterone level increased significantly after the first ($P=.001$) and last ($P<.001$) injections with no significant increase 3 months after the last injection ($P>.05$). No major side effect was found following the administration of HCG, with no need for open surgical bladder neck reconstruction. Conclusion Our preliminary results suggest the role of low-dose HCG in boys with BEEC suffering from urinary incontinence. The data also reveal the role of prostate enlargement in the improvement of urinary incontinence. Chronic treatment with HCG increases bladder capacity that may facilitate future reconstructive surgery.

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Institution

(Sabetkish, Eftekharzadeh, Talab, Kajbafzadeh) Pediatric Urology Research Center, Department of Pediatric Urology, Pediatrics Center of Excellence, Tehran University of Medical Sciences, Tehran, Iran, Islamic Republic of (Elmi) Department of Radiology, Massachusetts General Hospital, Harvard Medical School, Boston, MA, United States

Publisher

Elsevier Inc. (E-mail: usjcs@elsevier.com)

Year of Publication

2017

535.

Psychosocial outcome in adult men born with hypospadias.

Ortqvist L., Andersson M., Strandqvist A., Nordenstrom A., Frisen L., Holmdahl G., Nordenskjold A.

Embase

Journal of Pediatric Urology. 13 (1) (pp 79.e1-79.e7), 2017. Date of Publication: 01 Feb 2017.

[Article]

AN: 614105500

Introduction Hypospadias, which is a surgically treated congenital malformation of the male urethra, may have a negative impact on quality of life. This aspect has previously been subject to limited research. This study examined the long-term psychosocial outcome of a large cohort of adult males born with hypospadias. **Objective** The purpose of this case-control study was to assess a possible negative influence on the psychosocial outcome in adult males with hypospadias. **Study design** Males with hypospadias treated in Sweden and aged ≥ 18 years old participated in this follow-up study. Age-matched men and university students were recruited as controls. The participants answered a questionnaire designed to reflect the subjective quality of life, social factors, need of support and follow-up, and the perceived impact of the disease upon upbringing. It also looked at the validated Psychological General Well-Being (PGWB) questionnaire and Relationship Questionnaire (RQ). **Results and discussion** A total of 167 patients (median age 34 years, 63% distal, 24% mid, and 13% proximal hypospadias) and 169 controls (median age 33 years) participated in the study. Patients had their first operation at 4 years of age (median) and the median follow-up time was 29 years following the first surgery. Men with hypospadias had a comparable total quality of life level with a mean total PGWB score of 82 (normal range 78-83) compared with 85.6 in controls. Scores on wellbeing and vitality were lower, even if the differences were small. Hypospadias did not affect marital status, presence of children in the family, frequency of employment or experience of bullying. These men more often lived at home with their parents ($P=0.001$) and had a lower level of education ($P=0.004$), even if the educational level in both patients and controls was high compared with the general Swedish population. Patients with proximal hypospadias were shorter compared with controls ($P=0.003$), which was consistent with the prenatal growth restriction associated with hypospadias. The group with proximal hypospadias expressed a greater need for medical (45.5%) follow-up compared with mid (28.2%) and distal (18.1%) cases ($P=0.001$). Patients with proximal hypospadias tended to avoid close relationships because of fear of being hurt. **Conclusions** The findings suggested that patients treated for hypospadias have a good HRQoL, can be expected to have a normal psychosocial life, and marry and have children. Repeated follow-up and psychological support during childhood/adolescence is however of great importance for patients with more proximal hypospadias. [Table presented]

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Status

Embase

Institution

(Ortqvist, Strandqvist, Nordenstrom, Nordenskjold) Department of Women's and Children's Health, Karolinska Institutet, Stockholm S-171 76, Sweden (Ortqvist, Nordenskjold) Center for Molecular Medicine, Karolinska Institutet, CMM Q2:02, Stockholm S-17176, Sweden

(Ortqvist, Nordenskjold) Department of Pediatric Surgery, Astrid Lindgren Children's Hospital, Karolinska University Hospital, Stockholm S-17176, Sweden

(Andersson, Holmdahl) Sahlgrenska Academy, Women's and Children's Health, Gothenburg S-413 90, Sweden

(Andersson, Holmdahl) Department of Paediatric Surgery, Queen Silvia's Children's Hospital, Gothenburg S-41685, Sweden

(Nordenstrom) Department of Paediatric Endocrinology, Astrid Lindgren Children's Hospital, Karolinska University Hospital, Stockholm S-17176, Sweden

(Frisen) Department of Clinical Neuroscience, Karolinska Institutet, Administration Tomtebodavagen 18A, 5th floor, Stockholm S-171 77, Sweden

(Strandqvist, Frisen) Child and Adolescent Psychiatry Research Center, Karolinska Institutet, Norra stationsgatan 69, Plan 7, Stockholm S-113 64, Sweden

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2017

536.

Prevalence of preoperative penile abnormalities among voluntary male medical circumcision patients in Swaziland.

Oddo A.R., Ruedrich E., Zust C., Marugg L., VanderWal E., VanderWal H., Sartori R., Markert R., McCarthy M.C.

Embase

Journal of Pediatric Surgery. 52 (8) (pp 1332-1334), 2017. Date of Publication: August 2017.

[Article]

AN: 614102635

Background Circumcision has been found to be an effective strategy for lowering the transmission of HIV in Africa. The Luke Commission, a mobile hospital outreach program, has

used this information to decrease the rate of HIV in Swaziland by performing voluntary male medical circumcisions throughout the country. During many of these circumcisions, genital medical conditions and penile abnormalities are simultaneously discovered and corrected.

Purpose The goal of our study was to evaluate the prevalence of penile abnormalities discovered and treated during voluntary male medical circumcisions performed by The Luke Commission (TLC) throughout rural Swaziland.

Basic procedures We completed a retrospective analysis of all male patients who underwent voluntary male medical circumcision performed by TLC during a period from June-August, 2014. The penile abnormalities included: phimosis, paraphimosis, epispadias, hypospadias, ulcers, balanitis, torsion, and foreskin adherent to the glans.

Main findings Of 929 total circumcisions, 771 (83%) patients had at least one pre-existing penile abnormality identified during their examinations and procedures, totaling 1110 abnormalities. Three specific abnormalities were detected - phimosis, adherent foreskin, and hypospadias. The 6-12 and 13-19 age groups had adequate sample sizes to yield precise estimates of prevalence (age group 6-12: 87% (95% confidence interval [CI] = 84-90%; age group 13-19: 79% (95% CI = 74-84%).

Principle conclusions The Luke Commission is improving the lives of children and adults with limited access to healthcare through regular preoperative evaluations during male circumcision, and the organization is setting an example for other international healthcare groups.

Level of evidence Type of Study: Prognostic Study, Level II.

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Embase

Institution

(Oddo, Ruedrich, Zust, Marugg, Markert, McCarthy) Wright State University, Boonshoft School of Medicine, 3640 Colonel Glenn, Highway, Dayton, OH 45435, United States (VanderWal, VanderWal, Sartori) The Luke Commission, Miracle Campus, Box 41, Sidvokodvo, Swaziland

Publisher

W.B. Saunders

Year of Publication

2017

Can regional anesthesia have an effect on surgical outcomes in patients undergoing distal hypospadias surgery?.

Saavedra-Belaunde J.A., Soto-Aviles O., Jorge J., Escudero K., Vazquez-Cruz M., Perez-Brayfield M.

Embase

Journal of Pediatric Urology. 13 (1) (pp 45.e1-45.e4), 2017. Date of Publication: 01 Feb 2017.

[Article]

AN: 613978120

Background Caudal and penile blocks are the most popular regional anesthetic techniques used in infants and children undergoing urological surgery. A recent report has suggested that penile venous pooling resulting from caudal blocks could affect surgical outcomes after hypospadias operations. **Objective** The aim was to report our experience in patients with distal hypospadias undergoing repair with caudal versus penile block. **Study design** A retrospective clinical database was constructed for patients who underwent distal hypospadias repair by a single surgeon (M.P.B.) at our sponsoring institutions for the time period 2008-2013 (n = 192). Collected data included hypospadias classification (glanular, coronal, subcoronal), chordee status, perioperative anesthesia (caudal vs. penile), and assessment of postoperative complications (fistula and meatal stenosis). **Results** Risk ratio (RR) analysis for all distal hypospadias cases revealed that there is a higher risk of developing complications in patients who underwent caudal anesthesia than in patients who underwent penile block RR for a complication was 3.70 (95% CI 1.05-13.03; $p < 0.04$) (Figure). **Discussion** Similar to other papers in the literature, we found that patients who underwent caudal anesthesia had more complications than those who underwent penile block. The limitations of this study include not adjusting the results according to the severity of hypospadias. **Conclusion** The main goal of this study was accomplished by demonstrating that, in our series, caudal anesthesia is associated with a higher risk of fistula formation after undergoing distal hypospadias repair than penile block.[Figure presented]

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Status

Embase

Author NameID

Saavedra-Belaunde, Jose A.; ORCID: <http://orcid.org/0000-0001-6913-1752> Soto-Aviles, Omar;

ORCID: <http://orcid.org/0000-0001-8835-9971>

Escudero, Karina; ORCID: <http://orcid.org/0000-0002-0686-3160>

Perez-Brayfield, Marcos; ORCID: <http://orcid.org/0000-0003-1676-530X>

Institution

(Saavedra-Belaunde, Soto-Aviles, Jorge, Perez-Brayfield) Department of Urology, University of Puerto Rico School of Medicine, San Juan, PR, United States (Escudero, Vazquez-Cruz, Perez-Brayfield) HIMA San Pablo, Bayamon, PR, United States

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538.

Single-stage Female Epispadias Repair by Combined Infrasympheal Bladder Neck Plication and Urethrogenitoplasty: A Novel Technique.

Yadav S.S., Agarwal N., Kumar S., Tomar V., Vyas N., Teli R., Badgurjar M.K., Bamaniya M.K.

Embase

Urology. 100 (pp 240-245), 2017. Date of Publication: 01 Feb 2017.

[Article]

AN: 613732103

Objective To evaluate the role of infrasympheal bladder neck plication and suspension from the pubic bone along with urethrogenitoplasty as a single-stage procedure for treating isolated cases of female epispadias. **Materials and Methods** Six patients with female epispadias having grade 3 urinary incontinence were treated through bladder neck plication and suspension from the pubic bone along with urethrogenitoplasty from December 2013 to March 2016. **Results** Of the 6 patients with a mean age of 5.91 years (3-10 years) and a mean postoperative follow-up of 13.17 months (4-29 months), 5 patients stopped wearing diapers and were fully continent with a dry period of more than 3hours from the first week of catheter removal. One patient experienced urine leakage while playing or crying in the initial postoperative period. However, this patient became continent with a dry period of more than 3hours after 4 months of follow-up. All patients had an excellent cosmetic outcome without any considerable postoperative complications. All patients were voiding well without significant postvoid residual urine. **Conclusion** The present technique is simple, safe, and effective for achieving urinary continence in patients with female epispadias.

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Institution

(Yadav, Agarwal, Kumar, Tomar, Vyas, Teli, Bamaniya) Department of Urology, S.M.S Medical College, Jaipur, India (Badgurjar) Department of Surgery, S.M.S Medical College, Jaipur, India

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Year of Publication

2017

539.

Shorter anogenital and anoscrotal distances correlate with the severity of hypospadias: A prospective study.

Cox K., Kyriakou A., Amjad B., O'Toole S., Flett M.E., Welsh M., Ahmed S.F., Cascio S.

Embase

Journal of Pediatric Urology. 13 (1) (pp 57.e1-57.e5), 2017. Date of Publication: 01 Feb 2017.

[Article]

AN: 613422080

Introduction Anogenital distance (AGD) is a recognised marker of in utero androgen action.

Objective This study aimed to evaluate the relationship between severity of hypospadias and AGD. Study design Boys undergoing hypospadias repair in a single tertiary centre between May 2012 and February 16 were included in the study. Anogenital distance was measured from the centre of the anus to the base of the penis, and anoscrotal distance (ASD) from the centre of the anus to the junction between the smooth perineal skin and scrotal skin. Trained paediatric urologists made all measurements using digital callipers. Results Fifty-nine boys with hypospadias and 31 age-matched controls undergoing circumcision (median age 1.37 years, range 1.01-1.96) had AGD and ASD measured under anaesthetic. The patients were divided into two groups, according to hypospadias severity: group 1 - distal penile/subcoronal/glandular (n = 40); and group 2 - perineal/penoscrotal/midshaft (n = 19). The median AGD for controls was 74.0 mm (range 53.2-87.8) and for hypospadias it was 72.3 mm (range 50.7-90.0) (P = 0.816). The

median ASD for controls was 42.3 mm (range 31.0-56.1) and for hypospadias it was 39.4 mm (range 20.7-77.0) ($P = 0.224$). Considering severity of hypospadias, the median AGD for group 1 and group 2 was 73.7 mm (range 50.7-90.0) and 63.3 mm (range 53.6-77.0), respectively ($P < 0.001$). The median ASD was also higher in group 1, at 41.3 mm (range 20.7-65.0), compared to 35.2 mm (range 23.5-77.0) in group 2 ($P = 0.119$) (Summary Fig.). Discussion This study showed that more severe forms of hypospadias are associated with shorter AGD and ASD. These findings agree with two previous studies that identified reduced AGD in boys with hypospadias. However, these studies did not investigate an association with severity of hypospadias. As hypospadias is multifactorial, only a small proportion of cases are thought to be associated with impaired in utero androgen exposure. The shorter AGD in boys with severe hypospadias compared with mild hypospadias would indicate that AGD is a marker of the severity of androgen production. This may also suggest that less severe forms of hypospadias have a different aetiology involving a later stage of development, and that they are not the result of reduced androgen exposure in the male programming window between the 8-14 weeks gestation. Conclusion This study identified that boys with more severe hypospadias are more likely to have a shorter AGD and ASD than boys with mild hypospadias. This may indicate that there is a more profound impairment of in utero androgen action in severe hypospadias.[Figure presented]

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Status

Embase

Author NameID

Cox K.; ORCID: <http://orcid.org/0000-0003-3342-4291> Flett M.E.; ORCID: <http://orcid.org/0000-0001-5296-0423>

Welsh M.; ORCID: <http://orcid.org/0000-0003-1469-0201>

Institution

(Cox, Kyriakou, Ahmed) Developmental Endocrinology Research Group, Child Health, University of Glasgow, Royal Hospital for Children, 1345 Govan Street, Glasgow G51 4TF, United Kingdom

(Amjad, O'Toole, Flett, Cascio) Department of Paediatric Urology, Royal Hospital for Children, 1345 Govan Street, Glasgow G51 4TF, United Kingdom

(Welsh) School of Life Sciences, West Medical Building, University of Glasgow, Glasgow G12 8QQ, United Kingdom

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540.

Qualitative questionnaire on the psychosocial wellbeing of mothers of children with BEEC.

Di Grazia M., Pellizzoni S., Tonegatti L.G., Rigamonti W.

Embase

Journal of Pediatric Urology. 13 (1) (pp 55.e1-55.e6), 2017. Date of Publication: 01 Feb 2017.

[Article]

AN: 612772246

Introduction The bladder exstrophy-epispadias complex (BEEC) represents a spectrum of malformations that affect the anatomical and functional structure of the urogenital system. The parents of patients affected by this condition are subject to particularly stressful situations, such as worrying about their child's health, long hospital stays, concerns about the health and constant need for personal care for their children, that can profoundly compromise the quality of family life. **Objective** The objective of this explorative qualitative study is to evaluate the social situation and the psychological strategies implemented by the mothers of children between 6 and 10 years of age who are affected by BEEC. **Study design** Fourteen mothers of children aged 6-10 years and affected by BEEC (9 boys and 5 girls) were interviewed. Data on the mothers' experiences were collected through semi-structured interviews (Table). **Results** The qualitative analysis of the interviews showed that participants described experiences that were characterised by emotions such as fear and anger. Each mother had implemented a different and, sometimes, dysfunctional strategy in order to cope with the complex situation of the son/daughter. The aspects that most clearly emerged from mothers' descriptions were (1) the traumatic situation at the birth of the baby, (2) the sense of embarrassment concerning the pathological condition as the child was growing and the consequent sense of isolation of the mother, and (3) the fluctuation of feelings towards the multidisciplinary staff, which was sometimes seen as an important source of help and some other times as too destabilising and not helpful at all. **Discussion** The study provided some insight into the psychological and social conditions experienced by mothers of children with BEEC, which could serve as a basis for developing multidisciplinary teams with greater awareness about families living with this condition and better timing in addressing their needs. **Conclusions** Mothers of children with BEEC show emotional and social difficulties. This is a crucial aspect to consider when planning a multidisciplinary approach to the treatment/therapy,

especially considering that children examined in this study are approaching adolescence.[Table presented]

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Status

Embase

Institution

(Di Grazia, Rigamonti) Department of Medical, Surgical and Health Science, University of Trieste, Trieste, Italy (Pellizzoni, Tonegatti) Department of Life Science, University of Trieste, Trieste, Italy

(Rigamonti) Institute for Maternal and Child Health - IRCCS "Burlo Garofolo", Trieste, Italy

Publisher

Elsevier Ltd

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541.

Demographics and co-occurring conditions in a clinic-based cohort with Down syndrome in the United Arab Emirates.

Corder J.P., Al Ahbabi F.J.S., Al Dhaheri H.S., Chedid F.

Embase

American Journal of Medical Genetics, Part A. 173 (9) (pp 2395-2407), 2017. Date of Publication: September 2017.

[Article]

AN: 617259192

The majority of studies describing demographics and co-occurring conditions in cohorts with Down syndrome come from regions outside of the Middle East, mainly from Europe and North America. This paper describes demographics and co-occurring conditions in a hospital-based cohort of individuals with Down syndrome living in the Middle Eastern country of the United Arab Emirates (UAE). The first dedicated Down syndrome clinic in the UAE was established in 2012 at Tawam Hospital in Al Ain. This paper describes a clinic-based cohort of 221 participants over 4

years from the Gulf Down Syndrome Registry, a new Down syndrome database and contact registry created at Tawam Hospital. Key demographic findings include mean maternal age of 37 years, among the highest described in the literature. Sixty-two percent of mothers are >35 years. Over 90% of mothers received post-natal diagnosis of Down syndrome. High sex ratio, parental consanguinity, and large family size also characterize the group. The spectrum of many co-occurring conditions mirrors that of previously described populations, with some notable differences. Cardiovascular malformations are well represented, however, atrioventricular canal is not the most common. Genitourinary conditions are common, as evidenced by 12% of males with hypospadias and 15% with undescended testes. Glucose-6-phosphate dehydrogenase deficiency, alpha thalassemia trait, hypovitaminosis D, and dental caries are common in our cohort. This study describes a large hospital-based group with Down syndrome presenting to a new dedicated Down syndrome clinic in the UAE, highlighting unique demographic and co-occurring conditions found in that population.

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Status

Embase

Author NameID

Corder, Jennifer Price; ORCID: <http://orcid.org/0000-0002-3629-566X>

Institution

(Corder, Al Ahbabi, Al Dhaheri) Department of Pediatrics, Tawam Hospital in Affiliation With Johns Hopkins International, Al Ain, United Arab Emirates (Chedid) Department of Neonatology, Al Jalila Children's Specialty Hospital, Dubai, United Arab Emirates

Publisher

Wiley-Liss Inc. (E-mail: info@wiley.com)

Year of Publication

2017

542.

Results of tubularized urethral plate urethroplasty in Megameatus Intact Prepuce.

Bhat M., Bhat A., Singh V.

Embase

Indian Journal of Urology. 33 (4) (pp 315-318), 2017. Date of Publication: October-December 2017.

[Article]

AN: 618518132

Objective: The megameatus variant of anterior hypospadias with an intact complete foreskin occurs in approximately 1%-3% of hypospadias. Hence, the objective of the study was to evaluate the results of tubularized urethral plate urethroplasty (TUPU) in megameatus intact prepuce (MIP).

Material(s) and Method(s): A retrospective study (June 1996-June 2015) of MIP from our hypospadias registry was conducted. All patients with megameatus, either with an intact prepuce or with one previously removed, were included in the study. Case sheets of clinical records, investigations, clinical photographs, and videos were reviewed. Patients were classified into, glanular, coronal, subcoronal, and distal penile. TUPU were done. Patients were called for follow-up at 1, 3, 6, and 12 months postoperatively, and then yearly for the assessment of the cosmetic appearance and fistula, meatal stenosis, or other complications.

Result(s): Of 1026 patients with hypospadias, we identified 13 cases of megameatus variant of hypospadias; three of the 13 had been circumcized previously. Glanular approximation was done for the one patients of the glanular variant, and another had frenuloplasty. These two patients were excluded from the study. Incision in the inner preputial skin was closed in 10 patients to have an intact prepuce. Follow-up period varied from 6 months to 4 years (median follow-up 2 1/2 years). None of the patients developed complications such as fistula, meatal stenosis, and/or wound dehiscence.

Conclusion(s): Surgical correction of MIP in the era of increased cosmetic awareness is justified. Excellent results are obtained with TUPU and along with spongioplasty and frenuloplasty because of availability of wide urethral plate and well-developed spongiosum in these patients. TUPU should be the preferred procedure in cases of MIP.

Status

Embase

Institution

(Bhat, Singh) Department of Urology, Dr. S.N. Medical College, Jodhpur, Rajasthan, India (Bhat)

Department of Surgery, M.G. Medical College, Jaipur, Rajasthan, India

(Bhat) Department of Surgery, S.P. Medical College, Bikaner, Rajasthan, India

Publisher

Medknow Publications (B9, Kanara Business Centre, off Link Road, Ghatkopar (E), Mumbai 400 075, India)

Year of Publication

543.

Comparison of lingual mucosa and buccal mucosa grafts used in inlay urethroplasty in failed hypospadias of pre-pubertal boys in a Chinese group.

Hongyong J., Shuzhu C., Min W., Weijing Y., Yidong L.

Embase

PLoS ONE. 12 (8) (no pagination), 2017. Article Number: e0182803. Date of Publication: August 2017.

[Article]

AN: 617804039

Objective The purpose of this study was to compare the outcomes of the buccal mucosa and lingual mucosa used in children who received multiple failed hypospadias surgeries. **Method** We conducted a retrospective study of 62 children who received buccal or lingual mucosa graft urethroplasty in our hospital between 2012 and 2015. The ages ranged from 3.5-11 y. All cases included multiple failed hypospadias procedures, and the subjects received previous operations 2-3 times. All patients underwent one-stage operations. Thirty-three cases were treated with lingual mucosa grafts. The patient ages ranged from 3.5 to 11 y (median 7.5 y), and they had previous operations 2-3 times (mean 2.8+/-0.7). Grafts ranged from lengths of 2-6 cm (mean 5.1+/-0.46 cm) and widths of 0.5-1.5 cm (mean 1.2+/- 0.16 cm). Our follow-up was 5 to 12 m (mean 8.3+/-1.2 m). Twenty-nine cases were treated with buccal mucosa grafts. The patient ages ranged from 4 to 9.2 y (median 7.0 y), and they had previous operations 2-3 times (mean 2.5+/- 0.2). Grafts ranged from lengths of 2-5.3 cm (mean 4.9+/- 0.28 cm) and widths of 0.5-1.5 cm (mean 1.0+/-0.11 cm). Our follow-up was 5 to 12 m (mean 7.9+/-0.5 m). The results were tested with SPSS 18.0. The rates of complications were compared by a chi-square test, and pre-operative conditions were compared by t test. **Results** For the outcomes of the two groups, there was no significant difference between the groups in terms of age, preoperative surgery time, and the length and width of the grafts ($p>0.05$). For the lingual mucosa graft group, fistula: 2/33 (6.0%), stricture: 1/33(3.0%), ventral curvature: 2/33(6.0%), complications: 5/33(15.0%), success rate: 28/33(84.8%), Hose score: 14.34+/-0.95, peak flow: 6.5 ml/s-12.0 ml/s, and mean peak flow: 9.3+/-0.4 ml/s. For the buccal mucosa graft group, fistula: 2/29(6.8%), stricture: 2/29(6.8%), ventral curvature: 1/29 (3.4%), complication rate: 5/29(17.0%), success rate: 24/29 (83.0%), Hose

score: 14.28 +/-1.03, peak flow: 6.5 ml/s-12.0 ml/s, and mean peak flow: 9.2+/-0.2 ml/s. There were no differences between the two groups for overall success, complication rates, peak flow, and the Hose scores(P>0.05). Conclusion The lingual mucosal graft and the buccal mucosa graft both achieved good outcomes, and the lingual mucosa graft made up for the shortcomings of the buccal mucosa graft, which provided a reliable way to treat the multiple failed hypospadias surgeries in pre-pubertal boys.

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Status

Embase

Institution

(Hongyong) Urology Center, XinJiang Uygur Autonomous Region People's Hospital, Urumqi, China (Shuzhu, Min, Weijing, Yidong) Department of Urology, Ren Ji Hospital, School of Medicine, Shanghai Jiao Tong University, Shanghai, China

Publisher

Public Library of Science (E-mail: plos@plos.org)

Year of Publication

2017

544.

Maternal Stressors and Social Support and Risks of Delivering Babies with Gastroschisis or Hypospadias.

Carmichael S.L., Ma C., Tinker S., Shaw G.M.

Embase

American Journal of Epidemiology. 185 (12) (pp 1240-1246), 2017. Date of Publication: 15 Jun 2017.

[Article]

AN: 617272906

We examined the association of maternal stressful life events and social support with risks of gastroschisis and hypospadias, using data from the National Birth Defects Prevention Study, a population-based case-control study of US births taking place in 2006-2011. We examined maternal self-reports of 7 life events and 3 sources of social support during the periconceptional period among mothers of 593 gastroschisis cases, 1,142 male hypospadias cases, and 4,399 nonmalformed controls. Responses to the questions on stressful life events were summed to form an index (higher is worse), as were responses to questions on social support (higher is better). We used logistic regression to estimate adjusted odds ratios and 95% confidence intervals. The adjusted odds ratios for gastroschisis for a 4-point increase in the stress index were 3.5 (95% confidence interval (CI): 2.6, 4.8) among nonteenage mothers (age ≥ 20 years) and 1.0 (95% CI: 0.5, 1.7) among teenage mothers (age < 20 years). The odds ratio for hypospadias (among all mothers) was 0.8 (95% CI: 0.7, 1.1). Adjusted odds ratios for a social support score of 3 (versus 0) in the 3 respective groups were 0.6 (95% CI: 0.4, 1.0), 1.0 (95% CI: 0.5, 2.3), and 0.6 (95% CI: 0.4, 0.9). Given the lack of prior research on these outcomes and stress, results should be interpreted with caution.

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Institution

(Carmichael, Ma, Shaw) Department of Pediatrics, School of Medicine, Stanford University, 1265 Welch Road, Stanford, CA 94305-5415, United States (Tinker) National Center on Birth Defects and Developmental Disabilities, Centers for Disease Control and Prevention, Atlanta, GA, United States

Publisher

Oxford University Press (E-mail: jnl.info@oup.co.uk)

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2017

Comparative safety of anti-epileptic drugs during pregnancy: A systematic review and network meta-analysis of congenital malformations and prenatal outcomes.

Veroniki A.A., Cogo E., Rios P., Straus S.E., Finkelstein Y., Kealey R., Reynen E., Soobiah C., Thavorn K., Hutton B., Hemmelgarn B.R., Yazdi F., D'Souza J., MacDonald H., Tricco A.C.

Embase

BMC Medicine. 15 (1) (no pagination), 2017. Article Number: 95. Date of Publication: 05 May 2017.

[Article]

AN: 615882610

Background: Pregnant women with epilepsy frequently experience seizures related to pregnancy complications and are often prescribed anti-epileptic drugs (AEDs) to manage their symptoms. However, less is known about the comparative safety of AED exposure in utero. We aimed to compare the risk of congenital malformations (CMs) and prenatal outcomes of AEDs in infants/children who were exposed to AEDs in utero through a systematic review and Bayesian random-effects network meta-analysis.

Method(s): MEDLINE, EMBASE, and Cochrane CENTRAL were searched from inception to December 15, 2015. Two reviewers independently screened titles/abstracts and full-text papers for experimental and observational studies comparing mono- or poly-therapy AEDs versus control (no AED exposure) or other AEDs, then abstracted data and appraised the risk of bias. The primary outcome was incidence of major CMs, overall and by specific type (cardiac malformations, hypospadias, cleft lip and/or palate, club foot, inguinal hernia, and undescended testes).

Result(s): After screening 5305 titles and abstracts, 642 potentially relevant full-text articles, and 17 studies from scanning reference lists, 96 studies were eligible ($n = 58,461$ patients). Across all major CMs, many AEDs were associated with higher risk compared to control. For major CMs, ethosuximide (OR, 3.04; 95% CrI, 1.23-7.07), valproate (OR, 2.93; 95% CrI, 2.36-3.69), topiramate (OR, 1.90; 95% CrI, 1.17-2.97), phenobarbital (OR, 1.83; 95% CrI, 1.35-2.47), phenytoin (OR, 1.67; 95% CrI, 1.30-2.17), carbamazepine (OR, 1.37; 95% CrI, 1.10-1.71), and 11 polytherapies were significantly more harmful than control, but lamotrigine (OR, 0.96; 95% CrI, 0.72-1.25) and levetiracetam (OR, 0.72; 95% CrI, 0.43-1.16) were not.

Conclusion(s): The newer generation AEDs, lamotrigine and levetiracetam, were not associated with significant increased risks of CMs compared to control, and were significantly less likely to be associated with children experiencing cardiac malformations than control. However, this does not mean that these agents are not harmful to infants/children exposed in utero. Counselling is advised concerning teratogenic risks when the prescription is written for a woman of childbearing age and before women continue with these agents when considering pregnancy, such as switching from polytherapy to monotherapy with evidence of lower risk and avoiding AEDs, such

as valproate, that are consistently associated with CMs. These decisions must be balanced against the need for seizure control. Systematic Review Registration: PROSPERO

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Institution

(Veroniki, Cogo, Rios, Straus, Kealey, Reynen, Soobiah, Yazdi, D'Souza, MacDonald, Tricco) St. Michael's Hospital, Knowledge Translation Program, Li Ka Shing Knowledge Institute, 209 Victoria Street, East Building, Toronto, ON M5B 1W8, Canada (Straus) University of Toronto, Department of Geriatric Medicine, 27 King's College Circle, Toronto, ON M5S 1A1, Canada (Finkelstein) The Hospital for Sick Children, 555 University Avenue, Toronto, ON M5G 1X8, Canada

(Finkelstein) University of Toronto, Department of Paediatrics, 172 St. George Street, Toronto, ON M5R 0A3, Canada

(Finkelstein) University of Toronto, Department of Pharmacology and Toxicology, Medical Sciences Building, 1 King's College Circle, Toronto, ON M5S 1A8, Canada

(Soobiah) University of Toronto, Institute for Health Policy Management and Evaluation, 4th Floor, 155 College Street, Toronto, ON M5T 3M6, Canada

(Thavorn, Hutton) University of Ottawa, School of Epidemiology, Public Health and Preventive Medicine, Faculty of Medicine, Roger-Guindon Building, 451 Smyth Road, Ottawa, ON K1H 8M5, Canada

(Thavorn) The Ottawa Hospital, Clinical Epidemiology Program, Ottawa Hospital Research Institute, 501 Smyth Road, Ottawa, ON K1H 8L6, Canada

(Thavorn) Institute of Clinical and Evaluative Sciences (ICES uOttawa), 1053 Carling Ave, Ottawa, ON K1Y 4E9, Canada

(Hutton) Center for Practice Changing Research, The Ottawa Hospital - General Campus, Ottawa Hospital Research Institute, 501 Smyth Road, PO Box 201B, Ottawa, ON K1H 8L6, Canada

(Hemmelgarn) University of Calgary, Departments of Medicine and Community Health Sciences, TRW Building, 3rd Floor, 3280 Hospital Drive NW, Calgary, AB T2N 4Z6, Canada

(Tricco) University of Toronto, Epidemiology Division, Dalla Lana School of Public Health, 6th Floor, 155 College Street, Toronto, ON M5T 3M7, Canada

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BioMed Central Ltd. (E-mail: info@biomedcentral.com)

Year of Publication

2017

546.

Placental pathology and hypospadias.

Chen Y., Sun L., Geng H., Lei X., Zhang J.

Embase

Pediatric Research. 81 (3) (pp 489-495), 2017. Date of Publication: 01 Mar 2017.

[Article]

AN: 615090458

Background: Studies have shown that hypospadias is associated with placenta-mediated pregnancy complication (PMPC). The role of placental lesions is still unclear. We aimed to examine the association between hypospadias and placental pathology, and the effect of PMPC.

Method(s): Using data from the US Collaborative Perinatal Project in 1959-1966, we identified 15,780 male subjects (167 hypospadias) for analysis. Detailed placental examinations were conducted following a standard protocol. Subjects were divided into two groups according to whether they had PMPC, including small-for-gestational-age, pre-eclampsia/eclampsia or placental abruption. Logistic regression models were used to explore the association.

Result(s): The prevalence of hypospadias was two times higher in subjects with PMPC than those without. Compared to pregnancies with PMPC but no hypospadias, those with both PMPC and hypospadias had significant higher prevalence of placental lesions, such as low placental weight, vascular lesions, villous lesions, and membranous insertion of cord (adjusted odds ratio (OR) ranging from 2.6 to 5.2) after adjusting for potential confounders. In subjects without PMPC, no significant difference of placental pathology was found between those with or without hypospadias.

Conclusion(s): About one third of hypospadias cases were complicated with PMPC and had a higher risk of placental lesions, suggesting heterogeneity of hypospadias etiology and mechanisms.

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Embase

Institution

(Chen, Lei, Zhang) MOE-Shanghai Key Laboratory of Children's Environmental Health, Xinhua Hospital Affiliated to Shanghai Jiao Tong University, School of Medicine, Shanghai, China

(Chen) Department of Neonatology, Xinhua Hospital Affiliated to Shanghai Jiao Tong University, School of Medicine, Shanghai, China

(Sun) Department of Fetal Medicine, First Maternity and Infant Hospital Affiliated to Shanghai Tongji University, Shanghai, China

(Geng) Department of Pediatric Surgery, Xinhua Hospital Affiliated to Shanghai Jiao Tong University, School of Medicine, Shanghai, China

(Zhang) Shanghai Jiao Tong University School of Public Health, Shanghai, China

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2017

547.

High prevalence of same-sex twins in patients with cloacal exstrophy: Support for embryological association with monozygotic twinning.

Fullerton B.S., Sparks E.A., Hall A.M., Velazco C.S., Modi B.P., Lund D.P., Jaksic T., Hendren W.H.

Embase

Journal of Pediatric Surgery. 52 (5) (pp 807-809), 2017. Date of Publication: May 2017.

[Article]

AN: 614395601

Purpose Previous studies have hypothesized that cloacal exstrophy may be caused by errors early in embryological development related to monozygotic twinning. This study reports the prevalence of twins in a large cohort of patients with cloacal exstrophy. **Methods** Patients with cloacal exstrophy treated 1974-2015 were reviewed for reports of multiple gestation or conjoined twinning. The genetic sex of the patient and their twin, and any mention of anomaly in the twin were recorded. Neither placental exam nor genetic testing results were available to definitively determine zygosity. **Results** Of 71 patients, 10 had a live born twin (14%), all of whom were of the same genetic sex as the affected patient. One additional patient's twin suffered intrauterine fetal

demise, and another patient had a conjoined heteropagus twin. None of the twins were affected by exstrophy-epispadias complex. The rate of twin birth in this cohort was 4.4-7.7 higher than that reported by the Centers for Disease Control in the general population time period ($P < 0.001$), with a striking preponderance of same-sex pairs. Conclusions The highly significant prevalence of same-sex twin pairs within this cohort supports the hypothesis that the embryogenesis of cloacal exstrophy may be related to errors in monozygotic twinning. Level of evidence 2b

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PMID

28202184 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=28202184>]

Status

Embase

Institution

(Fullerton, Sparks, Hall, Velazco, Modi, Jaksic, Hendren) Department of Surgery, Boston Children's Hospital, 300 Longwood Ave, Boston, MA 02115, United States (Fullerton, Sparks, Velazco, Modi, Jaksic) Center for Advanced Intestinal Rehabilitation, Boston Children's Hospital, 300 Longwood Ave, Boston, MA 02115, United States

(Lund) Stanford University Department of Surgery and Lucile Packard Children's Hospital, 725 Welch Road, Palo Alto, CA 94304, United States

Publisher

W.B. Saunders

Year of Publication

2017

548.

Sexuality and fertility in men with hypospadias; improved outcome.

Ortqvist L., Fossum M., Andersson M., Nordenstrom A., Frisen L., Holmdahl G., Nordenskjold A.

Embase

Andrology. 5 (2) (pp 286-293), 2017. Date of Publication: 01 Mar 2017.

[Article]

AN: 613858875

The aim of this study was to investigate sexual function and fertility in adult men born with hypospadias. Patients born with hypospadias, age-matched controls, and a group of circumcised

men completed a questionnaire constructed to reflect their psychosexual situation and fertility. Core gender identity, sexual orientation, and gender role behavior was also assessed. 167 patients [63% distal, 24% mid shaft and 13% proximal, mean age 34 (19-54) years], 169 controls from the general population [mean age 33 (19-48) years] and 47 controls circumcised because of phimosis (mean age 26 [19-44]) participated and completed the questionnaire. There were no differences in having a partner, reported fertility, age at sexarche (mean age 17.8), number of sex partners or sexual interest between the patients and controls. More patients than controls reported anejaculation. Reported glanular sensitivity was lower in hypospadias patients and circumcised controls compared with non-circumcised controls. The odds of being satisfied with their sexual life increased with a higher penile perception score in patients (OR = 1.54, p = 0.01). There was no association with penile length. Sexual orientation, core gender identity and gender role behavior were sex-typical in both patients and controls. Patients with proximal hypospadias had a lower reported fertility, experienced anejaculation more often, and were less satisfied with their sexual life. Men born with hypospadias have a good long-term outcome concerning sexual function and fertility. Men born with proximal hypospadias have a more impaired outcome concerning both sexual function and fertility. As satisfaction with genital appearance is important for sexual life satisfaction, clinical, and psychological follow-up into adulthood is especially important in boys born with proximal hypospadias.

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Institution

(Ortqvist, Fossum, Nordenstrom, Nordenskjold) Department of Women's and Children's Health and Center for Molecular Medicine, Karolinska Institutet, Stockholm, Sweden (Ortqvist, Fossum, Nordenskjold) Unit of Pediatric Surgery, Astrid Lindgren Children's Hospital, Karolinska University Hospital, Stockholm, Sweden

(Andersson, Holmdahl) Sahlgrenska Academy, Women's and Children's Health, Gothenburg, Sweden

(Andersson, Holmdahl) Department of Pediatric Surgery, Queen Silvia's Children's Hospital, Gothenburg, Sweden

(Nordenstrom) Department of Pediatric Endocrinology, Astrid Lindgren Children's Hospital, Karolinska University Hospital, Stockholm, Sweden

(Frisen) Department of Clinical Neuroscience, Karolinska Institutet, Stockholm, Sweden

(Frisen) Child and Adolescent Psychiatry Research Center, Karolinska Institutet, Stockholm, Sweden

Publisher

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Year of Publication

2017

549.

Are carriers of CYP21A2 mutations less vulnerable to psychological stress? A population-based national cohort study.

Nordenstrom A., Butwicka A., Linden Hirschberg A., Almqvist C., Nordenskjold A., Falhammar H., Frisen L.

Embase

Clinical Endocrinology. 86 (3) (pp 317-324), 2017. Date of Publication: 01 Mar 2017.

[Article]

AN: 613255095

Background: Congenital adrenal hyperplasia (CAH) is one of the most common monogenic autosomal recessive disorders with an incidence of one in 15 000. About one in 70 individuals in the general population are carriers of a severe CYP21A2 mutation. It has been suggested that this confers a survival advantage, perhaps as a result of increased activity in the hypothalamic-pituitary-adrenal axis. We investigated vulnerability to psychological stress in obligate carriers. Method(s): The Swedish CAH Registry encompasses more than 600 patients. Parents, that is obligate carriers of the CYP21A2 mutation, were identified through the Multigeneration Register. The diagnosis of the child was used as the psychological stressor. Psychiatric diagnoses before and after the birth of a child with CAH were compared to those of controls derived from (i) the general population, (ii) parents of children with hypospadias and (iii) parents of children with diabetes mellitus type 1 (T1DM).

Result(s): Parents of children with CAH had less risk of being diagnosed with any psychiatric disorder (OR, 0.6), an affective disorder (OR, 0.5) or substance misuse (OR, 0.5) after the diagnosis of the child, compared to the general population. Their risk was also decreased compared to parents of a child with hypospadias (OR, 0.6, 0.4 and 0.2, respectively) and parents of a child with T1DM (OR 0.7, 0.6 and 0.2, respectively). The CYP21A2 carriers had a lower risk of developing mood and stress-related disorders after the diagnosis of the child.

Conclusion(s): Obligate CYP21A2 carriers had a reduced risk of a psychiatric diagnosis and were less vulnerable to a psychologically stressful situation, at least with respect to receiving a psychiatric diagnosis. This indicates a better ability to cope with psychological stress among heterozygous carriers of severe CYP21A2 mutations, which may contribute to the apparent survival advantage.

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Institution

(Nordenstrom, Linden Hirschberg, Nordenskjold) Department of Women's and Children's Health, Karolinska Institutet, Stockholm, Sweden (Nordenstrom) Department of Paediatric Endocrinology, Astrid Lindgren Children's Hospital, Karolinska University Hospital, Stockholm, Sweden

(Butwicka, Almqvist) Department of Medical Epidemiology and Biostatistics, Karolinska Institutet, Stockholm, Sweden

(Butwicka) Department of Child Psychiatry, Medical University of Warsaw, Warsaw, Poland

(Linden Hirschberg) Department of Obstetrics and Gynaecology, Karolinska University Hospital, Stockholm, Sweden

(Almqvist) Lung and Allergy Unit, Astrid Lindgren Children's Hospital, Karolinska University Hospital, Stockholm, Sweden

(Nordenskjold) Centre for Molecular Medicine, Karolinska Institutet, Stockholm, Sweden

(Nordenskjold) Paediatric Surgery, Astrid Lindgren Children's Hospital, Karolinska University Hospital, Stockholm, Sweden

(Falhammar) Department of Endocrinology, Metabolism and Diabetes, Karolinska University Hospital, Stockholm, Sweden

(Falhammar) Department of Molecular Medicine and Surgery, Karolinska Institutet, Stockholm, Sweden

(Frisen) Department of Clinical Neuroscience, Karolinska Institutet, Stockholm, Sweden

(Frisen) Child and Adolescent Psychiatry Research Centre, Stockholm, Sweden

Publisher

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Year of Publication

2017

550.

Maternal exposure to domestic hair cosmetics and occupational endocrine disruptors is associated with a higher risk of hypospadias in the offspring.

Haraux E., Braun K., Buisson P., Stephan-Blanchard E., Devauchelle C., Ricard J., Boudailliez B., Tourneux P., Gouron R., Chardon K.

Embase

International Journal of Environmental Research and Public Health. 14 (1) (no pagination), 2017.
Article Number: 27. Date of Publication: January 2017.

[Article]

AN: 613922305

Pregnant women are exposed to various chemical products at home and at work. Some of these products contain endocrine-disrupting chemicals (EDCs) such as cosmetics, pesticides, industrial chemicals, heavy metals, plastics or medications that could alter sexual differentiation and increase the risk of hypospadias. We evaluated maternal occupational and household exposures that could constitute risk factors for hypospadias. From 2011 to 2014, we enrolled 57 full-term newborns with hypospadias and three randomly selected controls per case (162 control newborns), matched for gestational age, from 11 maternity units in Picardy, France. Neonatal and parental data were collected at birth (personal characteristics, maternal lifestyle, and medical history). Maternal occupational exposure was assessed by a job-exposure matrix for EDCs from a job history questionnaire completed by mothers. Odds ratios (OR) and 95% confidence intervals (CI) were calculated with univariate and multivariable logistic regression, and adjusted for relevant covariates. Multivariate analysis showed a strong association between hypospadias and potential maternal occupational exposure to EDCs and maternal household use of hair cosmetics (OR 6.1, 95% CI: 1.1-34.9, OR: 9.6, 95% CI: 1.4-66.1, respectively). Our results suggest that maternal occupational exposure to EDCs is a risk factor for hypospadias and suggests a possible influence of household use of hair cosmetics during early pregnancy on the incidence of hypospadias in the offspring. A larger study with more accurate exposure assessment should evaluate the impact of EDCs in hair cosmetics on the incidence of hypospadias.

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Status

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Institution

(Haraux, Buisson, Ricard, Gouron) Department of Paediatric Surgery, Amiens University Hospital, Amiens 80054, France (Braun, Boudailliez) Department of Paediatrics, Amiens University

Hospital, Amiens 80054, France

(Stephan-Blanchard, Chardon) PeriTox-INERIS Laboratory, Jules Verne University of Picardy, Amiens 80054, France

(Devauchelle) Department of Paediatrics, Creil Hospital, Creil 60100, France

(Tourneux) Department of Paediatric Intensive Care Unit, Amiens University Hospital, Amiens 80054, France

Publisher

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Year of Publication

2017

551.

The role of tunica vaginalis flap in staged repair of hypospadias.

Kadian Y.S., Singh M., Rattan K.N.

Embase

Asian Journal of Urology. 4 (2) (pp 107-110), 2017. Date of Publication: April 2017.

[Article]

AN: 614252200

Objective The surgical repair of hypospadias is done in two stages in a select group of patients with severe anomaly. The first stage (I) procedure consists of correction of penile shaft curvature and second stage (II) repair involves the creation of a neourethra. This neourethra needs a cover of an intermediate layer in order to have good functional and cosmetic results. Among the various local flaps, tunica vaginalis flap is a good option for the use as an intermediate layer. **Methods** We have managed 22 patients of chordee with hypospadias by staged repair. In Stage I, chordee correction was done by dividing the urethral plate and covering the penile shaft with dorsal prepuce flaps. In Stage II, a neourethra was created and covered with tunica vaginalis flap either through the same incision (14/22) or via a subcutaneous tunnel (8/22). An indwelling catheter was kept for 10 to 12 days. **Results** Eighteen (81.8%) patients had successful functional and cosmetic

repair. Two patients (9.1%) had urethrocutaneous fistula of which one healed on subsequent dilatation while the other one (4.5%) needed repair. Overall fistula formation rate was 4.5%. In two patients, the external urinary meatus could be made upto subglanular or coronal level. Conclusion Staged repair of chordee with hypospadias is valuable in selected group of patients and tunica vaginalis flap is an excellent intermediate layer to cover the neourethra. However preoperative counseling is particularly essential in patients where the external urinary meatus can be created at coronal or subglanular level.

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Institution

(Kadian, Rattan) Department of Paediatric Surgery, Pt B D Sharma Post Graduate Institute of Medical Sciences, Rohtak, Haryana, India (Singh) Department of Surgery, Pt B D Sharma Post Graduate Institute of Medical Sciences, Rohtak, Haryana, India

Publisher

Editorial Office of Asian Journal of Urology (E-mail: ajurology@smmu.edu.cn)

Year of Publication

2017

552.

Management of 220 adolescents and adults with complications of hypospadias repair during childhood.

Howe A.S., Hanna M.K.

Embase

Asian Journal of Urology. 4 (1) (pp 14-17), 2017. Date of Publication: 01 Jan 2017.

[Review]

AN: 613898277

Objective The goal of hypospadias repair is to achieve normal voiding and good penile cosmesis with minimal complications. Some urethroplasties deteriorate from childhood to adolescence and late stage failures have been reported. We report our experience with adult patients who have had a previous repair during childhood and present with a late complication. Methods We reviewed the records of 220 patients aged 15-39 years old with a history of hypospadias repair

who presented to our clinic. Forty-five patients with chordee, 39 with urethral strictures, 11 urethral fistulae, five with hairy urethras, three with urethral diverticula, and 117 patients with an abnormal glans or subterminal meatus were repaired. Results Median follow-up was 14 months. Two patients had persistent chordee. Island skin flap urethroplasty afforded one patient with a urethral fistula and another with a recurrent urethral stricture, while the buccal mucosa group had one fistula which healed spontaneously and two recurrent strictures. For the patients undergoing glanular repairs, seven had dehiscence or breakdown of the repair. All other operations were successful. Conclusion Complications of childhood hypospadias repair may present later in life as some urethroplasties deteriorate with time. We now recommend to parents of children with repaired proximal hypospadias to come for follow-up as their child transitions to adolescence.

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Embase

Institution

(Howe, Hanna) Division of Pediatric Urology, Cohen Children's Medical Center of New York, Hofstra Northwell School of Medicine, Long Island, NY, United States (Hanna) Department of Urology, Weill Cornell Medical Center, New York, NY, United States

Publisher

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Year of Publication

2017

553.

History and evolution of the use of oral mucosa for urethral reconstruction.

Barbagli G., Balo S., Montorsi F., Sansalone S., Lazzeri M.

Embase

Asian Journal of Urology. 4 (2) (pp 96-101), 2017. Date of Publication: April 2017.

[Review]

AN: 613625089

We report here the history and evolution of the use of oral mucosa in reconstructive urethral surgery since it was first used for urethroplasty in 1894. Since that time, many authors have contributed to develop, improve and popularize the use of oral mucosa as a substitute material.

Paediatric urologists should be considered pioneers on the use of oral mucosa as they used it to repair primary and failed hypospadias. The use of oral mucosa to repair penile and bulbar urethral strictures was described, for the first time, in 1993. Important evolutions in the technique for harvesting oral mucosa from the cheek were reported in 1996. Today, oral mucosa is considered the gold standard material for any type of anterior urethroplasty in a one- or two-stage repair due to its biological and structural characteristics that make it a highly versatile that is adaptable to any environment required by the reconstructive urethral surgery. As the future approaches, tissue engineering techniques will provide patients with new materials originating from the oral epithelial mucosal cells, which are cultured and expanded into a scaffold. However, the path to reach this ambitious objective is still long and many difficulties must be overcome along the way.

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Institution

(Barbagli, Balo) Centro Chirurgico Toscano, Arezzo, Italy (Montorsi) Division of Oncology/Unit of Urology, IRCCS Ospedale San Raffaele, Milan, Italy

(Sansalone) Department of Experimental Medicine and Surgery, University of Tor Vergata, Rome, Italy

(Lazzeri) Department of Urology, Istituto Clinico Humanitas IRCCS, Clinical and Research Hospital, Rozzano, Italy

Publisher

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Year of Publication

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554.

Late surgical correction of hypospadias increases the risk of complications: a series of 501 consecutive patients.

Garnier S., Maillet O., Cereda B., Ollivier M., Broussous S., Lopez C., Paris F., Philibert P., Amouroux C., Jeandel C., Coffy A., Gaspari L., Daures J.P., Sultan C., Kalfa N.

Embase

BJU International. 119 (6) (pp 942-947), 2017. Date of Publication: June 2017.

[Article]

AN: 614328000

Objectives: To evaluate the outcomes of hypospadias surgery according to age and to determine if some complications are age-related.

Patients and Methods: This retrospective study was based on 722 boys with hypospadias undergoing primary repair. A total of 501 boys underwent urethroplasty and were included in the study. Complications requiring an additional procedure (stenosis, fistula, dehiscence, relapse of curvature, urethrocele) were included in the analysis, as well as healing problems, infections, haematomas and detrusor-sphincter dyssynergy. Logistic regression analysis was performed.

Result(s): Hypospadias was anterior in 63.1%, mid-penile in 20.5%, posterior in 8.4% and scrotal in 7.9% of the boys. The median (range) age was 4 (1-16) years. The overall rates of re-intervention and complications were 22.8% and 36.2%, respectively. Age >2 years was a significant predictor of complications ($P = 0.002$, odds ratio 1.98 [95% confidence interval 1.26-3.13]). Some periods of time appeared to be associated with a specific complication: dyssynergy was more common between the ages of 24 and 36 months (12.5 vs 3.6%; $P = 0.01$) and healing problems were more common in boys aged >13 years (1.5 vs 28.5%; $P = 0.06$).

Conclusion(s): Delayed surgery may be detrimental for patients. Factors related to age may influence the rate of complications. After the age of 2 years, urethral surgery may interfere with the normal toilet-training process. During puberty, endogenous testosterone may alter healing. Even if no specific data exist for severe hypospadias, it may be prudent to continue to advocate early surgery in patients with disorders of sex development.

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Institution

(Garnier, Maillet, Cereda, Ollivier, Jeandel, Broussous, Lopez, Kalfa) Department of Paediatric Surgery and Urology, Lapeyronie Hospital, CHU Montpellier, Montpellier, France (Paris,

Amouroux, Jeandel, Sultan) Department of Paediatric Endocrinology, Arnaud de Villeneuve Hospital, CHU Montpellier, Montpellier, France

(Paris, Philibert, Gaspari, Sultan, Kalfa) Department of Endocrinology, Lapeyronie Hospital, CHU Montpellier, Montpellier, France

(Coffy, Daures) Institute of Clinical Research, University of Montpellier, Montpellier, France

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Year of Publication

2017

555.

Experimentally induced testicular dysgenesis syndrome originates in the masculinization programming window.

van den Driesche S., Kilcoyne K.R., Wagner I., Rebourcet D., Boyle A., Mitchell R., McKinnell C., Macpherson S., Donat R., Shukla C.J., Jorgensen A., Meyts E.R.-D., Skakkebaek N.E., Sharpe R.M.

Embase

JCI insight. 2 (6) (pp e91204), 2017. Date of Publication: 23 Mar 2017.

[Article]

AN: 626580861

The testicular dysgenesis syndrome (TDS) hypothesis, which proposes that common reproductive disorders of newborn and adult human males may have a common fetal origin, is largely untested. We tested this hypothesis using a rat model involving gestational exposure to dibutyl phthalate (DBP), which suppresses testosterone production by the fetal testis. We evaluated if induction of TDS via testosterone suppression is restricted to the "masculinization programming window" (MPW), as indicated by reduction in anogenital distance (AGD). We show that DBP suppresses fetal testosterone equally during and after the MPW, but only DBP exposure in the MPW causes reduced AGD, focal testicular dysgenesis, and TDS disorders (cryptorchidism, hypospadias, reduced adult testis size, and compensated adult Leydig cell failure). Focal testicular dysgenesis, reduced size of adult male reproductive organs, and TDS disorders and their severity were all strongly associated with reduced AGD. We related our findings to human TDS cases by demonstrating similar focal dysgenetic changes in testes of men with preinvasive germ cell neoplasia (GCNIS) and in testes of DBP-MPW animals. If our results are translatable to humans, they suggest that identification of potential causes of human TDS disorders should focus on exposures during a human MPW equivalent, especially if negatively associated with offspring AGD.

PMID

28352662 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=28352662>]

Institution

(van den Driesche, Kilcoyne, Wagner, Rebourcet, Boyle, Mitchell, McKinnell, Macpherson, Sharpe) MRC Centre for Reproductive Health, Queen's Medical Research Institute, University of Edinburgh, Edinburgh, United Kingdom (Donat, Shukla) Edinburgh Urological Cancer Group, Department of Urology, Western General Hospital, Edinburgh, United Kingdom (Jorgensen, Meyts, Skakkebaek) Department of Growth & Reproduction, Copenhagen University Hospital (Rigshospitalet), Copenhagen, Denmark

Publisher

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Year of Publication

2017

556.

Comparacion entre la uretroplastia tubularizada con incision de la placa uretral y las tecnicas de Uretroplastia con colgajo pediculado para la reparacion del hipospadias proximal primario, Comparison of tubularized incised plate urethroplasty and onlay island flap urethroplasty techniques in the repair of primarily proximal hypospadias.

Aydogmus Y., Bagbanci S., Demirbas A., Hascicek A.M.

Embase

Archivos espanoles de urologia. 70 (8) (pp 740-745), 2017. Date of Publication: 01 Oct 2017.

[Article]

AN: 627015241

PMID

28976350 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=28976350>]

Institution

(Aydogmus) Department of Urology. Etimesgut Military Hospital. Ankara. Turkey (Bagbanci) Department of Urology. Ahi Evran University School of Medicine. Kirsehir. Turkey (Demirbas, Hascicek) Department of Urology. Ankara Training and Research Hospital. Ankara. Turkey

Publisher

NLM (Medline)

Year of Publication

2017

557.

Influence of Dartos Tissue and the Androgen Receptor in Congenital Penile Malformations:
Opening New Horizons in Research with Clinical Relevance [Figure presented].

Spinoit A.-F., Buelens S., Van Praet C., Van Laecke E., Praet M., Hoebeke P.

Embase

European Urology, Supplements. 16 (8) (pp 177-181), 2017. Date of Publication: November 2017.

[Review]

AN: 618184563

Background Congenital penile malformations (CPMs) are multifactorial in origin, and most factors remain unexplored. Initial findings indicate that CPMs are associated with abnormal histological development of dartos tissue (DT). The role of potentially aberrant androgen receptor (AR) expression in CPMs remains to be explored. Methods Preputial samples from children undergoing nonmedical circumcision and children undergoing primary penile surgery for CPM were immunohistochemically analyzed. The pathologist was blinded to the indication for surgery. Results DT disorganization strongly correlated with the clinical severity of the CPM condition. Both AR levels and DT patterns significantly differed in controls compared to cases. Higher age at surgery corresponded to higher AR levels in controls, which was not observed in cases. Conclusions DT disorganization strongly correlates with CPM clinical severity. AR expression is significantly lower in children presenting with a CPM than in healthy children. AR levels increase with age in healthy children, which is not observed in children presenting with a CPM. Congenital penile malformations (CPMs) are associated with abnormal histological development of dartos tissue. Androgen receptor (AR) expression is significantly lower in children presenting with a CPM than in healthy children. AR levels increase with age in healthy children, which is not observed in children with a CPM.

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Author NameID

Spinoit, Anne-Francoise; ORCID: <http://orcid.org/0000-0002-9045-9127>

Institution

(Spinoit, Buelens, Van Praet, Van Laecke, Praet, Hoebeke) Department of Urology, Ghent University Hospital, Ghent, Belgium

Publisher

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Year of Publication

2017

558.

Long-term Outcome of Common Congenital Problems Surgically Treated in Childhood [Figure presented].

Tekgul S.

Embase

European Urology, Supplements. 16 (8) (pp 189-193), 2017. Date of Publication: November 2017.

[Review]

AN: 618265744

Many of the common congenital pathologies surgically treated early in childhood may have long-term clinical and surgical complications when they come to adulthood. Surgically treated obstructive congenital pathologies will need follow-up for the resolution of obstruction and improvement of the upper tract. Of the children treated for vesicoureteric reflux, especially those with bilateral high grade, 5-20% may have a risk of hypertension and renal failure in the long term. Although bladder augmentation with an ileal segment in children is a procedure that can provide good results in terms of preservation of the upper urinary tract and renal function, and achieving continence, there are many long-term clinical and surgical complications. Many of the reconstructive procedures employed on the genitalia may have functional and cosmetic problems, which may require additional surgery in the long term. The rate of additional surgery is as high as 25% within 2-5 yr of follow-up after hypospadias surgery. Therefore, there is an obvious need of long-term follow-up for these children and urologists with expertise to recognize these complications and treat them. Many of the common congenital pathologies surgically treated early in childhood may have long-term clinical and surgical complications when they come to

adulthood. Therefore, there is an obvious need of long-term follow-up for these children and urologists with expertise to recognize these complications and treat them.

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Institution

(Tekgul) Division of Pediatric Urology, Department of Urology, Hacettepe University, Ankara, Turkey

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2017

559.

Functional and cosmetic outcome of partial penile disassembly repair in isolated male epispadias patients.

Bhat A., Bhat M., Kumar R., Kumar V.

Embase

African Journal of Urology. 23 (3) (pp 240-244), 2017. Date of Publication: September 2017.

[Article]

AN: 617136746

Introduction Epispadias is a rare congenital anomaly and requires a carefully constructed and well-planned approach for the management. Modified Cantwell-Ransley technique and Mitchell's complete penile disassembly are commonly used technique and these may require multiple surgeries in majority of the patients to achieve the goals of cosmesis and continence. **Objective** To evaluate the functional and cosmetic outcome of single stage partial penile disassembly repair in isolated male epispadias. **Patients and methods** A retrospective analysis of 43 cases of primary epispadias repair, performed during July 1998 to March 2013. Patients were classified on the basis of type of epispadias, urinary incontinence, presence/degree of chordee and penile rotation. **Technique** Penile de-gloving with mobilization of urethral plate from ventral to dorsal aspect with preservation of blood supply at both ends, distally up to the level of mid-glans and proximally up to pubic symphysis with division of peno-pubic ligament to lengthen the penis and position the

urethra ventrally. Tubularization of urethral plate followed by spongioplasty, corporoplasty with medial rotation of corporeal bodies (without any corporotomy) and glanuloplasty with meatoplasty is done to bring the meatus ventrally. Skin cover with rotation of ventral flaps and z-plasty when required. Results Age of the patients varied from 6 months to 26 years with a mean of 9 years. Ninety three percent of the patients had excellent cosmetic outcome while seven percent had minimal residual chordee/torque but did not require any surgery. None of the patients developed complications like fistula or stricture. All the 12 patients in the postpubertal group reported normal erections and successful ejaculations after the surgery. Postoperative follow up ranged from 2 to 10 years with a mean of 4 years. Conclusions The technique incorporates all the benefits of Cantwell-Ransley repair, can be done with less extensive dissection than total penile disassembly. Both functional and cosmetic results are good with low complication rate. Spongioplasty reconstructs near normal urethra and corporoplasty with spongioplasty also helps in prevention of urethral fistula.

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Institution

(Bhat, Bhat, Kumar, Kumar) Department of Urology, Preventive and Social Medicine, S.P. Medical College, Bikaner 334003, India

Publisher

Pan African Urological Surgeons Association(PAUSA) (E-mail: sunnydoodu@yahoo.com)

Year of Publication

2017

560.

Establishing disability weights for congenital pediatric surgical conditions: A multi-modal approach.

Poenaru D., Pemberton J., Frankfurter C., Cameron B.H., Stolk E.

Embase

Population Health Metrics. 15 (1) (no pagination), 2017. Article Number: 8. Date of Publication: 04 Mar 2017.

[Article]

AN: 614636414

Background: Burden of disease (BoD) as measured by Disability-Adjusted Life Years (DALYs) is one of the criteria for priority-setting in health care resource allocation. DALYs incorporate disability weights (DWs), which are currently expert-derived estimates or non-existent for most pediatric surgical conditions. The objective of this study is to establish DWs for a subset of key pediatric congenital anomalies using a range of health valuation metrics with caregivers in both high- and low-resource settings.

Method(s): We described 15 health states to health professionals (physicians, nurses, social workers, and therapists) and community caregivers in Kenya and Canada. The health states summaries were expert- and community-derived, consisting of a narrated description of the disease and a functional profile described in EQ-5D-5 L style. DWs for each health state were elicited using four health valuation exercises (preference ranking, visual analogue scale (VAS), paired comparison (PC), and time trade-off (TTO)). The PC data were anchored internally to the TTO and externally to existing data to yield DWs for each health state on a scale from 0 (health) to 1 (dead). Any differences in DWs between the two countries were analyzed.

Result(s): In total, 154 participants, matched by profession, were recruited from Kijabe, Kenya (n = 78) and Hamilton, Canada (n = 76). Overall calculated DWs for 15 health states ranged from 0.13 to 0.77, with little difference between countries (intra-class coefficient 0.97). However, DWs generated in Kenya for severe hypospadias and undescended testes were higher than Canadian-derived DWs ($p = 0.04$ and $p < 0.003$, respectively).

Conclusion(s): We have derived country-specific DWs for pediatric congenital anomalies using several low-cost methods and inter-professional and community caregivers. The TTO-anchored PC method appears best suited for future use. The majority of DWs do not appear to differ significantly between the two cultural contexts and could be used to inform further work of estimating the burden of global pediatric surgical disease. Care should be taken in comparing the DWs obtained in the current study to the existent list of DWs because methodological differences may impact on their compatibility.

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Status

Embase

Institution

(Poenaru, Pemberton, Frankfurter, Cameron) McMaster University, McMaster Pediatric Surgery Research Collaborative, Dept. of Surgery, Hamilton, Canada (Poenaru) MyungSung Medical College, Addis Ababa, Ethiopia

(Poenaru) Montreal Children's Hospital, McGill University, Department of Surgery, 16-4200
Sherbrooke West, Montreal, Westmount, QC H3Z 1C4, Canada
(Pemberton, Cameron) McMaster University, Dept. of Clinical Epidemiology and Biostatistics,
Hamilton, Canada
(Stolk) Erasmus Medical Center Rotterdam, Institute for Medical Technology Assessment,
Rotterdam, Netherlands
Publisher
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Year of Publication
2017

561.

Bladder continent catheterizable conduit (the Mitrofanoff procedure): Long-term issues that
should not be underestimated.

Faure A., Cooksey R., Bouty A., Woodward A., Hutson J., O'Brien M., Heloury Y.

Embase

Journal of Pediatric Surgery. 52 (3) (pp 469-472), 2017. Date of Publication: 01 Mar 2017.

[Article]

AN: 613179474

Background Effective bladder emptying by clean intermittent catheterization for children with
severe bladder dysfunction is critical for renal preservation and social integration. Use of a
continent catheterizable conduit (CCC) as urethral alternative procedure provides effective
bladder drainage. However, it brings a substantive maintenance. Methods Retrospective review
of the indications and long-term outcomes of 54 patients with a Mitrofanoff procedure in a single
center over a 20-year period (1995-2015). Results Indications of CCC include 21 neurogenic
bladders, 12 patients with epispadias/exstrophy, 13 bladder outlet obstruction, 6 malignancies
and 2 cloaca. Median age at surgery was 8.3 years (4 months-20 years). The appendix was used
in 76% of cases. Most frequently encountered complication was stomal stenosis (n = 17/34,
50%), occurring at median time of 9 months (2 months-13 years). The other complications were:
leakage in 9 (26.5%); conduit stricture in 5 (14.7%), angulation of the conduit in 2 (5.8%) and
prolapse in one (3%). Operative revision was encountered by 33 (61%) patients, the majority in
the first 2 years. Median follow-up was 4.3 years (3 months-16 years). Conclusions CCC has a

high incidence of complication. It has to be used only when the native urethra is not suitable for catheterization. Carers, patients and families must be prepared to deal with both the complexity of index conditions and the complications of this procedure.

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Status

Embase

Institution

(Faure, Cooksey, Bouty, Woodward, Hutson, O'Brien, Heloury) Royal Children's Hospital, Department of Paediatric Urology, Melbourne, Australia (Hutson) FD Stephens Surgical Research Group, Murdoch Childrens Research Institute, Department of Paediatric Urology, Melbourne, Australia

Publisher

W.B. Saunders

Year of Publication

2017

562.

Long-term sexual health outcomes in men with classic bladder exstrophy.

Baumgartner T.S., Lue K.M., Sirisreetreerux P., Metzger S., Everett R.G., Reddy S.S., Young E., Anele U.A., Alexander C.E., Gandhi N.M., Di Carlo H.N., Gearhart J.P.

Embase

BJU International. 120 (3) (pp 422-427), 2017. Date of Publication: September 2017.

[Article]

AN: 615896803

Objectives: To identify the long-term sexual health outcomes and relationships in men born with classic bladder exstrophy (CBE).

Material(s) and Method(s): A prospectively maintained institutional database comprising 1248 patients with exstrophy-epispadias was used. Men aged ≥ 18 years with CBE were included in the study. A 42-question survey was designed using a combination of demographic information and previously validated questionnaires.

Result(s): A total of 215 men met the inclusion criteria, of whom 113 (53%) completed the questionnaire. The mean age of the respondents was 32 years. Ninety-six (85%) of the respondents had been sexually active in their lifetime, and 66 of these (58%) were moderately to very satisfied with their sex life. The average Sexual Health Inventory for Men score was 19.8. All aspects of assessment using the Penile Perception Score questionnaire were on average between 'very dissatisfied' and 'satisfied'. Thirty-two respondents (28%) had attempted to conceive with their partner. Twenty-three (20%) were successful in conceiving, while 31 (27%) reported a confirmed fertility problem. A total of 31 respondents (27%) reported undergoing a semen analysis or post-ejaculatory urine analysis. Of these, only four respondents reported azoospermia.

Conclusion(s): Patients with CBE have many of the same sexual and relationship successes and concerns as the general population. This is invaluable information to give to both the parents of boys with CBE, and to the boys themselves as they transition to adulthood.

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Author NameID

Gearhart, John P.; ORCID: <http://orcid.org/0000-0003-0213-171X>

Institution

(Baumgartner, Lue, Sirisreetreerux, Metzger, Everett, Reddy, Young, Anele, Alexander, Gandhi, Di Carlo, Gearhart) Division of Pediatric Urology, James Buchanan Brady Urological Institute, Johns Hopkins University School of Medicine, Baltimore, MD, United States

Publisher

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Year of Publication

2017

Does the choice of suture material affect the incidence of urethrocutaneous fistulae after hypospadias repair? - A comparison of polyglactin 910 with polydioxanone.

Khalid A., Zaidi S.H., Baloch M., Saleem F.

Embase

International Journal of Toxicological and Pharmacological Research. 9 (3) (pp 189-193), 2017.

Date of Publication: 2017.

[Article]

AN: 617945440

Background: We aimed to assess the incidence of developing postoperative urethrocutaneous fistulae (UCF) while using either polyglactin 910 or polydioxanone for the repair of subcoronal hypospadias in paediatric patients.

Method(s): A multicenter, two-group posttest-only randomized experimental design was adopted for the study. The study was conducted at PNS Shifa Hospital Karachi, Combined Military Hospital Malir, Military Hospital Rawalpindi and Combined Military Hospital Multan from 2009 to 2016. Boys between the ages of 1 - 10 years with confirmed diagnosis of subcoronal hypospadias suitable for single stage repair with the Snodgrass technique and also completing a minimum follow up of 6 months were targeted for the study. The subjects were later randomized into PG group (those undergoing urethral repair with polyglactin 901) and PD group (those in whom polydioxanone was used). Both descriptive and inferential statistics were used for data analysis. SPSS v. 21.0 was used for data analysis with $p < 0.05$ taken as significant value.

Result(s): Two hundred patients with confirmed diagnosis of subcoronal hypospadias were recruited for the study in the proposed study period. Twenty-nine patients (29%) in the PG and 26 (26%) in the PD group developed UCF. There were seven (7%) cases of wound infection in the PG group compared to four cases (4%) in the PD Group; all eleven of the said were among those who developed UCF. Meatal stenosis was observed in six patients (6%) in the PG group and in nine patients (9%) of the PD group. No significant difference in the incidence of postoperative urethral fistula formation with the use of polyglactin 910 versus polydioxanone was however reported by the study findings.

Conclusion(s): As there was no significant difference in the incidence UCF between Polyglactin 910 and Polydioxanone, the choice of suture material should be based on economical variations and availability of the two products.

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Embase

Institution

(Khalid) Combined Military Hospital, Quetta, Pakistan (Zaidi) Combined Military Hospital,
Multan, Pakistan
(Baloch) Department of Pharmacolgy, Faculty of Pharmacy & Health Sciences, University of
Balochistan, Quetta, Pakistan
(Saleem) Department of Pharmacy Practice, Faculty of Pharmacy & Health Sciences, University
of Balochistan, Quetta, Pakistan
Publisher
International Journal of Toxicological and Pharmacological Research (E-mail: info@ijtpr.com)
Year of Publication
2017

564.

Human exposure to environmental contaminants and congenital anomalies: a critical review.

Foster W.G., Evans J.A., Little J., Arbour L., Moore A., Sauve R., Andres Leon J., Luo W.

Embase

Critical Reviews in Toxicology. 47 (1) (pp 59-84), 2017. Date of Publication: 02 Jan 2017.

[Review]

AN: 614413436

Congenital anomalies are an important cause of infant mortality and disability. Developmental exposure to environmental contaminants is thought to increase the risk for congenital anomalies. Herein, we describe a critical review of the literature conducted between February and March 2014 yielding 3057 references from which 97 unique relevant articles published from 2003 through 2014 were evaluated. Common congenital anomalies including hypospadias, cryptorchidism, anogenital distance (AGD), congenital heart defects and oral clefts were well represented in the literature whereas other outcomes such as neural tube defects, limb deficiency defects and gastroschisis were rarely described. While definitions used for congenital anomalies and methods of ascertainment were usually consistent across studies, inconsistencies were frequently found in grouping of different congenital heart defects. Despite strong links between some congenital anomalies and parental occupation, these studies are unable to provide clear insight into the specific chemicals responsible owing to lack of direct measures of exposure. In comparison, data are mixed for contaminant exposures at concentrations representative of results from contemporary biomonitoring studies. Of the environmental contaminants studied, the

association between phthalate exposures and developmental abnormalities of the male reproductive tract received the greatest attention. Important limitations of the literature studied relate to adequacy of sample size, absence of or weaknesses in exposure assessment methodologies, failure to account for biological plausibility and grouping of congenital anomalies with divergent mechanisms. We conclude that the literature is inadequate at this time to support a conclusion that exposure to environmental contaminants are or are not associated with increased risks for congenital anomalies in the general population.

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Status

Embase

Institution

(Foster) Department of Obstetrics & Gynecology, McMaster University, Hamilton, ON, Canada

(Evans) Department of Biochemistry and Medical Genetics, University of Manitoba, Winnipeg, MB, Canada

(Little) School of Epidemiology, Public Health and Preventive Medicine, Faculty of Medicine, University of Ottawa, Ottawa, ON, Canada

(Arbour) Department of Medical Genetics, University of British Columbia, Vancouver, BC, Canada

(Moore) Department of Paediatrics, University of Toronto and the Hospital for Sick Children, Toronto, ON, Canada

(Sauve) Department of Community Health Sciences, University of Calgary, Calgary, AB, Canada

(Andres Leon, Luo) Maternal, Child and Youth Health Surveillance and Epidemiology Division, Centre for Chronic Disease Prevention, Public Health Agency of Canada, Ottawa, ON, Canada

Publisher

Taylor and Francis Ltd (E-mail: healthcare.enquiries@informa.com)

Year of Publication

2017

565.

Nucleotide Transition 390C-T in the Wilms' Tumor 1 Gene: A Risk Factor of Hypospadias?.

Buglyo G., Magyar A., Biro S., Csizy I., Beyer D., Molnar K., Olah E.

Embase

Current Urology. 10 (3) (pp 136-139), 2017. Date of Publication: 01 Aug 2017.

[Article]

AN: 617914718

Introduction: The gene Wilms' tumor 1 (WT1) encodes a unique transcription factor. Its defects are known to cause a wide range of complex genitourinary malformations and may contribute to non-syndromic forms of hypospadias.

Material(s) and Method(s): We performed WT1 mutation analysis and copy number analysis of WT1-interacting protein in 13 Hungarian patients diagnosed with isolated hypospadias.

Result(s): Sequencing of WT1 revealed a high frequency of heterozygosity for transition 390C-T (5 heterozygotes out of 13 patients, including 2 brothers). WT1-interacting protein had a normal copy number in all patients.

Conclusion(s): Nucleotide substitution 390C-T may play a role in the pathogenesis of non-syndromic hypospadias. The genotype-phenotype correlation should be confirmed by a larger-scale analysis.

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Embase

Institution

(Buglyo, Biro, Beyer, Molnar) Department of Human Genetics, University of Debrecen, 98 Nagyerdei korut, 4032, Debrecen, Hungary (Magyar, Csizy, Olah) Department of Pediatrics, University of Debrecen, Debrecen, Hungary

Publisher

S. Karger AG

Year of Publication

2017

566.

Evaluation and treatment for ovotesticular disorder of sex development (OT-DSD) - experience based on a Chinese series.

Mao Y., Chen S., Wang R., Wang X., Qin D., Tang Y.

Embase

BMC Urology. 17 (1) (no pagination), 2017. Article Number: 21. Date of Publication: 28 Mar 2017.
[Article]

AN: 615038893

Background: The aim of this study is to review and present the clinical features and process of evaluation and treatment for OT-DSD in a single center in recent years in China.

Method(s): Sixteen patients with OT-DSD during the past 4 years underwent the evaluation and treatment in a single center. The clinical characteristics and outcomes of surgery were analyzed.

Result(s): The surgical age ranged from 17 months to 66 months with a mean age of 20 months, and the mean follow-up was 30 months (4 months to 56 months). The presentation in 11 patients was ambiguous genitalia, and the rest 5 patients were suspected to have DSD in preoperative examination before hypospadias repair. The karyotypes were 46, XX in 11 patients, 46, XX/46, XY in 3, 46, XX/47, XXY in 1, and 46, XY in 1. Initial reared sex was male in 14 patients, female in 1, and undetermined in 1. After surgery, genders were reassigned in 3 patients, while 15 patients were raised as male with testicular tissue left. Only 1 patient with ovarian tissue left was raised as female. Repair was completed in 11 males and 1 female, and stage I urethroplasty was done in 4 males. No further surgery to remove the gonads was needed for inconsonance of gender assignment. No gonadal tumors were detected.

Conclusion(s): OT-DSD is a rare and complex deformity with few systematic reports in China. It's important to establish a regular algorithm for evaluation and treatment of OT-DSD.

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Status

Embase

Institution

(Mao, Chen, Wang, Qin, Tang) Department of Pediatric Surgery of Children's Medical Center, Sichuan Academy of Medical Sciences, Sichuan Provincial People's Hospital, Chengdu, China

(Wang) Department of Burn and Plastic Surgery, West China Hospital of Sichuan University, Chengdu, China

Publisher

BioMed Central Ltd. (E-mail: info@biomedcentral.com)

Year of Publication

2017

567.

Lingual mucosal graft two-stage Bracka technique for redo hypospadias repair.

Sakr A., Elkady E., Abdalla M., Fawzi A., Kamel M., Desoky E., Seleem M., Omran M., Elsayed E., Khalil S.

Embase

Arab Journal of Urology. 15 (3) (pp 236-241), 2017. Date of Publication: September 2017.

[Article]

AN: 617423170

Objectives To report our initial experience in redo hypospadias repair with a lingual mucosal graft (LMG) using a two-stage Bracka technique. **Patients and methods** This study was prospectively conducted and included 26 patients with hypospadias with failed previous repairs. All the patients had a LMG using a two-stage Bracka technique. In the first stage, the harvested LMG, from the ventro-lateral surface of the tongue, was implanted in a well-prepared vascularised bed in the ventral aspect of the penis. After 6 months, tubularisation of the well-taken graft was completed. Tunica vaginalis or a dartos flap was used as second-layer coverage of the neourethra. Success was defined as acceptable aesthetic and functional outcomes without any additional surgical interventions. **Results** The mean (SD) patient age was 5.15 (1.6) years. The mean (SD) LMG length was 3.82 (0.9) cm and the width was 1.5 (0.5) cm. The mean (SD) number of previous repairs was 2.76 (1.1). The mean (SD) follow-up was 12 (2) months. Donor-site complications included: pain in all patients, with a pain score of >3 on the visual analogue pain scale (0-10) in 10 (38%); and speech problems in 19 (73%). First-stage complications were graft loss (n = 2) and contracture (n = 1). The second stage was completed in 23 patients resulting in the following significant complications: meatal stenosis plus fistula (n = 2), breakdown (n = 1). Successful hypospadias repair was achieved in 77% (20/26) of the patients. **Conclusion** Lingual mucosa is a reliable and versatile graft material in the armamentarium of two-stage Bracka hypospadias repair with the merits of easy harvesting and minor donor-site complications.

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Institution

(Sakr, Elkady, Abdalla, Fawzi, Kamel, Desoky, Seleem, Omran, Elsayed, Khalil) Department of Urology, Zagazig University, Faculty of Medicine, Zagazig, Egypt

Publisher

Arab Association of Urology (E-mail: araburo@yahoo.com)

Year of Publication

2017

568.

Mathieu technique with incision of the urethral plate versus standard tubularised incised-plate urethroplasty in primary repair of distal hypospadias: A prospective randomised study.

Khalil M., Gharib T., El-shaer W., Sebaey A., Elmohamady B., Elgamal K.

Embase

Arab Journal of Urology. 15 (3) (pp 242-247), 2017. Date of Publication: September 2017.

[Article]

AN: 616321648

Objective To compare outcomes of the Mathieu incised-plate (Mathieu-IP) technique vs the standard tubularised incised-plate (TIP) technique for primary management of distal hypospadias.

Patients and methods Between April 2012 and August 2015, 66 patients (aged 15-60 months) with primary distal hypospadias were randomly allocated to two groups. Group 1 included 34 patients who underwent Mathieu-IP repair and Group 2 included 32 patients managed using the TIP technique for primary management of distal hypospadias. Postoperatively, all patients were examined weekly up to 1 month and then at 3 and 6 months. Perioperative data, complications and outcomes of both procedures were statistically analysed and compared. **Results** There were no statistically significant differences in patient demographics between the groups at baseline.

There was no statistically significant difference in the mean (SD) operative time between Groups 1 and 2, at 95 (7.6) and 91.2 (8.1) min, respectively. There was no statistically significant difference in the shape of the urine stream at micturition or the neomeatus between the groups postoperatively. The rate of postoperative fistula was significantly higher in Group 2 (TIP) compared to Group 1 (Mathieu-IP), at 18.7% vs 2.9% ($P = 0.004$). There was no postoperative meatal stenosis in Group 1, which did occur in five patients (15.6%) in Group 2 ($P = 0.002$).

Conclusion The Mathieu-IP technique appeared to be better than the standard TIP technique with regard to postoperative fistula formation and meatal stenosis, and with acceptable cosmesis.

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Institution

(Khalil, Gharib, El-shaer, Sebaey, Elmoahady, Elgamal) Department of Urology, Faculty of Medicine, Benha University, Benha, Egypt

Publisher

Arab Association of Urology (E-mail: araburo@yahoo.com)

Year of Publication

2017

569.

Comparing the bulking effect of calcium hydroxyapatite and Deflux injection into the bladder neck for improvement of urinary incontinence in bladder exstrophy-epispadias complex.

Eftekharzadeh S., Sabetkish N., Sabetkish S., Kajbafzadeh A.-M.

Embase

International Urology and Nephrology. 49 (2) (pp 183-189), 2017. Date of Publication: 01 Feb 2017.

[Article]

AN: 613475623

Objectives: The aim of this study was to evaluate the efficacy of the endoscopic injection of calcium hydroxyapatite (CaHA) into the bladder neck (BN) region of patients with urinary incontinence and bladder exstrophy-epispadias complex (BEEC).

Patients and Methods: We designed a retrospective cohort study in which we retrospectively studied medical charts of female and male patients of BEEC who had undergone CaHA or Deflux injection for continence improvement between 2009 and 2014. Sixteen incontinent patients with a mean $\pm$ SD age of 8.09 $\pm$ 3.5 years received an endoscopic submucosal injection of 5.4 ml of pure CaHA powder with autologous plasma (group A). Patients in group B (N = 21), control group, with a mean $\pm$ SD age of 7.51 $\pm$ 2.8 years received Deflux injection (5.1 ml). The mean follow-up after injection was 38 $\pm$ 5.2 and 33 $\pm$ 4.1 months in groups A and B, respectively.

Result(s): No post-injection complication was detected in none of the patients during the follow-up. Eleven patients (68.75%) in group A became socially dry following 1-2 injections, the degree of incontinence was improved in 4 patients (25%), and there was no change in one patient (6.25%). However, Deflux injection resulted in complete dryness in 14 (66.66%), improvement in the degree of incontinence in 5 (23.81%) and no change in 2 patients (9.52%), leading to no significant difference in continence achievement between CaHA and Deflux groups ($p = 0.9$). The

statistical analysis was not significantly different in terms of bladder capacity ($p = 0.7$) or Q max ($p = 0.8$).

Conclusion(s): The preliminary results of this study revealed that CaHA may be applied as an affordable bulking agent in treatment of urinary incontinence in BEEC.

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Status

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Institution

(Eftekharzadeh, Sabetkish, Sabetkish, Kajbafzadeh) Section of Tissue Engineering and Stem Cells Therapy, Pediatric Urology and Regenerative Medicine Research Center, Children's Medical Center, Pediatric Center of Excellence, Tehran University of Medical Sciences, No. 62, Dr. Qarib's St, Keshavarz Blvd, Tehran 14194 33151, Iran, Islamic Republic of

Publisher

Springer Netherlands

Year of Publication

2017

570.

A rare case of genital malformation with omphalocele, exstrophy of bladder, imperforate anus and spinal defect complex-autopsy findings.

Mamatha K., Yelikar B.R., Deshpande V.R., Disha B.S.

Embase

Journal of Clinical and Diagnostic Research. 11 (7) (pp ED37-ED38), 2017. Date of Publication: 01 Jul 2017.

[Article]

AN: 617532170

Omphalocele, Exstrophy of cloaca, Imperforate anus, and Spinal defects (OEIS) is a severe manifestation of exstrophy-epispadias sequence with a combination of defects including OEIS. It results from improper closure of anterior abdominal wall and defective development of cloaca and urogenital septum due to defect in blastogenesis during the 4th week of gestation. Identification of

this complex is important through foetal autopsy as this condition can recur in siblings. Prenatal diagnosis also helps to prevent foetal death with appropriate management in the less severe cases. In severe cases, termination of pregnancy is considered. A primigravida with 28 weeks of gestation had delivered a live baby with multiple congenital anomalies; baby died after 10 minutes. These anomalies were grouped under OEIS complex.

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Status

Embase

Institution

(Mamatha, Yelikar, Deshpande, Disha) Department of Pathology, BLDEU's Shri B.M. Patil Medical College, Vijayapur, Karnataka, India

Publisher

Journal of Clinical and Diagnostic Research (No 3, 1/9 Roop Nagar,GT Karnal Road, Delhi 110007, India)

Year of Publication

2017

571.

Reproductive function in the sons of women who experienced stress due to bereavement before and during pregnancy: a nationwide population-based cohort study.

Plana-Ripoll O., Li J., Kesmodel U.S., Parner E., Olsen J., Basso O.

Embase

Fertility and Sterility. 107 (1) (pp 189-197.e8), 2017. Date of Publication: 01 Jan 2017.

[Article]

AN: 613819931

Objective To estimate the association between prenatal exposure to maternal stress and reproductive disorders in Danish men, where prenatal stress exposure was defined as the mother's loss of a close relative during pregnancy or in the 12 months before conception. Design Population-based cohort study. Setting Not applicable. Patient(s) All males born in Denmark between 1973 and 2008 (n = 1,217,576) and observed for up to 39 years. Intervention(s) None. Main Outcome Measure(s) Male reproductive function, defined using a composite outcome including congenital malformations of genital organs, testicular cancer, diagnosis of male

infertility, or assisted conception use due to male factor infertility. Result(s) In total, 28,986 men (2.4%) had been exposed to prenatal stress, and 62,929 (5.2%) experienced the composite outcome during the follow-up period. Prenatal exposure to stress was associated with an elevated risk of reproductive problems (hazard ratio [HR] 1.09; 95% CI, 1.04-1.15). The association was stronger when the exposure occurred during the first trimester of pregnancy, and for congenital malformations of genital organs. When focusing on infertility alone, we saw no evidence of increased risk (HR 0.90; 95% CI, 0.77-1.06). In addition, the probability of marrying a woman was lower for exposed men (HR 0.93; 95% CI, 0.89-0.98). Conclusion(s) Prenatal stress in the form of the mother's bereavement during the first trimester of pregnancy is associated with a higher risk of reproductive disorders from congenital malformations of the genital organs in the male offspring. The lack of an association between maternal bereavement and later infertility in the exposed male offspring may be due in part to the men's lower probability of attempting to have children.

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Status

Embase

Institution

(Plana-Ripoll) Section of Epidemiology, Department of Public Health, Aarhus University, Aarhus, Denmark (Li, Olsen) Department of Clinical Epidemiology, Aarhus University Hospital, Aarhus, Denmark

(Kesmodel) Department of Obstetrics and Gynecology, Herlev and Gentofte Hospital, Herlev, Denmark

(Parner) Section of Biostatistics, Department of Public Health, Aarhus University, Aarhus, Denmark

(Olsen) Department of Epidemiology, Fielding School of Public Health, University of California, Los Angeles, California, United States

(Basso) Department of Obstetrics and Gynecology, Research Institute of the McGill University Health Centre (MUHC), Montreal, Quebec, Canada

(Basso) Department of Epidemiology, Biostatistics and Occupational Health, McGill University, Montreal, Quebec, Canada

Publisher

Elsevier Inc. (E-mail: usjcs@elsevier.com)

Year of Publication

2017

572.

Etiology and clinical presentation of birth defects: Population based study.

Feldkamp M.L., Carey J.C., Byrne J.L.B., Krikov S., Botto L.D.

Embase

BMJ (Online). 357 (no pagination), 2017. Article Number: 2249. Date of Publication: 2017.

[Article]

AN: 616511064

Objective To assess causation and clinical presentation of major birth defects. **Design** Population based case cohort. **Setting** Cases of birth defects in children born 2005-09 to resident women, ascertained through Utah's population based surveillance system. All records underwent clinical re-review. **Participants** 5504 cases among 270 878 births (prevalence 2.03%), excluding mild isolated conditions (such as muscular ventricular septal defects, distal hypospadias). **Main outcome measures** The primary outcomes were the proportion of birth defects with a known etiology (chromosomal, genetic, human teratogen, twinning) or unknown etiology, by morphology (isolated, multiple, minors only), and by pathogenesis (sequence, developmental field defect, or known pattern of birth defects). **Results** Definite cause was assigned in 20.2% (n=1114) of cases: Chromosomal or genetic conditions accounted for 94.4% (n=1052), teratogens for 4.1% (n=46, mostly poorly controlled pregestational diabetes), and twinning for 1.4% (n=16, conjoined or acardiac). The 79.8% (n=4390) remaining were classified as unknown etiology; of these 88.2% (n=3874) were isolated birth defects. Family history (similarly affected first degree relative) was documented in 4.8% (n=266). In this cohort, 92.1% (5067/5504) were live born infants (isolated and non-isolated birth defects): 75.3% (4147/5504) were classified as having an isolated birth defect (unknown or known etiology). **Conclusions** These findings underscore the gaps in our knowledge regarding the causes of birth defects. For the causes that are known, such as smoking or diabetes, assigning causation in individual cases remains challenging. Nevertheless, the ongoing impact of these exposures on fetal development highlights the urgency and benefits of population based preventive interventions. For the causes that are still unknown, better strategies are needed. These can include greater integration of the key elements of etiology, morphology, and pathogenesis into epidemiologic studies; greater collaboration between researchers (such as developmental biologists), clinicians (such as medical geneticists), and epidemiologists; and better ways to objectively measure fetal exposures (beyond maternal self reports) and closer (prenatally) to the critical period of organogenesis.

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PMID

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Status

Embase

Institution

(Feldkamp, Carey, Byrne, Krikov, Botto) Department of Pediatrics, University of Utah, School of Medicine, 295 Chipeta Way, Salt Lake City, UT, United States (Byrne) Department of Obstetrics and Gynecology, University of Utah, School of Medicine, Salt Lake City, UT, United States

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2017

573.

Evaluation and outcome of M plasty for the management of doughnut scrotum.

Mylarappa P., Puvvada S., Arvind Nayak K., Ramesh D.

Embase

Journal of Clinical Urology. 10 (4) (pp 364-367), 2017. Date of Publication: 01 Jul 2017.

[Article]

AN: 616853736

Introduction: Penoscrotal transposition is a rare anomaly of the external genitalia. They can be classified as complete or incomplete based on degrees of positional exchange between the penis and the scrotum. Both forms are known to be associated with hypospadias and multiple surgeries are required for complete correction. Most surgeries performed for the correction of penoscrotal transposition involve making a complete circular incision around the root of the scrotum, which often results in massive penile lymphoedema and often delays the correction of hypospadias. The M plasty technique can prevent the incidence of lymphoedema by preserving the dorsal penile skin.

Objective(s): To evaluate the effectiveness of M plasty for the correction of penoscrotal transposition.

Material(s) and Method(s): Sixteen patients underwent M plasty for incomplete penoscrotal transposition. An 'M'-shaped incision was made at the base of the scrotum and the scrotal halves were dissected and brought down posterior and caudal to the penis and sutured primarily.

Result(s): All patients showed excellent cosmetic results. There was minimal postoperative oedema with no vascular compromise to penile or scrotal skin.

Conclusion(s): M plasty is an excellent technique for the correction of penoscrotal transposition. The low incidence of penile lymphoedema could be attributed to the preservation of the dorsal penile skin. This procedure provides an excellent cosmetic appearance and also allows for early correction of hypospadias.

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Embase

Institution

(Mylarappa, Puvvada, Arvind Nayak, Ramesh) M.S. Ramaiah Hospital, Bangalore, India

Publisher

SAGE Publications Ltd (E-mail: info@sagepub.co.uk)

Year of Publication

2017

574.

Single penile incision for combined hypospadias and inguinal surgery: A comparative study.

Chua M.E., Alqarni N., Ming J.M., Alyami F., Dos Santos J., Lopes R.I., Farhat W.A., Koyle M.A.

Embase

Canadian Urological Association Journal. 11 (5) (pp E192-E196), 2017. Date of Publication: May 2017.

[Article]

AN: 616804065

Introduction: We sought to compare the surgical outcomes of hypospadias repair with correction of inguinal pathology using a single penile incision vs. conventional approach using two incisions.

Method(s): This is a retrospective study that reviewed all patients who underwent concurrent surgical repair for both hypospadias and inguinal pathologies between January 2003 and November 2015. Patients were classified into Group A, conventional (inguinal or scrotal and

penile incision) approach; or Group B, single penile incision approach. Baseline characteristics, including age, degree of hypospadias, type and laterality of inguinal pathology, operative time, and surgical outcomes, were collected. Between groups, variable comparisons were analyzed using Mann-Whitney U-Test and Fisher-exact test. Statistical significant set at <0.05 .

Result(s): Seventy-six patients (Group A: 40; Group B: 36) were eligible for study. Baseline characteristics of both groups were comparable, with no significant statistical difference. Overall mean operative time for Group A was 139.3 +/- 56.2 minutes, while Group B was 107.8 +/- 46.7 minutes ($Z=2.6$; $U=470.5$; $p=0.009$). Two patients in Group A and two patients in Group B had testicular ascension, all of which also had hypospadias-related complications ($p=1.0$).

Hypospadias-related complications in Group A included seven urethrocutaneous fistulae and two repair dehiscence. Eight urethrocutaneous fistulae, one urethral stricture, and two repair dehiscence occurred in Group B ($p=0.448$). Surgical outcome appearance in both groups were comparable, with no statistically significant difference ($p=0.466$).

Conclusion(s): Single penile incision for both hypospadias repair and correction of inguinal pathology is a feasible technique and comparable to the conventional approach, with similar surgical outcomes and shorter overall operative time.

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Status

Embase

Institution

(Chua, Ming, Dos Santos, Lopes, Farhat, Koyle) Division of Urology, Department of Surgery, Hospital for Sick Children, Toronto, ON, Canada (Alqarni, Alyami) King Saud University, King Saud University Medical City, Riyadh, Saudi Arabia

Publisher

Canadian Urological Association (E-mail: josephine.sciortino@cua.org)

Year of Publication

2017

575.

The clinical analysis of small supernumerary marker chromosomes in 17 children with mos 45,X/46,X,+mar karyotype.

Wang H., Wang T., Yang N., He Y., Chen L., Hong L., Shao X., Zhu H., Li H.

Embase

Oncology Letters. 13 (6) (pp 4385-4389), 2017. Date of Publication: June 2017.

[Article]

AN: 616496595

Small supernumerary marker chromosome (sSMC) is a type of structurally abnormal chromosome. In order to identify the origin, morphology and other characteristics of sSMCs in children with mos 45,X/46,X,+mar karyotype, 17 patients (16 females and 1 male) were analyzed. All patients underwent general physical examination, gonadal imaging and molecular cytogenetic analyses, including Giemsa banding, dual-color fluorescence in situ hybridization and detection of the sex-determining region Y gene by polymerase chain reaction. Cytogenetic analyses indicated sSMCs in 14/17 cases were derived from the X chromosome, of which 8 individuals presented with ring-shaped sSMCs and 6 with centric minute-shaped sSMCs. The remaining 3 cases were derived from the Y chromosome, and all presented with minute-shaped sSMCs. All female patients exhibited short stature, gonadal dysgenesis and other typical features of Turner syndrome. The male patient exhibited short stature, hypospadias and bilateral cryptorchidism. In conclusion, the majority of the sSMCs in patients with a mos 45,X/46,X,+mar karyotype were derived from the sex chromosomes. The molecular cytogenetic features of sSMCs may provide useful information for genetic counseling, prenatal diagnosis and individualized treatment.

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Status

Embase

Institution

(Wang, Yang, He, Chen, Hong, Shao, Zhu) Department of Clinical Laboratory, Children's Hospital of Soochow University, Suzhou, Jiangsu 215003, China (Wang, Li, Li) Center for Reproduction and Genetics, Suzhou Hospital Affiliated to Nanjing Medical University, Suzhou, Jiangsu 215002, China

Publisher

Spandidos Publications (10 Vriaxidos Street, Athens 116 10, Greece. E-mail: subscriptions@spandidos-publications.com)

Year of Publication

2017

Preliminary experience using a tunica vaginalis flap as the dorsal component of Bracka's urethroplasty.

Harper L., Michel J.-L., Sauvat F.

Embase

BJU International. 119 (3) (pp 470-473), 2017. Date of Publication: 01 Mar 2017.

[Article]

AN: 614465388

Objective: To evaluate clinical use of a tunica vaginalis flap as the dorsal component of a two-stage urethroplasty in boys with cripple hypospadias.

Patients and Methods: We performed the first stage of a Bracka two-stage urethroplasty, using a tunica vaginalis flap as the dorsal component in six boys with cripple hypospadias. We analysed their clinical characteristics and the results of this technique.

Result(s): The mean (range) age of the boys was 57 (34-120) months. The mean (range) number of previous procedures the boys had undergone was 4 (3-5). At the 6-month follow-up, all the boys presented significant fibrosis of the dorsal graft rendering it unusable for tubularisation.

Conclusion(s): Exposure to the external environment seems to induce retraction and fibrosis of the tunica vaginalis. We believe one should be very cautious about using tunica vaginalis as the dorsal component of a two-stage urethroplasty, as significant fibrosis might well render the flap unusable.

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Status

Embase

Institution

(Harper, Michel, Sauvat) Department of Pediatric Surgery, CHU F Guyon, Bellepierre, Saint-Denis de La Reunion, Reunion Island, France

Publisher

Blackwell Publishing Ltd (E-mail: customerservices@oxonblackwellpublishing.com)

Year of Publication

2017

577.

Paediatric urology.

Undre S., Cherian A.

Embase

Surgery (United Kingdom). 35 (6) (pp 324-330), 2017. Date of Publication: June 2017.

[Review]

AN: 616132599

Paediatric urology is a subject that covers the urological aspects of care in children some of which are seen also in adults, but may have specific diagnostic methods and treatments that are quite different. Additionally, it covers a range of congenital anomalies either on their own or in combination with a spectrum of disorders that need more complex management available at only specialized centres. For the purposes of a broad and basic understanding of the subject, this article will cover relevant topics and up to date guidelines.

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Institution

(Undre, Cherian) Lister Hospital, Stevenage, United Kingdom

Publisher

Elsevier Ltd

Year of Publication

2017

578.

Management of the exstrophy-epispadias complex in adolescents and adults.

Jana A., Maiti K., Mondal T.K., Majhi T.K.

Embase

African Journal of Urology. 23 (2) (pp 100-104), 2017. Date of Publication: June 2017.

[Article]

AN: 616322286

Objectives To study the presentation and management of the exstrophy-epispadias complex in adolescents and adults and to evaluate the outcome of the repair. **Subjects and methods** This retrospective review of our medical records of adult patients with an exstrophy-epispadias complex managed between January 2010 and June 2015 included one female and 7 male patients. Only patients aged above 10 years who had not undergone any surgical treatment during childhood were included in the study. All patients were subjected to preoperative biopsy from the urinary bladder plate. None of them underwent cystectomy. Seven patients underwent urinary bladder augmentation using ileocystoplasty, bladder neck reconstruction with para-urethral tissue closure using a wedge of pubic bone, and single-stage epispadias repair. In one patient epispadias repair was not done due to intraoperative hemodynamic instability. **Results** All patients are continent but one in whom epispadias repair was not done and who is waiting for epispadias repair. One patient has developed a urethro-cutaneous fistula at the base of the penis. Two patients developed urinary bladder stones, and both of them were managed by minimally invasive methods. All the patients are on clean intermittent self-catheterization. They are all doing well socially and psychologically. **Conclusion** Though exstrophy-epispadias in adolescents and adults is a rare entity, the described modified surgical technique provides good results and improves the patients' social and psychological well being.

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Institution

(Jana, Maiti, Mondal, Majhi) Nilratan Sircar Medical College and Hospital, Kolkata, West Bengal, India

Publisher

Pan African Urological Surgeons Association(PAUSA) (E-mail: sunnydoodu@yahoo.com)

Year of Publication

2017

579.

Bhat's modifications of Glassberg-Duckett repair to reduce complications in management severe hypospadias with curvature.

Bhat M., Sabharwal K., Bhat A., Kumar R.

Embase

African Journal of Urology. 23 (2) (pp 94-99), 2017. Date of Publication: June 2017.

[Article]

AN: 616322282

Objective Disadvantages of two-stage hypospadias repair are the necessity of 2 or 3 surgeries, loss of time/money, complications like splaying of the stream, dribbling of urine or ejaculate and milking of the ejaculate due to a poor-quality urethra. The current article details our modifications of flap repair allowing to manage such patients in one stage and reducing the complications.

Subjects and methods Twenty one patients (aged 2-23 years, between January 2006 and June 2012 mean 11.5 years) of severe hypospadias were managed with flap tube urethroplasty combined with TIP since June 2006 and June 2012. Curvature was corrected by penile de-gloving, mobilization of urethral plate/urethra with corpus spongiosum and transecting urethral plate at corona. Buck's fascia was dissected between the corporeal bodies and superficial corporotomies were done as required. Mobilized urethral plate was tubularized to reconstruct proximal urethra up to peno-scrotal junction and distal tube was reconstructed with raised inner preputial flap after measuring adequacy of skin width. Both neo-urethrae were anastomosed in elliptical shape and covered with spongiosum. Distal anastomosis was done 5-8 mm proximal to tip of glans preventing protrusion of skin on glans. Tubularized urethral plate was covered by spongioplasty. Skin tube was covered by dartos pedicle and fixed to corpora. Scrotoplasty was done in layers, covering the anastomosis. Results Type of hypospadias was scrotal 10, perineo-scrotal 5, penoscrotal 4 and proximal penile in 2 cases. Chordee (severe 15 and moderate 6) correction was possible penile de-gloving with mobilization of urethral plate with spongiosum after dividing urethral plate at corona 8, next 5 cases required dissection of corporal bodies, superficial corporotomy 5 and 3 cases lateral dissection of Buck's fascia. Length of tubularized urethral plate varied from 3 to 5 cm and flap tube varied from 5.5 to 13 cm (average 7.5 cm). Complications were fistula 2, meatal stenosis 1, and dilated distal urethra 1 with overall success rate of 81%. None of them had residual curvature, torsion, splaying or dribbling urine in follow up of 10-36 (average 18) months. Conclusions TIPU with spongioplasty of proximal urethra and dartos cover on skin tube reconstructs functional urethra. Distal end skin sutured to glans mucosa 5-8 mm proximal to the tip of glans reconstructs a cosmetically normal looking meatus. An exact measurement of the width and length of the stretched dartos, fixation of the skin tube to the corpora and covering the skin tube with dartos helps in prevention of diverticula. Elliptical anastomosis covered with spongiosum prevents fistula and stricture at anastomotic site.

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Institution

(Bhat) Department of Urology, Dr S.N. Medical College, India (Bhat, Sabharwal, Kumar)

Department of Urology, S.P. Medical College, India

(Bhat) Department of Surgery, S.P. Medical College, India

Publisher

Pan African Urological Surgeons Association(PAUSA) (E-mail: sunnydoodu@yahoo.com)

Year of Publication

2017

580.

To Finish the Cut or Not: Should Neonatal Circumcisions Be Completed or Aborted in Patients with Unrecognized Glanular Hypospadias.

Zamilpa I., Patel A., Booth J., Canon S.

Embase

Clinical Pediatrics. 56 (2) (pp 157-161), 2017. Date of Publication: 01 Feb 2017.

[Article]

AN: 614329789

We retrospectively evaluated the management of patients with unrecognized glanular hypospadias and a completed (group 1) or aborted (group 2) neonatal circumcision. The rate and type of subsequent surgeries performed were analyzed. Penile curvature, urinary stream deviation, and their impact on management were evaluated. Surgery was done in 55% of patients - 40% of group 1 and 86% of group 2. Completion of the circumcision was done in 63% of group 2. Hypospadias repair was performed in 56% of group 1 and in 34% of group 2. Penile curvature rate did not affect the rate or type of surgery performed. Urinary stream deviation did not affect the rate of repair, but was a significant factor leading to hypospadias repair. We concluded that providers performing neonatal circumcisions do not have to abort the procedure when a glanular hypospadias is noticed. Most patients will require circumcision completion only.

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Institution

(Zamilpa, Patel) Arkansas Children's Hospital, 1 Children's Way, Little Rock, AR 72202-3500,
United States (Booth) Arkansas Children's Hospital, Fayetteville, AR, United States
(Canon) University of Arkansas for Medical Sciences, Little Rock, AR, United States
Publisher
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Year of Publication
2017

581.

Seasonal trends in the prevalence of hypospadias: Aetiological implications.

Mamoulakis C., Avgenakis G., Gkatzoudi C., Duijker G., Zisis I.E., Heretis I., Antypas S., Sofikitis N., Spandidos D.A., Tsatsakis A.M., Tzonou A.

Embase

Experimental and Therapeutic Medicine. 13 (6) (pp 2960-2968), 2017. Date of Publication: June 2017.

[Article]

AN: 616123249

The aim of the present study was to examine the seasonality of hypospadias in Greece in an attempt to elucidate the aetiology. All boys born between 1991-1998, who underwent hypospadias repair at 'Aghia Sophia' Children's Hospital, Athens (n=542) were analysed. All Greek live-born males during the same period (population at risk; m=421,175) served as the controls. Seasonality by month of birth was evaluated with specific statistical tools. Meteorological parameters were also analysed. All tests yielded significant results, suggesting a simple harmonic prevalence pattern (highest/lowest: autumn, peak in October/spring, trough in April). Therefore, the first trimester of hypospadiac gestations coincides more frequently with winter. Meteorological parameters varied seasonally (maximal sunlight; air temperature in summer/minimal in winter, maximal rainfall in winter/minimal in summer) and were strongly associated pairwise. Hypospadiac birth prevalence follows a simple harmonic seasonal pattern and is associated with that of cryptorchidism in Greece. The coincidence of the first or third trimester of a potentially genetically influenced gestation with winter could lead to the phenotypic expression of hypospadias or cryptorchidism, respectively. The potential role of a cyclic-varied androgen-production stimulator, such as human chorionic gonadotrophin may be speculated. The

seasonality of a common environmental factor acting directly/indirectly may contribute to these patterns, and possibly to the common pathogenesis of these congenital malformations.

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Institution

(Mamoulakis, Avgenakis, Zisis, Heretis) Department of Urology, University General Hospital of Heraklion, University of Crete Medical School, Heraklion 71003, Greece (Gkatzoudi, Duijker) Clinic of Social and Family Medicine, Faculty of Medicine, University of Crete, Heraklion 71003, Greece

(Antypas) First Pediatric Surgery Clinic, Aghia Sophia Children's Hospital, Athens 11527, Greece

(Sofikitis) Department of Urology, University of Ioannina, Medical School, Ioannina 45110, Greece

(Spandidos) Laboratory of Clinical Virology, Medical School, Faculty of Medicine, University of Crete, Heraklion 71003, Greece

(Tsatsakis) Department of Forensic Sciences and Toxicology, Faculty of Medicine, University of Crete, Heraklion 71003, Greece

(Tzonou) Department of Hygiene, Epidemiology and Medical Statistics, University of Athens, Medical School, Athens 11527, Greece

Publisher

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Year of Publication

2017

582.

Disorders of sex development (DSD): not only babies with ambiguous genitalia. A practical guide for surgeons.

Kearsey I., Hutson J.M.

Embase

Pediatric Surgery International. 33 (3) (pp 355-361), 2017. Date of Publication: 01 Mar 2017.

[Review]

AN: 613774214

Introduction and methods: In this review, we describe the common clinical scenarios that may be present to a paediatric surgeon when a patient has a disorder of sex development (DSD). Our aim was to prepare surgeons so that they can respond with correct approaches to diagnose and manage the given situations.

Result(s): DSD present in three distinct clinical situations: in the neonate with some abnormality of the external genitalia; in the child undergoing surgical treatment for inguinal hernia or during open or laparoscopic orchidopexy or during hypospadias correction; and at or after puberty, which may be precocious or delayed or in an adolescent girl with masculinisation at puberty. We describe the clinical features, likely diagnoses and the recommended management pathway in these scenarios.

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Status

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Institution

(Hutson) Department of Urology, The Royal Children's Hospital, Flemington Road, Parkville, Melbourne, VIC 3052, Australia (Kearsey, Hutson) Department of Paediatrics, University of Melbourne, Melbourne, Australia

(Kearsey, Hutson) Surgical Research Laboratory, Murdoch Childrens Research Institute, Melbourne, Australia

Publisher

Springer Verlag (E-mail: service@springer.de)

Year of Publication

2017

583.

Role of tubularization of urethral plate in development of urethrocutaneous fistula post hypospadias repair.

Alsaywid B.S., Mohammedkhalil A.K., Mesawa A.A., Alzahrani S.Y., Askar A.H., Abuznadah W.T., Alrimawi M.H.

Embase

Urology Annals. 9 (2) (pp 141-144), 2017. Date of Publication: April-June 2017.

[Article]

AN: 615631162

Introduction: This study aimed to demonstrate the outcome of hypospadias repair in the pediatric population using the tubularized urethral plate technique and to compare the incidence of fistula between incised and unincised urethral plate. **Methodology:** This is a retrospective cohort study of pediatric patients who had hypospadias repair in a tertiary hospital in Jeddah, Saudi Arabia, between January 2000 and December 2012. They were divided into two main groups according to the status of the urethral plate: Group A included patients who had incision of the urethral plate just before tubularization, and Group B included patients who underwent tubularization without incision.

Result(s): After reviewing 310 medical records, 106 patients were included in the final analysis, with a median age of 2 years (interquartile range = 3 years). There was no statistically significant relationship between types of hypospadias and the development of fistula ($P = 0.26$). In Group A, we identified 87 patients (82%), and in Group B, there were 19 patients (18%). The overall incidence of fistula was 34.9% ($n = 37$). The incidence of fistula in Groups A and B was 36% ($n = 31$) and 32% ($n = 6$), respectively. This difference was not statistically significant. Despite a high fistula rate, only 12 patients (11%) were required to repeat surgery.

Conclusion(s): Incision of the urethral plate did not affect the fistula rate. In comparison to international literature, the incidence of fistula was significantly higher which could be explained by the fact that one-third of those patients had a previous hypospadias repair.

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Institution

(Alsaywid, Mesawa, Alrimawi) Department of Surgery, Urology Section, King Khalid National Guard Hospital, King Abdulaziz Medical City, Saudi Arabia (Alsaywid, Mohammedkhalil,

Abuznadah) Department of Surgery, Urology Section, College of Medicine, King Saud Bin Abdulaziz University for Health Sciences, P.O. Box 2407, Jeddah 21451, Saudi Arabia

(Alzahrani) Department of Surgery, Urology Section, College of Medicine, Umm Al Qura University, Makkah, Saudi Arabia

(Askar) Department of Surgery, Urology Section, College of Medicine, Ibn Sina College, Jeddah, Saudi Arabia

Publisher

Medknow Publications (B9, Kanara Business Centre, off Link Road, Ghatkopar (E), Mumbai 400 075, India)

Year of Publication
2017

584.

Technical tip: Identifying the boundaries of the urethral plate in TIP Urethroplasty.

McGee P., Hough M.

Embase

JPRAS Open. 12 (pp 44-46), 2017. Date of Publication: 01 Jun 2017.

[Article]

AN: 615065773

Status

Embase

Institution

(McGee, Hough) Department of Plastic Surgery, Ninewells Hospital, Dundee DD1 9SY, United Kingdom

Publisher

Elsevier Ltd

Year of Publication

2017

585.

Antiepileptic drugs prescribed in pregnancy and prevalence of major congenital malformations: Comparative prevalence studies.

Petersen I., Collings S.-L., McCrea R.L., Nazareth I., Osborn D.P., Cowen P.J., Sammon C.J.

Embase

Clinical Epidemiology. 9 (pp 95-103), 2017. Date of Publication: 16 Feb 2017.

[Article]

AN: 614854547

Objective: The aim of this study was to examine the prevalence of major congenital malformations associated with antiepileptic drug (AED) treatment in pregnancy.

Patients and Methods: Using data from The Health Improvement Network, we identified women who have given live birth and their offspring. Four subgroups were selected based on the AED treatment in early pregnancy, valproate, carbamazepine, lamotrigine and women not receiving AED treatment. We compared the prevalence of major congenital malformations within children of these four groups and estimated prevalence ratios (PRs) using Poisson regression adjusted for maternal age, sex of child, quintiles of Townsend deprivation score and indication for treatment.

Result(s): In total, 240,071 women were included in the study. A total of 229 women were prescribed valproate in pregnancy, 357 were prescribed lamotrigine and 334 were prescribed carbamazepine and 239,151 women were not prescribed AEDs. Fifteen out of 229 (6.6%) women prescribed valproate gave birth to a child with a major congenital malformation. The figures for lamotrigine, carbamazepine and women not prescribed AEDs were 2.7%, 3.3% and 2.2%, respectively. The prevalence of major congenital malformation was similar for women prescribed lamotrigine or carbamazepine compared to women with no AED treatment in pregnancy. For women prescribed valproate in polytherapy, the prevalence was fourfold higher. After adjustments, the effect of estimates attenuated, but the prevalence remained two- to threefold higher in women prescribed valproate.

Conclusion(s): The results of our study suggest that lamotrigine and carbamazepine are safer treatment options than valproate in pregnancy and should be considered as alternative treatment options for women of childbearing potential and in pregnancy.

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Status

Embase

Institution

(Petersen, Collings, McCrea, Nazareth, Sammon) Department of Primary Care and Population Health, University College London, London, United Kingdom (Petersen) Department of Clinical Epidemiology, Aarhus University, Aarhus N, Denmark

(Collings) OXON Epidemiology, London, United Kingdom

(Osborn) Division of Psychiatry, University College London, London, United Kingdom

(Cowen) University Department of Psychiatry, Warneford Hospital, Oxford, United Kingdom

Publisher

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Year of Publication

2017

586.

Modified PATIO repair for urethrocutaneous fistula post-hypospadias repair: operative technique and outcomes.

Rathod K., Loyal J., More B., Rajimwale A.

Embase

Pediatric Surgery International. 33 (1) (pp 109-112), 2017. Date of Publication: 01 Jan 2017.

[Article]

AN: 612449124

Background: To describe a modification of PATIO repair for urethrocutaneous fistula repair and evaluate its outcome.

Method(s): We studied 15 boys who underwent modified PATIO repair from Jan 2010 to Sept 2015. Parameters studied included age, type of hypospadias, age at first urethroplasty, hypospadias repair technique, number of urethroplasties required, location of fistula, time gap between urethroplasty and fistula repair, method of fistula repair, and outcome of fistula repair.

Result(s): Mean age of the studied patients was 67.6 months (38-139). Type of hypospadias was Coronal = 3, subcoronal = 8, mid-penile = 2, prominal penile = 1, and penoscrotal = 1. Ten patients had single urethroplasty, while two patients had two urethroplasties, details not available for three patients. Average age at urethroplasty was 43.4 months (18-110). 12 patients had Snodgrass repair, Mathieu = 1 patient, tubularised plate repair = 1 patient, and unknown = 1 patient. Location of fistula was coronal in nine patients and subcoronal in six patients. Average operative time was 47.2 min (30-68). Follow-up is available for 12 patients out of which 2 patients had recurrent fistula, one of which was successfully treated by the redo modified PATIO method. Conclusion(s): Modified PATIO method is technically easy method for urethrocutaneous fistula repair, with less operating time and good postoperative outcomes.

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Status

Embase

Author NameID

Rathod, Kirtikumar; ORCID: <http://orcid.org/0000-0002-2031-8812>

Institution

(Rathod, Loyal, More, Rajimwale) University Hospitals of Leicester NHS Trust, Leicester, United Kingdom

Publisher

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Year of Publication

2017

587.

A simple dressing for hypospadias surgery in children.

Mendez-Gallart R., Garcia-Palacios M., Rodriguez-Barca P., Estevez-Martinez E., Carril A.L., Bautista-Casasnovas A.

Embase

Canadian Urological Association Journal. 11 (1-2) (pp E58-E59), 2017. Date of Publication: January-February 2017.

[Article]

AN: 614678112

One of the most controversial aspects of hypospadias surgery is the election of an appropriate wound dressing. In fact, there may be as many different types of dressing as there are types of surgical repair. Here, we describe a new, simple method for hypospadias dressing in children that minimizes painful removal.

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Institution

(Mendez-Gallart, Garcia-Palacios, Rodriguez-Barca, Estevez-Martinez, Carril, Bautista-Casasnovas) Department of Pediatric Surgery and Urology, University Hospital of Santiago, Santiago de Compostela, Spain

Publisher

Canadian Urological Association (E-mail: josephine.sciortino@cua.org)

Year of Publication

2017

588.

Efficacy of using Zaontz urethral stent in hypospadias repair by the Face, Legs, Activity, Cry, Consolability (FLACC) scale: A prospective study.

Ozcan S., Bagcioglu M., Karakan T., Diri M.A., Demirbas A.

Embase

Canadian Urological Association Journal. 11 (1-2) (pp E15-E18), 2017. Date of Publication: January-February 2017.

[Article]

AN: 614678056

Introduction: The developments in hypospadias surgical techniques and materials are intended to improve surgery outcomes and patient comfort. The aim of this study is to determine the effect of the Zaontz urethral stent (ZUS) (Cook Medical) on patient comfort and surgical success rates in children undergoing hypospadias surgery.

Method(s): A feeding tube was used to repair 46 cases of primary distal hypospadias, and ZUS (6F, 8F, and 10F in diameter) was used to repair 31 cases of primary distal hypospadias between December 2009 and June 2011 in our clinic. ZUS was compared with the feeding tube in terms of surgical success rates and patient comfort in assessments made during postoperative periods.

Result(s): The patients with ZUS were followed with a stent for seven days postoperatively, as were the patients with the feeding tube. There was no statistical difference between the two groups in terms of fistula formation ($p>0.05$). Patient comfort was evaluated by the Face, Legs, Activity, Cry, Consolability (FLACC) scale on the first and third postoperative days, and a statistically significant difference was observed in favour of ZUS on the third postoperative day ($p<0.05$).

Conclusion(s): Compared with a feeding tube in hypospadias repair, ZUS does not make any contribution to the urinary fistula rates. However, ZUS may have an advantage in terms of patient comfort in the postoperative followup. On the other hand, the small number of patients and the high price of the ZUS were the most important limitations. Prospective, randomized trials are needed to assess efficacy and cost.

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Status

Embase

Institution

(Ozcan) Artvin State Hospital, Urology Department, Artvin, Turkey (Bagcioglu) Kafkas

University, Faculty of Medicine, Urology Department, Kars, Turkey

(Karakan, Diri, Demirbas) Ankara Training and Research Hospital, Urology Department, Ankara, Turkey

Publisher

Canadian Urological Association (E-mail: josephine.sciortino@cua.org)

Year of Publication

2017

589.

Direct visual internal urethrotomy: Is it a durable treatment option?.

Pal D., Kumar S., Ghosh B.

Embase

Urology Annals. 9 (1) (pp 18-22), 2017. Date of Publication: January-March 2017.

[Article]

AN: 614260902

Objective: To evaluate the long-term success rate of direct vision internal urethrotomy as a treatment for anterior urethral strictures.

Material(s) and Method(s): We retrospectively analyzed the results for patients who underwent internal urethrotomy from January 2009 to January 2014 for anterior urethral strictures. Patients were followed till January 2016. Patients with complicated urethral strictures with a history of previous urethroplasty, hypospadias repair, or previous radiation were excluded from the study, as anticipated low success rate of direct visual internal urethrotomy (DVIU) in these patients. The Kaplan-Meier method was used to analyze stricture-free probability after the first, second, and third urethrotomy.

Result(s): A total of 186 patients were included in this study. Stricture-free rates after first, second, and third urethrotomy were 29.66%, 22.64%, and 13.33%, respectively.

Conclusion(s): Although DVIU may be a management option for anterior urethral stricture disease, it seems that long-term results are disappointing.

Copyright © 2017 Urology Annals.

Status

Embase

Institution

(Pal, Kumar, Ghosh) Department of Urology, Institute of Post Graduate Medical Education and Research, 244, A.J.C. Bose Road, Kolkata, West Bengal 700 020, India

Publisher

Medknow Publications (B9, Kanara Business Centre, off Link Road, Ghatkopar (E), Mumbai 400 075, India)

Year of Publication

2017

590.

Hypospadias reconstruction: 11-year follow-up study of outcomes and patient satisfaction.

Harb A., Page F., Nassimizadeh M., Park A.

Embase

European Journal of Plastic Surgery. 40 (1) (pp 23-28), 2017. Date of Publication: 01 Feb 2017.

[Article]

AN: 611707357

Background: Hypospadias is the most common congenital penile malformation, with ventral and proximal displacement of the urethral meatus. Relevant data on long-term outcomes and satisfaction following hypospadias reconstruction are scarce. The purpose of this study was to evaluate the 11-year surgical results and patient satisfaction ratings in a group of patients with hypospadias who underwent repair over a 6-year period.

Method(s): Retrospective analysis was performed on 122 hypospadias patients operated on by a single surgeon at a single centre between August 2000 and December 2006. Information was collected through a review of patient notes, postal questionnaires and telephone contact with patients. Data collection included the patients' functional results, cosmetic outcomes and satisfaction ratings. Satisfaction was rated on a 1-10 scale and responses were collected over a 2-month period.

Result(s): A total of 122 patients underwent hypospadias repair. The average age was 38 months (range 1-360 months). Two stage procedures were done in 56 patients, with 66 patients undergoing single stage repair. There was a total of 22 complications reported in separate

patients (18 %). Functional and cosmetic outcomes were generally good. Overall patient satisfaction was high (8.7/10).

Conclusion(s): This is one of the largest outcome and patient satisfaction studies following hypospadias repair. The results provide a reliable indicator of complication rates and long-term outcomes and satisfaction rates. Hypospadias repair is associated with high patient satisfaction. The chief complaint among hypospadias patients is of penile shape and inadequate size. Longer prospective follow-up would be beneficial.

Level of Evidence: Level IV, therapeutic study

Copyright © 2016, Springer-Verlag Berlin Heidelberg.

Status

Embase

Institution

(Harb, Nassimizadeh) Plastic & Reconstructive Surgery, Queen Elizabeth Hospital, Mindelsohn Way, Birmingham B152TH, United Kingdom (Page) Plastic & Reconstructive Surgery, John Radcliffe Hospital, Headley Way, Oxford OX39DU, United Kingdom (Park) University Hospital Coventry & Warwickshire, Clifford Bridge Road, Coventry, West Midlands CV22DX, United Kingdom

Publisher

Springer Verlag (E-mail: service@springer.de)

Year of Publication

2017

591.

Double breasting spongioplasty in tubularized/tubularized incise plate urethroplasty: A new technique.

Bhat M., Kumar R., Bhat A.

Embase

Indian Journal of Urology. 33 (1) (pp 58-63), 2017. Date of Publication: January-March 2017.

[Article]

AN: 614049689

Introduction: The main disadvantage of currently described techniques of spongioplasty is superimposition of 3 suture lines (neourethra, spongioplasty, and skin closure) which is likely to

increase the chances of a fistula. We describe and evaluate the results of a double breasting spongioplasty in urethroplasty.

Method(s): A prospective study of 60 primary hypospadias was undertaken by double breasting spongioplasty from August 2012 to March 2014. Mobilization of the urethral plate and the spongiosum is done by creating a plane just proximal to the meatus. Double breasting spongioplasty is done after tubularization of urethral plate. First layer of spongiosum is sutured toward lateral side of the neourethra covering the suture line. A second double breasting layer is sutured over the first layer with its suture line toward the opposite side covering the suture line of the first layer; thus avoiding overlapping of suture lines of all the three layers.

Result(s): Age of the patients varied from 10 months to 16 years with a mean and median of 3.73 and 3.50 years, respectively. Hypospadias was distal, mid, and proximal in 38, 10, and 12 cases, respectively. Chordee was noticed in 35 cases and torque in 28 cases. Overall complication rate was 5% and fistula rate was 1.66%.

Conclusion(s): Double breasting spongioplasty avoids superimposition of suture line and adds two layers of spongiosum over neourethra, thus decreases the chances of urethral fistula and gives cylindrical shape to neourethra.

Status

Embase

Institution

(Bhat) Department of Urology, Dr. S.N. Medical College, Jodhpur, India (Bhat, Bhat, Kumar)

Department of Urology, S.P. Medical College, Bikaner, Rajasthan, India

(Bhat) Department of Sugery, S.P. Medical College, Bikaner, Rajasthan, India

Publisher

Medknow Publications (B9, Kanara Business Centre, off Link Road, Ghatkopar (E), Mumbai 400 075, India)

Year of Publication

2017

592.

Patient Satisfaction with Surgical Outcome after Hypospadias Correction.

Dokter E.M.J., Moues C.M., van Rooij I.A.L.M., van der Biezen J.J.

Embase

European Urology, Supplements. 16 (1) (pp 16-22), 2017. Date of Publication: 01 Jan 2017.

[Review]

AN: 613193343

Background Hypospadias is a congenital malformation in which surgical correction is indicated in most cases. Postoperative patient satisfaction is important because of its influence on the child's psychological development. **Objective** To evaluate patient satisfaction with surgical outcome after hypospadias correction, comparison with physician satisfaction, and the influence of patient and treatment characteristics on satisfaction. **Design, setting, and participants** Seventy-four patients who had hypospadias surgery between 1996 and 2010 in Medical Centre Leeuwarden participated in the study. **Measurements** Patient/parent and physician satisfaction scores were measured using a standardised hypospadias satisfaction questionnaire (maximum score 32), and clinical outcome using the Hypospadias Objective Scoring Evaluation (HOSE; maximum score 16). Patient and treatment characteristics recorded were: preoperative meatal location, preoperative chordee, number of planned surgeries, reconstructive type and timing, patient age during the study, complications, and repeat operations. **Results and limitations** Patients (mean age 10.5 yr) had a lower overall satisfaction score (27.1) than the physicians (30.6). Patients were least satisfied with overall genital appearance (3.1), penile length (3.3), and scars (3.3), whereas physician satisfaction was lowest for scars (3.5). The mean HOSE was 15.4 (standard deviation 0.9). Patients with acceptable HOSE (85%) had higher patient and physician satisfaction compared to patients with unacceptable HOSE. Patient satisfaction was lower among patients with a preoperative proximal meatal location or chordee, and with correction techniques other than the Mathieu approach. Physician satisfaction decreased with increasing patient age and was lower for patients with preoperative chordee, postoperative complications, or repeat operations. **Conclusions** Overall patient and physician satisfaction and clinical outcome scores were relatively high. Patient satisfaction was lower and based on different factors compared to physician satisfaction. Patient satisfaction seems more influenced by aesthetic appearance, but both patients and physicians appear to incorporate clinical characteristics and outcome in their opinion on satisfaction. **Patient summary** Different factors seem to influence patient and physician satisfaction with hypospadias correction, and there is only low correlation between the two. Therefore, patient satisfaction should be evaluated properly instead of making assumptions based on physician satisfaction or clinical outcome only.

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Status

Embase

Institution

(Dokter, Moues, van der Biezen) Department of Plastic Surgery and Hand Surgery, Medical Centre Leeuwarden, Leeuwarden, Netherlands (Dokter, van Rooij) Department for Health Evidence, Radboud University Medical Centre, Nijmegen, Netherlands

Publisher

Elsevier B.V.

Year of Publication

2017

593.

Late Diagnosis of 5-alpha-Reductase Type 2 Deficiency in an Adolescent Girl with Primary Amenorrhoea.

Hummadi A.-R.A., Yahya A.O., Al-Qahtani A.M.

Embase

Sultan Qaboos University medical journal. 17 (2) (pp e218-e220), 2017. Date of Publication: 01 May 2017.

[Article]

AN: 625644694

Deficiency of the 5-alpha-reductase enzyme has been found to affect male sexual development. We report an 18-year-old patient who was referred to an endocrinology clinic in Jizan, Saudi Arabia, in April 2014 with primary amenorrhoea, virilisation and a lack of secondary sex characteristics. As female external genitalia were present at birth, she had been raised as a female. Magnetic resonance imaging revealed no uterine or ovarian tissue in the pelvis and the presence of a scrotal sac. She was diagnosed with 5-alpha-reductase type 2 deficiency, a 46,XY disorder of sexual development. Typically, affected males have pseudovaginal perineoscrotal hypospadias and ambiguous genitalia at birth. Individuals who have been raised as female manifest characteristics of virilisation at puberty, including deepening of the vocal tone, phallus enlargement, scrotal hyperpigmentation and increased muscle mass.

PMID

28690896 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=28690896>]

Institution

(Hummadi, Yahya) Department of Endocrinology & Internal Medicine, King Fahad Central Hospital, Jizan, Saudi Arabia (Al-Qahtani) Department of Family & Community Medicine, College of Medicine, Najran University, Najran, Saudi Arabia

Year of Publication

2017

594.

Hypospadias in male infants - a review.

Zhu X.-Y., Feng D.-C., Han T.

Embase

European review for medical and pharmacological sciences. 21 (4 Supplement) (pp 1-3), 2017.

Date of Publication: 01 Oct 2017.

[Review]

AN: 623178176

The external genitalia problems are cumbersome problems for both doctors and parents, as these abnormalities have a consequent impact on future generations. However, the affected young infants are unaware of the consequences due to immature emotional state. Further, the feeling of being different and inferior in affected young patients could give rise to negative emotions including depression, insecurity, anxiety, powerlessness, etc. These all factors collectively could cause a mental imbalance in the affected children. The present review article is focused on the latest updates in the area.

PMID

29165774 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=29165774>]

Institution

(Zhu) Department of Pediatric Surgery, Xuzhou Children's Hospital, Xuzhou, Jiangsu, China

Year of Publication

2017

595.

Risk Factors for Urethrocutaneous Fistula Repair After Hypospadias Surgery: A Retrospective Study.

Feng J., Yang Z., Tang Y., Chen W., Zhao M.-X., Ma N., Wang W.-X., Xu L.-S., Li Y.-Q.

Embase

Annals of plastic surgery. 79 (6) (pp e41-e44), 2017. Date of Publication: 01 Dec 2017.

[Article]

AN: 623060593

OBJECTIVES: We review our experience in urethrocutaneous fistula (UCF) repair after hypospadias surgery to investigate the risk factors for unsuccessful outcome. **CONCLUSIONS:** Our study suggested that UCF repairs after hypospadias surgery were easier to fail if one of the UCFs was larger than 2 mm or it had been repaired repeatedly. But when both factors existed, the increase of the risk was not statistically significant. The age of patients, site and number of UCFs, complications other than UCF, and postoperative infection were not significantly related to the success rate of UCF repair.

RESULTS: Urethrocutaneous fistula repairs failed in 38 patients (20.5%) at first attempt. In the univariate analysis, size of UCFs ($P = 0.012$), times of UCF repair ($P = 0.008$), and postoperative infection ($P = 0.044$) were statistically related with the outcome of surgery. In the multivariate analysis, only the size of UCFs ($P = 0.030$; adjusted OR, 2.42; 95% CI, 1.09-5.36) and times of repair ($P = 0.008$; adjusted OR, 3.09; 95% CI, 1.35-7.07) were identified as risk factors for unsuccessful outcome. We had consistent results in the stratified analysis. No additive or multiplicative interaction between the 2 risk factors was found.

METHODS: Two hundred eleven patients had undergone UCF repair in our department from January 2005 to December 2015. This study included 185 patients who were followed up for more than 6 months. The age of patients, size, site and number of UCFs, number of UCF repairs, urethral complications other than UCF, and postoperative infection were included as potential risk factors. Binary logistic regression analysis was used for multivariate analysis. Odds ratio (OR) and 95% confidence intervals (CIs) were calculated. Stratified analysis and assessment of additive interaction were performed to have a better understanding of the relation between the risk factors.

PMID

28570466 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=28570466>]

Institution

(Feng) From the 2nd Department, Plastic Surgery Hospital, Chinese Academy of Medical Sciences and Peking Union Medical College, Beijing, China

Year of Publication

2017

596.

Under-reporting of major birth defects in Northwest Russia: a registry-based study.

Kovalenko A.A., Brenn T., Odland J.O., Nieboer E., Krettek A., Anda E.E.

Embase

International journal of circumpolar health. 76 (1) (pp 1366785), 2017. Date of Publication: 2017.

[Article]

AN: 622787497

The objective was to assess the prevalence of selected major birth defects, based on data from two medical registries in Murmansk County, and compare the observed rates with those available for Norway and Arkhangelsk County, Northwest Russia. It included all newborns (≥ 22 completed weeks of gestation) registered in the Murmansk County Birth Registry (MCBR) and born between 1 January 2006 and 31 December 2009 ($n=35,417$). The infants were followed-up post-partum for 2 years through direct linkage to the Murmansk Regional Congenital Defects Registry (MRCDR). Birth defects identified and confirmed in both registries constituted the "cases" and corresponded to one or more of the 21 birth defect types reportable to health authorities in Moscow. The overall prevalence of major birth defects recorded in the MRCDR was 50/10,000 before linkage and 77/10,000 after linkage with the MCBR. Routine under-reporting to the MRCDR of 40% cases was evident. This study demonstrates that birth registry data improved case ascertainment and official prevalence assessments and reduced the potential of under-reporting by physicians. The direct linkage of the two registries revealed that hypospadias cases were the most prevalent among the major birth defects in Murmansk County. ABBREVIATIONS: ICD-10, International Classification of Diseases, 10th revision; MCBR, Murmansk County Birth Registry; MRCDR, Murmansk Regional Congenital Defects Registry; MGC, Murmansk Genetics Center.

PMID

28853333 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=28853333>]

Institution

(Kovalenko, Brenn, Odland, Krettek, Anda) a Department of Community Medicine , UiT The Arctic University of Norway , Tromsø , Norway (Kovalenko) b International School of Public Health , Northern State Medical University , Arkhangelsk , Russia

(Nieboer) c Department of Biochemistry and Biomedical Sciences , McMaster University ,
Hamilton , Canada

(Krettek) d Department of Biomedicine and Public Health, School of Health and Education ,
University of Skovde , Skovde , Sweden

(Krettek) e Department of Internal Medicine and Clinical Nutrition , Institute of Medicine,
Sahlgrenska Academy at University of Gothenburg , Gothenburg , Sweden

Year of Publication

2017

597.

Can We Preserve the Urethral Plate in Proximal Hypospadias Repair?.

Acimi S., Acimi M.A.

Embase

Annals of plastic surgery. 79 (1) (pp 68-72), 2017. Date of Publication: 01 Jul 2017.

[Article]

AN: 621743250

PURPOSE: To know the limits of the preservation of the urethral plate in the correction of the curvature associated with proximal hypospadias. **METHODS:** From January 2002 to January 2015, we treated 224 patients of proximal hypospadias. In 182 cases, we used of the lateral pictures taken during the successive saline erection tests to measure the correction obtained after each stage.

RESULTS: The analysis of the lateral pictures taken at the successive saline erection tests clearly demonstrated that release of the skin and dartos fascia provides an important correction of the chordee. This correction is complete of all curvature lower than 45 degrees and in 3 fourths of cases for curvatures between 45 and 90 degrees. However, for angles 90 degree and more, the release of the skin and dartos fascia was insufficient in more than 73.7% of cases. And the mobilization of the urethral plate with resection of the underlying fibrous tissue realized gives a very low correction of the chordee (0-20 degrees).

CONCLUSIONS: The essential factor responsible of curvature associated with proximal hypopsadias is the fibrosis tissue present on the ventral side of the penis. However, in the severe forms, a short urethral plate must be transected to obtain a complete correction of the chordee.

PMID

28328642 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=28328642>]

Institution

(Acimi) From the Visceral Surgery, Children's Hospital Canastel, Faculty of medicine, University Of Oran, Oran Algeria

Year of Publication

2017

598.

Anterior Urethral Advancement as a Single-Stage Technique for Repair of Anterior Hypospadias: Our Experience.

Gite V.A., Nikose J.V., Bote S.M., Patil S.R.

Embase

Urology journal. 14 (4) (pp 4034-4037), 2017. Date of Publication: 02 Jul 2017.

[Article]

AN: 621714798

PURPOSE: Many techniques have been described to correct anterior hypospadias with variable results. Anterior urethral advancement as one stage technique was first described by Ti Chang Shing in 1984. It was also used for the repair of strictures and urethrocutaneous fistulae involving distal urethra. We report our experience of using this technique with some modification for the repair of anterior hypospadias. **MATERIALS AND METHODS:** In the period between 2013-2015, 20 cases with anterior hypospadias including 2 cases of glanular, 3 cases of coronal, 12 cases of subcoronal and 3 cases of distal penile hypospadias were treated with anterior urethral advancement technique. Patients' age groups ranged from 18 months to 10 years.

Postoperatively, patients were passing urine from tip of neomeatus with satisfactory stream during follow up period of 6 months to 2 years.

RESULTS: There were no major complications in any of our patients except in one patient who developed meatal stenosis which was treated by periodic dilatation. Three fold urethral mobilization was sufficient in all cases.

CONCLUSION: Anterior urethral advancement technique is a single-stage procedure with good cosmetic results and least complications for anterior hypospadias repair in properly selected cases.

PMID

28670672 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=28670672>]

Institution

(Gite, Nikose, Bote, Patil) Department of Urology, Grant Govt. medical college and Sir J.J. group of hospitals, Mumbai, Maharashtra, India

Year of Publication

2017

599.

Variants in congenital hypogonadotrophic hypogonadism genes identified in an Indonesian cohort of 46,XY under-virilised boys.

Ayers K.L., Bouty A., Robevska G., van den Bergen J.A., Juniarto A.Z., Listyasari N.A., Sinclair A.H., Faradz S.M.

Embase

Human genomics. 11 (1) (pp 1), 2017. Date of Publication: 16 Feb 2017.

[Article]

AN: 620722713

BACKGROUND: Congenital hypogonadotrophic hypogonadism (CHH) and Kallmann syndrome (KS) are caused by disruption to the hypothalamic-pituitary-gonadal (H-P-G) axis. In particular, reduced production, secretion or action of gonadotrophin-releasing hormone (GnRH) is often responsible. Various genes, many of which play a role in the development and function of the GnRH neurons, have been implicated in these disorders. Clinically, CHH and KS are heterogeneous; however, in 46,XY patients, they can be characterised by under-virilisation phenotypes such as cryptorchidism and micropenis or delayed puberty. In rare cases, hypospadias may also be present. **RESULTS:** Here, we describe genetic mutational analysis of CHH genes in Indonesian 46,XY disorder of sex development patients with under-virilisation. We present 11 male patients with varying degrees of under-virilisation who have rare variants in known CHH genes. Interestingly, many of these patients had hypospadias.

CONCLUSIONS: We postulate that variants in CHH genes, in particular PROKR2, PROK2, WDR11 and FGFR1 with CHD7, may contribute to under-virilisation phenotypes including hypospadias in Indonesia.

PMID

28209183 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=28209183>]

Institution

(Ayers, Bouty, Robevska, van den Bergen, Sinclair) Murdoch Childrens Research Institute,
Melbourne, Victoria, Australia (Ayers, Sinclair) Department of Paediatrics, University of
Melbourne, Melbourne, Victoria, Australia

(Bouty) The Royal Children's Hospital, Melbourne, Victoria, Australia

(Juniarto, Listyasari, Faradz) Division of Human Genetics, Centre for Biomedical Research,
Faculty of Medicine, Diponegoro University (FMDU), JL. Prof. H. Soedarto, SH, Tembalang,
Semarang, 50275, Central Java, Indonesia

Year of Publication

2017

600.

Association of MAMLD1 single-nucleotide polymorphisms with hypospadias in Chinese Han
population.

Liu Y., Ye W., Wu M., Huang Y.

Embase

Frontiers in bioscience (Landmark edition). 22 (pp 1173-1176), 2017. Date of Publication: 01 Mar
2017.

[Article]

AN: 620112136

Hypospadias is one of the most common congenital malformations among children. Both gene
mutations and environmental factors are thought to be involved in the development of
hypospadias. The mastermind-like domain-containing 1 gene (MAMLD1, formerly CXorf6) is a
new candidate gene and its mutation has been shown in some cases of hypospadias. Here, by
direct sequencing of PCR products, we assessed and found mutations that occur in 220 sporadic
cases of hypospadias. The mutations p.N589S (c.1766A>G) was found at a significantly higher
rate among patients with hypospadias.

PMID

28199199 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=28199199>]

Institution

(Liu, Ye, Wu) Department of Urology, Renji Hospital, School of Medicine, Shanghai Jiaotong University, Shanghai, 200127, China (Huang) Department of Urology, Renji Hospital, School of Medicine, Shanghai Jiaotong University, Shanghai, 200127, China, Hyr88955@yeah.net

Year of Publication

2017

601.

Analysis of preoperative antibiotic prophylaxis in stented, distal hypospadias repair.

Smith J., Patel A., Zamilpa I., Bai S., Alliston J., Canon S.

Embase

The Canadian journal of urology. 24 (2) (pp 8765-8769), 2017. Date of Publication: 01 Apr 2017.

[Article]

AN: 620010580

MATERIALS AND METHODS: We retrospectively reviewed consecutive patients treated with stented, distal hypospadias repair from 2011 to 2014 by three surgeons and compared two groups: patients who received preoperative antibiotics and patients who did not. Patients with a history of previous hypospadias repair were excluded from the study. **RESULTS:** Two hundred twenty-four subjects were identified. Group 1 (135) received preoperative antibiotic and Group 2 (89) did not receive preoperative antibiotics. There was no statistically significant difference in SSI prevalence with 0 patients in Group 1 and 1 patient in Group 2 having a SSI.

CONCLUSION: Although prophylactic antibiotics prior to hypospadias repair are most often used by pediatric urologists, this study demonstrates further evidence that antibiotics prior to this procedure do not appear to lower the rate of SSI. This study is limited by its retrospective nature and disparate mean follow up in the two cohorts. Surgical site infection does not appear to be decreased by prophylactic antibiotic therapy before distal hypospadias repair.

INTRODUCTION: Surgical site infection [SSI] is a risk for any surgical procedure, including hypospadias repair. Prophylactic antibiotic therapy for patients having surgery is often effective in preventing SSIs, but with increasing rates of antibiotic resistance, this practice has been questioned. The objectives of this study are 1) to assess the incidence of SSIs in patients following stented, distal hypospadias repair and 2) to observe for any potential difference in the incidence of SSIs for patients with and without preoperative antibiotic utilization in this setting.

PMID

28436366 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=28436366>]

Institution

(Smith) Pediatric Urology Division, Arkansas Children's Hospital, Little Rock, Arkansas, USA

Year of Publication

2017

602.

Patient-reported long-term outcome after primary hypospadias repair.

Nozohoor Ekmark A., Arnbjornsson E., Svensson H., Hansson E.

Embase

Journal of plastic surgery and hand surgery. 51 (3) (pp 172-177), 2017. Date of Publication: 01 Jun 2017.

[Article]

AN: 620008033

PURPOSE: Long-term evaluation of the surgical outcome after puberty, particularly patient reported outcome is rare in the literature. The aims of this study were to investigation the patients' satisfaction with the long-term results and their views and memories of their childhood surgery and follow-up. METHODS: A modified version of previously used questionnaire was sent to 134 patients ages 18 years or older previously primarily repaired due to hypospadias by one of the authors (HS) between 1989 -2009.

RESULTS: Thirty-nine patients responded. Eighty-two per cent were satisfied with the appearance of their penis, 87% were satisfied with their ability to urinate and their sexual function and 92% were satisfied with the overall surgical results. Ninety per cent of patients were positive to the current duration of our post-pubertal follow-up program or would have preferred an even longer follow-up.

CONCLUSIONS: The majority of patients were satisfied with the long-term surgical results and the duration of follow-up. Despite having problems patients does not always contact the health care system spontaneously, which warrants long-term follow-up.

PMID

27687614 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=27687614>]

Institution

(Nozohoor Ekmark, Svensson, Hansson) a Department of Clinical Sciences in Malmo , Lund University , Lund , Sweden (Nozohoor Ekmark, Arnbjornsson) b Department of Paediatric Surgery , Skane University Hospital , Lund , Sweden (Arnbjornsson) c Department of Clinical Sciences Lund , Lund University , Lund , Sweden (Svensson, Hansson) d Department of Plastic and Reconstructive Surgery , Skane University Hospital , Lund , Sweden
Year of Publication
2017

603.

Factors associated with reoperation in hypospadias surgery - A nationwide, population-based study.

Lu Y.-C., Huang W.-Y., Chen Y.-F., Chang H.-C., Pong Y.-H., Shih T.-H., Huang K.-H.

Embase

Asian journal of surgery. 40 (2) (pp 116-122), 2017. Date of Publication: 01 Apr 2017.

[Article]

AN: 619697547

CONCLUSION: The type of hypospadias and the surgeon caseload volume were significantly associated with the need for secondary hypospadias surgery. The findings of this study provide important information on the outcomes of hypospadias repair for parents and specialists.

BACKGROUND/OBJECTIVE: To analyze the preoperative factors associated with the need for secondary surgery following primary urethroplasty.

METHODS: This study utilized a subset of the National Health Insurance Research Database, which includes the data on all paid medical benefit claims from 1997 to 2007, for 1 million beneficiaries in 2005. We analyzed the claims data for all patients with hypospadias who had undergone primary urethroplasty. The characteristics of the patients, surgeons, and hospitals associated with surgical outcomes were analyzed to investigate possible associations with the need for secondary surgery.

RESULTS: Among 52,705 live male newborn babies, 218 were diagnosed with hypospadias, of whom 89 received repair surgery. A total of 75 (84.3%) male newborn babies received single hypospadias surgery, and 14 (15.7%) underwent more than two surgical procedures. Univariate analysis demonstrated that the type of hypospadias and the surgeon caseload volume were

significantly associated with the need for additional hypospadias surgery ($p = 0.02$ and $p = 0.03$, respectively). In multivariate analysis, the type of hypospadias (distal vs. proximal, odds ratio, 0.25; $p = 0.03$) and the surgeon caseload volume (high vs. low, odds ratio, 0.04; $p = 0.05$) were significantly correlated with secondary operation.

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PMID

26825262 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=26825262>]

Institution

(Lu, Chang, Pong, Huang) Department of Urology, National Taiwan University Hospital, College of Medicine, National Taiwan University, Taipei, Taiwan (Huang) Institute of Health and Welfare Policy, National Yang-Ming University, Taipei, Taiwan; National Health Insurance Committee, Ministry of Health and Welfare, Executive Yuan, Taipei, Taiwan

(Chen) Institute of Health and Welfare Policy, National Yang-Ming University, Taipei, Taiwan

(Shih) Graduate Institute of Clinical Medical Sciences, College of Medicine, Chang Gung University, Taoyuan, Taiwan

Year of Publication

2017

604.

An unanswered question in pediatric urology: the post pubertal persistence of prepubertal congenital penile curvature correction by tunical plication.

Ozkuvanci U., Ziyilan O., Donmez M.I., Yucel O.B., Oktar T., Ander H., Nane I.

Embase

International braz j urol : official journal of the Brazilian Society of Urology. 43 (5) (pp 925-931), 2017. Date of Publication: 01 Sep 2017.

[Article]

AN: 619170659

OBJECTIVE: The aim of this study is to analyze post pubertal results of pre pubertal tunica albuginea plication with non-absorbable sutures in the correction of CPC. **MATERIALS AND**

METHODS: The files of patients who underwent tunica albuginea plication without incision (dorsal/lateral) were retrospectively reviewed. Patients younger than 13 years of age at the time of operation and older than 14 years of age in November 2015 were included. Patients with a

penile curvature of less than 30 degrees & more than 45 degrees and penile/urethral anomalies were excluded. All of the patients underwent surgery followed by circumcision.

RESULTS: The mean age of patients at the time of the operation was 9.7 years (range, 6-13 years). The mean degree of ventral penile curvature measured during the operation was 39 degrees while it was 41 degrees in the lateral curvatures. All of the patients were curvature-free at the end of the operation. At the time of the follow-up examination, the mean age was 16.7 years (range, 14-25 years). Six patients had a straight (0-10 degrees) penis during erection and seven patients had recurrent penile curvatures ranging from 30 to 50 degrees.

CONCLUSION: Pre pubertal tunica albuginea plication of congenital penile curvature (30-45 degrees) with non-absorbable sutures performed without incision is a minimal invasive method especially when performed during circumcision. However, recurrence might be observed in half of the patients after puberty.

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PMID

28727375 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=28727375>]

Institution

(Ozkuvanci, Ziylan, Donmez, Yucel, Oktar, Ander, Nane) Department of Urology, Istanbul Faculty of Medicine, Istanbul University, Istanbul, Turkey

Year of Publication

2017

605.

ECLAMC Study: Prevalence patterns of hypospadias in South America: Multi-national analysis over a 24-year period.

Fernandez N., Perez J., Monterrey P., Poletta F.A., Bagli D.J., Lorenzo A.J., Zarante I.

Embase

International braz j urol : official journal of the Brazilian Society of Urology. 43 (2) (pp 325-334), 2017. Date of Publication: 01 Mar 2017.

[Article]

AN: 617983559

OBJECTIVE: To evaluate prevalence trends of hypospadias in South-America it is essential to perform multicenter and multinational studies with the same methodology. Herein we present

systematic data as part of an international multicenter initiative evaluating congenital malformations in South America over a 24-year period. MATERIALS AND METHODS: A nested case-control study was conducted using the Latin American Collaborative Study of Congenital Malformations (ECLAMC), between January 1989 and December 2012. Cases were stratified as isolated (IH) and non-isolated hypospadias (NIH). Global prevalence was calculated and discriminated by country. Associations between birth weight and gestational age, and NIH distribution by associated abnormality and severity of hypospadias, were analyzed.

RESULTS: A total of 159 hospitals from six countries participated, reporting surveillance on 4.020.384 newborns. A total of 4.537 hypospadias cases were detected, with a global prevalence of 11.3/10.000 newborns. Trend analyses showed in Chile, Brazil and Uruguay a statistically significant increase in prevalence. Analysis of severity and associated anomalies did not find an association for distal cases, but did for proximal (RR=1.64 [95% CI=1.33-2.03]).

CONCLUSION: This is one of only a few Latin American multicenter studies reporting on the epidemiology of hypospadias in South America in the last two decades. Our data adds to evidence suggesting an increase in some countries in the region at different times. There were also variations in prevalence according to severity. This study adds to literature describing associated anomalies at a hospital-based level.

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27802003 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=27802003>]

Institution

(Fernandez, Perez) Departamento de Urologia, Pontificia Universidad Javeriana, Departamento de Urologia, Hospital Universitario San Ignacio, Bogota, Colombia (Fernandez, Zarante)

Instituto de Genetica Humana, Pontificia Universidad Javeriana, Bogota, Colombia

(Monterrey) Departamento de matematicas, Rosario University, Bogota, Colombia

(Poletta) ECLAMC (Estudio Colaborativo Latinoamericano de Malformaciones Congenitas) at Centro de Educacion Medica e Investigaciones Clinicas (CEMIC-CONICET), Buenos Aires, Argentina e Instituto Nacional de Genetica Medica Populacional (INaGeMP), Rio de Janeiro, Brasil

(Bagli, Lorenzo) Division of Urology, Department of Surgery, Hospital for Sick Children and University of Toronto, Toronto, Ontario

Year of Publication

2017

606.

Prenatal Anogenital Distance Is Shorter in Fetuses With Hypospadias.

Gilboa Y., Perlman S., Kivilevitch Z., Messing B., Achiron R.

Embase

Journal of ultrasound in medicine : official journal of the American Institute of Ultrasound in Medicine. 36 (1) (pp 175-182), 2017. Date of Publication: 01 Jan 2017.

[Article]

AN: 617651739

OBJECTIVES: Recent research provides evidence that anogenital distance may serve as a novel metric to assess reproductive potential in men. In children, a shorter anogenital distance was linked with cryptorchidism, hypospadias, and micropenis. Scarce data exist in the literature regarding anogenital distance measurement in the fetus. The aim of our study was to assess whether intrauterine measurement of fetal anogenital distance could assist in the differential diagnosis of male genital anomalies. **METHODS:** Anogenital distance was prospectively measured in all cases referred for suspected isolated abnormal male genitalia. Final diagnoses, confirmed by a pediatric urologist, were compared with anogenital distance prenatal measurements.

RESULTS: Fifty-two cases were referred for evaluation because of suspected male external genital malformation during a 12-month period. Cases with normal-appearing genitalia, associated major malformations, and early severe fetal growth restriction were excluded from the study. Postnatal examination revealed 14 cases of hypospadias in varying severity and 8 cases of a buried penis. All fetuses with hypospadias had an anogenital distance measurement below the fifth percentile. Statistical analysis revealed a significant difference between the normal mean anogenital distance for gestational age versus those with hypospadias (mean $\pm$ -SD, 16.90 $\pm$ -4.08 and 11.68 $\pm$ -3.31 mm, respectively; $P=.001$). No significant difference was found between the normal mean anogenital distance for gestational age versus those with a buried penis (18.85 $\pm$ -2.76 and 19.46 $\pm$ -3.41 mm; $P=.700$).

CONCLUSIONS: Fetuses with hypospadias have a statistically significant shorter anogenital distance compared with the general population. Therefore, anogenital distance may serve as a complementary objective sonographic parameter in the prenatal assessment and counseling of male external genital anomalies.

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Institution

(Gilboa, Perlman, Kivilevitch, Messing, Achiron) Prenatal Diagnostic Unit, Department of Obstetrics and Gynecology, Haim Sheba Medical Center, Tel Hashomer, Israel, affiliated with the Sackler Faculty of Medicine, Tel Aviv University, Tel-Aviv, Israel

Year of Publication

2017

607.

EUROmediCAT signal detection: an evaluation of selected congenital anomaly-medication associations.

Given JE; Loane M; Luteijn JM; Morris JK; de Jong van den Berg LT; Garne E; Addor MC; Barisic I; de Walle H; Gatt M; Klungsoyr K; Khoshnood B; Latos-Bielenska A; Nelen V; Neville AJ; O'Mahony M; Pierini A; Tucker D; Wiesel A; Dolk H.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

British Journal of Clinical Pharmacology. 82(4):1094-109, 2016 10.

[Evaluation Study. Journal Article]

UI: 27028286

AIMS: To evaluate congenital anomaly (CA)-medication exposure associations produced by the new EUROmediCAT signal detection system and determine which require further investigation.

METHODS: Data from 15 EUROCAT registries (1995-2011) with medication exposures at the chemical substance (5th level of Anatomic Therapeutic Chemical classification) and chemical subgroup (4th level) were analysed using a 50% false detection rate. After excluding antiepileptics, antidiabetics, antiasthmatics and SSRIs/psycholeptics already under investigation, 27 associations were evaluated. If evidence for a signal persisted after data validation, a literature review was conducted for prior evidence of human teratogenicity.

RESULTS: Thirteen out of 27 CA-medication exposure signals, based on 389 exposed cases, passed data validation. There was some prior evidence in the literature to support six signals (gastroschisis and levonorgestrel/ethinylestradiol (OR 4.10, 95% CI 1.70-8.53; congenital heart disease/pulmonary valve stenosis and nucleoside/tide reverse transcriptase inhibitors (OR 5.01, 95% CI 1.99-14.20/OR 28.20, 95% CI 4.63-122.24); complete absence of a limb and pregnen (4) derivatives (OR 6.60, 95% CI 1.70-22.93); hypospadias and pregnadien derivatives (OR 1.40, 95% CI 1.10-1.76); hypospadias and synthetic ovulation stimulants (OR 1.89, 95% CI 1.28-2.70).

Antipropulsives produced a signal for syndactyly while the literature revealed a signal for hypospadias. There was no prior evidence to support the remaining six signals involving the ordinary salt combinations, propulsives, bulk-forming laxatives, hydrazinophthalazine derivatives, gonadotropin releasing hormone analogues and selective serotonin agonists.

CONCLUSION: Signals which strengthened prior evidence should be prioritized for further investigation, and independent evidence sought to confirm the remaining signals. Some chance associations are expected and confounding by indication is possible.

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1

Status

MEDLINE

Authors Full Name

Given, Joanne E; Loane, Maria; Luteijn, Johannes M; Morris, Joan K; de Jong van den Berg, Lolkje T W; Garne, Ester; Addor, Marie-Claude; Barisic, Ingeborg; de Walle, Hermien; Gatt, Miriam; Klungsoyr, Kari; Khoshnood, Babak; Latos-Bielenska, Anna; Nelen, Vera; Neville, Amanda J; O'Mahony, Mary; Pierini, Anna; Tucker, David; Wiesel, Awi; Dolk, Helen.

Institution

Given, Joanne E. Centre for Maternal, Fetal and Infant Research, Institute of Nursing and Health Research, Ulster University, United Kingdom. Loane, Maria. Centre for Maternal, Fetal and Infant Research, Institute of Nursing and Health Research, Ulster University, United Kingdom. Luteijn, Johannes M. Wolfson Institute of Preventive Medicine, Queen Mary University of London, United Kingdom.

Morris, Joan K. Wolfson Institute of Preventive Medicine, Queen Mary University of London, United Kingdom.

de Jong van den Berg, Lolkje T W. Department of Pharmacy, University of Groningen, the Netherlands.

Garne, Ester. Paediatric Department, Hospital Lillebaelt, Kolding, Denmark.

Addor, Marie-Claude. Division of Medical Genetics, CHUV, Lausanne, Switzerland.

Barisic, Ingeborg. Department of Medical Genetics and Reproductive Health, Children's University Hospital Zagreb, Croatia.

de Walle, Hermien. Eurocat Northern Netherlands, University of Groningen, University Medical Center Groningen, Department of Genetics, Groningen, the Netherlands.

Gatt, Miriam. Department of Health Information and Research, Guardamangia, Malta.

Klungsoyr, Kari. Medical Birth Registry of Norway, the Norwegian Institute of Public Health and Department of Global Public Health and Primary Care, University of Bergen, Norway.

Khoshnood, Babak. Paris Registry of Congenital Anomalies, Obstetrical, Perinatal and Pediatric Epidemiology Research Team, Center for Biostatistics and Epidemiology, INSERM U1153, Maternite de Port-Royal, PARIS, France.

Latos-Bielenska, Anna. Polish Registry of Congenital Malformations, Department of Medical Genetics, Poznan, Poland.

Nelen, Vera. Provinciaal Instituut voor Hygiene (PIH), Antwerp, Belgium.

Neville, Amanda J. IMER Registry (Emilia Romagna Registry of Birth Defects), Centre for Clinical and Epidemiological Research, University of Ferrara and Azienda Ospedaliero Univerisitarion di Ferrara, Italy.

O'Mahony, Mary. Health Service Executive, Cork, Ireland.

Pierini, Anna. Epidemiology and Health Promotion Macro-Area Working Group, Unit of Environmental Epidemiology and Disease Registries, CNR Institute of Clinical Physiology, Pisa, Italy.

Tucker, David. CARIS - Congenital Anomaly Register and Information Service for Wales, Public Health Wales, Swansea, United Kingdom.

Wiesel, Awi. Mainz Model Birth Registry, University Children's Hospital Mainz, Germany.

Dolk, Helen. Centre for Maternal, Fetal and Infant Research, Institute of Nursing and Health Research, Ulster University, United Kingdom. h.dolk@ulster.ac.uk.

PMID

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2016

608.

Familial forms of disorders of sex development may be common if infertility is considered a comorbidity.

Brauner R; Picard-Dieval F; Lottmann H; Rouget S; Bignon-Topalovic J; Bashamboo A; McElreavey K.

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BMC Pediatrics. 16(1):195, 2016 11 29.

[Journal Article]

UI: 27899089

BACKGROUND: Families with 46,XY Disorders of Sex Development (DSD) have been reported, but they are considered to be exceptionally rare, with the exception of the familial forms of disorders affecting androgen synthesis or action. The families of some patients with anorchia may include individuals with 46,XY gonadal dysgenesis. We therefore analysed a large series of patients with 46,XY DSD or anorchia for the occurrence in their family of one of these phenotypes and/or ovarian insufficiency and/or infertility and/or cryptorchidism.

METHODS: A retrospective study chart review was performed for 114 patients with 46,XY DSD and 26 patients with 46,XY bilateral anorchia examined at a single institution over a 33 year period.

RESULTS: Of the 140 patients, 25 probands with DSD belonged to 21 families and 7 with anorchia belonged to 7 families. Familial forms represent 22% (25/114) of the 46,XY DSD and 27% (7/26) of the anorchia cases. No case had disorders affecting androgen synthesis or action or 5 alpha-reductase deficiency. The presenting symptom was genital ambiguity (n = 12), hypospadias (n = 11) or discordance between 46,XY karyotyping performed in utero to exclude trisomy and female external genitalia (n = 2) or anorchia (n = 7). Other familial affected individuals presented with DSD and/or premature menopause (4 families) or male infertility (4 families) and/or cryptorchidism. In four families mutations were identified in the genes SRY, NR5A1, GATA4 and FOG2/ZFPM2. Surgery discovered dysgerminoma or gonadoblastoma in two cases with gonadal dysgenesis.

CONCLUSIONS: This study reveals a surprisingly high frequency of familial forms of 46,XY DSD and anorchia when premature menopause or male factor infertility are included. It also demonstrates the variability of the expression of the phenotype within the families. It highlights the need to the physician to take a full family history including fertility status. This could be important to identify familial cases, understand modes of transmission of the phenotype and eventually understand the genetic factors that are involved.

Version ID

1

Status

MEDLINE

Authors Full Name

Brauner, Raja; Picard-Dieval, Flavia; Lottmann, Henri; Rouget, Sebastien; Bignon-Topalovic, Joelle; Bashamboo, Anu; McElreavey, Ken.

Institution

Brauner, Raja. Fondation Ophtalmologique Adolphe de Rothschild and Universite Paris Descartes, Paris, France. raja.brauner@wanadoo.fr Picard-Dieval, Flavia. Fondation Ophtalmologique Adolphe de Rothschild and Universite Paris Descartes, Paris, France.

Lottmann, Henri. Assistance Publique-Hopitaux de Paris, Hopital Necker-Enfants Malades, Service de chirurgie viscerale pediatrique, Paris, France.

Rouget, Sebastien. Fondation Ophtalmologique Adolphe de Rothschild and Universite Paris Descartes, Paris, France.

Bignon-Topalovic, Joelle. Human Developmental Genetics, Institut Pasteur, Paris, France.

Bashamboo, Anu. Human Developmental Genetics, Institut Pasteur, Paris, France.

McElreavey, Ken. Human Developmental Genetics, Institut Pasteur, Paris, France.

PMID

<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5129225>

Year of Publication

2016

609.

Circumcision-related tragedies seen in children at the Komfo Anokye Teaching Hospital, Kumasi, Ghana.

Appiah KA; Gyasi-Sarpong CK; Azorliade R; Aboah K; Laryea DO; Otu-Boateng K; Baah-Nyamekye K; Maison PO; Arthur D; Antwi IO; Frimpong-Twumasi B; Yenli EM; Togbe SK; Amoah G.

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BMC Urology. 16(1):65, 2016 Nov 08.

[Journal Article. Observational Study]

UI: 27825332

BACKGROUND: Circumcision is a common minor surgical procedure and it is performed to a varying extent across countries and religions. Despite being a minor surgical procedure, major complications may result from it. In Ghana, although commonly practiced, circumcision-related injuries have not been well documented. This study is to describe the scope of circumcision-related injuries seen at the Komfo Anokye Teaching Hospital in Kumasi, Ghana.

METHODS: The study was conducted at the Urology Unit of the Komfo Anokye Teaching Hospital in Kumasi. Consecutive cases of circumcision-related injuries seen at the unit over an 18 month period were identified and included in the study. Data was collected using a structured

questionnaire. Data was entered and analysed using SPSS version 16. Charts and tables were generated using Microsoft Excel.

RESULTS: A total of 72 cases of circumcision-related injuries were recorded during the 18 month period. Urethrocuteaneous fistula was the commonest injury recorded, accounting for 77.8 % of cases. Other injuries recorded were glans amputations (6.9 %); iatrogenic hypospadias (5.6 %), and epidermal inclusion cysts (2.8 %). The majority of children were circumcised in health facilities (75 %) and nurses were the leading providers (77.8 %). The majority of circumcisions were conducted in the neonatal period (94.7 %).

CONCLUSION: Circumcision-related injuries commonly occurred in the neonatal period. Most of the injuries happened in health facilities. The most common injury recorded was urethrocuteaneous fistula but the most tragic was penile amputation. There is the need for education and training of providers to minimise circumcision-related injuries in Ghana.

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1

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MEDLINE

Author Initials

Appiah, Kwaku Addai Arhin; ORCID: <http://orcid.org/0000-0002-7583-8938>

Authors Full Name

Appiah, Kwaku Addai Arhin; Gyasi-Sarpong, Christian Kofi; Azorliade, Roland; Aboah, Ken; Laryea, Dennis Odai; Otu-Boateng, Kwaku; Baah-Nyamekye, Kofi; Maison, Patrick Opoku Manu; Arthur, Douglas; Antwi, Isaac Opoku; Frimpong-Twumasi, Benjamin; Yenli, Edwin Mwintiereh; Togbe, Samuel Kodzo; Amoah, George.

Institution

Appiah, Kwaku Addai Arhin. Department of Surgery, Komfo Anokye Teaching Hospital, Kumasi, Ghana. addaiarhin@yahoo.com. Gyasi-Sarpong, Christian Kofi. Department of Surgery, School of Medical Sciences-KNUST, Kumasi, Ghana.

Azorliade, Roland. Department of Surgery, Komfo Anokye Teaching Hospital, Kumasi, Ghana.

Aboah, Ken. Department of Surgery, School of Medical Sciences-KNUST, Kumasi, Ghana.

Laryea, Dennis Odai. Public Health Unit, Komfo Anokye Teaching Hospital, Kumasi, Ghana.

Otu-Boateng, Kwaku. Department of Surgery, Komfo Anokye Teaching Hospital, Kumasi, Ghana.

Baah-Nyamekye, Kofi. Department of Surgery, Komfo Anokye Teaching Hospital, Kumasi, Ghana.

Maison, Patrick Opoku Manu. Department of Surgery, Komfo Anokye Teaching Hospital, Kumasi, Ghana.

Arthur, Douglas. Department of Surgery, Komfo Anokye Teaching Hospital, Kumasi, Ghana.

Antwi, Isaac Opoku. Department of Surgery, Komfo Anokye Teaching Hospital, Kumasi, Ghana.

Frimpong-Twumasi, Benjamin. Department of Surgery, Komfo Anokye Teaching Hospital, Kumasi, Ghana.

Yenli, Edwin Mwintierih. Department of Surgery, Tamale Teaching Hospital, Tamale, Ghana.

Togbe, Samuel Kodzo. Department of Surgery, Komfo Anokye Teaching Hospital, Kumasi, Ghana.

Amoah, George. Department of Surgery, Komfo Anokye Teaching Hospital, Kumasi, Ghana.

PMID

<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5101822>

Year of Publication

2016

610.

Psychosexual Outcome Among Iranian Individuals With 5alpha-Reductase Deficiency Type 2 and Its Relationship With Parental Sexism.

Khorashad BS; Aghili Z; Kreukels BP; Hiraifar M; Roshan GM; Afkhamizadeh M; Abbaszadegan MR; Ghaemi N; Khazai B; Cohen-Kettenis PT.

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MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Journal of Sexual Medicine. 13(11):1629-1641, 2016 11.

[Journal Article]

UI: 27693263

INTRODUCTION: Few studies exist on the psychosexual outcome of homogeneous groups of individuals with 5alpha-reductase deficiency type 2 (5alpha-RD-2) and the relation between gender changes and parental hostile and benevolent sexism, which are two components of ambivalent sexism that assume a stereotypical approach toward women in an overtly negative way or a chivalrous, seemingly positive way.

AIM: To report on the psychosexual outcome of individuals with 5alpha-RD-2 and to investigate its relation to the level of parental sexism in a relatively large sample of Iranians with 5alpha-RD-2.

METHODS: Twenty participants (mean age = 19.5 years, SD = 6.345) with a molecularly confirmed diagnosis of 5alpha-RD-2 who were assigned the female gender at birth and raised as female were included in the study. Participants and their parents were interviewed and their

medical records were assessed. Parents also completed the Ambivalent Sexism Inventory (ASI), which includes hostile and benevolent sexism subscales.

MAIN OUTCOME MEASURES: Psychosexual outcome and parental hostile and benevolent sexism measurements.

RESULTS: Twelve of 20 participants (60%) were diagnosed with gender identity disorder not otherwise specified (Diagnostic and Statistical Manual of Mental Disorders, Fourth Edition, Text Revision). Ten of these transitioned to the male gender. The other 10 participants (50%), including the two diagnosed with gender identity disorder not otherwise specified, continued living in a female gender role. When comparing the ASI subscale scores between families of participants who changed their gender and those who did not, no significant difference was found for ASI total and hostile sexism scores, but there was a difference for benevolent sexism ($P = .049$): those whose daughters had changed their gender had higher benevolent sexism scores.

CONCLUSION: The high prevalence of gender change and gender dysphoria reported in the literature was confirmed in this relatively large and homogeneous sample of Iranians with 5-alpha-RD-2 raised as female. Prenatal exposure to testosterone is hypothesized to play a role in the development of gender identity and sexual orientation, but parental attitudes also might be important. Although gender change in individuals with 5-alpha-RD-2 is often attributed to high levels of hostile sexism in some cultures, our findings show this to be associated with benevolent sexism.

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1

Status

MEDLINE

Authors Full Name

Khorashad, Behzad S; Aghili, Zahra; Kreukels, Baudewijntje P C; Hiraifar, Mehran; Roshan, Ghasem M; Afkhamizadeh, Mozhgan; Abbaszadegan, Mohammad Reza; Ghaemi, Nosrat; Khazai, Behnaz; Cohen-Kettenis, P T.

Institution

Khorashad, Behzad S. Evolution and Human Behavior Group, Psychiatry and Behavioral Sciences Research Center, Mashhad University of Medical Sciences, Mashhad, Iran. Aghili, Zahra. Psychiatry and Behavioral Sciences Research Center, Department of Psychiatry, Mashhad University of Medical Sciences, Mashhad, Iran.

Kreukels, Baudewijntje P C. Department of Medical Psychology, VU University Medical Center, Amsterdam, the Netherlands.

Hiradfar, Mehran. Department of Pediatric Surgery, School of Medicine, Mashhad University of Medical Sciences, Mashhad, Iran.

Roshan, Ghasem M. Evolution and Human Behavior Group, Psychiatry and Behavioral Sciences Research Center, Mashhad University of Medical Sciences, Mashhad, Iran.

Afkhamizadeh, Mozghan. Endocrine Research Center, Imam Reza Hospital, School of Medicine, Mashhad University of Medical Sciences, Mashhad, Iran.

Abbaszadegan, Mohammad Reza. Division of Human Genetics, Immunology Research Center, Avicenna Research Institute, Mashhad University of Medical Sciences, Mashhad, Iran.

Ghaemi, Nosrat. Department of Pediatrics, School of Medicine, Mashhad University of Medical Sciences, Mashhad, Iran.

Khazai, Behnaz. Evolution and Human Behavior Group, Psychiatry and Behavioral Sciences Research Center, Mashhad University of Medical Sciences, Mashhad, Iran. Electronic address: behnazkhazai@icloud.com.

Cohen-Kettenis, P T. Department of Medical Psychology, VU University Medical Center, Amsterdam, the Netherlands.

Year of Publication

2016

611.

Quality of life in female patients with bladder exstrophy-epispadias complex: Long-term follow-up.

Bujons A; Lopategui DM; Rodriguez N; Centeno C; Caffaratti J; Villavicencio H.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid

MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Journal of pediatric urology. 12(4):210.e1-6, 2016 Aug.

[Journal Article]

UI: 27290615

INTRODUCTION: Bladder exstrophy-epispadias complex (BEEC) is a congenital malformation that requires multiple surgeries during childhood and life-long follow-up. It often presents with conditions that have the potential to impact quality-of-life (QoL) and psychosocial functioning of affected patients, such as incontinence and sexual dysfunction. The aim of this study is to examine the QoL, urinary continence, sexual function, and overall health in a long-term series of female patients with BEEC.

METHOD: A retrospective review was performed of female patients with BEEC born between 1964 and 1996. Thirty-three patients were asked to complete four validated questionnaires to evaluate their QoL regarding urinary continence and sexual activity (ICIQ, Potenziani-14, and PISQ-12 questionnaires). Nineteen patients completed and returned the questionnaires. The overall QoL was assessed with the SF-36 questionnaire, and demographics were evaluated. Statistical analysis was performed to compare the general QoL with that of the general population.

RESULTS: The median age of the patients was 26 years (range 18-50) (Table). A low to moderate impact of urinary incontinence on QoL was reported by 30% of patients in the ICIQ. Also as a result of urinary incontinence, 84% of patients reported a moderate to severe impact on their sexual lives. Twelve patients got married with eight gestations and five births. SF-36 reported general QoL comparable with that of the general population in five out of eight items. Differences were seen in the mental health, emotional role, and physical functioning items ($p < 0.001$). The main factors for the differences were poor body image, anxiety, and urinary incontinence. A satisfactory social life was reported by 70% of patients.

CONCLUSION: Questionnaire studies on BEEC consistently report a high rate of patients not answering, 43% in the present study. The rarity of the disease determines a small sample size, which diminishes statistical power and could potentially conceal small differences with controls. Despite these limitations, the present findings are consistent with previous studies on BEEC with validated QoL questionnaires: adult women with BEEC suffer psychosocial impact mainly from incontinence, and also from gynecological complications, but their resilience and coping mechanisms allow them to achieve a quasi-normal QoL. Female patients with BEEC reported a normal QoL in five of eight items in the SF-36 questionnaire. Urinary incontinence was the main factor for the moderately decreased QoL according to specific questionnaires.

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1

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Authors Full Name

Bujons, Anna; Lopategui, Diana M; Rodriguez, Nelly; Centeno, Clara; Caffaratti, Jorge; Villavicencio, Humberto.

Institution

Bujons, Anna. Pediatric Urology Department, Fundacio Puigvert, Barcelona, Spain. Electronic address: abujons76@gmail.com. Lopategui, Diana M. Pediatric Urology Department, Fundacio Puigvert, Barcelona, Spain.

Rodriguez, Nelly. Pediatric Urology Department, Fundacio Puigvert, Barcelona, Spain.
Centeno, Clara. Pediatric Urology Department, Fundacio Puigvert, Barcelona, Spain.
Caffaratti, Jorge. Pediatric Urology Department, Fundacio Puigvert, Barcelona, Spain.
Villavicencio, Humberto. Pediatric Urology Department, Fundacio Puigvert, Barcelona, Spain.
Year of Publication
2016

612.

Factors influencing waiting time in hypospadias repair surgery.
Idiodi-Thomas HO; Ademuyiwa AO; Elebute OA; Alakaloko FM; Bode CO.
OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid
MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present
Nigerian Postgraduate Medical Journal. 23(1):21-4, 2016 Jan-Mar.
[Journal Article]
UI: 27098945

AIMS: Hypospadias is a common congenital anomaly of the urethra and phallus, which is not life threatening. It is thus less prioritised in a resource-limited setting. The aim of this study was to evaluate the management of hypospadias by our paediatric surgery unit and determine the factors affecting the delay between presentation and surgical repair while proffering possible solutions to such delay in hypospadias repair surgery.

PATIENTS AND METHODS: This was a retrospective review of all hypospadias repair surgeries carried out by our paediatric surgery unit over a 38-month period, evaluating the period between presentation and first surgery for each patient. Data were analysed using IBM SPSS Statistics for Windows, Version 20.0 (IBM Corp., Released 2011, Armonk, NY, USA). Chi-square test was used to compare categorical variables and $P \leq 0.05$ was considered significant.

RESULTS: In 38 months, 47 operations for hypospadias were carried out on 42 boys. Thirty-seven patients (88.1%) had >3 months delay to surgery. The most frequent contributory factor to delay was unavailable theatre space (13 patients, 31%). Surgical outcome was good in only 16 patients (44%). Of the 16 patients with good outcome, 10 (63%) were operated between the ages of 2-4 years ($P > 0.05$).

CONCLUSIONS: Multiple factors are responsible for delays in carrying out hypospadias surgery in resource-limited environments, notably securing a functional operating theatre suite in the light

of more urgent conditions. To combat these delays, we recommend having dedicated hypospadias repair sessions and surgeons dedicated to hypospadias repair. Hypospadias outreach camps are also proposed.

Version ID

1

Status

MEDLINE

Authors Full Name

Idiodi-Thomas, Hestia O; Ademuyiwa, Adesoji O; Elebute, Olumide A; Alakaloko, Felix M; Bode, Christopher O.

Institution

Ademuyiwa, Adesoji O. Department of Surgery, Paediatric Surgery Unit, Lagos University Teaching Hospital, Idi-Araba; Department of Surgery, Paediatric Surgery Unit, College of Medicine of the University of Lagos, Lagos, Nigeria.

Year of Publication

2016

613.

Commentary to 'Results of distal hypospadias repair after pediatric urology fellowship training: A comparison of junior surgeons to their mentor'.

Palmer LS.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid

MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Journal of pediatric urology. 12(3):163, 2016 Jun.

[Journal Article]

UI: 27012164

Version ID

1

Status

MEDLINE

Authors Full Name

Palmer, Lane S.

Institution

Palmer, Lane S. Cohen Children's Medical, Center of New York, Lake Success, NY, USA.

Electronic address: lpalmermd@gmail.com.

Year of Publication

2016

614.

Kelly procedure for exstrophy or epispadias patients: Anatomical description of the pudendal neurovasculature.

Ben-Chaim J; Hidas G; Wikenheiser J; Landau EH; Wehbi E; Kelly MS; McLorie GA; Khoury AE.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid

MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Journal of pediatric urology. 12(3):173.e1-6, 2016 Jun.

[Journal Article]

UI: 26947891

INTRODUCTION: Adequate penile length in males with bladder exstrophy or epispadias is a major challenge. Kelly previously described a surgical technique of a single stage reconstruction for patients with exstrophy or epispadias that potentially achieves significant penile lengthening by completely detaching the insertion of the corpora cavernosa from the ischiopubic rami.

However, because of the possibility of damage to the pudendal neurovascular supply that may lead to partial or complete penile loss, this technique has not gained popularity. The aim of this study is to describe the surgical anatomic relationship of the pudendal neurovascular bundle (NVB) to the ischiopubic rami and to determine a safer approach to dissection during the Kelly procedure.

METHODS: We performed meticulous dissection in three formalin-fixed and one fresh adult male cadavers to demonstrate the anatomical relationships between the pudendal neurovascular supply of the penis and the cavernosal insertion to the ischiopubic ramus.

RESULTS AND DISCUSSION: We demonstrated the relationships and distance between the NVB and the area of separation between the crus and the ischiopubic ramus at the level of the periosteum. The insertion of the crus to the ischiopubic ramus is inferior lateral, whereas the NVB lies at a superior medial position. This anatomical relationship is best visualized when the dissection is carried out starting from the distal portion of the NVB and proceeding proximally.

This area of the periosteum is avascular and the NVB can be preserved safely as long as the dissection is conducted at that subperiosteal level. Based on this cadaver dissection study, we suppose that detaching the corporal cavernosa from the pubic bones at the subperiosteal level allows for a safe distance to be maintained from the pudendal nerve at all times. We believe that if a surgeon performs the dissection inferiorly and laterally, the corpora cavernosa can be safely detached from the ischiopubic ramus and injury to the pudendal vessels and nerve can be avoided. However, it must be noted that there are limitations to applying the results from this study of normal, adult cadavers to the anatomy of children and adolescents with exstrophy or epispadias, who form the largest proportion of patients who are candidates for this procedure.

CONCLUSION: This anatomical study demonstrates the relationship between the pudendal NVB, the crus, and the ischiopubic ramus. We demonstrated how the separation of the crus from the ischiopubic periosteum might be performed more safely.

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Version ID

1

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Authors Full Name

Ben-Chaim, Jacob; Hidas, Guy; Wikenheiser, Jamie; Landau, Ezekiel H; Wehbi, Elias; Kelly, Maryellen S; McLorie, Gordon A; Khoury, Antoine E.

Institution

Ben-Chaim, Jacob. Hadassah Medical Center, Department of Urology, Jerusalem, Israel. Hidas, Guy. Hadassah Medical Center, Department of Urology, Jerusalem, Israel.

Wikenheiser, Jamie. University of California, Irvine, Department of Urology, Orange, CA, USA.

Landau, Ezekiel H. Hadassah Medical Center, Department of Urology, Jerusalem, Israel.

Wehbi, Elias. University of California, Irvine, Department of Urology, Orange, CA, USA.

Kelly, Maryellen S. University of California, Irvine, Department of Urology, Orange, CA, USA.

McLorie, Gordon A. University of California, Irvine, Department of Urology, Orange, CA, USA.

Khoury, Antoine E. University of California, Irvine, Department of Urology, Orange, CA, USA.

Electronic address: aekhoury@uci.edu.

PMID

<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5922430>

Year of Publication

2016

615.

Diethylstilbestrol-induced mouse hypospadias: "window of susceptibility". [Review]

Sinclair AW; Cao M; Baskin L; Cunha GR.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid

MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Differentiation. 91(1-3):1-18, 2016 Jan-Mar.

[Journal Article. Research Support, N.I.H., Extramural. Review]

UI: 26810244

This review presents published and novel results that define the programming window for diethylstilbestrol (DES)-induced abnormal development of the mouse penis. These data indicate that DES has its greatest effect during the period of most intense penile morphogenesis, namely postnatal days 0-15 (P0-P15). Pregnant mice and their neonatal pups were injected subcutaneously with 200 ng/gbw DES every other day from embryonic day 12-18 (DES E12-E18), postnatal day 0-10 (DES P0-P10), embryonic day 12 to postnatal day 10 (DES E12-P10), postnatal day 5-15 (DES P5-P15), and postnatal day 10-20 (DES P10-P20). Aged-matched controls received sesame oil vehicle. After euthanasia at 10, 15, 20 and 60 days, penises were analyzed by gross morphology, histology and morphometry. Penises of all 5 groups of DES-treated mice were reduced in size, which was confirmed by morphometric analysis of internal penile structures. The most profound effects were seen in the DES E12-P10, DES P0-P10, and DES P5-P15 groups, thus defining a DES "programming window". For all parameters, DES treatment from P10 to P20 showed the most mild of effects. Adverse effects of DES on the MUMP cartilage and erectile bodies observed shortly after the last DES injection reverted to normality in the DES P5-P15, but not in the E12-P10 and P0-P10 groups, in which MUMP cartilage and erectile body malformations persisted into adulthood, again emphasizing a "window of susceptibility" in the early neonatal period.

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Version ID

1

Status

MEDLINE

Authors Full Name

Sinclair, Adriane Watkins; Cao, Mei; Baskin, Laurence; Cunha, Gerald R.

Institution

Sinclair, Adriane Watkins. Department of Urology, University of California San Francisco, 400 Parnassus Avenue, Box A610, San Francisco, CA 94143, United States. Cao, Mei. Department of Urology, University of California San Francisco, 400 Parnassus Avenue, Box A610, San Francisco, CA 94143, United States.

Baskin, Laurence. Department of Urology, University of California San Francisco, 400 Parnassus Avenue, Box A610, San Francisco, CA 94143, United States.

Cunha, Gerald R. Department of Urology, University of California San Francisco, 400 Parnassus Avenue, Box A610, San Francisco, CA 94143, United States. Electronic address: gerald.cunha@ucsf.edu.

PMID

<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4803596>

Year of Publication

2016

616.

Is approximated de-epithelized glanuloplasty beneficial for hypospadiologist?. Es la glanduloplastia con desepitelizacion beneficiosa para el hipospadiologo? <Es la glanduloplastia con desepitelizacion beneficiosa para el hipospadiologo?>

ZakiEldahshoury M; Gamal W; Salem E; Rashed E; Mamdouh A.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Actas Urologicas Espanolas. 40(4):258-62, 2016 May.

[Journal Article]

UI: 26781549

OBJECTIVE: Further evaluation of the cosmetic and functional results of approximated de-epithelized glanuloplasty in different degree of hypospadias.

MATERIAL AND METHODS: This study included 96 male patients (DPH=68 & MPH=28).

Patients selected for repair with glans approximation should have wide urethral plate & grooved glans. All cases were repaired with the classic TIP and glans approximation technique. Follow up was for one year by clinical examination of the meatal shape, size & site, glans shape, skin covering, suture line, urethral catheter, edema & fistula in addition to parent satisfaction.

RESULTS: Mean operative time was 49+/-9minutes. As regards the functional and cosmetic outcomes, success was reported in 95.8%, while failure was in 4.16% in the form of glanular disruption in two patients and subcoronal urethrocuteaneous fistula in another two patients.

CONCLUSION: Glans approximation has many advantages, good cosmetic and functional results, short operative time, less blood loss, no need for tourniquet. Study of a large number of cases and comparing glans approximation with the classic TIP technique.

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Version ID

1

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Authors Full Name

ZakiEldahshoury, M; Gamal, W; Salem, E; Rashed, E; Mamdouh, A.

Institution

ZakiEldahshoury, M. Departamento de Urologia, Sohag University Hospital, Sohag, Egipto.

Electronic address: m.eldahshoury@gmail.com. Gamal, W. Departamento de Urologia, Sohag University Hospital, Sohag, Egipto.

Salem, E. Departamento de Urologia, Sohag University Hospital, Sohag, Egipto.

Rashed, E. Departamento de Urologia, Sohag University Hospital, Sohag, Egipto.

Mamdouh, A. Departamento de Urologia, Sohag University Hospital, Sohag, Egipto.

Year of Publication

2016

617.

Onset Manifestations of Spinal and Bulbar Muscular Atrophy (Kennedy's Disease). [Review]

Finsterer J; Soraru G.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid

MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Journal of Molecular Neuroscience. 58(3):321-9, 2016 Mar.

[Journal Article. Meta-Analysis. Review]

UI: 26482145

Spinal and bulbar muscular atrophy (SBMA) is regarded as a disorder with adult onset between third and fifth decade of life. However, there is increasing evidence that SBMA may start already before adulthood. The present study investigated the following: (1) Which clinical manifestations have been described so far in the literature as initial manifestations? (2) Which was the age at onset of these manifestations? and (3) Is age at onset dependent on the CAG-repeat length if non-motor manifestations are additionally considered? Data for this review were identified by searches of MEDLINE using appropriate search terms. Onset manifestations in SBMA can be classified as frequent, rare, motor, non-motor, or questionable. Frequent are muscle weakness, cramps, fasciculations/twitching, tremor, dysarthria, dysphagia, or gynecomastia. Rare are myalgia, easy fatigability, exercise intolerance, polyneuropathy, hyper-CKemia, under-masculinized genitalia, scrotal hypospadias, microphallus, laryngospasm, or oligospermia. Questionable manifestations include sensory disturbances, cognitive impairment, increased pituitary volume, diabetes, reduced tongue pressure, elevated creatine-kinase, or low androgens/high estrogens. Age at onset is highly variable ranging from 4-76 years. Non-motor manifestations develop usually before motor manifestations. Age at onset depends on what is considered as an onset manifestation. Considering non-motor onset manifestations, age at onset is independent of the CAG-repeat size. In conclusion, age at onset of SBMA depends on what is regarded as onset manifestation. If non-motor manifestations are additionally considered, age at onset is independent of the CAG-repeat length. Since life expectancy is hardly reduced in SBMA, re-investigation of patients from published studies with regard to their initial disease profiles is recommended.

Version ID

1

Status

MEDLINE

Authors Full Name

Finsterer, Josef; Soraru, Gianni.

Institution

Finsterer, Josef. Krankenanstalt Rudolfstiftung, Postfach 20, 1180, Vienna, Austria.

fifigs1@yahoo.de. Soraru, Gianni. Department of Neurosciences, University of Padova, Padova, Italy.

Year of Publication

2016

618.

Prevalence of Congenital Cutaneous Anomalies in 1000 Newborns and a Review of the Literature.

Sarikaya Solak S; Kivanc Altunay I; Tukenmez Demirci G; Can B.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid

MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

American Journal of Perinatology. 33(1):79-83, 2016 Jan.

[Journal Article. Observational Study]

UI: 26171599

OBJECTIVE: There are limited reports studying on congenital cutaneous anomalies in newborns, particularly in Turkey. Some of congenital cutaneous anomalies serve as an important clue for accompanying syndromes or other medical conditions. This study aimed to determine the prevalence of congenital cutaneous anomalies in newborns and to discuss their clinical significance with a brief review of literature.

STUDY DESIGN: A total of 1,000 newborns were examined by a dermatologist in a hospital-based, cross-sectional, prospective study between October 2011 and April 2012.

RESULTS: We observed 11 different congenital cutaneous anomalies in 48 newborns of 1,000 (4.8%). The most commonly seen anomalies were sacral dimple, accessory nipple, acrochordon, hypospadias, open spinal dysraphism, and accessory tragus. None of the newborns with cutaneous anomalies had any association.

CONCLUSIONS: Although congenital cutaneous anomalies are rare in newborns, clinicians should be aware of them as they may be in association with syndromes and other medical conditions. It is also important to give appropriately provided information to avoid parents concerns.

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Version ID

1

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MEDLINE

Authors Full Name

Sarikaya Solak, Sezgi; Kivanc Altunay, Ilknur; Tukenmez Demirci, Gulsen; Can, Burce.

Institution

Sarikaya Solak, Sezgi. Department of Dermatology, Sisli Etfal Training and Research Hospital, Istanbul, Turkey. Kivanc Altunay, Ilknur. Department of Dermatology, Sisli Etfal Training and Research Hospital, Istanbul, Turkey.

Tukenmez Demirci, Gulsen. Department of Dermatology, Faculty of Medicine, Baskent University, Istanbul Hospital, Istanbul, Turkey.

Can, Burce. Department of Dermatology, Istanbul Medeniyet University, Goztepe Training and Research Hospital, Istanbul, Turkey.

Year of Publication

2016

619.

A Simple Method for Closure of Urethrocutaneous Fistula after Tubularized Incised Plate Repair: Preliminary Results.

Shirazi M; Ariafar A; Babaei AH; Ashrafzadeh A; Adib A.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid

MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Nephrourology Monthly. 8(6):e40371, 2016 Nov.

[Journal Article]

UI: 27933278

BACKGROUND: Urethrocutaneous fistula (UCF) is the most prevalent complication after hypospadias repair surgery. Many methods have been developed for UCF correction, and the best technique for UCF repair is determined based on the size, location, and number of fistulas, as well as the status of the surrounding skin.

OBJECTIVES: In this study, we introduced and evaluated a simple method for UCF correction after tubularized incised plate (TIP) repair.

METHODS: This clinical study was conducted on children with UCFs ≤ 4 mm that developed after TIP surgery for hypospadias repair. The skin was incised around the fistula and the tract was released from the surrounding tissues and the dartos fascia, then ligated with 5 - 0 polydioxanone (PDS) sutures. The dartos fascia, as the second layer, was covered on the fistula tract with PDS thread (gauge 5 - 0) by the continuous suture method. The skin was closed with 6 - 0 Vicryl sutures. After six months of follow-up, surgical outcomes were evaluated based on fistula relapse and other complications.

RESULTS: After six months, relapse occurred in only one patient, a six-year-old boy with a single 4-mm distal opening, who had undergone no previous fistula repairs. Therefore, in 97.5% of the cases, relapse was non-existent. Other complications, such as urethral stenosis, intraurethral

obstruction, and epidermal inclusion cysts, were not seen in the other patients during the six-month follow-up period.

CONCLUSIONS: This repair method, which is simple, rapid, and easily learned, is highly applicable, with a high success rate for the closure of UCFs measuring up to 4 mm in any location.

Version ID

1

Status

PubMed-not-MEDLINE

Authors Full Name

Shirazi, Mehdi; Ariafar, Ali; Babaei, Amir Hossein; Ashrafzadeh, Abdosamad; Adib, Ali.

Institution

Shirazi, Mehdi. Department of Urology, School of Medicine, Shiraz University of Medical Sciences, Shiraz, IR Iran. Ariafar, Ali. Urology Oncology Research Center, Shiraz University of Medical Sciences, Shiraz, IR Iran.

Babaei, Amir Hossein. Student Research Committee, Shiraz University of Medical Sciences, Shiraz, IR Iran.

Ashrafzadeh, Abdosamad. Department of Urology, School of Medicine, Shiraz University of Medical Sciences, Shiraz, IR Iran.

Adib, Ali. Student Research Committee, Shiraz University of Medical Sciences, Shiraz, IR Iran.

PMID

<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5135729>

Year of Publication

2016

620.

Maternal inflammatory bowel disease and hypospadias in male offspring: a population-based study in Denmark.

Andersen AB; Ehrenstein V; Erichsen R; Froslev T; Sorensen HT.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid

MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

BMJ Open Gastroenterology. 3(1):e000121, 2016.

[Journal Article]

UI: 27933203

BACKGROUND: The occurrence of inflammatory bowel disease (IBD) and hypospadias has been concurrently increasing, possibly through shared environmental risk factors such as endocrine disrupting compounds. Also, maternal IBD may disturb the normal development of the fetal reproductive tract. However, whether maternal IBD increases the risk of hypospadias in male offspring is unknown. We compared hypospadias risk in sons of mothers with and without IBD.

METHODS: We used Danish nationwide population-based registries to conduct a longitudinal prevalence study including all live-born boys from 1979 through 2009. We computed HRs, as estimates of prevalence ratios (PRs), with 95% CIs for hypospadias, using Cox proportional hazards regression, while adjusting for measured confounding.

RESULTS: Among 966 038 live-born boys, 4688 (0.5%) had a mother with a history of IBD diagnosis before the relevant childbirth. Among the boys with maternal IBD, 36 (0.8%) were diagnosed with hypospadias any time after birth, whereas 6112 (0.6%) sons of mothers without IBD diagnosis had hypospadias (adjusted PR: 1.20, (95% CI 0.86 to 1.67). Adjusted PRs for maternal Crohn's disease and ulcerative colitis were 1.38 (95% CI 0.83 to 2.29) and 1.10 (95% CI 0.71 to 1.68), respectively. Analyses defining hypospadias diagnosis recorded <6 months postpartum showed similar results.

CONCLUSIONS: We found no convincing evidence of an association between maternal IBD and hypospadias.

Version ID

1

Status

PubMed-not-MEDLINE

Authors Full Name

Andersen, Ane Birgitte Telen; Ehrenstein, Vera; Erichsen, Rune; Froslev, Trine; Sorensen, Henrik Toft.

Institution

Andersen, Ane Birgitte Telen. Department of Clinical Epidemiology, Aarhus University Hospital, Aarhus N, Denmark. Ehrenstein, Vera. Department of Clinical Epidemiology, Aarhus University Hospital, Aarhus N, Denmark.

Erichsen, Rune. Department of Clinical Epidemiology, Aarhus University Hospital, Aarhus N, Denmark.

Froslev, Trine. Department of Clinical Epidemiology, Aarhus University Hospital, Aarhus N, Denmark.

Sorensen, Henrik Toft. Department of Clinical Epidemiology, Aarhus University Hospital, Aarhus N, Denmark.

PMID

<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5128952>

Year of Publication

2016

621.

Accurate Diagnosis of Severe Hypospadias Using 2D and 3D Ultrasounds.

Rodriguez Fernandez V; Lopez Ramon Y Cajal C; Marin Ortiz E; Sarmiento Carrera N.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Case Reports in Obstetrics and Gynecology. 2016:2450341, 2016.

[Journal Article]

UI: 27774326

The hypospadias is the most common urogenital anomaly of male neonates but the prenatal diagnosis of this is often missed before birth. We present the prenatal diagnosis of a severe penoscrotal hypospadias using 2D and 3D ultrasounds. 3D sonography allowed us the best evaluation of the genitals and their anatomical relations. This ample detailed study allowed us to show the findings to the parents and the pediatric surgeon and to configure the best information about the prognosis and surgical treatment.

Version ID

1

Status

PubMed-not-MEDLINE

Author Initials

Rodriguez Fernandez, Vanesa; ORCID: <https://orcid.org/0000-0001-8890-0617>

Authors Full Name

Rodriguez Fernandez, Vanesa; Lopez Ramon Y Cajal, Carlos; Marin Ortiz, Elena; Sarmiento Carrera, Nerea.

Institution

Rodriguez Fernandez, Vanesa. Service of Obstetrics and Gynecology, Alvaro Cunqueiro Hospital, Vigo, Spain. Lopez Ramon Y Cajal, Carlos. Service of Obstetrics and Gynecology, Alvaro Cunqueiro Hospital, Vigo, Spain.

Marin Ortiz, Elena. Service of Obstetrics and Gynecology, Alvaro Cunqueiro Hospital, Vigo, Spain.

Sarmiento Carrera, Nerea. Service of Pediatrics, Alvaro Cunqueiro Hospital, Vigo, Spain.

PMID

<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5059589>

Year of Publication

2016

622.

Proximal hypospadias: we aren't always keeping our promises. [Review]

Long CJ; Canning DA.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid

MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

F1000Research. 5, 2016.

[Review. Journal Article]

UI: 27746897

Hypospadias surgery is a humbling art form. The evolution of surgical techniques has made distal hypospadias outcomes favorable, but recent publications suggest that our complication rates for proximal hypospadias are much higher than previously reported. To explain these shortcomings, we examine the literature and focus on the lack of standardized documentation, the subsequent inability to objectify the severity of the phenotype, and the underestimation of complications due to lack of long-term follow up. The variability in surgical technique and the fact that the literature abounds with small case series from single institutions also limits our ability to compare outcomes. We believe that the use of standardized and scored phenotype assessments from diagnosis through the extended postoperative period will allow for improved scientific assessment of outcomes. This will facilitate multi-institution collaboration and tabulation of outcomes, allowing rapid data accumulation and assessment for this rare disorder. As surgeons, we must follow boys through puberty into adulthood and must honestly report our results in order to advance our surgical approach to this complicated problem.

Version ID

1

Status

PubMed-not-MEDLINE

Authors Full Name

Long, Christopher J; Canning, Douglas A.

Institution

Long, Christopher J. Division of Urology, The Children's Hospital of Philadelphia, Perelman School of Medicine at the University of Pennsylvania, Philadelphia, PA, USA. Canning, Douglas A. Division of Urology, The Children's Hospital of Philadelphia, Perelman School of Medicine at the University of Pennsylvania, Philadelphia, PA, USA.

PMID

<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5040151>

Year of Publication

2016

623.

The long-term outcomes after staged repair of exstrophy-epispadias complex.

Khemchandani SI.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Journal of Indian Association of Pediatric Surgeons. 21(4):158-163, 2016 Oct-Dec.

[Journal Article]

UI: 27695206

INTRODUCTION: Classic bladder exstrophy (BE) is a rare malformation of the genito-urinary tract affecting 1:50,000 to 1:100,000 live births. The surgical reconstruction of the BE-epispadias complex is challenging for the most experienced pediatric urologists, surgeons, and orthopedists.

PURPOSE: To assess the success of staged reconstruction of the BE and long-term effects on the upper urinary tract, renal function, and continence.

MATERIALS AND METHODS: This is retrospective study; between 1994 and 2013, 30 patients with BE have undergone stage 1 repair at the institute. Eighteen male patients have been operated for epispadias repair and thirteen patients have undergone Guy Leadbetter bladder neck reconstruction. Three patients required augmentation cystoplasty one child is continent after epispadias repair only and one child attained continence after single-stage repair.

RESULTS: Hence, out of 17 patients, 14 are socially continent, four patients require clean intermittent self-catheterization for bladder emptying. Four patients, who are coming for regular follow-up, are awaiting continence procedure. Two patients who underwent augmentation cystoplasty are on hemodialysis for renal failure and one child has altered renal function. CONCLUSION: In our experience, the modern staged repair offers a low risk of renal scarring with acceptable continence opportunity with acceptable cosmetic appearance of external genitalia in the males and females.

Version ID

1

Status

PubMed-not-MEDLINE

Authors Full Name

Khemchandani, Sajni I.

Institution

Khemchandani, Sajni I. Department of Urology, Institute of Kidney Diseases and Research Center and Dr. H L Trivedi Institute of Transplantation Sciences, Civil Hospital Campus, Ahmedabad, Gujarat, India.

PMID

<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4980875>

Year of Publication

2016

624.

Female hypospadias presenting with urinary retention and renal failure in an adolescent: uncommon and late presentation with significant hidden morbidity.

Prakash G; Singh M; Goel A; Jhanwar A.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

BMJ Case Reports. 2016, 2016 Sep 22.

[Journal Article]

UI: 27659910

An 11-year-old girl presented with poor urinary stream with vaginal voiding since childhood. She had palpable bladder and raised serum creatinine (671 micro mol/L). Suprapubic catheterisation was performed after a failed urethral attempt. Following catheterisation she improved symptomatically. She underwent cystoscopy through the suprapubic portal as a guide wire could be negotiated through bladder neck; external urethral meatus was found as a dimple higher up in anterior vaginal wall. Female hypospadias is usually an incidental diagnosis during catheterisation for various reasons. Diagnosis could be established only after antegrade cystoscopy. Anecdotal cases have been reported in the literature with similar presentation. Early diagnosis is essential as renal failure could be a late presentation if patients continue to void with stenosed meatus. Long-term follow-up is needed as these patients may need definitive management later on.

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Version ID

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Status

PubMed-not-MEDLINE

Authors Full Name

Prakash, Gaurav; Singh, Manmeet; Goel, Apul; Jhanwar, Ankur.

Institution

Prakash, Gaurav. Department of Urology, King George's Medical University, Lucknow, Uttar Pradesh, India. Singh, Manmeet. Department of Urology, King George's Medical University, Lucknow, Uttar Pradesh, India.

Goel, Apul. Department of Urology, King George's Medical University, Lucknow, Uttar Pradesh, India.

Jhanwar, Ankur. Department of Urology, King George's Medical University, Lucknow, Uttar Pradesh, India.

PMID

<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5051457>

Year of Publication

2016

Preoperative Testosterone Therapy Prior to Surgical Correction of Hypospadias: A Review of the Literature. [Review]

Krishnan A; Chagani S; Rohl AJ.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid

MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Cureus. 8(7):e677, 2016 Jul 08.

[Journal Article. Review]

UI: 27551655

Hypospadias is a congenital anomaly of the male urethra that causes significant morbidity in the pediatric male population. The mainstay of treatment is hypospadias repair surgery. However, given the common co-occurrence of microphallus in these patients, surgery remains difficult without adequate tissue for proper reconstruction of the urethra. The use of preoperative testosterone therapy, parenterally or topically, has been a topic of study for several years in urologic literature. This literature review aims to summarize the different studies that have been conducted to address whether preoperative testosterone therapy is beneficial, inconsequential, or detrimental to the surgical and cosmetic outcomes of hypospadias repair as well as to address the differences in routes of administration.

Version ID

1

Status

PubMed-not-MEDLINE

Authors Full Name

Krishnan, Arvind; Chagani, Sean; Rohl, Austin J.

Institution

Krishnan, Arvind. University of Central Florida College of Medicine. Chagani, Sean. University of Central Florida College of Medicine.

Rohl, Austin J. University of Central Florida College of Medicine.

PMID

<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4977217>

Year of Publication

2016

The success of tubularized-incised plate urethroplasty in adults and children.

Polat H; Gulacti U; Gok A; Yucel MO; Cift A; Lok U; Benlioglu C.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid

MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Springerplus. 5(1):689, 2016.

[Journal Article]

UI: 27350923

PURPOSE: Hypospadias repair is rarely performed in adults. It is believed that the success rate is lower in adulthood. We aimed to compare the success rate of primary hypospadias repair with tubularized-incised plate (TIP) urethroplasty in adults and children.

PATIENTS AND METHODS: The databases of consecutive boys and adults who were treated with TIP urethroplasty for primary hypospadias between 2012 and 2015 were evaluated. All operations in the boys and adult patients were performed by a single surgeon. We considered urethroplasty complications to include a urethrocutaneous fistula, neourethral stricture, meatal stenosis, diverticulum, and glans dehiscence. Urine flow was also evaluated using uroflowmetry.

RESULTS: Seventy-seven consecutive patients underwent surgery by a single surgeon in the last 3 years for hypospadias repair. Nineteen of these patients were adults. Urethrocutaneous fistulae developed in 2 of the 19 (10.5 %) adults, and 3 of the 58 (5.2 %) boys. In addition, there were urinary tract infections in 2 (3.4 %) children, meatal stenosis in 1 (1.7 %) child, and glans dehiscence in 1 (5.3 %) adult. Uroflowmetry was normal in all patients. There was no difference in outcomes between boys and adults.

CONCLUSION: Our data showed that the success rate of hypospadias repair with TIP urethroplasty is similar in adults and children. TIP urethroplasty is associated with good results in adults and boys.

Version ID

1

Status

PubMed-not-MEDLINE

Authors Full Name

Polat, Haci; Gulacti, Umut; Gok, Alper; Yucel, Mehmet Ozgur; Cift, Ali; Lok, Ugur; Benlioglu, Can.

Institution

Polat, Haci. Department of Urology, Faculty of Medicine, Adiyaman University, Adiyaman, Turkey.

Gulacti, Umut. Department of Emergency, Faculty of Medicine, Adiyaman University, Adiyaman, Turkey.

Gok, Alper. Department of Urology, Faculty of Medicine, Adiyaman University, Adiyaman, Turkey.

Yucel, Mehmet Ozgur. Department of Urology, Faculty of Medicine, Adiyaman University, Adiyaman, Turkey.

Cift, Ali. Department of Urology, Faculty of Medicine, Adiyaman University, Adiyaman, Turkey.

Lok, Ugur. Department of Emergency, Faculty of Medicine, Adiyaman University, Adiyaman, Turkey.

Benlioglu, Can. Department of Urology, Faculty of Medicine, Adiyaman University, Adiyaman, Turkey.

PMID

<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4899396>

Year of Publication

2016

627.

Age of the mother as a risk factor and timing of hypospadias repair according to severity.

Jorge JC; Perez-Brayfield MR; Torres CM; Pineyro-Ruiz C; Torres N.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid

MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

SOJ Urology and Nephrology Open Access. 2(1), 2016.

[Journal Article]

UI: 27331196

BACKGROUND & OBJECTIVES: Hypospadias is characterized by a displacement of the urethral opening in males that can change from the typical position within the glans penis to a subcoronal position (Type I), to anywhere along the ventral shaft (Type II), to penoscrotal, scrotal, or perineal positions (Type III). We and others have previously reported that age of the mother (≥ 40 years old) is a risk factor for having a child with hypospadias, but there is a scarcity of reports on whether such risk is higher for having a child with the mild (Type I) or the more severe forms (Types II and III). In addition, we aimed to assess the timing of hypospadias repair according to severity.

METHODS: Parents of children with hypospadias were interviewed by using a series of questionnaires ($n = 128$ cases). Severity was confirmed in the clinic and age of the mother was self-reported. Number of surgeries, age of child by the first and the last intervention was also assessed. Ordered logistic regression and the Brant test were employed to calculate risk between

mild (Type I) and severe cases (Types II and III), and the assumption of proportional odds, respectively. The Mann-Whitney U Test was used to compare number of surgeries and age by the last repair between mild and severe cases. One-way ANOVA was employed to compare age of the child at the time of first surgery across severities (Types I - III).

RESULTS: Women ≥ 40 years of age are 3.89 times [95% CI: 1.20-12.64] at a higher risk for having a child with the more severe forms of the condition than younger women. Repair of Type I was accomplished with 1 intervention whereas more severe cases required 1 - 4 (2 $\pm$ 0.5) surgical interventions. The timing for hypospadias repair of Type I cases occurred at an average age of 16.2 $\pm$ 4.88 months, of Type II cases occurred at an average age of 20.3 $\pm$ 8.15 months whereas the average age of the first hypospadias repair among Type III cases was 12.68 $\pm$ 2.52 months. Number of surgeries according to severity ($p \leq 0.0018$, z-ratio = 2.91) and age difference for the timing of last repair ($p \leq 0.045$, z-ratio = 1.69) were statistically different, but not the age difference for the first repair.

CONCLUSIONS: Increased maternal age is associated with the most severe forms of hypospadias. There is room for improvement for the timing of hypospadias repair according to severity.

Version ID

1

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PubMed-not-MEDLINE

Authors Full Name

Jorge, Juan Carlos; Perez-Brayfield, Marcos Raymond; Torres, Camille M; Pineyro-Ruiz, Coriness; Torres, Naillil.

Institution

Jorge, Juan Carlos. Department of Anatomy and Neurobiology, Officer of Diversity and Inclusion, School of Medicine, University of Puerto Rico, San Juan, Puerto Rico. Perez-Brayfield, Marcos Raymond. Division of Urology, School of Medicine, University of Puerto Rico, San Juan, Puerto Rico.

Torres, Camille M. MD program, School of Medicine, University of Puerto Rico, San Juan, Puerto Rico.

Pineyro-Ruiz, Coriness. Department of Anatomy and Neurobiology, PhD Program, School of Medicine, University of Puerto Rico, San Juan, Puerto Rico.

Torres, Naillil. Biology Program, Rio Piedras Campus, University of Puerto Rico, San Juan, Puerto Rico.

PMID

<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4908969>

Year of Publication

628.

Does Preputial Reconstruction Increase Complication Rate of Hypospadias Repair? 20-Year Systematic Review and Meta-Analysis.

Castagnetti M; Gnech M; Angelini L; Rigamonti W; Bagnara V; Esposito C.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Frontiers in Pediatrics. 4:41, 2016.

[Journal Article]

UI: 27200322

INTRODUCTION: We performed a systematic review of the literature on preputial reconstruction (PR) during hypospadias repair to determine the cumulative risk of preputial skin complications and the influence of PR on urethroplasty complications, namely, fistula formation and overall reoperation rate of the repair.

MATERIALS AND METHODS: A systematic search of the literature published after 06/1995 was performed in 06/2015 using the keyword "hypospadias." Only studies on the outcome of PR in children, defined as dehiscence of the reconstructed prepuce or secondary phimosis needing circumcision, were selected. A meta-analysis of studies comparing PR vs. circumcision was performed for the outcomes "hypospadias fistula formation" and "reoperation rate."

RESULTS: Twenty studies were identified. Nineteen reported the outcome of PR in 2115 patients. Overall, 95% (2016/2115) of patients undergoing PR had distal hypospadias. The cumulative rate of PR complications was 7.7% (163/2115 patients), including 5.7% (121/2115 patients) preputial dehiscences and 1.5% (35/2117 reported patients) secondary phimoses needing circumcision. A meta-analysis of seven studies comparing patients undergoing PR vs. circumcision showed no increased risk of urethral fistula formation associated with PR, odds ratio (OR) (Mantel-Haenszel, Fixed effect, 95% CI), 1.25 (0.80-1.97). Likewise, two studies comparing the overall reoperation rate did not show an increased risk of reoperation associated with PR, OR (Mantel-Haenszel, Random effect, 95% CI), 1.27 (0.45-3.58).

CONCLUSION: PR carries an 8% risk of specific complications (dehiscence of reconstructed prepuce or secondary phimosis needing circumcision), but does not seem to increase the risk of urethroplasty complications, and the overall reoperation rate of hypospadias repair.

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1

Status

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Authors Full Name

Castagnetti, Marco; Gnech, Michele; Angelini, Lorenzo; Rigamonti, Waifro; Bagnara, Vincenzo; Esposito, Ciro.

Institution

Castagnetti, Marco. Section of Paediatric Urology, Urology Unit, University Hospital of Padova, Padua, Italy. Gnech, Michele. Section of Paediatric Urology, Urology Unit, University Hospital of Padova, Padua, Italy.

Angelini, Lorenzo. Section of Paediatric Urology, Urology Unit, University Hospital of Padova, Padua, Italy.

Rigamonti, Waifro. Pediatric Surgery, Institute for Maternal and Child Health, IRCCS "Burlo Garofolo", Trieste, Italy.

Bagnara, Vincenzo. Department of Paediatric Surgery, Policlinico "G.B. Morgagni", Catania, Italy.

Esposito, Ciro. Department of Paediatrics, Federico II University of Naples, Naples, Italy.

Comments

Comment in (CIN)

PMID

<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4848293>

Year of Publication

2016

629.

Bladder exstrophy-epispadias complex and the role of methylenetetrahydrofolate reductase C677T polymorphism: A case control study.

Raman VS; Bajpai M; Ali A.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid

MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Journal of Indian Association of Pediatric Surgeons. 21(1):28-32, 2016 Jan-Mar.

[Journal Article]

UI: 26862292

PURPOSE: The Bladder Exstrophy-Epispadias Complex (BEEC) is the most serious form of midline abdominal malformation. The etiology of BEEC is unknown and is thought to be multifactorial. Methylenetetrahydrofolate reductase (MTHFR) polymorphism C677T is strongly associated with other midline abnormalities such as neural tube defects. No proper case-control study existed comparing MTHFR polymorphism with BEEC. We sought to find an association with MTHFR polymorphism and patients with bladder exstrophy (BE).

MATERIALS AND METHODS: The design of the study was a case-control study, involving 50 children with BEEC and 50 normal healthy school children. Genetic analysis for MTHFR 677 polymorphism was carried out after DNA extraction and polymerase chain reaction amplification. Epidemiological analysis was done by using the birth defect questionnaire on parents of BEEC.

RESULTS: Forty-two classical BE, two cloacal exstrophies (CE), four epispadias, and two exstrophy variant patients were a part of this study. Severe variety of BE had a significant association with C667T MTHFR polymorphism as compared to the normal control population ($P = 0.01$).

CONCLUSION: C677T MTHFR polymorphism has a strong association with severe variety (CE) of BEEC occurrence.

Version ID

1

Status

PubMed-not-MEDLINE

Authors Full Name

Raman, Venkat Shankar; Bajpai, Minu; Ali, Abid.

Institution

Raman, Venkat Shankar. Department of Pediatric Surgery, Army Hospital R&R, Dhaula Kuan, India. Bajpai, Minu. All India Institute of Medical Sciences, New Delhi, India.

Ali, Abid. All India Institute of Medical Sciences, New Delhi, India.

PMID

<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4721125>

Year of Publication

2016

Concordance of Expert and Parental Opinion about Hypospadias Surgical Outcome Is Severity Dependent.

Perez-Brayfield MR; Jorge JC; Aviles LA; Diaz J; Ortiz V; Morales-Cosme W.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present
Frontiers in Pediatrics. 4:2, 2016.

[Journal Article]

UI: 26835440

INTRODUCTION: Hypospadias is a male congenital condition where the opening of the urethral meatus is not located in the typical anatomical position. It has been a challenge for empirical studies to ascertain the level of concordance of opinion among parents and urologists with regard to surgical outcomes according to hypospadias severity.

MATERIALS AND METHODS: Parents of children who had undergone hypospadias repair were recruited for this study (n = 104). A set of questionnaires that included some items with Likert scale were created to evaluate postsurgical satisfaction by parents and urologists. SAHLISA-50, a validated instrument for adult Spanish-speaking adults, was used to assess health literacy.

Cohen's kappa (kappa) coefficient was used to assess interobserver agreement and Chi Square "Goodness of Fit" Test was used to determine probability of satisfaction.

FINDINGS: Concordance on the level of satisfaction with surgical outcomes for Type cases I was slight (kappa = 0.20; CI 95% 0-0.60), for Type II cases was moderate (kappa = 0.54; CI 95% 0.13-0.94), and for Type III cases was substantial (kappa = 0.62; CI 95% 0-0.56). However, the probability of satisfaction did not change according to severity (Chi Square "Goodness of Fit" Test; parents, p = 0.84; pediatric urologists, p = 0.92). These results cannot be explained by parental health literacy according to SAHLISA-50 test scores.

CONCLUSION: The level of concordance of opinion among parents and urologists with regard to their level of satisfaction with surgical outcomes is related to hypospadias severity, whereby the greatest level of concordance of opinion was achieved among most severe cases. This study underscores the need for longer follow-up to properly assess satisfaction with hypospadias repair, especially for the less severe forms of the condition.

Version ID

1

Status

PubMed-not-MEDLINE

Authors Full Name

Perez-Brayfield, Marcos Raymond; Jorge, Juan Carlos; Aviles, Luis A; Diaz, Joanne; Ortiz, Vanessa; Morales-Cosme, Wilfredo.

Institution

Perez-Brayfield, Marcos Raymond. Division of Urology, School of Medicine, University of Puerto Rico, San Juan, PR, USA. Jorge, Juan Carlos. Department of Anatomy and Neurobiology, School of Medicine, University of Puerto Rico, San Juan, PR, USA.

Aviles, Luis A. Department of Social Sciences, School of Public Health, University of Puerto Rico, San Juan, PR, USA.

Diaz, Joanne. MD Program, School of Medicine, University of Puerto Rico, San Juan, PR, USA.

Ortiz, Vanessa. Division of Urology, School of Medicine, University of Puerto Rico, San Juan, PR, USA.

Morales-Cosme, Wilfredo. MPH Program, School of Public Health, University of Puerto Rico, San Juan, PR, USA.

PMID

<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4725097>

Year of Publication

2016

631.

Should Surgery for Hypospadias Be Performed Before An Age of Consent?.

Carmack A; Notini L; Earp BD.

OVID Medline Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid

MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Journal of Sex Research. 53(8):1047-1058, 2016 Oct.

[Journal Article]

UI: 26479354

Hypospadias is a relatively common genital condition in which the urethral opening forms on the underside of the penis, as opposed to at the tip of the glans. Patients with hypospadias are typically referred for surgery during infancy or early childhood. Recent evidence, however, indicates that many individuals with hypospadias do not experience the functional or psychosocial difficulties commonly attributed to the condition, and that surgical intervention for hypospadias carries substantial risk of adverse outcomes. In this article, we review published outcomes data and conduct an in-depth analysis of the typical rationales for hypospadias surgery, taking into consideration both the potential benefits and harms of the procedure, as well as the existence of

nonsurgical alternatives. We argue, first, that most childhood surgeries for hypospadias are performed for anticipated future problems concerning function and cosmesis, rather than extant physical and/or psychosocial problems that are adversely affecting the child's well-being. Second, we contend that the surgery can be safely performed after an age of consent without increasing the absolute risk of surgical complications to an ethically meaningful degree. We conclude, therefore, that surgery for hypospadias should be performed only if requested by the affected individual, under conditions of informed consent.

Version ID

1

Status

PubMed-not-MEDLINE

Authors Full Name

Carmack, Adrienne; Notini, Lauren; Earp, Brian D.

Institution

Carmack, Adrienne. a MD, Diplomate, American Board of Urology, Bastrop, Texas. Notini, Lauren. b Doctoral Candidate in Bioethics, Centre for Health Equity, Melbourne School of Population and Global Health and Department of Paediatrics, University of Melbourne. Earp, Brian D. c Research Associate, Oxford Uehiro Centre for Practical Ethics, University of Oxford.

Year of Publication

2016

632.

Interaction between MTHFR 677C>T and periconceptional folic acid supplementation in the risk of Hypospadias.

Dokter E.M., van Rooij I.A., Wijers C.H., Groothuisink J.M., van der Biezen J.J., Feitz W.F., Roeleveld N., van der Zanden L.F.

Embase

Birth Defects Research Part A - Clinical and Molecular Teratology. (no pagination), 2016. Date of Publication: 2016.

[Article In Press]

AN: 608674596

Background: Hypospadias is a congenital malformation with both environmental factors and genetic predisposition involved in the pathogenesis. The role of maternal periconceptional folic acid supplement use in the development of hypospadias is unclear. As folate levels may also be influenced by the C677T polymorphism in the methylenetetrahydrofolate reductase (MTHFR) gene, we hypothesize that a gene-environment interaction between this polymorphism and folic acid use is involved in the etiology of hypospadias.

Method(s): We conducted a case-control study among 855 hypospadias cases and 713 population-based controls from the AGORA data- and biobank. Folic acid supplement use was derived from maternal questionnaires and infant and maternal DNA was used to determine the MTHFR C677T polymorphism using Taqman assays. We performed separate analyses for different hypospadias phenotypes (anterior/middle/posterior).

Result(s): Hypospadias was neither associated with folic acid use or the MTHFR C677T polymorphism, nor with their interaction. However, we did find an association with middle hypospadias when no supplements were used (odds ratio = 1.6; 95% confidence interval, 1.1-2.4), especially in infants carrying the CT/TT genotype (odds ratio = 2.5; 95% confidence interval, 1.4-4.7). In addition, more infants with these genotypes seemed to have posterior hypospadias, regardless of folic acid use.

Conclusion(s): Our study does not suggest a major role for folic acid supplements or the MTHFR C677T polymorphism in the etiology of hypospadias in general, but not using folic acid and/or carrying the MTHFR C677T polymorphism may be associated with middle and posterior hypospadias. Therefore, we stress the importance of studying gene-environment interactions preferably in stratified analyses for different hypospadias phenotypes.

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Status

Article-In-Press

Institution

(Dokter, van Rooij, Wijers, Roeleveld, van der Zanden) Radboud Institute for Health Sciences, Department for Health EvidenceRadboud University Medical CenterNijmegen The Netherlands
(Groothuismink) Radboud Institute for Health Sciences, Department of Human GeneticsRadboud University Medical CenterNijmegen The Netherlands
(van der Biezen) Department of Plastic Surgery and Hand SurgeryMedical Centre LeeuwardenLeeuwarden The Netherlands
(Feitz) Department of Urology, Paediatric UrologyRadboudumc Amalia Children's HospitalNijmegen The Netherlands

(Roeleveld) Department of PaediatricsRadboudumc Amalia Children's HospitalNijmegen The Netherlands

Publisher

John Wiley and Sons Inc. (P.O.Box 18667, Newark NJ 07191-8667, United States)

Year of Publication

2016

633.

Do caudal blocks cause complications following hypospadias surgery in children?.

Ayob F., Arnold R.

Embase

Anaesthesia. (no pagination), 2016. Date of Publication: 2016.

[Article In Press]

AN: 610351894

Status

Article-in-Press

Institution

(Ayob, Arnold) Department of Anaesthesia University Hospital Lewisham London UK

Publisher

Blackwell Publishing Ltd (E-mail: customerservices@oxonblackwellpublishing.com)

Year of Publication

2016

634.

Bladder-Exstrophy-Epispadias-Complex risk and C677T polymorphism in MTHFR gene: Case-control study among Indian children.

Ali A., Pandey R.K., Gayan S., Bajpai M.

Embase

Molecular Cytogenetics. (no pagination), 2016. Date of Publication: January 21, 2014.

[Article In Press]

AN: 609058114

Status

Article-in-Press

Institution

(Ali, Pandey, Gayan, Bajpai) Department of Pediatric Surgery, All India Institute of Medical Sciences, New Delhi, 110029 India

Publisher

BioMed Central Ltd. (E-mail: info@biomedcentral.com)

Year of Publication

2016

635.

Erratum: Quality of Life in Female Epispadias (European Journal of Pediatric Surgery (2015) 27 DOI: 10.1055/s-0035-1554793).

Amesty M.V., Chocarro G., Lobato R., Monsalve S., Martinez-Urrutia M.J., Lopez-Pereira P.C., Jaureguizar E.

Embase

European Journal of Pediatric Surgery. 26 (3) (pp e1), 2016. Date of Publication: 01 Jun 2016.

[Erratum]

AN: 607000603

Status

Embase

Institution

(Amesty, Chocarro) Service of Pediatric Surgery, Hospital Universitario la Paz, Paseo de la Castellana 261, Madrid 28046, Spain (Lobato, Monsalve, Martinez-Urrutia, Lopez-Pereira, Jaureguizar) Section of Pediatric Urology, Hospital Universitario la Paz, Madrid, Spain

Publisher

Georg Thieme Verlag (E-mail: iaorl@iaorl.org)

Year of Publication

2016

636.

Injection Therapy for Peyronie's Disease.

Shaw E., Yafi F.A., Sangkum P., Hellstrom W.J.G.

Embase

Contemporary Endocrinology. (pp 301-313), 2016. Date of Publication: 2016.

[Chapter]

AN: 627404163

Peyronie's disease (PD) is an underdiagnosed condition that affects up to 9 % of the male population. Intralesional injection therapy (ILI) is one of many modalities in the treatment of PD. Corticosteroids were the first known employed ILI agent but is presently not recommended due to its unproven efficacy. ILI of verapamil, a calcium channel blocker, and interferon have displayed some promising results with regard to improvements in plaque size, penile curvature and erectile function. Their use for the treatment of PD, however, remains off-label. Based on the positive results of the IMPRESS 1 and 2 trials, ILI of collagenase clostridium histolyticum became the first FDA-approved drug for the treatment of PD in 2013. When combined with ILI, penile traction therapy may prevent PD-related loss of penile length, if used diligently. Finally, ILI of stem cells, while still in its infancy, offers hope for a more targeted treatment of the disease in the future.

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Status

Embase

Institution

(Shaw, Yafi, Hellstrom) Department of Urology, Tulane Medical Center, 1430 Tulane Avenue, 86-42, New Orleans, LA 70112, United States (Sangkum) Department of Urology, Ramathibodi Hospital, 270 Rama VI Road, Bangkok 10400, Thailand

Publisher

Humana Press Inc. (E-mail: humana@humanapr.com)

Year of Publication

2016

637.

Fertility and sexual dysfunction issues in adults with genitourinary congenital anomalies.

Trofimenko V., Brant W.O.

Embase

Current Opinion in Urology. 26 (4) (pp 357-362), 2016. Date of Publication: 01 Jul 2016.

[Review]

AN: 610260704

Purpose of review As growing numbers of adolescents with a history of congenital genitourinary anomalies successfully enter adulthood, their spectrum of urologic concerns broadens to include sexual function and reproduction. Recent findings In hypospadias repair, preoperative testosterone was found to reduce rates of postoperative complications of urethrocutaneous fistula formation and meatal stenosis. Following hypospadias correction, dissatisfaction with surgical outcomes has been observed to correlate with psychological outcomes, rather than objective measurements such as location of meatus degree of curvature. In women with a congenital absence of a vagina, sigmoid vaginoplasty and dilation yield similar sexual outcomes, however, vaginoplasty was associated with a 20% rate of reoperation. Ilioinguinal-to-dorsal neurotomy for restoration of penile sensation in myelomeningocele has shown success in a small pilot study. Both sexual activity and paternity rates are higher in women, compared with men who are born with bladder exstrophy. Summary The extent and complexity of issues related to sexual function and fertility in the population of patients with a history of genitourinary malformation requires a thoughtful approach to timely surgical management and consistent care through their transition from childhood to adulthood.

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27139192 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=27139192>]

Status

Embase

Institution

(Trofimenko) Division of Urology, University of Utah Medical Center, Salt Lake City, UT, United States (Brant) Center for Reconstructive Urology and Men's Health, University of Utah, 30 North 1900 East, Salt Lake City, UT 84132, United States

Publisher

Lippincott Williams and Wilkins (E-mail: agents@lww.com)

Year of Publication
2016

638.

Adult hypospadias: Urethral and penile reconstruction.

Vricella G.J., Coplen D.E.

Embase

Current Opinion in Urology. 26 (4) (pp 363-368), 2016. Date of Publication: 01 Jul 2016.

[Review]

AN: 610259893

Purpose of review This article provides a subjective, concise review of contemporary advances in reconstructive urology as it pertains to adult hypospadias repair. Herein, we highlight the most important and interesting articles among the many published within the past 12 months. Recent findings The main themes in the recent literature covered herein, include management of postoperative complications, long-term follow-up, and penile cosmesis. Summary Recent literature would suggest beauty is in the eye of the beholder when it comes to cosmesis after hypospadias repair. Long-term data are reassuring regarding uroflow improvement through puberty and into adulthood for common distal shaft hypospadias repairs.

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PMID

27152923 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=27152923>]

Status

Embase

Institution

(Vricella, Coplen) Department of Urologic Surgery, Washington University, St. Louis, MI, United States

Publisher

Lippincott Williams and Wilkins (E-mail: agents@lww.com)

Year of Publication

2016

639.

Managing adult urinary incontinence from the congenitally incompetent bladder outlet.

Carrasco A., Vemulakonda V.M.

Embase

Current Opinion in Urology. 26 (4) (pp 351-356), 2016. Date of Publication: 01 Jul 2016.

[Review]

AN: 610067900

Purpose of review Treatment of urinary incontinence in adult patients with congenital incompetent bladder outlet (exstrophyepispadias complex, cloacal anomalies, or neurogenic bladder secondary to myelomeningocele) is a challenging surgical problem. In this review article, we summarize and highlight recent studies in the management of incontinence in this patient population. Recent findings The literature regarding management of urinary incontinence in this patient population is scarce. Injection of bulking agents to the bladder neck, artificial/autologous slings, artificial urinary sphincters, bladder neck reconstruction, bladder neck closure, or a combination of these are the cornerstone of management. Augmentation cystoplasty is a major adjunct procedure that can help increase continence rate and success of surgery in select patients. The level of evidence on bladder neck procedures for this patient population is low because of significant limitations, including small sample, heterogeneity of primary diagnosis/surgical techniques, variable definitions of continence, and the retrospective nature of most studies in this field. Summary Standard options for treatment of urinary incontinence in the congenitally incompetent bladder outlet procedure remain unchanged. There is no single reproducible procedure to accomplish the goal of renal preservation and continence in these patients, and often patients require multiple procedures to achieve continence. Most importantly, the pediatric and adult urologist should continue to work toward achieving a well tolerated and efficient transition of care. There is a need to standardize data acquisition and reporting of outcomes. Although randomized control studies would be ideal, because of the small number of patients with these conditions, this may not be practical. Collaboration and continued discussion among experts in the field is needed to gain a better understanding of the optimal management strategy in this growing patient population.

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Status

Embase

Institution

(Carrasco, Vemulakonda) Department of Pediatric Urology, Children's Hospital Colorado, Aurora, Colorado 80045, United States

Publisher

Lippincott Williams and Wilkins (E-mail: agents@lww.com)

Year of Publication

2016

640.

Cerebro-costo-mandibular syndrome: Clinical, radiological, and genetic findings.

Tooley M., Lynch D., Bernier F., Parboosingh J., Bhoj E., Zackai E., Calder A., Itasaki N., Wakeling E., Scott R., Lees M., Clayton-Smith J., Blyth M., Morton J., Shears D., Kini U., Homfray T., Clarke A., Barnicoat A., Wallis C., Hewitson R., Offiah A., Saunders M., Langton-Hewer S., Hilliard T., Davis P., Smithson S.

Embase

American Journal of Medical Genetics, Part A. 170 (5) (pp 1115-1126), 2016. Date of Publication: 01 May 2016.

[Article]

AN: 609058190

Cerebro-Costo-Mandibular syndrome (CCMS) is a rare autosomal dominant condition comprising branchial arch-derivative malformations with striking rib-gaps. Affected patients often have respiratory difficulties, associated with upper airway obstruction, reduced thoracic capacity, and scoliosis. We describe a series of 12 sporadic and 4 familial patients including 13 infants/children and 3 adults. Severe micrognathia and reduced numbers of ribs with gaps are consistent findings. Cleft palate, feeding difficulties, respiratory distress, tracheostomy requirement, and scoliosis are common. Additional malformations such as horseshoe kidney, hypospadias, and septal heart defect may occur. Microcephaly and significant developmental delay are present in a small minority of patients. Key radiological findings are of a narrow thorax, multiple posterior rib gaps and abnormal costo-transverse articulation. A novel finding in 2 patients is bilateral accessory ossicles arising from the hyoid bone. Recently, specific mutations in SNRPB, which encodes components of the major spliceosome, have been found to cause CCMS. These mutations

cluster in an alternatively spliced regulatory exon and result in altered SNRPB expression. DNA was available from 14 patients and SNRPB mutations were identified in 12 (4 previously reported). Eleven had recurrent mutations previously described in patients with CCMS and one had a novel mutation in the alternative exon. These results confirm the specificity of SNRPB mutations in CCMS and provide further evidence for the role of spliceosomal proteins in craniofacial and thoracic development.

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Status

Embase

Institution

(Tooley, Smithson) Department of Clinical Genetics, St. Michael's Hospital, Bristol, United Kingdom (Lynch, Bernier, Parboosingh) Department of Medical Genetics, University of Calgary, Calgary, Canada

(Bhoj, Zackai) Department of Clinical Genetics, Children's Hospital of Philadelphia, Philadelphia, PA, United States

(Calder) Department of Radiology, Great Ormond Street Children's Hospital, London, United Kingdom

(Itasaki) Centre for Comparative and Clinical Anatomy, University of Bristol, Bristol, United Kingdom

(Wakeling) North West Thames Regional Genetic Service, North West London Hospitals NHS Trust, London, United Kingdom

(Scott, Lees, Barnicoat) Department of Clinical Genetics, Great Ormond Street Children's Hospital, London, United Kingdom

(Clayton-Smith) Department of Clinical Genetics, St. Mary's Hospital, Manchester, United Kingdom

(Blyth) Department of Clinical Genetics, Chapel Allerton Hospital, Leeds, United Kingdom

(Morton) Department of Clinical Genetics, Birmingham Women's Hospital, United Kingdom

(Shears, Kini) Department of Clinical Genetics, Churchill Hospital, Oxford, United Kingdom

(Homfray) Department of Clinical Genetics, St. George's Hospital, London, United Kingdom

(Clarke) Department of Clinical Genetics, University Hospital Wales, Cardiff, United Kingdom

(Wallis, Hewitson) Department of Paediatric Respiratory Medicine, Great Ormond Street Children's Hospital, London, United Kingdom

(Offiah) Academic Unit of Child Health, Sheffield Children's NHS Foundation Trust, Sheffield, United Kingdom

(Saunders) Department of Otolaryngology, St. Michael's Hospital, Bristol, United Kingdom

(Langton-Hewer, Hilliard) Department of Paediatric Respiratory Medicine, Bristol Royal Hospital for Children, London, United Kingdom

(Davis) Department of Paediatric Intensive Care, Bristol Royal Hospital for Children, Bristol, United Kingdom

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641.

The epidemiologic evidence linking prenatal and postnatal exposure to endocrine disrupting chemicals with male reproductive disorders: A systematic review and meta-analysis.

Bonde J.P., Flachs E.M., Rimborg S., Glazer C.H., Giwercman A., Ramlau-Hansen C.H., Hougaard K.S., Hoyer B.B., Haervig K.K., Petersen S.B., Rylander L., Specht I.O., Toft G., Brauner E.V.

Embase

Human Reproduction Update. 23 (1) (pp 104-125), 2016. Date of Publication: 2016.

[Article]

AN: 621789462

BACKGROUND: More than 20 years ago, it was hypothesized that exposure to prenatal and early postnatal environmental xenobiotics with the potential to disrupt endogenous hormone signaling might be on the causal path to cryptorchidism, hypospadias, low sperm count and testicular cancer. Several consensus statements and narrative reviews in recent years have divided the scientific community and have elicited a call for systematic transparent reviews. We aimed to fill this gap in knowledge in the field of male reproductive disorders.

OBJECTIVE AND RATIONALE: The aim of this study was to systematically synthesize published data on the risk of cryptorchidism, hypospadias, low sperm counts and testicular cancer following in utero or infant exposure to chemicals that have been included on the European Commission's list of Category 1 endocrine disrupting chemicals defined as having documented adverse effects due to endocrine disruption in at least one intact organism.

SEARCH METHOD(S): A systematic literature search for original peer reviewed papers was performed in the databases PubMed and Embase to identify epidemiological studies reporting

associations between the outcomes of interest and exposures documented by biochemical analyses of biospecimens including maternal blood or urine, placenta or fat tissue as well as amnion fluid, cord blood or breast milk; this was followed by meta-analysis of quantitative data. OUTCOME(S): The literature search resulted in 1314 references among which we identified 33 papers(28 study populations) fulfilling the eligibility criteria. These provided 85 risk estimates of links between persistent organic pollutants and rapidly metabolized compounds (phthalates and Bisphenol A) and male reproductive disorders. The overall odds ratio (OR) across all exposures and outcomes was 1.11 (95% CI 0.91-1.35). When assessing four specific chemical subgroups with sufficient data for meta-analysis for all outcomes, we found that exposure to one of the four compounds, p,p'-DDE, was related to an elevated risk: OR 1.35 (95% CI 1.04-1.74). The data did not indicate that this increased risk was driven by any specific disorder.

WIDER IMPLICATION(S): The current epidemiological evidence is compatible with a small increased risk of male reproductive disorders following prenatal and postnatal exposure to some persistent environmental chemicals classified as endocrine disruptors but the evidence is limited. Future epidemiological studies may change the weight of the evidence in either direction. No evidence of distortion due to publication bias was found, but exposure-response relationships are not evident. There are insufficient data on rapidly metabolized endocrine disruptors and on specific exposure-outcome relations. A particular data gap is evident with respect to delayed effects on semen quality and testicular cancer. Although high quality epidemiological studies are still sparse, future systematic and transparent reviews may provide pieces of evidence contributing to the narrative and weight of the evidence assessments in the field.

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Embase

Institution

(Bonde, Flachs, Glazer, Hoyer, Haervig, Petersen, Specht, Brauner) Department of Occupational and Environmental Medicine, Bispebjerg University Hospital, Copenhagen NV DK-2400, Denmark (Bonde) Institute of Public Health, University of Copenhagen, Copenhagen K DK-1016, Denmark

(Rimborg) The Royal Library/ University of Copenhagen Library, Copenhagen N DK-2200, Denmark

(Giwerzman) Molecular Reproductive Medicine, Department of Translational Medicine, Lund University, Lund, Sweden

(Ramlau-Hansen) Institute of Public Health, Aarhus University, Aarhus C DK-8000, Denmark

(Hougaard) National Research Centre for the Working Environment, Copenhagen O DK-2100, Denmark

(Hoyer, Toft) Department of Clinical Epidemiology, Aarhus University Hospital, Aarhus N DK-8200, Denmark

(Rylander) Department of Occupational and Environmental Medicine, University of Lund, Lund SE-221 85, Sweden

(Brauner) Research Center for Prevention and Health (RCPH), University of Copenhagen, Glostrup DK-2600, Denmark

Publisher

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642.

The Efficacy of Congenital Penile Curvature Repair in Preadolescent Males: Early Outcomes.

McQuaid J.W., Johnson E.K., Andrews E., Rosoklija I., Cendron M.

Embase

Urology. 92 (pp 95-99), 2016. Date of Publication: 01 Jun 2016.

[Article]

AN: 609431910

Objective To characterize the spectrum of congenital penile curvature without hypospadias evaluated at our institution, and to assess the efficacy of surgical repair in prepubescent boys.

Methods The study group included 82 boys with a diagnosis of congenital penile curvature without hypospadias who elected for surgical repair from 2008 to 2010. We retrospectively reviewed clinical and operative characteristics and recorded surgical outcomes during the length of patients' follow-up at our institution. **Results** Of the patients electing surgery, 32 of 82 (39%) underwent penile degloving alone for class I (skin) chordee. Ten of 82 patients (12.2%) required further excision of class II (dysgenic fascial) chordee, and 40 of 82 (48.8%) patients required some combination of techniques to correct varying degrees of class III (dysgenic fascial) chordee. A total of 22.2% of patients returning for follow-up with corporal disproportion had persistent penile curvature compared with 14.3% of boys with fascial chordee and 11.5% of boys with skin chordee. Those children treated with tunical plications and incisions alone fared worse than those

children treated with some combination of the Nesbit or modified Nesbit procedure. Conclusion Our study shows not only that the proportion of patients with a hyposplastic urethra may be smaller than previously reported but also that corporal disproportion may be more prevalent. Attention should be paid to this later group as plication without the use of Nesbit techniques may not be enough to ensure an appropriate repair.

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Embase

Institution

(McQuaid, Andrews, Cendron) Department of Urology, Boston Children's Hospital, Harvard Medical School, 300 Longwood Avenue, Boston, MA 02468, United States (Johnson, Rosoklija) Department of Urology, Lurie Children's Hospital, Northwestern University, Chicago, IL, United States

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643.

When the right (Drug) should be left: Prenatal drug exposure and heterotaxy syndrome.

van Veenendaal N.R., Kusters C.D.J., Oostra R.-J., Bergman J.E.H., Cobben J.-M.

Embase

Birth Defects Research Part A - Clinical and Molecular Teratology. 106 (7) (pp 573-579), 2016.

Date of Publication: 01 Jul 2016.

[Article]

AN: 609161072

Background: Recent studies reported an association between prenatal propylthiouracil exposure and birth defects, including abnormal arrangement across the left-right body axis, suggesting an association with heterotaxy syndrome.

Method(s): This case-control and case-finding study used data from 1981 to 2013 from the EUROCAT birth defect registry in the Northern Netherlands. First, we explored prenatal exposures in heterotaxy syndrome (cases) and Down syndrome (controls). Second, we describe the specific birth defects in offspring of mothers using propylthiouracil (PTU) prenatally.

RESULT(S): A total of 66 cases with heterotaxy syndrome (incidence 12.1 per 100,000 pregnancies) and 783 controls with Down syndrome (143.3 per 100,000 pregnancies) were studied. No differences in intoxication use during pregnancy were found between cases and controls, including smoking (28.0% vs. 22.7%; $p = 0.40$), alcohol (14.0% vs. 26.9%; $p = 0.052$), and recreational drugs (0 vs. 0.3%; $p = 1.00$). We found an association between heterotaxy syndrome and prenatal drug exposure to follitropin-alfa (5.6% vs. 1.1%; $p = 0.04$), and drugs used in nicotine dependence (3.7% vs. 0.2%; $p = 0.02$). Five mothers used PTU during pregnancy and gave birth to a child with trisomy 18, renal abnormalities, or hypospadias and cardiac defects.

Conclusion(s): This study identified follitropin-alfa and drugs used in nicotine dependence as possible teratogens of heterotaxy syndrome. Our data suggest the possibility that there is an increased risk of birth defects (including renal, urological, and cardiac abnormalities) in children born among mothers taking PTU prenatally, but not for heterotaxy syndrome. Birth Defects Research (Part A) 106:573-579, 2016. © 2016 Wiley Periodicals, Inc.

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Institution

(van Veenendaal, Cobben) Department of Pediatrics and Clinical Genetics, University of Amsterdam, Academic Medical Center, Amsterdam, Netherlands (van Veenendaal) Department of Pediatrics, Onze Lieve Vrouwe Gasthuis, Amsterdam, Netherlands

(Kusters, Bergman) Eurocat Northern Netherlands, Department of Genetics, University of Groningen, University Medical Center Groningen, Groningen, Netherlands

(Oostra) Department of Anatomy, Embryology and Physiology, University of Amsterdam, Academic Medical Center, Amsterdam, Netherlands

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644.

Male reproductive disorders and fertility trends: Influences of environment and genetic susceptibility.

Skakkebaek N.E., Rajpert-De Meyts E., Buck Louis G.M., Toppari J., Andersson A.-M., Eisenberg M.L., Jensen T.K., Jorgensen N., Swan S.H., Sapra K.J., Ziebe S., Priskorn L., Juul A.

Embase

Physiological Reviews. 96 (1) (pp 55-97), 2016. Date of Publication: 18 Nov 2015.

[Article]

AN: 606942117

It is predicted that Japan and European Union will soon experience appreciable decreases in their populations due to persistently low total fertility rates (TFR) below replacement level (2.1 child per woman). In the United States, where TFR has also declined, there are ethnic differences. Caucasians have rates below replacement, while TFRs among African-Americans and Hispanics are higher. We review possible links between TFR and trends in a range of male reproductive problems, including testicular cancer, disorders of sex development, cryptorchidism, hypospadias, low testosterone levels, poor semen quality, childlessness, changed sex ratio, and increasing demand for assisted reproductive techniques. We present evidence that several adult male reproductive problems arise in utero and are signs of testicular dysgenesis syndrome (TDS). Although TDS might result from genetic mutations, recent evidence suggests that it most often is related to environmental exposures of the fetal testis. However, environmental factors can also affect the adult endocrine system. Based on our review of genetic and environmental factors, we conclude that environmental exposures arising from modern lifestyle, rather than genetics, are the most important factors in the observed trends. These environmental factors might act either directly or via epigenetic mechanisms. In the latter case, the effects of exposures might have an impact for several generations post-exposure. In conclusion, there is an urgent need to prioritize research in reproductive physiology and pathophysiology, particularly in highly industrialized countries facing decreasing populations. We highlight a number of topics that need attention by researchers in human physiology, pathophysiology, environmental health sciences, and demography.

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Institution

(Skakkebaek, Rajpert-De Meyts, Buck Louis, Toppari, Andersson, Eisenberg, Jensen, Jorgensen, Swan, Sapra, Ziebe, Priskorn, Juul) Department of Growth and Reproduction and EDMaRC, Rigshospitalet, University of Copenhagen, Copenhagen, Denmark (Skakkebaek, Rajpert-De Meyts, Buck Louis, Toppari, Andersson, Eisenberg, Jensen, Jorgensen, Swan, Sapra, Ziebe, Priskorn, Juul) Division of Epidemiology, Statistics and Prevention Research, Eunice Kennedy Shriver National Institute of Child Health and Human Development, National Institutes of Health, Bethesda, MD, United States

(Skakkebaek, Rajpert-De Meyts, Buck Louis, Toppari, Andersson, Eisenberg, Jensen, Jorgensen, Swan, Sapra, Ziebe, Priskorn, Juul) Department of Physiology and Pediatrics, University of Turku, Turku University Hospital, Turku, Finland

(Skakkebaek, Rajpert-De Meyts, Buck Louis, Toppari, Andersson, Eisenberg, Jensen, Jorgensen, Swan, Sapra, Ziebe, Priskorn, Juul) Stanford University, Stanford, CA, United States

(Skakkebaek, Rajpert-De Meyts, Buck Louis, Toppari, Andersson, Eisenberg, Jensen, Jorgensen, Swan, Sapra, Ziebe, Priskorn, Juul) Icahn School of Medicine at Mount Sinai, New York, NY, United States

(Skakkebaek, Rajpert-De Meyts, Buck Louis, Toppari, Andersson, Eisenberg, Jensen, Jorgensen, Swan, Sapra, Ziebe, Priskorn, Juul) The Fertility Clinic, Rigshospitalet, Copenhagen, Denmark

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645.

Foreskin and penile problems in childhood.

Abbas T., McCarthy L.

Embase

Surgery (United Kingdom). 34 (5) (pp 221-225), 2016. Date of Publication: 01 May 2016.

[Article]

AN: 610081365

This article outlines the embryology, natural history, and management of different conditions of the foreskin and penis in children. Although the classification of hypospadias is included, the management of this condition is not. Epispadias is not covered at all.

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Institution

(Abbas) Hamad General Hospital, Doha, Qatar (McCarthy) Birmingham Children's Hospital, Birmingham, United Kingdom

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646.

Tubularized incised plate urethroplasty repair in adult hypospadias patients. Are results similar to those reported in the pediatric age group? A prospective study.

Bhat A., Bhat M., Upadhaya R., Kumar V., Kumar R., Mittal R.

Embase

African Journal of Urology. 22 (2) (pp 86-91), 2016. Date of Publication: 01 Jun 2016.

[Article]

AN: 608821777

Objective: Most studies published in the literature report on the results of tubularized incised plate urethroplasty (TIPU) for hypospadias repair in children. Hence, the objective of this study was to evaluate the results of TIPU repair in adults.

Patients and Methods: The records of 60 adult patients with primary hypospadias treated with TIPU between April 2009 and May 2012 were reviewed. All the procedures were done by the same surgeon under similar conditions and using the same kind of instruments and suture material. On clinical examination, the meatal location, as well as the presence/absence of chordee and penile torsion was assessed. The quality of the spongiosum and the width of the urethral plate were evaluated intraoperatively. The postoperative complications and results were recorded and the data were analyzed.

Result(s): The patients' age ranged from 16 to 27 years with a mean of 21 years. Out of the 60 cases, 43 (72%) had distal penile, 7 (11%) mid-penile and 10 (17%) proximal hypospadias. Penile torsion was present in 10 (17%) cases with 80% having a torsion $\leq 45^\circ$ and 20% having a torsion of $45-90^\circ$. Ventral chordee ranging from 30° to 90° was present in 14 (23%) cases. Chordee correction was possible by penile de-gloving in 4 (29%) patients (2 with distal and 2 with mid-penile hypospadias), by further mobilization of the urethral plate with the corpus spongiosum in 3 (21%) and by proximal urethral mobilization in another 6 (42%) patients with proximal hypospadias. One (7%) patient also required tunica albuginea plication. The urethral plate was wide in 22 (37%), average in 26 (43%) and narrow in 12 (20%) patients. Fifty percent of the patients with a narrow urethral plate developed complications, compared to 15% of the patients whose urethral plate had an average width and none of the patients with a wide urethral plate. The spongiosum was well developed in 38 (63%) patients, while in 11 (18%) patients each the spongiosum was moderately and poorly developed. The complication rates were significantly higher (55%) in patients with a poorly developed spongiosum as compared to those with a well-developed spongiosum. The overall complication rate was 17% including fistula in 10% of the patients (2 patients with mid-penile and 4 patients with proximal hypospadias). Meatal stenosis was found in 4 patients with distal hypospadias (7%) who responded well to meatal dilatation. The fistula cases required surgical repair with a success rate of 100%. Mean hospitalization and follow-up were 9 days and 6-24 (median 37) months, respectively.

Conclusion(s): Complications encountered in the present study were urethral fistula and meatal stenosis with a higher incidence in patients with proximal hypospadias than reported in the literature. The important factors for the outcome of TIPU were the severity of hypospadias, the degree of curvature and the development of the spongiosum and urethral plate. Proximal hypospadias with a poor urethral plate and severe curvature in adults is not suitable for TIPU. In such cases, single-stage flap urethroplasty or two-stage buccal mucosal urethroplasty should be considered instead.

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Embase

Author NameID

Bhat A.; ORCID: <http://orcid.org/0000-0002-2141-4726> Bhat M.; ORCID: <http://orcid.org/0000-0002-2141-4726>

Institution

(Bhat, Bhat, Upadhaya, Kumar, Kumar, Mittal) Department of Urology, S.P. Medical College Bikaner, Rajasthan 334003, India

Publisher

Pan African Urological Surgeons Association(PAUSA) (E-mail: sunnydoodu@yahoo.com)

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2016

647.

A Systematic Review of Children's Environmental Health in Brazil.

Froes Asmus C.I.R., Camara V.M., Landrigan P.J., Claudio L.

Embase

Annals of Global Health. 82 (1) (pp 132-148), 2016. Date of Publication: 01 Jan 2016.

[Review]

AN: 612634372

In the region of the Americas, approximately 100,000 children under the age of 5 years die each year due to environmental hazards. Brazil, due to its large size and wide range of environmental challenges, presents numerous hazards to children's health. The aim of this study was to systematically review the scientific literature that describes children's exposures to environmental pollutants in Brazil and their effects on Brazilian children's health. A systematic review of the scientific literature was performed without language restrictions and time of publication (years). The literature search was conducted in the following key resources: PubMed (MEDLINE), Scopus and Web of Science with the MeSH Terms: Environmental exposure AND Brazil (filters: Human, Child [birth to 18 years] and Affiliation Author). The Virtual Health Library was also employed to access the databases Scielo and Lilacs. The search strategy was [DeCS Terms]: Child OR adolescent AND Environmental exposure AND Brazil. Health effects in children associated with exposure to environmental pollutants in Brazil were reported in 74 studies, during the period between 1995 and 2015. The most frequently cited effect was hospital admission for respiratory causes including wheezing, asthma, and pneumonia among children living in areas with high concentrations of air pollutants. A broad spectrum of other health effects possibly linked to pollutants also was found such as prematurity, low birth weight, congenital abnormality (cryptorchidism, hypospadias, micropenis), poor performance in tests of psychomotor and mental development, and behavioral problems. Exposure to pesticides in utero and postnatally was associated with a high risk for leukemia in children <2 years old. These results show that there is a need in Brazil for stricter monitoring of pollutant emissions and for health surveillance programs especially among vulnerable populations such as pregnant women and young children.

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Status

Embase

Institution

(Froes Asmus, Camara) Public Health Institute, School of Medicine, Federal University of Rio de Janeiro, Rio de Janeiro, Brazil (Froes Asmus, Landrigan, Claudio) Preventive Medicine

Department, Icahn School of Medicine of Mount Sinai, New York, NY, United States

(Landrigan) Arnhold Global Health Institute, Icahn School of Medicine of Mount Sinai, New York, NY, United States

Publisher

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Year of Publication

2016

648.

Quality of Life in Female Epispadias.

Amesty M.V., Chocarro G., Lobato R., Monsalve S., Martinez-Urrutia M.J., Lopez-Pereira P.C., Jaureguizar E.

Embase

European Journal of Pediatric Surgery. 26 (3) (pp 277-281), 2016. Date of Publication: 01 Jun 2016.

[Article]

AN: 604642257

Introduction and Aim Female epispadias is a rare congenital anomaly associated with significant urinary incontinence. This study aims to evaluate the results of its treatment in terms of continence, sexuality, and impact of incontinence on quality of life. **Materials and Methods** We retrospectively reviewed nine patients (5-39 years) treated of female epispadias in our hospital during the period 1976-2013. Urinary continence (ICIQ-SF), sexuality, impact of incontinence on quality of life (Potenziani-14-CI-IO-2000-QOL), and overall quality of life (SF-36) were evaluated. **Results** All nine women were treated by bladder neck reconstruction (Young-Dees-Leadbetter)

and genitoplasty; four received 2.5 (R = 1-5) endoscopic bladder neck injections; one required enterocystoplasty, urinary diversion, and bladder neck closure because of persistent incontinence; five performed pelvic floor rehabilitation; and two took anticholinergics. Three achieved continence; five had mild urinary incontinence; and 1 had moderate urinary incontinence and was awaiting an endoscopic injection. Of the seven who were older than 18 years, five answered the questionnaires of quality of life and sexuality. All had a general quality of life (SF-36) that does not differ from the normal population and had a slight impact (7 [0-15] points) of incontinence on quality of life (Potenziani-14-CI-IO-QOL-2000). Four had a normal sex life, and the fifth had lack of self-confidence due to her incontinence. Conclusion Patients with female epispadias have good long-term results regarding quality of life and sexuality, despite having some degree of urinary incontinence.

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Institution

(Amesty, Chocarro) Service of Pediatric Surgery, Hospital Universitario la Paz, Paseo de la Castellana 261, Madrid 28046, Spain (Lobato, Monsalve, Martinez-Urrutia, Lopez-Pereira, Jaureguizar) Section of Pediatric Urology, Hospital Universitario la Paz, Madrid, Spain

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Year of Publication

2016

649.

Geometrical modified nesbit corporoplasty to correct different types of penile curvature:

Description of the surgical procedure based on geometrical principles and long-term results.

Vicini P., Di Nicola S., Antonini G., De Berardinis E., Gentile V., De Marco F.

Embase

International Journal of Impotence Research. 28 (6) (pp 209-215), 2016. Date of Publication: 01 Nov 2016.

[Article]

AN: 611644743

We present the use of a modified corporoplasty, based on geometrical principles, to determine the exact site for the incision in the tunica or plaque and the exact amount of albuginea for overlaying to correct with extreme precision the different types of congenital or acquired penile curvature due to Peyronie's disease. To describe our experience with a new surgical procedure for the enhancement of penile curvature avoiding any overcorrection or undercorrection. Between March 2004 and April 2013, a total of 74 patients underwent the geometrical modified corporoplasty. All patients had congenital curvature until 90degree or acquired stable penile curvature 'less' than 60degree, that made sexual intercourse very difficult or impossible, normal erectile function, absence of hourglass or hinge effect. Preoperative testing included a physical examination, 3 photographs (frontal, dorsal and lateral) of penis during erection, a 10 mcg PGE1-induced erection and Doppler ultrasound, administration of the International Index of Erectile Function (IIEF-15) questionnaire. A follow-up with postoperative evaluation at 12 weeks, 12 and 24 months, included the same preoperative testing. Satisfaction rates were better assessed with the use of validated questionnaire such as the International Erectile Dysfunction Inventory of the Treatment Satisfaction (EDITS). Statistical analysis with Student's t-test was performed using commercially available, personal computer software. A total of 25 patients had congenital penile curvature with a mean deviation of 46.8degree (range 40-90), another 49 patients had Peyronie's disease with a mean deviation of 58.4 (range 45-60). No major complications were reported. Postoperative correction of the curvature was achieved in all patients (100%). Neither undercorrection nor overcorrection were recorded. No significant relapse (curvature>15degree) occurred in our patients. Shortening of the penis was reported by 74% but did not influence the high overall satisfaction of 92% (patients completely satisfied with their sexual life). The erectile function was analyzed in both groups, Student's t-test showed a significant improvement in erectile function, preoperative average IIEF-15 scores were 17.43+/-4.67, whereas postoperatively it was 22.57+/-4.83 (P=0.001). This geometrical modified Nesbit corporoplasty is a valid therapy which allows penile straightening. The geometric principles make the technique reproducible in multicentre studies.

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Status

Embase

Institution

(Vicini, Di Nicola, De Marco) Department of Urology, 'I.N.I.' Italian Neurotraumatologic Institute
Grottaferrata, Via S' Anna snc, Grottaferrata, Rome 00046, Italy (Antonini, De Berardinis,
Gentile) Department of Urology, 'Sapienza' Rome University, Rome, Italy

Publisher

Nature Publishing Group (Houndmills, Basingstoke, Hampshire RG21 6XS, United Kingdom)

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650.

Evaluation of onlay island flap technique in shallow urethral plate hypospadiasis.

Mohajerzadeh L., Mirshemirani A., Rouzrokh M., Sadeghian N., Khaleghnejad-Tabari A., Mahdavi
A., Poorhasan S.

Embase

Iranian Journal of Pediatrics. 26 (1) (no pagination), 2016. Article Number: e660. Date of
Publication: 30 Jan 2016.

[Article]

AN: 608197374

Background: Hypospadias is one of the most common congenital genital anomalies in males that necessitates to be operated early in infancy (when 6 to 9 months old). On the other hand, hypospadias is a challenging field of pediatric urology with multiple reconstruction techniques. A perfect hypospadias repair is supposed to return urethral continuity with sufficient caliber, eradicate phallus curvature, and supply an acceptable appearance with low complications.

Objective(s): This study aimed to evaluate the outcomes of using onlay island flap technique in the repair of hypospadias with shallow urethral plate.

Patients and Methods: In this prospective study within June 2012 to December 2013, we performed onlay island flap procedure to repair hypospadias with shallow urethral plate measuring less than 6 millimeter. This technique was selected for all types of hypospadiasis except subcoronal type. Nesbit's dorsal plication procedure was established for chordee. In cases with very small glans, urethroplasty was performed without glansplasty.

Result(s): Twenty three patients with mean age of 30 (range 10 -60) months underwent onlay island flap repair; all had a shallow urethral plate < 6 mm, 3 had a very small glans, and 18 had chordee. Meatus was located in distal shaft in 5 cases, mid shaft in 8, proximal in 6 and

penoscrotal type in 4 patients. Chordee was corrected with Nesbit's dorsal plication in 16 cases. Complications were: meatal stenosis in 2 cases and urethrocutaneous fistula in 2 patients, all of which were repaired surgically. Mean follow up time was 13 (3 -20) months. All cases that had glansplasty have excellent esthetic appearance.

Conclusion(s): This technique offers acceptable results regarding meatal stenosis, urethrocutaneous fistula and esthetic outcome.

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Institution

(Mohajerzadeh, Mirshemirani, Rouzrokh, Sadeghian, Khaleghnejad-Tabari, Mahdavi, Poorhasan)
Pediatric Sugery Department, Mofid Children's Hospital, Iran University of Medical Sciences,
Tehran, Iran, Islamic Republic of

Publisher

Tehran University of Medical Sciences (TUMS) (E-mail: editor@anesthpain.com)

Year of Publication

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651.

Maternal inflammatory bowel disease and hypospadias in male offspring: a population-based study in Denmark.

Andersen A.B.T., Ehrenstein V., Erichsen R., Froslev T., Sorensen H.T.

Embase

BMJ Open Gastroenterology. 3 (1) (no pagination), 2016. Article Number: e000121. Date of Publication: 31 Dec 2016.

[Article]

AN: 616545970

Background: The occurrence of inflammatory bowel disease (IBD) and hypospadias has been concurrently increasing, possibly through shared environmental risk factors such as endocrine disrupting compounds. Also, maternal IBD may disturb the normal development of the fetal reproductive tract. However, whether maternal IBD increases the risk of hypospadias in male offspring is unknown. We compared hypospadias risk in sons of mothers with and without IBD.

Method(s): We used Danish nationwide populationbased registries to conduct a longitudinal prevalence study including all live-born boys from 1979 through 2009. We computed HRs, as estimates of prevalence ratios (PRs), with 95% CIs for hypospadias, using Cox proportional hazards regression, while adjusting for measured confounding.

Result(s): Among 966 038 live-born boys, 4688 (0.5%) had a mother with a history of IBD diagnosis before the relevant childbirth. Among the boys with maternal IBD, 36 (0.8%) were diagnosed with hypospadias any time after birth, whereas 6112 (0.6%) sons of mothers without IBD diagnosis had hypospadias (adjusted PR: 1.20, (95% CI 0.86 to 1.67). Adjusted PRs for maternal Crohn's disease and ulcerative colitis were 1.38 (95% CI 0.83 to 2.29) and 1.10 (95% CI 0.71 to 1.68), respectively. Analyses defining hypospadias diagnosis recorded <6 months postpartum showed similar results.

Conclusion(s): We found no convincing evidence of an association between maternal IBD and hypospadias.

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Status

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Institution

(Andersen, Ehrenstein, Erichsen, Froslev, Sorensen) Department of Clinical Epidemiology, Aarhus University Hospital, Aarhus N, Denmark

Publisher

BMJ Publishing Group (E-mail: subscriptions@bmjgroup.com)

Year of Publication

2016

652.

Hormone therapy in hypospadias surgery: A survey on the current practice in Turkey.

Gollu Bahadir G., Ergun E., Telli O., Khanmammadov F., Cakmak A.M.

Embase

Turkish Journal of Medical Sciences. 46 (6) (pp 1624-1628), 2016. Date of Publication: 2016.

[Article]

AN: 613807098

Background/aim: Nowadays surgical intervention is possible in smaller phalluses and younger children with hypospadias disease. Different hormone treatments with different doses, modalities, indications, and treatment times come along with some disputes. The aim of this study is to evaluate the management approaches in hypospadias surgery of surgeons in regards to hormone preparations.

Material(s) and Method(s): Questionnaires were sent via e-mail to 110 actively working pediatric surgeons and urologists. The answers of 99 surgeons were evaluated (90%). Two surgeons declared that they did not perform hypospadias surgery.

Result(s): When testosterone usage in penile surgery was questioned, 44.4% of participants (n = 44) answered positively. Small-short penis glans, narrow urethral plate, chordee, disorders of sexual development, buccal mucosa-graft operations, slight tissue, and defective ventral skin were the indications for usage. Forty of forty-four surgeons stated usage in proximal hypospadias, 18 of them in penile hypospadias, and 15 of them in distal hypospadias. The most common form was dihydrotestosterone (62%). According to the respondents, fistulas (83%), infections (78%), and wound dehiscence (77%) were reduced. Fifty-six percent of the surgeons stated that bleeding was increased and 39% stated easier dissection.

Conclusion(s): As a result of this questionnaire we can understand that there is no standard usage of testosterone in Turkey. Optimal points of usage can be introduced by increasing prospective randomized trials and education programs can ensure similar effective usage.

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Embase

Institution

(Gollu Bahadır, Ergun, Khanmammadov, Cakmak) Department of Pediatric Surgery, Faculty of Medicine, Ankara University, Ankara, Turkey (Telli) Department of Pediatric Urology, Faculty of Medicine, Ankara University, Ankara, Turkey

Publisher

Turkiye Klinikleri Journal of Medical Sciences (Talapapa Bulvary no. 102, Hamammonu 1 06230, Turkey)

Year of Publication

2016

653.

Advantages of Reduced Prophylaxis after Tubularized Incised Plate Repair of Hypospadias.

Zeiai S., Nordenskjold A., Fossum M.

Embase

Journal of Urology. 196 (4) (pp 1244-1249), 2016. Date of Publication: 01 Oct 2016.

[Article]

AN: 612993372

Purpose Concerns about antibiotic resistance, adverse drug reactions and questionable medical benefits have led to changes in prophylactic antibiotic management in hypospadias repair at our clinic. In March 2010 our guidelines were changed from continuous prophylaxis for 14 days to 1 dose preoperatively and another at removal of the stent. We analyze the effects of this new regimen. **Materials and Methods** We performed a prospective journal cohort study of all our hypospadias operations from June 2008 to December 2011. We collected data from consecutive patients undergoing primary tubularized incised plate repair and postoperative stent. Patients operated on before March 2010 were compared to those operated on later. End points were postoperative infection requiring antibiotics and any complication that required redo surgery.

Results The study included 113 primary tubularized incised plate repairs with postoperative stents. Patient distribution was the same in both groups. Of 58 patients in the group receiving continuous antibiotic prophylaxis 17 had a complication and/or infection, compared to 9 of 55 patients receiving 2-dose prophylaxis. The infection rate was 5% in the continuous prophylaxis group and 4% in the 2-dose group. In contrast to our expectations, a lower complication rate was observed in the group with lower antibiotic dose without an increased risk of infection.

Conclusions There is little documented evidence concerning benefits of antibiotic prophylaxis for postoperative complications, which gives rise to large variations in clinical practice. In our study lower antibiotic dose did not increase the number of infections, but rather decreased complication rates. We advocate antibiotic prophylaxis with only a 2-dose regimen.

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Embase

Institution

(Zeiai, Nordenskjold, Fossum) Department of Women's and Children's Health, Center for Molecular Medicine, Karolinska Institutet, Stockholm, Sweden (Zeiai, Nordenskjold, Fossum)

Department of Pediatric Surgery, Astrid Lindgren Children's Hospital, Karolinska University Hospital, Stockholm, Sweden

Publisher

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Year of Publication

2016

654.

The Effect of Staged Transverse Preputial Island Flap Urethroplasty for Proximal Hypospadias with Severe Chordee.

Chen C., Yang T.-Q., Chen J.-B., Sun N., Zhang W.-P.

Embase

Journal of Urology. 196 (5) (pp 1536-1540), 2016. Date of Publication: 01 Nov 2016.

[Article]

AN: 612934817

Purpose We compare the effects of staged tranverse preputial island flap urethroplasty and the Byars 2-stage procedure in patients with proximal hypospadias and severe chordee. **Materials and Methods** We studied 87 consecutive children referred for proximal hypospadias with severe chordee between March 2011 and March 2014. Of the cases 42 were repaired with staged tranverse preputial island flap (group 1) and 45 were managed by 2-stage Byars urethroplasty (group 2). Mean $\pm$ SD age at first stage surgery was 26.6 $\pm$ 13.3 months in group 1 and 24.8 $\pm$ 14.7 months in group 2. Postoperative complications in both groups were assessed regarding fistulas, urethral strictures, diverticula, meatal stenosis and glanular dehiscence. **Results** After the second stage 2 patients (4.8%) in group 1 and 10 (23.2%) in group 2 had urethrocutaneous fistulas ($p < 0.05$). One patient (2.4%) in group 1 and 2 patients (4.4%) in group 2 had urethral strictures ($p > 0.05$). All patients with stricture were cured by repeated dilation and no patient required reoperation. One patient (2.4%) in group 1 and no patient in group 2 had diverticulum ($p > 0.05$). No patient in either group had signs or symptoms of meatal stenosis or residual chordee. Three patients (7.1%) in group 1 and 12 (26.7%) in group 2 needed reoperation ($p < 0.05$). **Conclusions** Two-stage urethroplasty, particularly tranverse preputial island flap partial urethroplasty, is appropriate for treating patients with proximal hypospadias and severe chordee.

Use of the tranverse preputial island flap can decrease complications associated with the second stage and significantly improve the success rate.

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Embase

Institution

(Chen, Yang, Chen) Department of Pediatric Surgery, First Affiliated Hospital of Guangxi Medical University, NanNing, China (Sun, Zhang) Department of Pediatric Urology, Beijing Children's Hospital, Affiliated to Capital Medical University, Beijing, China

Publisher

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2016

655.

Maternal first trimester serum levels of free-beta human chorionic gonadotrophin and male genital anomalies.

Schneuer F.J., Bower C., Holland A.J.A., Tasevski V., Jamieson S.E., Barker A., Lee L., Majzoub J.A., Nassar N.

Embase

Human Reproduction. 31 (8) (pp 1895-1903), 2016. Date of Publication: 01 Aug 2016.

[Article]

AN: 611633714

Study Question Are maternal first trimester levels of serum free-beta hCG associated with the development of hypospadias or undescended testis (UDT) in boys? Summary Answer Overall, first trimester maternal levels of serum free-beta hCG are not associated with hypospadias or UDT. However, elevated levels were found in severe phenotypes (proximal hypospadias and bilateral UDT) suggesting an altered pathway of hormonal release in early pregnancy. What is Known Already Human chorionic gonadotrophin peaks in first trimester of pregnancy stimulating fetal testosterone production, which is key to normal male genital development. Endocrine-

disrupting insults early in pregnancy have been associated with increased risk of common genital anomalies in males such as hypospadias and UDT. One plausible etiological pathway is altered release of hCG.

Study Design, Size, Duration We conducted a record-linkage study of two separate populations of women attending first trimester aneuploidy screening in two Australian states, New South Wales (NSW) and Western Australia (WA), in 2006-2009 and 2001-2003, respectively.

Participants/Materials, Setting, Methods Included were women who gave birth to a singleton live born male infant. There were 12 099 boys from NSW and 10 518 from WA included, of whom 90 and 77 had hypospadias; and 107 and 109 UDT, respectively. Serum levels of free-beta hCG were ascertained from laboratory databases and combined with relevant birth outcomes and congenital anomalies via record linkage of laboratory, birth, congenital anomalies and hospital data. Median and quartile levels of gestational age specific free-beta hCG multiple of the median (MoM) were compared between affected and unaffected boys. Logistic regression was used to evaluate the association between levels of free-beta hCG MoM and hypospadias or UDT, stratified by suspected placental dysfunction and co-existing anomalies. Where relevant, pooled analysis was conducted.

Main Results and the Role of Chance There was no difference in median hCG levels amongst women with an infant with hypospadias (NSW = 0.88 MoM, $P = 0.83$; WA = 0.84 MoM, $P = 0.76$) or UDT (NSW = 0.89 MoM, $P = 0.54$; WA = 0.95 MoM, $P = 0.95$), compared with women with an unaffected boy (NSW = 0.92 MoM; WA = 0.88 MoM). Low (<25th centile) or high (>75th centile) hCG levels were not associated with hypospadias or UDT, nor when stratifying by suspected placental dysfunction and co-existing anomalies. However, there was a tendency towards high levels for severe types, although confidence intervals were wide. When combining NSW and WA results, high hCG MoM levels (>75th centile) were associated with increased risk of proximal hypospadias (odds ratio (OR) 4.34; 95% CI: 1.08-17.4) and bilateral UDT (OR 2.86; 95% CI: 1.02-8.03).

LIMITATIONS, REASONS FOR CAUTION There were only small numbers of proximal hypospadias and bilateral UDT in both cohorts and although we conducted pooled analyses, results reported on these should be interpreted with caution. Gestational age by ultrasound may have been inaccurately estimated in small and large for gestational age fetuses affecting hCG MoM calculation in those pregnancies. Despite the reliability of our datasets in identifying adverse pregnancy outcomes, we did not have pathology information to confirm tissue lesions in the placenta and therefore our composite outcome should be considered as a proxy for placental dysfunction.

Wider Implications of the Findings This is one of the largest population-based studies examining the association between maternal first trimester serum levels of free-beta hCG and genital anomalies-hypospadias and UDT; and the first to compare specific phenotypes by severity. Overall, our findings does not support the hypothesis that alteration in maternal hCG levels is associated with the development of male genital anomalies; however, high hCG free-beta levels found in severe types suggest different underlying etiology involving higher production and secretion of hCG. These findings require

further exploration and replication. STUDY FUNDING/COMPETING INTEREST(S) This work was funded by the National Health and Medical Research Council (NHMRC) grant APP1047263. N.N. is supported by a NHMRC Career Development Fellowship APP1067066. C.B. was supported by a NHMRC Principal Research Fellowship #634341. The funding agencies had no role in the design, analysis, interpretation or reporting of the findings. There are no competing interests. TRIAL REGISTRATION NUMBER Not applicable.

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Embase

Institution

(Schneuer, Nassar) Menzies Centre for Health Policy, School of Public Health, University of Sydney, Level 6 the Hub, Charles Perkins Centre D17, Sydney, NSW 2006, Australia (Bower, Jamieson) Telethon Kids Institute, University of Western Australia, Crawley, WA 6009, Australia (Holland) Discipline of Paediatrics and Child Health, Children's Hospital at Westmead, Sydney Medical School, University of Sydney, Sydney, NSW 2145, Australia (Tasevski) Pathology North, NSW Health Pathology, Royal North Shore Hospital, St Leonards, NSW 2065, Australia (Barker) Department of Paediatric Surgery, Princess Margaret Hospital for Children, Subiaco, WA 6008, Australia (Lee) Division of Maternal-Fetal Medicine, Department of Obstetrics and Gynecology, David Geffen School of Medicine at UCLA, Los Angeles, CA 90095, United States (Majzoub) Division of Endocrinology, Boston Children's Hospital, Harvard Medical School, CLSB 16028, 300 Longwood Avenue, Boston, MA 02115, United States

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Oxford University Press

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2016

656.

Gender Identity and Sex Role of Patients Operated on for Bladder Exstrophy-Epispadias.

Taskinen S., Suominen J.S., Mattila A.K.

Embase

Journal of Urology. 196 (2) (pp 531-535), 2016. Date of Publication: 01 Aug 2016.

[Article]

AN: 611153563

Purpose We evaluated whether genital deformity has an impact on gender identity and sex role in patients operated on for bladder exstrophy-epispadias complex. **Materials and Methods** A total of 62 adolescents and adults operated on for bladder exstrophy-epispadias complex were mailed questionnaires evaluating gender identity (Gender Identity/Gender Dysphoria Questionnaire for Adolescents and Adults) and sex role (Bem Sex Role Inventory). Of the patients 33 responded and the results were compared with 99 gender matched controls. **Results** On the gender identity questionnaire female patients had median scores similar to those of their gender matched controls (4.93 vs 4.89, $p = 0.412$) but in males the score was lower compared to controls (4.87 vs 4.96, $p = 0.023$), indicating somewhat more conflicted gender identity. However, no patient had gender dysphoria. Female sex role index was higher in female patients vs controls (5.9 vs 5.3, $p = 0.003$) but was comparable between male patients and controls (5.2 vs 5.0, $p = 0.459$).

Masculine sex role indices were comparable between female patients and controls as well as between male patients and controls. Of 32 patients 17 were considered to have androgynous sex role, as were 24 of 97 controls ($p = 0.004$). The exact diagnosis (bladder exstrophy or epispadias) or dissatisfaction with appearance of the genitals had no impact on gender identity or on sex role indices. **Conclusions** Male patients had lower gender identity scores compared to controls and female sex role was enhanced among female patients. Androgynous sex role was more common in patients vs controls. Gender dysphoria was not noted in any patient.

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Status

Embase

Institution

(Taskinen, Suominen, Mattila) Department of Pediatric Surgery, Children's Hospital, Helsinki University Hospital and University of Helsinki, Helsinki, Finland (Taskinen, Suominen, Mattila) Department of Adult Psychiatry, Tampere University Hospital, Tampere, Finland

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657.

Shorter anogenital distance correlates with the severity of hypospadias in pre-pubertal boys.

Singal A.K., Jain V.G., Gazali Z., Shekhawat P.

Embase

Human Reproduction. 31 (7) (pp 1406-1410), 2016. Date of Publication: 2016.

[Article]

AN: 610980987

Study question: Do pre-pubertal boys with hypospadias have a shorter anogenital distance (AGD) than boys with normal genitalia? summary answer: AGD is significantly shorter in boys with hypospadias and decreases with the severity of hypospadias. what is known already: Animal studies have shown that androgen disruption and exposure to endocrine disrupting chemicals during a critical time period in early gestation, termed the male programming window(MPW), result in hypospadias and reduced AGD; and the severity of hypospadias correlates with the reduction in AGD. However, this correlation has not been established in humans. study design, size, duration: A prospective descriptive study involving measurement of AGD in pre-pubertal boys (n = 458) presenting to our pediatric urology clinic with hypospadias and normal genitalia was performed over a period of 3 years. participants/materials, setting, methods: AGD was measured in pre-pubertal boys from 5 months to 14 years of age presenting to our clinic with hypospadias (n = 180: four were excluded) and compared with randomly selected boys with normal genitalia (controls, n = 274). Three variants of AGD, from the midpoint of the anus to base of the scrotum (AGD-AS), to the anterior base of penis (AGD-1) and to the posterior base of penis (AGD-2), were measured and assessed for correlation with the severity of hypospadias. Severity of hypospadias was classified as anterior, middle and posterior according to the meatal location. main results and the role of chance: No significant difference in weight (P = 0.123), age (P = 0.162) or height (P = 0.591) between the two groups was observed. Only AGD-AS was significantly shorter in boys with hypospadias compared with controls (mean+SD: 40.6+/-9.7 mm versus 45.6+/-9.4 mm, P = 0.001). This relation persisted after adjusting AGD for weight, height and age (beta = 0.016, 95% confidence interval: 0.10-0.21; P < 0.001). The Spearman test showed a significant negative correlation for the severity of hypospadias with all the three AGD measures. Analysis of variance between anterior, middle and posterior subgroups showed a significant reduction in mean AGD-AS (P = 0.003) and AGD-2 (P = 0.008). limitations, reasons for caution: No data were collected pertaining to in utero exposure to endocrine disrupting chemicals

(EDCs) or cigarette smoke, or current diet and environmental exposure to EDCs, which may have influenced the AGD. Family history of genital malformation and use of IVF were not known. There may have been a selection bias as only boys presenting to our clinic were included. wider implications of the findings: The findings suggest that prenatal androgens during early gestation influence development of the male reproductive system and support the existence of aMPWin humans. Of the three AGDs, AGD-AS may be the most reliable biomarker of this in utero androgen action. However, no direct link to any specific exposure leading to shortened AGD in pre-pubertal boys with hypospadias could be determined. Further large scale multi-center studies are needed to understand this association better. study funding/competing interest(s): Funding was from the Hypospadias Foundation. No conflicts of interest to disclose.

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Institution

(Singal, Jain, Gazali) Hypospadias Foundation and MITR Hospital, RH-5, O-26, Sector-7, Vashi, Kharghar, Navi Mumbai, Maharashtra 400703, India (Singal) Division of Pediatric Urology, MGM Medical College, MGM University of Health Sciences, Kamothe, Navi Mumbai, India

(Jain, Shekhawat) Department of Pediatrics, MetroHealth Medical Centre, Case Western Reserve University, Cleveland, OH, United States

(Shekhawat) Division of Neonatology, MetroHealth Medical Centre, Case Western Reserve University, Cleveland, OH, United States

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Oxford University Press

Year of Publication

2016

658.

Placental weight and male genital anomalies: A nationwide danish cohort study.

Arendt L.H., Ramlau-Hansen C.H., Wilcox A.J., Henriksen T.B., Olsen J., Lindhard M.S.

Embase

American Journal of Epidemiology. 183 (12) (pp 1122-1128), 2016. Date of Publication: 15 Jun 2016.

[Article]

AN: 612907346

The most consistently reported risk indicators for the male genital anomalies cryptorchidism and hypospadias are prematurity and low birth weight. Placental dysfunction has been hypothesized as a possible underlying cause, and an association between placental weight at birth and hypospadias has been indicated. In a population-based cohort of 388,422 Danish singleton boys born alive (1997-2008), we studied the association between placental weight and cryptorchidism and hypospadias. Missing data were handled with multiple imputation, and we estimated hazard ratios by means of Cox regression models. During follow-up, 1,713 boys were diagnosed with hypospadias and 6,878 with cryptorchidism (3,624 underwent corrective surgery). We observed an association between low placental weight and risk of both genital anomalies. Boys with a placental weight in the lowest decile (<10%) had higher risks of both cryptorchidism (hazard ratio = 1.52, 95% confidence interval: 1.31, 1.76) and hypospadias (hazard ratio = 1.97, 95% confidence interval: 1.59, 2.45) than boys in the reference decile (50.0-59.9%). In conclusion, we found higher risks of both genital malformations in boys born with a low placental weight. The relationship seemed stronger for hypospadias than for cryptorchidism. Taken together, our data support a role for placental dysfunction in the etiology of these anomalies.

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Institution

(Arendt, Ramlau-Hansen, Wilcox, Henriksen, Olsen, Lindhard) Section for Epidemiology,
Department of Public Health, Faculty of Health, Aarhus University, Bartholins Alle 2, Aarhus C
8000, Denmark

Publisher

Oxford University Press

Year of Publication

2016

659.

Impact of caudal block on postoperative complications in children undergoing tubularised incised plate urethroplasty for hypospadias repair: a retrospective cohort study.

Kim M.H., Im Y.J., Kil H.K., Han S.W., Joe Y.E., Lee J.H.

Embase

Anaesthesia. 71 (7) (pp 773-778), 2016. Date of Publication: 01 Jul 2016.

[Article]

AN: 612628176

This study aimed to assess the association between caudal block and postoperative complications after tubularised incised plate urethroplasty. The medical records of 388 paediatric patients who underwent urethroplasty at a tertiary medical centre were analysed retrospectively. Among the 342 patients included, 216 patients received a caudal block and 72 (21.1%) patients suffered surgical complications. The number of patients having surgical complications was significantly greater among patients who received a caudal block than among patients who did not receive a caudal block (53 (24.5%) versus 19 (15.1%), respectively, $p = 0.04$). Based on multivariate logistic regression analysis, duration of surgery, caudal block and hypospadias types were independent risk factors for the surgical complications. Patients with caudal block had an odds ratio of 2.1 (95% CI, 1.14-3.81, $p = 0.018$) for the development of postoperative complications compared with patients without caudal block. This analysis demonstrates that caudal block is associated with surgical complications after tubularised incised plate urethroplasty.

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Institution

(Kim, Kil, Joe, Lee) Department of Anesthesiology and Pain Medicine, Yonsei University College of Medicine, Seoul, South Korea (Im) Department of Urology, Seoul National University Children's Hospital, Seoul, South Korea

(Han) Department of Urology, Severance Hospital, Yonsei University College of Medicine, Seoul, South Korea

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Year of Publication

660.

Hypospadias repair with onlay preputial graft: a 25-year experience with long-term follow-up.

Cambareri G.M., Yap M., Kaplan G.W.

Embase

BJU International. 118 (3) (pp 451-457), 2016. Date of Publication: 01 Sep 2016.

[Article]

AN: 611711476

Objective: To evaluate the long-term outcomes of hypospadias repair using an onlay preputial graft.

Patients and Methods: Patient records from 1989 to 2013 were retrospectively reviewed. One surgeon performed all cases and surgical technique was the same for all patients.

Result(s): There were 62 patients in the cohort, with a mean (range) follow-up of 47.4 (1-185) months. The meatal location was separated into distal (one patient), midshaft (19) and proximal (42). In all, 22 (35.5%) patients had complications. There were three main types of complications, including meatal stenosis in three (4.8%), stricture in three (4.8%), and fistula in 21 (33.9%). The mean (range) timing of presentation with a complication after surgery was 24.9 (1-127) months. In all, 54.5% of the patients with complications presented at ≥ 1 year after the initial surgery and 31.8% presented at ≥ 3 years. On univariable analysis age at the time of surgery, length of the graft, presence of chordee or meatal location (proximal or midshaft) did not predict a complication. The width of the graft was associated with a complication, with each 1 mm increase in width decreasing the odds of a complication by 56%. On multivariable analysis width remained statistically significant (odds ratio 0.44, 95% confidence interval 0.230-0.840; $P = 0.013$) for predicting a complication.

Conclusion(s): Hypospadias repair with onlay preputial graft is an option for single-stage repair, especially in cases of proximal hypospadias or where the urethral plate width and/or the glanular groove is insufficient for other types of repair. Compared with flaps, the use of grafts may decrease the risk of penile torsion and prevent less bulk around the urethra, improving skin and glans closure.

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Status

Embase

Institution

(Cambareri, Yap) Department of Urology, University of California San Diego (UCSD), San Diego, CA, United States (Kaplan) Department of Urology, Rady Children's Specialists of San Diego, San Diego, CA, United States

Publisher

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Year of Publication

2016

661.

Fetal growth restriction but not preterm birth is a risk factor for severe hypospadias.

Hashimoto Y., Kawai M., Nagai S., Matsukura T., Niwa F., Hasegawa T., Heike T.

Embase

Pediatrics International. 58 (7) (pp 573-577), 2016. Date of Publication: 01 Jul 2016.

[Article]

AN: 611437845

Background: Hypospadias has multifactorial causes and occurs at a high frequency among very low-birthweight infants. Placental insufficiency is hypothesized to be one cause of hypospadias; that is, decreased human chorionic gonadotropin (hCG) secretion caused by placental insufficiency is suspected to result in abnormal male external genitalia, but there is little direct evidence to support this. The aim of this study was therefore to identify the features of hypospadias and to clarify the male genital abnormalities caused by fetal growth restriction (FGR).

Method(s): We reviewed the clinical data of boys who underwent hypospadias repair between 2005 and 2011 at Kyoto University Hospital.

Result(s): Twenty boys were included in this study. Fifteen (75%) of the subjects were preterm or low-birthweight infants. Thirteen (65%) had FGR, 60% of whom had severe hypospadias regardless of gestational age. In addition, 92% of the FGR infants also had other genital

anomalies, such as cryptorchidism, bifid scrotum, or micropenis. In contrast, only 14% and 43% of the non-FGR infants had severe hypospadias or genital anomalies other than hypospadias, respectively. Placental histopathology was available in eight FGR infants, in seven of whom it was suggestive of blood flow deficiency such as infarction and single umbilical artery.

Conclusion(s): Infants with FGR have a high incidence of hypospadias. FGR caused by placental dysfunction, but not low birthweight, is a risk factor for severe hypospadias associated with multiple genital anomalies.

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Institution

(Hashimoto) Department of Pediatric Endocrinology and Metabolism, Children's Medical Center, Osaka City General Hospital, Osaka, Japan (Hashimoto, Kawai, Nagai, Matsukura, Niwa, Hasegawa, Heike) Department of Pediatrics, Graduate School of Medicine, Kyoto University, Kyoto, Japan

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662.

Anogenital distance as a marker of androgen exposure in humans.

Thankamony A., Pasterski V., Ong K.K., Acerini C.L., Hughes I.A.

Embase

Andrology. 4 (4) (pp 616-625), 2016. Date of Publication: 01 Jul 2016.

[Review]

AN: 611401518

Abnormal foetal testis development has been proposed to underlie common disorders of the male reproductive system such as cryptorchidism, hypospadias, reduced semen quality and testicular germ cell tumour, which are regarded as components of a 'testicular dysgenesis syndrome'. The

increasing trends and geographical variation in their incidence have been suggested to result from in utero exposure to environmental chemicals acting as endocrine disruptors. In rodents, the anogenital distance (AGD), measured from the anus to the base of genital tubercle, is a sensitive biomarker of androgen exposure during a critical embryonic window of testis development. In humans, several epidemiological studies have shown alterations in AGD associated with prenatal exposure to several chemicals with potential endocrine disrupting activity. However, the link between AGD and androgen exposure in humans is not well-defined. This review focuses on the current evidence for such a relationship. As in rodents, a clear gender difference is detected during foetal development of the AGD in humans which is maintained thereafter. Reduced AGD in association with clinically relevant outcomes of potential environmental exposures, such as cryptorchidism or hypospadias, is in keeping with AGD as a marker of foetal testicular function. Furthermore, AGD may reflect variations in prenatal androgen exposure in healthy children as shorter AGD at birth is associated with reduced masculine play behaviour in preschool boys. Several studies provide evidence linking shorter AGD with lower fertility, semen quality and testosterone levels in selected groups of adults attending andrology clinics. Overall, the observational data in humans are consistent with experimental studies in animals and support the use of AGD as a biomarker of foetal androgen exposure. Future studies evaluating AGD in relation to reproductive hormones in both infants and adults, and to gene polymorphisms, will help to further delineate the effect of prenatal and postnatal androgen exposures on AGD.

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Status

Embase

Institution

(Thankamony, Pasterski, Acerini, Hughes) Department of Paediatrics, Addenbrooke's Hospital, University of Cambridge, Cambridge, United Kingdom (Thankamony) Department of Growth and Reproduction, Rigshospitalet, University of Copenhagen, Denmark

(Pasterski) Department of Psychology, University of Cambridge, Cambridge, United Kingdom

(Ong) MRC Epidemiology Unit, Institute of Metabolic Science, University of Cambridge, Cambridge, United Kingdom

Publisher

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663.

Urethrocutaneous fistulae after hypospadias repair: When do they occur?.

Liao A.Y., Smith G.H.H.

Embase

Journal of Paediatrics and Child Health. 52 (5) (pp 556-560), 2016. Date of Publication: 01 May 2016.

[Article]

AN: 611043829

Aim: The aim is to determine the incidence and timing of urethrocutaneous fistula diagnosis after hypospadias surgery.

Method(s): A retrospective review of all patients who had both initial hypospadias surgery and subsequent fistula repair from 1995 to 2012. A comparison was made between patients who had an initial onlay island flap procedure and those who had a tubularised incised plate repair.

Result(s): Patient age at initial surgery ranged from 6 months to 16 years of age. The median time to fistula presentation was 8.5 months with a range of less than 1 month to 13.9 years post-hypospadias surgery. The median time to fistula repair was 17 months. The overall fistula rate was 8%. There was no significant difference between the rates of fistulae for onlay island flap (9%) versus tubularised incised plate procedure (7%).

Conclusion(s): Urethrocutaneous fistulae can present many years after the original hypospadias repair. The majority are diagnosed within the first year after surgery. Rates of fistulae are probably underreported due to short follow-up, but more importantly, due to patients transferring to other surgeons for fistula repair.

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Embase

Institution

(Liao, Smith) Department of Urology, The Children's Hospital at Westmead, Sydney, NSW, Australia

Publisher

Blackwell Publishing (E-mail: info@asia.blackpublishing.com.au)

Year of Publication
2016

664.

Requirement for basement membrane laminin alpha5 during urethral and external genital development.

Lin C., Werner R., Ma L., Miner J.H.

Embase

Mechanisms of Development. 141 (pp 62-69), 2016. Date of Publication: 01 Aug 2016.

[Article]

AN: 610469952

Hypospadias, a congenital malformation of the penis characteristic of an abnormal urethral orifice, affects 1 in every 125 boys, and its incidence is rising. Herein we test the hypothesis that the basement membrane protein laminin alpha5 (LAMA5) plays a key role in the development of the mouse genital tubercle, the embryonic anlage of the external genitalia. Using standard histological analyses and electron microscopy, we characterized the morphology of the external genitalia in Lama5 knockout (LAMA5-KO) mouse embryos during both androgen-independent genital tubercle development and androgen-mediated sexual differentiation. We compared regulatory gene expression between control and LAMA5-KO by in situ hybridization. We also examined the epithelial structure of the mutant genital tubercle using immunofluorescence staining and histological analyses of semi-thin sections. We found that Lama5 was expressed in both ectodermal and endodermal epithelia of the cloaca. The LAMA5-KO displayed a profound external genital malformation in which the genital tubercle was underdeveloped with a large ectopic orifice at the proximal end. In older embryos, the urethra failed to form a tubular structure and was left completely exposed. These defects were not associated with a significant alteration in regulatory gene expression, but rather with a defective ectodermal epithelium and an abnormal disintegration of the cloacal membrane. We conclude that LAMA5 is required in the basement membrane to maintain normal architecture of the ventral ectoderm during genital tubercle development, which is essential for the formation of a tubular urethra. Perturbation of LAMA5, and possibly other basement membrane components, may cause hypospadias in humans.

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Status

Embase

Author NameID

Miner, Jeffrey H.; ORCID: <http://orcid.org/0000-0002-1510-8714>

Institution

(Lin, Ma) Division of Dermatology, Washington University School of Medicine, St. Louis, MO 63110, United States (Werner) Division of Experimental Paediatric Endocrinology and Diabetes, Department of Paediatric and Adolescent Medicine, University of Luebeck, Luebeck 23538, Germany

(Miner) Division of Nephrology, Department of Internal Medicine, Washington University School of Medicine, St. Louis, MO 63110, United States

Publisher

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Year of Publication

2016

665.

Application of the STROBE statement to the hypospadias literature: Report of the international pediatric urology task force on hypospadias.

Braga L.H., Lorenzo A.J., Bagli D.J., Pippi Salle J.L., Caldamone A.

Embase

Journal of Pediatric Urology. 12 (6) (pp 367-380), 2016. Date of Publication: 01 Dec 2016.

[Article]

AN: 613760185

Introduction Observational studies, particularly case series, represent the majority of the current hypospadias research. As a result, this literature lacks standardization of surgical techniques, uniform definitions of hypospadias complications, and consistency of outcome reporting, which may make it difficult to compare results across studies. A modified version of the STROBE statement, containing 20 items, was presented at the International Pediatric Urology Task Force on Hypospadias meeting to assist with clear and transparent reporting of hypospadias studies. The adoption and implementation of this modified tool will allow investigators and health care

providers to critically evaluate quality and identify bias within the literature. In addition this instrument will ensure consistency of reporting, improving objective comparisons between studies, unification of results, and development of evidence-based clinical guidelines. Methods In this article, we have applied the modified STROBE statement to the hypospadias literature, aiming to create a guide on study reporting for pediatric urologists, and ultimately improve the quality of research in our field. We present itemized recommendations for adequate reporting of hypospadias studies and case series, ranging from drafting the abstract to addressing biases and potential sources of confounding. Included with each item is a brief explanation of its importance and potential effect on the study, as well as pertinent examples of hypospadias articles. Results A modified STROBE summary table containing 20 items is presented in (Supplementary Table 1). Conclusions If properly conducted and reported, hypospadias studies have the potential to provide useful information to clinicians and surgeons. However, authors should recognize the inherent limitations of these observational studies, especially in the form of bias, which may introduce invalid data or limit generalizability. Thus, we expect that the use of this guiding tool will not only improve transparency of hypospadias reporting, but also improve its methodological quality, allowing proper comparison and interpretation of data across different institutions.[Table presented]

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Status

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Institution

(Braga) Division of Urology, McMaster Children's Hospital, McMaster University, Hamilton, Ontario, Canada (Lorenzo, Bagli) Division of Urology, The Hospital for Sick Children, University of Toronto, Toronto, Ontario, Canada

(Pippi Salle) Division of Pediatric Urology, Sidra Medical and Research Center, Doha, Qatar

(Caldamone) Division of Urology, Hasbro Children's Hospital, Brown University, Providence, RI, United States

Publisher

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666.

Unexpected outcome of a modification of Bracka repair for proximal hypospadias: High incidence of diverticula with flaps.

Tiryaki S., elekberova V., Dokumcu Z., Ergun R., Tekin A., Yagmur I., Ulman I., Avanoglu A.

Embase

Journal of Pediatric Urology. 12 (6) (pp 395.e1-395.e6), 2016. Date of Publication: 01 Dec 2016.

[Article]

AN: 613457759

Introduction Various graft and flap techniques have been proposed for urethral reconstruction in proximal hypospadias repair. The Bracka repair involving the transfer of inner prepuce like a Wolfe graft mostly results in satisfactory results besides a high fistula rate. **Aim** The aim was to decrease the high fistula rate with Bracka repair; we wanted to use the advantages of vascularized skin in the Bracka method. **The aim of this study was to evaluate our results with this modification.** **Study design** Our modification involves using a flap instead of a graft. In the first stage, chordee was corrected by transection of the urethral plate and dorsal midline plication when necessary. Instead of a graft as suggested by Bracka, inner preputial skin with ample blood supply was transferred and stitched to the denuded ventral penile surface. In the second stage after 6 months, this flap was tubularized in the Thiersch-Duplay fashion. **Hospital records of patients who had undergone two stage modified Bracka repair between June 2007 and July 2012 were reviewed, including complaints, complications, and need for interventions.** **Results** Thirty-eight patients had undergone this operation. Four patients were lost to follow-up. The main complaint was obstructed urinary flow. Voiding symptoms were first attributed to urethral stenosis, but were, however, found to be due to diverticulum and vortex of the urine in the dilated urethra. Twenty-one patients (61%) had voiding problems and 10 patients (29%) had urinary tract infections. Fistula was observed in 23 and diverticula were observed in 24 patients. Of these, 16 patients had both fistula and diverticula. Only two patients (5%) were free of complications and totally satisfied with the operation, and 23 of the 34 patients had complications requiring intervention (Figure). **Discussion** Inner preputial flaps used in proximal hypospadias repairs are prone to diverticula formation. They become redundant in time requiring reoperation, thus decreasing the success rate. Careful fixation of the flap to the corpora and allowing time for additional attachment of the urethral plate substitution through fibrotic activity could not overcome this complication. **Conclusion** Our modification of the Bracka technique using a flap for the plate resulted in a high rate of complications (in particular diverticulum formation) and was therefore abandoned. We recommend careful use of flaps in hypospadias surgery and long-term follow-up studies to evaluate actual functional and cosmetic results.[Figure presented]

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Embase

Institution

(Tiryaki, Tekin, Yagmur, Ulman, Avanoğlu) Ege University, Faculty of Medicine, Department of Pediatric Surgery, Division of Pediatric Urology, İzmir, Turkey (İkbrova, Dokumcu) Ege University, Faculty of Medicine, Department of Pediatric Surgery, İzmir, Turkey (Ergun) Marmara University, Faculty of Medicine, Department of Pediatric Surgery, Division of Pediatric Urology, İstanbul, Turkey

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2016

667.

Familial forms of disorders of sex development may be common if infertility is considered a comorbidity.

Brauner R., Picard-Dieval F., Lottmann H., Rouget S., Bignon-Topalovic J., Bashamboo A., McElreavey K.

Embase

BMC Pediatrics. 16 (1) (no pagination), 2016. Article Number: 195. Date of Publication: 29 Nov 2016.

[Article]

AN: 613438649

Background: Families with 46,XY Disorders of Sex Development (DSD) have been reported, but they are considered to be exceptionally rare, with the exception of the familial forms of disorders affecting androgen synthesis or action. The families of some patients with anorchia may include individuals with 46,XY gonadal dysgenesis. We therefore analysed a large series of patients with 46,XY DSD or anorchia for the occurrence in their family of one of these phenotypes and/or ovarian insufficiency and/or infertility and/or cryptorchidism.

Method(s): A retrospective study chart review was performed for 114 patients with 46,XY DSD and 26 patients with 46,XY bilateral anorchia examined at a single institution over a 33 year period.

Result(s): Of the 140 patients, 25 probands with DSD belonged to 21 families and 7 with anorchia belonged to 7 families. Familial forms represent 22% (25/114) of the 46,XY DSD and 27% (7/26) of the anorchia cases. No case had disorders affecting androgen synthesis or action or 5 alpha-reductase deficiency. The presenting symptom was genital ambiguity (n = 12), hypospadias (n = 11) or discordance between 46,XY karyotyping performed in utero to exclude trisomy and female external genitalia (n = 2) or anorchia (n = 7). Other familial affected individuals presented with DSD and/or premature menopause (4 families) or male infertility (4 families) and/or cryptorchidism. In four families mutations were identified in the genes SRY, NR5A1, GATA4 and FOG2/ZFPM2. Surgery discovered dysgerminoma or gonadoblastoma in two cases with gonadal dysgenesis.

Conclusion(s): This study reveals a surprisingly high frequency of familial forms of 46,XY DSD and anorchia when premature menopause or male factor infertility are included. It also demonstrates the variability of the expression of the phenotype within the families. It highlights the need to the physician to take a full family history including fertility status. This could be important to identify familial cases, understand modes of transmission of the phenotype and eventually understand the genetic factors that are involved.

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Status

Embase

Institution

(Brauner, Picard-Dieval, Rouget) Fondation Ophtalmologique Adolphe de Rothschild and
Universite Paris Descartes, Paris, France (Lottmann) Assistance Publique-Hopitaux de Paris,
Hopital Necker-Enfants Malades, Service de chirurgie viscerale pediatrique, Paris, France
(Bignon-Topalovic, Bashamboo, McElreavey) Human Developmental Genetics, Institut Pasteur,
Paris, France

Publisher

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Year of Publication

2016

668.

Spontaneous bladder rupture in non-augmented bladder exstrophy.

Giutronich S., Scalabre A., Blanc T., Borzi P., Aigrain Y., O'Brien M., Mouriquand P.D.E., Heloury Y.

Embase

Journal of Pediatric Urology. 12 (6) (pp 400.e1-400.e5), 2016. Date of Publication: 01 Dec 2016.

[Article]

AN: 613176124

Objective Bladder perforation is not commonly described in bladder exstrophy patients without bladder augmentation. The goal of this study was to identify the risk factors of spontaneous perforation in non-augmented exstrophy bladders. **Methods** The study was a retrospective multi-institutional review of bladder perforation in seven male and two female patients with classic bladder exstrophy-epispadias (E-E). **Results** Correction of E-E was performed using Kelly repair in two and staged repair in seven (Table). Bladder neck repair was performed in eight patients at a mean age of 6 years. Three patients had additional urethral surgery. Before rupture, six patients were voiding only per urethra. Two patients were voiding urethrally but were also performing occasional CIC via a Mitrofanoff. One patient was performing CIC 3 hourly per urethra. Six were dry during the day. Six of the patients had lower urinary tract symptoms: five had frequency and four were straining to void. Two had suffered episodes of urinary retention. Pre-rupture ultrasound showed that the upper urinary tract was dilated in four patients. Micturating cystourethrogram was performed in six showing vesico-ureteral reflux in five. Two had urethral stenosis. Nuclear medicine was done in three patients with two abnormal differential function. Urodynamics was performed in two patients with low capacity (100 mL) and hypocompliant (<10) bladders. Both had high leak point pressures: 60 cmH₂O at 100 mL. The mean age at rupture was 11 years, with a range of 5-20 years. Patients presented with abdominal pain, associated with signs of intestinal obstruction in seven and fever in two. Eight patients underwent laparotomy and one prolonged drainage via SPC. Simple closure was performed in seven and bladder neck closure in one, because of extension of the rupture inferiorly. All patients recovered well. Following rupture, five underwent augmentation and Mitrofanoff. One of these suffered a recurrent rupture. Two other patients refused augmentation and Mitrofanoff and one of these has since had a subsequent rupture. **Conclusions** The limitations of this series include the small number of patients and its retrospective nature, without knowledge of the incidence. Bladder rupture is a risk even in non-augmented bladder exstrophy. It is potentially life-threatening and most often requires laparotomy. Rupture occurs because of poor bladder emptying and/or high pressure.

Urodynamics may identify those at risk. CIC with or without augmentation should not be delayed once poor bladder emptying and/or high pressure are identified.[Table presented]

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Status

Embase

Author NameID

Heloury, Yves; ORCID: <http://orcid.org/0000-0003-0843-3970> Borzi, Peter; ORCID:

<http://orcid.org/0000-0001-5197-2860>

Institution

(Giutronic, O'Brien, Heloury) Department of Paediatric Urology, Royal Children's Hospital, Flemington Road, Parkville, VIC, Australia (Scalabre, Mouriquand) Department of Paediatric Urology, Hopital Femme Mere Enfant, Hospices Civils de Lyon and Claude Bernard University, Lyon, France

(Blanc, Aigrain) Department of Paediatric Surgery, Hopital Necker Enfants Malades, Paris, France

(Borzi) Department of Paediatric Surgery, Lady Cilento Children's Hospital, South Brisbane, QLD, Australia

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669.

Does maternal exposure during pregnancy to higher ambient temperature increase the risk of hypospadias?.

Kilinc M.F., Cakmak S., Demir D.O., Doluoglu O.G., Yildiz Y., Horasanli K., Dalkilic A.

Embase

Journal of Pediatric Urology. 12 (6) (pp 407.e1-407.e6), 2016. Date of Publication: 01 Dec 2016.

[Article]

AN: 613175911

Introduction The association between ambient temperature that the mother is exposed to during pregnancy and hypospadias has not been investigated by the studies, although the recent studies showed the correlation between some congenital malformations (congenital heart disease, neural tube defect, etc.) and ambient temperature. **Objective** The aim was to investigate the relation between hypospadias and the ambient temperatures that the mother is exposed to during her pregnancy. **Methods** The data of patients with hypospadias that had their gestational periods in Ankara and Istanbul regions, and had other urological treatments (circumcision, urinary tract infection, pyeloplasty, nephrolithotomy, etc.) between January 2000 and November 2015 were analyzed retrospectively. The ambient temperature at 8-14 weeks of gestation was investigated for each patient by reviewing the data of the General Directorate of Meteorology, since this period was risky for development of hypospadias. The data including ambient temperature that the pregnant mother was exposed to, maternal age, parity, economical status, gestational age at birth, and birth weight were compared between two groups. The retrospective nature of the study may be a potential source for selection bias. **Results** The data of 1,709 children that had hypospadias repair and 4,946 children that had other urological treatments between 2000 and 2015 were retrospectively analyzed. There were no differences between the groups for maternal age, parity, economical status, gestational age at birth, and birth weight (Table). Analysis of exposed maximum and average ambient temperatures at 8-14 weeks of gestation revealed that July and August, hot periods in summer time, were more prevalent in the hypospadias group ($p = 0.01$). The average and maximum monthly ambient temperatures during summer increased the risk for hypospadias (OR, 1.32; 95% CI, 1.08-1.52; and OR, 1.22; 95% CI, 0.99-1.54, respectively). **Conclusions** In this paper, we evaluated the relation between hypospadias and the ambient temperatures that the mother is exposed during her pregnancy. The results of this study indicated that the high ambient temperatures the mother and fetus are exposed to at 8-14 weeks of gestation increased the risk of hypospadias in the offspring.[Table presented]

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Author NameID

Kilinc, Muhammet Fatih; ORCID: <http://orcid.org/0000-0002-2515-7106>

Institution

(Kilinc, Demir, Doluoglu, Yildiz) Department of Urology, Ankara Training and Research Hospital, Ankara, Turkey (Cakmak, Horasanli, Dalkilic) Department of Urology, Sisli Etfal Training and Research Hospital, Istanbul, Turkey

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670.

Concealed epispadias associated with a buried penis.

Sol Melgar R., Gorduza D., Demede D., Mouriquand P.

Embase

Journal of Pediatric Urology. 12 (6) (pp 347-351), 2016. Date of Publication: 01 Dec 2016.

[Review]

AN: 612772714

Objective The aim was to describe the clinical presentation and the surgical management of penile epispadias associated with a buried penis in five children. **Patients and methods** This is a 5-year retrospective review of patients presenting with a buried penis, a congenital defect of the penile skin shaft associated with an unretractable foreskin for whom a penile epispadias was found at the time of surgery. All had undergone surgery combining a Cantwell-Ransley procedure and refashioning of the penile skin following the authors' technique. **Results** Three children had a glanular epispadias and two had a midshaft epispadias. Four had a satisfactory outcome, and one required a complementary urethroplasty for glanular dehiscence. **Conclusion** Buried penis and epispadias are usually isolated congenital anomalies, although they can be associated. It is therefore recommended to warn parents about the possibility of underlying penile anomaly in children with buried penises and unretractable foreskin. Careful palpation of the dorsum of the glans through the foreskin looking for a dorsal cleft could indicate an associated epispadiac urethra. Surgical correction of both anomalies can be done at the same time. Parents of boys with buried penises should be warned that underlying penile anomaly may exist.

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Embase

Institution

(Sol Melgar, Gorduza, Demede, Mouriquand) Department of Paediatric Urology, Hopital Mere-Enfant, Universite Claude-Bernard, Bron, France

Publisher

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Year of Publication

2016

671.

Maternal hypertension and risk for hypospadias in offspring.

Agopian A.J., Hoang T.T., Mitchell L.E., Morrison A.C., Tu D., Nassar N., Canfield M.A.

Embase

American Journal of Medical Genetics, Part A. 170 (12) (pp 3125-3132), 2016. Date of Publication: 01 Dec 2016.

[Article]

AN: 611892902

Hypospadias is one of the most common birth defects in male infants. Maternal hypertension is a suspected risk factor; however, few previous studies have addressed the possibility of reporting bias, and several previous studies have not accounted for hypospadias severity. We analyzed data from the Texas Birth Defects Registry for 10,924 nonsyndromic cases and statewide vital records for deliveries during 1999-2009, using Poisson regression. After adjustment for potential confounders, hypospadias was associated with maternal hypertension (adjusted prevalence ratio: 1.5, 95% confidence interval: 1.4-1.7). Similar associations were observed with gestational and pregestational hypertension, including separate analyses restricted to the subset of cases with severe (second- or third-degree) hypospadias. All of these associations were also similar among the subset of cases with isolated hypospadias (without additional birth defects). To evaluate the potential for bias due to potential hypertension misclassification, we repeated our analyses using logistic regression, comparing the cases to controls with other birth defects. In these analyses, the associations with gestational hypertension were similar, but adjusted associations with pregestational hypertension were no longer observed. Our findings support an association between gestational hypertension and hypospadias in offspring, but also suggest that previously observed associations with pregestational hypertension may have been inflated due to differential misclassification of hypertension (e.g., reporting bias). As gestational hypertension is recognized

after hypospadias development, more research is needed to determine if this association reflects an increase in gestational hypertension risk secondary to hypospadias or if both conditions have shared risk factors (e.g., precursors of gestational hypertension). © 2016 Wiley Periodicals, Inc.

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Status

Embase

Institution

(Agopian, Hoang, Mitchell, Morrison) Department of Epidemiology, Human Genetics and Environmental Sciences, Human Genetics Center, University of Texas School of Public Health, Houston, TX, United States (Tu) Department of Urology, Baylor College of Medicine, Texas Children's Hospital, Houston, TX, United States

(Nassar) Menzies Centre for Health Policy, The University of Sydney, NSW, Australia

(Canfield) Birth Defects Epidemiology and Surveillance Branch, Texas Department of State Health Services, Austin, TX, United States

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Year of Publication

2016

672.

Gonadoblastoma in patients with 45,X/46,XY mosaicism: A 16-year experience.

Coyle D., Kutasy B., Han Suyin K., Antao B., Lynch S.A., McDermott M.B., O'Connell S.M., Quinn F.

Embase

Journal of Pediatric Urology. 12 (5) (pp 283.e1-283.e7), 2016. Date of Publication: 01 Oct 2016.

[Article]

AN: 611117981

Background It is recognised that individuals with a 45,X/46,XY karyotype, known as Turner mosaic syndrome with Y chromosome material (TMSY), have an increased risk of developing gonadoblastoma (GB), which may then devolve into one of a number of germ cell malignancies. Hence, children with TMSY are usually recommended to undergo prophylactic gonadectomy.

Objective We designed this study to describe the phenotypic features of our series of children with TMSY who underwent prophylactic gonadectomy in order to evaluate the prevalence of GB and germ cell malignancies in their resected specimens. **Study design** This is a retrospective case series wherein we comprehensively reviewed the clinical, histological, and cytogenetic features of all patients who underwent prophylactic gonadectomy at three tertiary paediatric referral centres over 16 years. Cases were identified from surgical logbooks and through the institutional histopathology database. Data were collected with particular reference to clinical phenotype, predominant karyotype cell line, operative management, anatomical findings and the presence of neoplastic changes. **Results** Fourteen children ranging in age at the time of surgery from 2 weeks to 17 years were included in the series. Eleven children were reared as females. The three children who were reared as males had severe penoscrotal hypospadias. The 46,XY cell line was the predominant cell line in seven (50%) cases in blood lymphocytes. The resected specimens from four patients (28.6%) contained GB, with three patients having bilateral GB. This sub-group of patients with GB were aged 5 months, 48 months, 71 months, and 13 years. GB arose in one patient with and three patients without genital virilisation. There was no focus of invasive germ cell tumour in any specimen. **Discussion** GB may be present in infants with TMSY as young as 5 months, even with low levels of Y chromosome material. The prevalence of GB in prophylactic gonadectomy specimens is similar to many previously reported series, although the absence of dysgerminoma in our series is reassuring. The exclusive presence of GB in intra-abdominal gonads is in keeping with the findings of several other series. **Conclusion** Owing to the presence of gonadoblastoma in the gonads of children with TMSY as young as 5 months, we recommend that all patients with intra-abdominal gonads in the context of TMSY should duly undergo prophylactic gonadectomy, although the timing of such surgery can be discussed with parents during counselling regarding the risk of malignancy.

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Status

Embase

Author NameID

Coyle, David; ORCID: <http://orcid.org/0000-0002-7685-6417>

Institution

(Coyle, Antao, Quinn) Department of Paediatric Surgery, Our Lady's Children's Hospital, Crumlin, Dublin, Ireland (Kutasy) Department of Paediatric Surgery, National Children's Hospital, Tallaght, Dublin, Ireland

(Han Suyin) Department of Histopathology, Temple Street Children's University Hospital, Dublin, Ireland

(Lynch) National Centre for Medical Genetics, Our Lady's Children's Hospital, Crumlin, Dublin, Ireland

(McDermott) Department of Histopathology, Our Lady's Children's Hospital, Crumlin, Dublin, Ireland

(O'Connell) Department of Paediatrics and Child Health, Cork University Hospital, Cork, Ireland
Publisher

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673.

Quality of life in young adult patients treated for bladder exstrophy.

da Cruz J.A.S., de Mattos B., Srougi M., Nguyen H., Bonan R., Denes F., Giron A., Passerotti C.

Embase

Central European Journal of Urology. 69 (2) (pp 221-224), 2016. Date of Publication: 2016.

[Article]

AN: 610888182

Introduction Bladder exstrophy (BE) is a rare condition that requires complex surgical corrections to achieve the goals of bladder functionality, normal sexual function, continence, and finally cosmesis. The purpose of this study was to identify clinical parameters that predict better quality of life (QOL) scores using a validated questionnaire (SF-36) with young adults after completing surgical reconstruction. Material and methods Forty-three young adults (mean age 22.35 years, 29 men and 14 women) treated for BE were evaluated using the Short Form 36 general health questionnaire (SF-36). Clinical assessment involved evaluation of the actual condition regarding continence, sexual function, genital satisfaction and overall cosmesis. Results Both genders presented similar QOL scores ($p = 0.36$). The QOL was not age-related ($p = 0.63$). Neither genders did not present any differences in the number of procedures ($p = 0.27$). Although no significant gender difference was found, clinical impairments - such as urinary fistula, incontinence, penile length and infertility - were associated with worse QOL scores and were male-related ($p < 0.01$). The most common complaint after complete surgical repair was about penile length (26/29 patients, 89.6%). Conclusions Age and gender were not predictors of better QOL scores. Any clinical impairment, such as urinary leakage due to incontinence or fistula,

penile length and infertility, tended to significantly decrease the overall QOL in male patients with BE. The male genitalia seems to be the most troublesome aspect post-adolescence in treated male patients with exstrophy-epispadias. It has an important impact on the overall QOL, mainly having a social affect on those patients.

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Status

Embase

Institution

(da Cruz, Giron, Passerotti) Center of Robotic Surgery, German Hospital Oswaldo Cruz, Sao Paulo, Brazil (de Mattos, Srougi, Bonan, Denes) Laboratory of Medical Research, Urology - LIM 55, University of Sao Paulo, Department of Urology, Sao Paulo, Brazil
(Nguyen) Cardon Children Medical Center, AZ, United States

Publisher

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Year of Publication

2016

674.

Altitude as a risk factor for the development of hypospadias. Geographical cluster distribution analysis in South America.

Fernandez N., Lorenzo A., Bagli D., Zarante I.

Embase

Journal of Pediatric Urology. 12 (5) (pp 307.e1-307.e5), 2016. Date of Publication: 01 Oct 2016.

[Article]

AN: 610628629

Objective Hypospadias is the most common congenital anomaly affecting the genitals. It has been established as a multifactorial disease with increasing prevalence. Many risk factors have been identified such as prematurity, birth weight, mother's age, and exposure to endocrine disruptors. In recent decades multiple authors using surveillance systems have described an increase in prevalence of hypospadias, but most of the published literature comes from developed countries in Europe and North America and few of the published studies have involved cluster analysis. Few large-scale studies have been performed addressing the effect of altitude

and other geographical aspects on the development of hypospadias. Acknowledging this limitation, we present novel results of a multinational spatial scan statistical analysis over a 30-year period in South America and an altitude analysis of hypospadias distribution on a continent level. Method A retrospective review was performed of the Latin American collaborative study of congenital malformations (ECLAMC). A total of 4,020,384 newborns was surveyed between 1982 and December 2011 in all participating centers. We selected all patients with hypospadias. All degrees of clinical severity were included in the analysis. Each participating center was geographically identified with its coordinates and altitude above sea level. A spatial scan statistical analysis was performed using Kulldorf's methodology and a prevalence trend analysis over time in centers below and above 2000 m. Results During the study period we found 159 hospitals in six different countries (Colombia, Bolivia, Brazil, Argentina, Chile, and Uruguay) with 4,537 cases of hypospadias and a global prevalence rate of 11.3/10,000 newborns. Trend analysis showed that centers below 2000 m had an increasing trend with an average of 10/10,000 newborns as opposed to those centers above 2000 m that showed a reducing trend with an average prevalence of 7.8 ($p = 0.1246$). We identified clusters with significant increases of prevalence in five centers along the coast at an average altitude of 219.8 m above sea level ($p > 0.0000$). Reduction in prevalence was found in clusters located in two centers on the Andes mountains. Altitude of 2,000 m was associated with hypospadias (Figure), with an OR 0.59 (0.5-0.69). There are ethnic arguments to support our results supported by protective polymorphism distribution in high lands. Conclusion Altitude above 2,000 m is suggested to have a protective effect for hypospadias. Specific clusters have been identified with increased risk for hypospadias. Environmental risk factors in these areas need to be further studied given the association seen between altitude and the distribution of more severe cases.[Figure presented]

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Embase

Author NameID

Fernandez, Nicolas; ORCID: <http://orcid.org/0000-0002-9675-5963>

Institution

(Fernandez, Zarante) Pontificia Universidad Javeriana, Bogota, Colombia (Lorenzo, Bagli)

Division of Urology, Hospital for Sick Children and University of Toronto, Toronto, Canada

Publisher

Elsevier Ltd

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2016

675.

PLAGL1 epimutation and bladder exstrophy: Coincidence or concurrent etiology?.

Kolarova J., Bens S., Ammerpohl O., Hilger A.C., Zhang R., Reutter H., Siebert R.

Embase

Birth Defects Research Part A - Clinical and Molecular Teratology. 106 (8) (pp 724-728), 2016.

Date of Publication: 01 Aug 2016.

[Article]

AN: 610573936

Background: The bladder exstrophy-epispadias complex (BEEC) is characterized by a spectrum of genitourinary malformations. Both classical bladder exstrophy and the most severe phenotype, exstrophy of the cloaca, display omphaloceles, a cardinal anomaly of some disorders caused by altered imprinting. Therefore, we hypothesized that BEEC in some patients could occur on the basis of an undiagnosed imprinting disorder. Such altered imprinting is associated with changes in the parent-of-origin-specific DNA methylation.

Method(s): We analyzed the DNA methylation of 54 imprinted loci in 23 selected patients with different BEEC subtypes (epispadias n = 1, classical bladder exstrophy n = 10, exstrophy of the cloaca n = 12) using the Infinium HumanMethylation450 BeadChip. A total of 471,722 not imprinted autosomal CpG loci and 891 imprinted CpG loci were investigated. Findings were corroborated by methylation-specific-multiplex ligation-dependent probe amplification (MS-MLPA) and microsatellite analysis.

Result(s): No significant differences in the DNA methylation of the not imprinted and imprinted CpG were observed depending on subtype of BEEC. Nevertheless, in 1 of the 23 patients who displayed a classical bladder exstrophy, we detected hypomethylation of the imprinted PLAGL1 locus in chromosome 6q24. We verified this hypomethylation by MS-MLPA and showed further the methylation loss to be caused most likely by a mosaic epimutation.

Conclusion(s): Considering that it is highly unlikely to detect a PLAGL1 epimutation among 23 individuals given the low incidence of this alteration in the population, our observations further support a link between BEEC and imprinting disorders. Birth Defects Research (Part A) 106:724-728, 2016. © 2016 Wiley Periodicals, Inc.

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Embase

Institution

(Kolarova, Bens, Ammerpohl, Siebert) Institute of Human Genetics, Christian-Albrechts-University
Kiel, Kiel, Germany (Kolarova, Bens, Siebert) Institute of Human Genetics, University of Ulm,
Ulm, Germany

(Hilger, Zhang, Reutter) Institute of Human Genetics, University of Bonn, Bonn, Germany

(Reutter) Department of Neonatology and Pediatric Intensive Care, Children's Hospital, University
of Bonn, Bonn, Germany

Publisher

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Year of Publication

2016

676.

AGORA, a data- and biobank for birth defects and childhood cancer.

van Rooij I.A.L.M., van der Zanden L.F.M., Bongers E.M.H.F., Renkema K.Y., Wijers C.H.W.,
Thonissen M., Dokter E.M.J., Marcelis C.L.M., de Blaauw I., Wijnen M.H.W.A., Hoogerbrugge
P.M., Bokkerink J.P.M., Schreuder M.F., Koster-Kamphuis L., Cornelissen E.A.M., Kapusta L.,
van Heijst A.F.J., Liem K.D., de Gier R.P.E., Kuijpers-Jagtman A.M., Admiraal R.J.C., Berge S.J.,
van der Biezen J.J., Verdonck A., Vander Poorten V., Hens G., Roosenboom J., Lilien M.R., de
Jong T.P., Broens P., Wijnen R., Brooks A., Franke B., Brunner H.G., Carels C.E.L., Knoers
N.V.A.M., Feitz W.F.J., Roeleveld N.

Embase

Birth Defects Research Part A - Clinical and Molecular Teratology. 106 (8) (pp 675-684), 2016.

Date of Publication: 01 Aug 2016.

[Article]

AN: 610299716

BACKGROUND: Research regarding the etiology of birth defects and childhood cancer is
essential to develop preventive measures, but often requires large study populations. Therefore,
we established the AGORA data- and biobank in the Netherlands. In this study, we describe its
rationale, design, and ongoing data collection.

METHOD(S): Children diagnosed with and/or treated for a structural birth defect or childhood cancer and their parents are invited to participate in the AGORA data- and biobank. Controls are recruited through random sampling from municipal registries. The parents receive questionnaires about demographics, family and pregnancy history, health status, prescribed medication, lifestyle, and occupational exposures before and during the index pregnancy. In addition, blood or saliva is collected from children and parents, while medical records are reviewed for diagnostic information.

RESULT(S): So far, we have collected data from over 6,860 families (3,747 birth defects, 905 childhood cancers, and 2,208 controls). The types of birth defects vary widely and comprise malformations of the digestive, respiratory, and urogenital tracts as well as facial, cardiovascular, kidney, skeletal, and central nervous system anomalies. The most frequently occurring childhood cancer types are acute lymphatic leukemia, Hodgkin and non-Hodgkin lymphoma, Wilms' tumor, and brain and spinal cord tumors. Our genetic and/or epidemiologic studies have been focused on hypospadias, anorectal malformations, congenital anomalies of the kidney and urinary tract (CAKUT), and orofacial clefts.

CONCLUSION(S): The large AGORA data- and biobank offers great opportunities for investigating genetic and nongenetic risk factors for disorders in children and is open to collaborative initiatives. Birth Defects Research (Part A) 106:675-684, 2016. © 2016 Wiley Periodicals, Inc.

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Status

Embase

Institution

(van Rooij, van der Zanden, Wijers, Dokter, Roeleveld) Department for Health Evidence, Radboud Institute for Health Sciences, Radboud university medical center, Nijmegen, Netherlands (Bongers, Marcelis, Franke, Brunner) Department of Human Genetics, Radboud university medical center, Nijmegen, Netherlands

(Renkema, Knoers) Department of Genetics, University Medical Center Utrecht, Netherlands

(Thonissen, Kuijpers-Jagtman, Carels) Department of Orthodontics and Craniofacial Biology, Radboud university medical center, Nijmegen, Netherlands

(Dokter, van der Biezen) Department of Plastic Surgery and Hand Surgery, Medical Centre Leeuwarden, Leeuwarden, Netherlands

(de Blaauw, Wijnen) Department of Surgery, Pediatric Surgery, Radboudumc Amalia Children's Hospital, Nijmegen, Netherlands

(de Blaauw, Wijnen) Department of Pediatric Surgery, Erasmus Medical Center, Rotterdam, Netherlands

(Hoogerbrugge, Bokkerink, Schreuder, Koster-Kamphuis, Cornelissen, Kapusta, van Heijst, Liem, Roeleveld) Department of Pediatrics, Radboudumc Amalia Children's Hospital, Nijmegen, Netherlands

(de Gier, Feitz) Department of Urology, Pediatric Urology, Radboudumc Amalia Children's Hospital, Nijmegen, Netherlands

(Admiraal) Department of Otorhinolaryngology, Radboud university medical center, Nijmegen, Netherlands

(Berge) Department of Oral and Maxillofacial surgery, Radboud university medical center, Nijmegen, Netherlands

(Verdonck, Carels) Department of Oral Health Sciences-Orthodontics and Leuven Cleft Lip and Palate Team, KU Leuven, University Hospitals KU Leuven, Belgium

(Vander Poorten, Hens) Department of Otorhinolaryngology, Head and Neck Surgery and Leuven Cleft Lip and Palate Team, University Hospitals KU Leuven, Belgium

(Roosenboom) Department of Neurosciences, Experimental Otorhinolaryngology, KU Leuven, Belgium

(Lilien) Department of Pediatric Nephrology, Wilhelmina Children's Hospital, University Medical Center Utrecht, Netherlands

(de Jong) Department of Pediatric Urology, Wilhelmina Children's Hospital, University Medical Center Utrecht, Netherlands

(de Jong) Department of Pediatric Urology, Emma Children's Hospital, Academic Medical Center Amsterdam, Netherlands

(Broens) Department of Surgery, Division of Pediatric Surgery, University Medical Center Groningen, Groningen, Netherlands

(Brooks) Department of Clinical Genetics, Erasmus Medical Centre, Rotterdam, Netherlands

(Franke) Department of Psychiatry, Radboud university medical center, Nijmegen, Netherlands
Publisher

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2016

Joint effects of genetic variants and residential proximity to pesticide applications on hypospadias risk.

Carmichael S.L., Yang W., Ma C., Roberts E., Kegley S., English P., Lammer E.J., Witte J.S., Shaw G.M.

Embase

Birth Defects Research Part A - Clinical and Molecular Teratology. 106 (8) (pp 653-658), 2016.

Date of Publication: 01 Aug 2016.

[Article]

AN: 610052822

Background: We examined risks associated with joint exposure of gene variants and pesticides.

Method(s): Analyses included 189 cases and 390 male controls born from 1991 to 2003 in California's San Joaquin Valley. We used logistic regression to examine risks associated with joint exposures of gene variants and pesticides that our previous work identified as associated with hypospadias. Genetic variables were based on variants in DGKK, genes involved in sex steroid synthesis/metabolism, and genes involved in genital tubercle development. Pesticide exposure was based on residential proximity to commercial agricultural pesticide applications.

Result(s): Odds ratios (ORs) were highest among babies with joint exposures, who had two- to fourfold increased risks; for example, the OR was 3.7 (95% confidence interval [CI], 0.8-16.5) among subjects with the risk-associated DGKK haplotype and pesticide exposure; OR, 1.5 (95% CI, 0.7-3.1) among subjects with the haplotype and no pesticide exposure; and OR, 0.9 (95% CI, 0.5-1.6) among subjects without the haplotype but with pesticide exposure, relative to subjects with neither. However, results did not provide statistical evidence that these risks were significantly greater than expected on an additive scale, relative to risks associated with one exposure at a time.

Conclusion(s): We observed elevated risks associated with joint exposures to selected pesticides and genetic variants but no statistical evidence for interaction. Birth Defects Research (Part A) 106:653-658, 2016. © 2016 Wiley Periodicals, Inc.

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Status

Embase

Institution

(Carmichael, Yang, Ma, Shaw) Department of Pediatrics, Division of Neonatology and Developmental Medicine, Stanford University School of Medicine, Stanford, CA, United States

(Roberts) Public Health Institute, Oakland, CA, United States

(Kegley) Pesticide Research Institute, Berkeley, CA, United States

(English) California Department of Public Health, Richmond, CA, United States

(Lammer) UCSF Benioff Children's Hospital Oakland, Oakland, CA, United States
(Witte) Department of Epidemiology and Biostatistics and Institute of Human Genetics, University of California, San Francisco, CA, United States

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Year of Publication

2016

678.

Two-stage graft urethroplasty for proximal and complicated hypospadias in children: A retrospective study.

Faure A., Bouty A., Nyo Y.L., O'Brien M., Heloury Y.

Embase

Journal of Pediatric Urology. 12 (5) (pp 286.e1-286.e7), 2016. Date of Publication: 01 Oct 2016.

[Article]

AN: 609242898

Introduction Although two-stage graft urethroplasty is widely used, the literature regarding the complication rates and functional characteristics of reconstructed neourethra is relatively modest. Objectives The aim was to analyze the complication rates and uroflow data of boys who have previously undergone a two-stage graft urethroplasty procedure for proximal and complicated hypospadias. Patients and methods We retrospectively reviewed the clinical outcomes of 52 boys with proximal (n = 44) and complicated (n = 8) hypospadias who underwent two-stage graft urethroplasty repair (median age of 15 months and 3 years respectively) between 2004 and 2015. Fifteen toilet-trained boys without fistulas underwent uroflowmetry. The uroflow data were plotted on age-volume-dependent normograms with normal controls. The median follow-up was 34 months (8 months-8 years). Results and complications Complications were identified in three patients (6%) after the first stage (i.e. contracture of the graft) and in 20 patients (38.4%) after the second stage, including meatal stenosis (n = 8, 15.3%), urethral stricture (n = 4, 7.6%), urethrocutaneous fistula (n = 8, 15.3%), glandular dehiscence (n = 1, 1.9%), and diverticulum (n = 1, 1.9%). The patients with failed hypospadias experienced fewer complications than those who underwent the two-stage procedure for primary repair (25% and 45%, respectively). The reoperation rate was 36.8%. Eleven of the 15 toilet-trained boys were asymptomatic but exhibited

flow rates below the normal range (median Qmax = 7 mL/s, range 3.5-16.7). Only one of the boys with a low flow rate was confirmed to have urethral stenosis under general anesthesia.

Discussion In our study, primary hypospadias repair requiring urethral plate transection elicited worse outcomes than those observed in the prior failed hypospadias cases. However, because of our study's retrospective design, we were unable to accurately assess the initial position of the meatus in the redo hypospadias cases. Our data also demonstrated that the majority of cases without any voiding symptoms exhibited flow rates that were below the normal range despite no urethral stricture under general anesthesia. These findings indicate that urethras reconstructed via two-stage graft urethroplasty repair are not functionally equivalent to normal urethras, at least prior to puberty. **Conclusion** Two-stage graft urethroplasty repair was successful in 62% of cases after the second-stage procedure, but one-third of the boys required a reoperation after the two-stage planned repair. We demonstrated that although we used a urethral tissue substitute, the urine flow patterns of the patients without strictures were abnormal.

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Embase

Institution

(Faure, Bouty, Nyo, O'Brien, Heloury) Royal Children's Hospital, Department of Urology, Parkville, VIC 3052, Australia

Publisher

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Year of Publication

2016

679.

International Consultation on Urological Diseases: Congenital Anomalies of the Genitalia in Adolescence.

Higuchi T., Holmdahl G., Kaefer M., Koyle M., Wood H., Woodhouse C., Wood D.

Embase

Urology. 94 (pp 288-310), 2016. Date of Publication: 01 Aug 2016.

[Article]

AN: 612748615

Objective To provide a comprehensive overview of genital anomalies encountered among adolescents, including late effects of problems addressed earlier in childhood. **Materials and Methods** The major congenital genital anomalies encountered in pediatric urology were identified. They include hypospadias, exstrophy-epispadias, cloacal malformations, disorders of sexual development, undescended testes, and some acquired penile anomalies seen in adolescence (priapism, adolescent varicocele). Recommendations of the International Consultation on Urological Diseases are provided on various aspects of these conditions, such as postpubertal cosmesis and function, fertility implications, and long-term nephrological considerations (when relevant). **Results** Specific recommendations for care, including strength of clinical recommendation, are provided in this paper. Whereas the basis of this paper is to discuss specific management recommendations as they relate to several heterogeneous conditions, general recommendations include patient-centered discussions regarding operative treatment be deferred until the patient is able to articulate goals and participate in shared decision-making and utilization of multidisciplinary teams for conditions where multiple organ systems may be involved. **Conclusion** Congenital abnormalities of the genitalia are common and widely heterogeneous. Late effects and concerns often emerge after puberty, and patients should be followed throughout their adult lives to address such concerns.

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Status

Embase

Institution

(Higuchi) University of Colorado, Aurora, CO, United States (Holmdahl) Department of Pediatric Surgery/Urology, The Queen Silvia Children's Hospital, Gothenburg, Sweden

(Kaefer) Riley Hospital for Children, Indianapolis, IN, United States

(Koyle) Hospital for Sick Children, Toronto, Ontario, Canada

(Wood) Cleveland Clinic, Cleveland, OH, United States

(Woodhouse) University College, London, United Kingdom

(Wood) University College London Hospitals, London, United Kingdom

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Year of Publication

2016

680.

Hypospadias and maternal exposure to atrazine via drinking water in the National Birth Defects Prevention study.

Winston J.J., Emch M., Meyer R.E., Langlois P., Weyer P., Mosley B., Olshan A.F., Band L.E., Luben T.J.

Embase

Environmental Health: A Global Access Science Source. 15 (1) (no pagination), 2016. Article Number: 76. Date of Publication: 15 Jul 2016.

[Article]

AN: 611209756

Background: Hypospadias is a relatively common birth defect affecting the male urinary tract. It has been suggested that exposure to endocrine disrupting chemicals might increase the risk of hypospadias by interrupting normal urethral development.

Method(s): Using data from the National Birth Defects Prevention Study, a population-based case-control study, we considered the role of maternal exposure to atrazine, a widely used herbicide and potential endocrine disruptor, via drinking water in the etiology of 2nd and 3rd degree hypospadias. We used data on 343 hypospadias cases and 1,422 male controls in North Carolina, Arkansas, Iowa, and Texas from 1998-2005. Using catchment level stream and groundwater contaminant models from the US Geological Survey, we estimated atrazine concentrations in public water supplies and in private wells. We assigned case and control mothers to public water supplies based on geocoded maternal address during the critical window of exposure for hypospadias (i.e., gestational weeks 6-16). Using maternal questionnaire data about water consumption and drinking water, we estimated a surrogate for total maternal consumption of atrazine via drinking water. We then included additional maternal covariates, including age, race/ethnicity, parity, and plurality, in logistic regression analyses to consider an association between atrazine and hypospadias.

Result(s): When controlling for maternal characteristics, any association between hypospadias and daily maternal atrazine exposure during the critical window of genitourinary development was found to be weak or null (odds ratio for atrazine in drinking water = 1.00, 95 % CI = 0.97 to 1.03 per 0.04 mug/day increase; odds ratio for maternal consumption = 1.02, 95 % CI = 0.99 to 1.05; per 0.05 mug/day increase).

Conclusion(s): While the association that we observed was weak, our results suggest that additional research into a possible association between atrazine and hypospadias occurrence, using a more sensitive exposure metric, would be useful.

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Status

Embase

Institution

(Winston) Carolina Population Center, University of North Carolina at Chapel Hill, Chapel Hill, NC, United States (Emch) Department of Geography, Carolina Population Center, University of North Carolina at Chapel Hill, Chapel Hill, NC, United States

(Meyer) North Carolina Birth Defects Monitoring Program, State Center for Health Statistics, Raleigh, NC, United States

(Meyer) Department of Maternal and Child Health, University of North Carolina at Chapel Hill, Chapel Hill, NC, United States

(Langlois) Texas Department of State Health Services, Birth Defects Epidemiology and Surveillance Branch, Austin, TX, United States

(Weyer) Center for Health Effects of Environmental Contamination, University of Iowa, Iowa City, IA, United States

(Mosley) Department of Pediatrics, Arkansas Children's Hospital, Little Rock, AR, United States

(Olshan) Department of Epidemiology, University of North Carolina at Chapel Hill, Chapel Hill, NC, United States

(Band) Department of Geography, Institute for the Environment, University of North Carolina at Chapel Hill, Chapel Hill, NC, United States

(Luben) National Center for Environmental Assessment, United States Environmental Protection Agency, Research Triangle Park, NC, United States

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BioMed Central Ltd. (E-mail: info@biomedcentral.com)

Year of Publication

2016

Meatal Mobilization and Glanuloplasty: A Viable Option for Coronal and Glanular Hypospadias Repair.

Moradi M., Kazemzadeh B., Hood B., Rezaee H., Kaseb K.

Embase

Urology. 94 (pp 204-207), 2016. Date of Publication: 01 Aug 2016.

[Article]

AN: 610649141

Objective To present the meatal mobilization with glanuloplasty inclusive (MMGPI) modification of meatal advancement and glanuloplasty inclusive. **Materials and Methods** A total of 120 patients with anterior hypospadias underwent MMGPI between September 2008 and October 2014 at Kermanshah University of Medical Sciences. Satisfactory outcomes were defined as continuous straight urinary flow and catheterization of new meatus without difficulty. Cosmetic outcomes were considered acceptable if patients maintained a slit-like meatus at the glanular tip. Patients were examined at 1 week, 1, 3, 6, 12, and 24 months. **Results** The 120 patients with anterior hypospadias underwent MMGPI. There were no hematoma, meatal necrosis, or other early complications. In patients with glanular hypospadias, there were no meatal regressions or stenosis, all voiding patterns were normal, and all patients maintained a slit-like meatus at the glanular tip. Two patients with coronal hypospadias had meatal stenosis and 2 patients had meatal regression. Five patients with sub-coronal hypospadias had 2-mm meatal regression with downward sloping urinary stream, and 2 patients had meatal stenosis. In all, meatus remained distal to the preoperative meatus with no necrosis. Small sample size was the major limitation of this study. **Conclusion** MMGPI represents a viable option for glanular and coronal hypospadias repair.

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Author NameID

Moradi, Mahmoudreza; ORCID: <http://orcid.org/0000-0003-3007-7888>

Institution

(Moradi, Kazemzadeh, Rezaee, Kaseb) Urology Department, Imam Reza Hospital, Kermanshah University of Medical Sciences, Iran, Islamic Republic of (Hood) Wake Forest University, Winston-Salem, NC, United States

Publisher

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Year of Publication

2016

682.

A New Modification of the Koyanagi Technique for the One-stage Repair of Severe Hypospadias.

Kang L., Huang G., Zeng L., Huang Y., Ma X., Zhang Y., Yuan M., Zhang J., Huang L.

Embase

Urology. 93 (pp 175-179), 2016. Date of Publication: 01 Jul 2016.

[Article]

AN: 610208896

Objective To describe a new modification of the Koyanagi technique for the one-stage repair of severe hypospadias and its short-term outcomes. **Patients and Methods** Our modified Koyanagi technique was performed in 24 patients with severe hypospadias between February 2012 and January 2015. The age of the patients ranged from 1.9 to 11.9 years (mean = 3.5 years). The flap design was similar to the Koyanagi technique, but our modified technique highlighted the following points: after the chordee was completely corrected, the urethral plate was recreated using foreskin, and then a U-shaped incision was made on the original and recreated urethral plate (as in the Duplay technique); a pedicled flap of the tunica vaginalis or scrotal dartos was used for additional coverage of the neourethra. **Results** The operation time lasted from 120 to 150 minutes (mean = 140 minutes). There were 5 patients (20.8%) who developed complications: 4 patients (16.7%) developed a fistula and 1 patient (4.2%) developed dehiscence of the urethra. There were no reported urethral strictures, meatal stenosis, or urethral diverticula. The complications in the 5 patients were successfully addressed with secondary repair, and all patients achieved satisfactory cosmetic and urethral functional results. **Conclusion** The modified Koyanagi technique simplified the operation and better preserved the blood supply to the flap. The additional coverage of the neourethra using a pedicled flap of the tunica vaginalis or scrotal dartos significantly decreased the rate of fistula formation. This technique is highly suitable for the one-stage repair of severe hypospadias with penoscrotal transposition.

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Embase

Institution

(Kang, Huang, Zeng, Huang, Ma, Zhang, Yuan, Zhang, Huang) Department of Pediatric Surgery, West China Hospital, Sichuan University, Chengdu 610041, China

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Year of Publication

2016

683.

The long-term outcome of boys with partial androgen insensitivity syndrome and a mutation in the androgen receptor gene.

Lucas-Herald A., Bertelloni S., Juul A., Bryce J., Jiang J., Rodie M., Sinnott R., Boroujerdi M., Lindhardt Johansen M., Hiort O., Holterhus P.M., Cools M., Guaragna-Filho G., Guerra-Junior G., Weintrob N., Hannema S., Drop S., Guran T., Darendeliler F., Nordenstrom A., Hughes I.A., Acerini C., Tadokoro-Cuccaro R., Ahmed S.F.

Embase

Journal of Clinical Endocrinology and Metabolism. 101 (11) (pp 3959-3967), 2016. Date of Publication: November 2016.

[Article]

AN: 613230705

Background: In boys with suspected partial androgen insensitivity syndrome (PAIS), systematic evidence that supports the long-term prognostic value of identifying a mutation in the androgen receptor gene (AR) is lacking.

Objective(s): To assess the clinical characteristics and long-term outcomes in young men with suspected PAIS in relation to the results of AR analysis.

Method(s): Through the International Disorders of Sex Development Registry, clinical information was gathered on young men suspected of having PAIS (n = 52) who presented before the age of 16 years and had genetic analysis of AR.

Result(s): The median ages at presentation and at the time of the study were 1 month (range, 1 day to 16 years) and 22 years (range, 16 to 52 years), respectively. Of the cohort, 29 men (56%) had 20 different AR mutations reported. At diagnosis, the median external

masculinization scores were 7 and 6 in cases with and without AR mutation, respectively ($P = .9$), and median current external masculinization scores were 9 and 10, respectively ($P = .28$). Thirty-five men (67%) required at least one surgical procedure, and those with a mutation were more likely to require multiple surgeries for hypospadias ($P=.004$). All cases with an AR mutation had gynecomastia, compared to 9% of those without an AR mutation. Of the six men who had a mastectomy, five (83%) had an AR mutation.

Conclusion(s): Boys with genetically confirmed PAIS are likely to have a poorer clinical outcome than those with XY DSD, with normal T synthesis, and without an identifiable AR mutation.

Routine genetic analysis of AR to confirm PAIS informs long-term prognosis and management. (J Clin Endocrinol Metab 101: 3959-3967, 2016).

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Institution

(Lucas-Herald, Bryce, Jiang, Rodie, Sinnott, Boroujerdi, Ahmed) University of Glasgow, School of Medicine, Royal Hospital for Children, 1345 Govan Road, Glasgow G51 4TF, United Kingdom

(Bertelloni) University Hospital Pisa, Pisa 56125, Italy

(Juul, Lindhardt Johansen) Copenhagen University Hospital, Copenhagen 2100, Denmark

(Hiort) University of Luebeck, Luebeck 23562, Germany

(Holterhus) Christian-Albrechts-University of Kiel, University Hospital of Schleswig-Holstein, Kiel 24105, Germany

(Cools) University Hospital Ghent, Ghent University, Ghent B-9000, Belgium

(Guaragna-Filho, Guerra-Junior) State University of Campinas, Campinas 13083-970, Brazil

(Weintrob) Dana Dwek Children's Hospital, Tel Aviv University, Tel Aviv 64239, Israel

(Hannema) Leids Universitair Medisch Centrum, Leiden 2333 ZA, Netherlands

(Hannema) Sophia Children's Hospital, Erasmus University Medical Center, Rotterdam 3015 CN, Netherlands

(Guran) Marmara University, Istanbul 34722, Turkey

(Darendeliler) Istanbul University, Istanbul 34452, Turkey

(Nordenstrom) Karolinska Institutet, Stockholm SE-171 77, Sweden

(Hughes, Acerini, Tadokoro-Cuccaro) University of Cambridge, Cambridge CB2 1TN, United Kingdom

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684.

Multivariate Analysis of the Factors Associated With Sexual Intercourse, Marriage, and Paternity of Hypospadias Patients.

Kanematsu A., Higuchi Y., Tanaka S., Hashimoto T., Nojima M., Yamamoto S.

Embase

Journal of Sexual Medicine. 13 (10) (pp 1488-1495), 2016. Date of Publication: 01 Oct 2016.

[Article]

AN: 612704386

Introduction Patients with hypospadias are treated surgically during childhood, which has the intention of enabling a satisfactory sexual life in adulthood. However, it is unclear whether patients with corrected hypospadias can lead a satisfactory sexual life and sustain a marital relationship and produce offspring. **Aim** To evaluate factors associated with achievement of sexual intercourse, marriage, and paternity in patients with hypospadias who have reached adulthood. **Methods** Self-completion questionnaires were mailed in April 2012 to patients with hypospadias at least 18 years old who had been treated at our institution during childhood from 1973 through 1998 by a single surgeon and the same surgical policy. Assessments included the International Prostate Symptom Score, the International Index for Erectile Function-5, and non-validated questions related to current social and physical status and sexual, marital, and paternity experiences. Candidate factors were extracted from patients' neonatal data, surgical findings and results, and current physical and social status obtained by the questionnaires. **Main Outcome Measures** Candidate factors associated with heterosexual intercourse, marriage, and paternity experiences were analyzed using univariate and multivariate proportional hazard models and log-rank test of Kaplan-Meier curves. **Results** Of the 518 patients contacted, 108 (age = 18-50 years, median = 28 years) met the inclusion criteria. Two- and one-stage repairs were performed as the initial treatment in 79 and 12, respectively, and 17 of the analyzed cases were reoperations for patients initially treated elsewhere. Fifty-seven patients had the milder type (31 glandular, 26 penile), 36 had the proximal type (13 penoscrotal, 23 scrotal-perineal), and 15 had an unknown type. Multivariate analyses by Cox proportional hazard model and log-rank tests confirmed that experience of sexual intercourse was associated with the milder type of hypospadias ($P = .025$ and $.0076$ respectively), marriage was associated with stable employment ($P = .020$ and $.026$, respectively), and paternity was associated with the absence of additional surgery after

completion of the initial repair ($P = .013$ by multivariate analysis). Conclusion There was scant overlap of factors associated with the three events. The present findings provide reference information for surgeons and parents regarding future sexual and marriage experiences of children treated for hypospadias.

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Institution

(Kanematsu, Higuchi, Hashimoto, Nojima, Yamamoto) Department of Urology, Hyogo College of Medicine, Nishinomiya, Japan (Tanaka) Department of Pharmacoepidemiology, Graduate School of Medicine and Public Health, Kyoto University, Kyoto, Japan

Publisher

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685.

Adverse health effects in children of women exposed in utero to diethylstilbestrol (DES). Effets indésirables chez les enfants de femmes exposées au diéthylstilbestrol (DES) in utero <Effets indésirables chez les enfants de femmes exposées au diéthylstilbestrol (DES) in utero.>

Tournaire M., Epelboin S., Devouche E., Viot G., Le Bidois J., Cabau A., Dunbavand A., Levadou A.

Embase

Therapie. 71 (4) (pp 395-404), 2016. Date of Publication: 01 Sep 2016.

[Article]

AN: 612700980

Objective Exposure to diethylstilbestrol (DES) in utero is associated with adverse health effects, including genital anomalies in women and men, and cancers in women. Animal studies showed birth defects and tumors in the offspring of DES exposed mice, revealing transgenerational transmission of DES effects. In humans, birth defects, such as hypospadias were observed in

children of prenatally exposed women. The aim of this research was to further assess the health effects in children of prenatally exposed women. Methods In a retrospective cohort study, the reports of women exposed to DES in utero on their 4409 children were compared with those of unexposed women on their 6203 children. Comparisons used odd ratios (OR) between children of exposed and unexposed women and standardized incidence rate (SIR) with the general population. These cohorts were recruited on a voluntary basis to answer questionnaires. Results There was a global increase of defects in children born to exposed women when compared with those born to unexposed (OR 2.29, 95% CI: 1.80-2.79, $P < 0.001$) and with the general population (SIR 2.39, 95% CI: 2.11-2.68). Increased defects were observed in male genital tract, esophagus, lip or palate, musculoskeletal and circulatory systems. For female genital tract anomalies, there was no significant increase. However, this cohort being relatively young, further follow-up is needed. An increase of cerebral palsy was revealed. The incidence of cancers was not increased, in particular for breast, uterus and ovary. Conclusion Our results confirmed a transgenerational transmission of defects in male genital tract. With caution due to possible bias associated with this method, our data suggest an increase of defects for esophagus, lip or palate, musculoskeletal and circulatory system in children of exposed women.

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Status

Embase

Institution

(Tournaire, Cabau) Assistance publique-Hopitaux de Paris, Paris 75012, France (Tournaire, Devouche, Viot, Le Bidois) Descartes University, Paris 75006, France

(Epelboin) Hopital Bichat-Claude-Bernard, Assistance publique-Hopitaux de Paris, Paris 75018, France

(Epelboin) Diderot University, Paris 75013, France

(Devouche) Laboratoire psychopathologie et processus de sante, Research Unit 4057, Boulogne 92100, France

(Viot) Hopital Cochin, Assistance publique-Hopitaux de Paris, Paris 75014, France

(Le Bidois) Pediatric Cardiology Unit, hopital Necker, Assistance publique-Hopitaux de Paris, Paris 75015, France

(Dunbavand) Mutualite francaise, Paris 75015, France

(Tournaire, Levadou) Association Reseau DES France, Mont-de-Marsan 40000, France

Publisher

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686.

First trimester phthalate exposure and male newborn genital anomalies.

Sathyanarayana S., Grady R., Barrett E.S., Redmon B., Nguyen R.H.N., Barthold J.S., Bush N.R., Swan S.H.

Embase

Environmental Research. 151 (pp 777-782), 2016. Date of Publication: 01 Nov 2016.

[Article]

AN: 612690975

Background Anti-androgenic phthalates are environmental chemicals that affect male genital development in rodents leading to genitourinary birth defects. We examined whether first trimester phthalate exposure may exert similar effects in humans leading to an increased incidence of newborn male genital anomalies in a multi-center cohort study. Methods We recruited first trimester pregnant women within The Infant Development and the Environment Study (TIDES) from 2010 to 2012 from four study centers and limited analyses to all mother/male infant dyads who had complete urinary phthalate and birth exam data (N=371). We used multivariate logistic regression to determine the odds of having a genital anomaly in relation to phthalate exposure. Results Hydrocele was the primary abnormality observed in the cohort (N=30) followed by undescended testes (N=5) and hypospadias (N=3). We observed a statistically significant 2.5 fold increased risk (95% CI 1.1, 5.9) of having any anomaly and 3.0 fold increased risk (95% CI 1.2, 7.6) of isolated hydrocele in relation to a one log unit increase in the sum of di-ethylhexyl phthalate (DEHP) metabolites. Conclusions First trimester urinary DEHP metabolite concentrations were associated with increased odds of any newborn genital anomaly, and this association was primarily driven by isolated hydrocele which made up the majority of anomalies in newborn males. The association with hydrocele has not been previously reported and suggests that it may be an endpoint affected by prenatal phthalate exposures in the first trimester of development. Future human studies should include hydrocele assessment in order to confirm findings.

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Embase

Institution

(Sathyanarayana, Grady) Seattle Children's Research Institute, Seattle, WA, United States

(Sathyanarayana) Department of Environmental and Occupational Health Sciences, University of Washington, Seattle, WA, United States

(Sathyanarayana) Department of Pediatrics, University of Washington School of Medicine, Seattle, WA, United States

(Barrett) Department of Obstetrics and Gynecology, University of Rochester School of Medicine and Dentistry, Rochester, NY, United States

(Redmon) Department of Endocrinology, University of Minnesota Medical School, Minneapolis, MN, United States

(Nguyen) Division of Epidemiology & Community Health, University of Minnesota, Minneapolis, MN, United States

(Barthold) Division of Urology, Department of Surgery, Nemours/Alfred I DuPont Hospital for Children, DE, United States

(Bush) Departments of Psychiatry and Pediatrics, University of California, San Francisco, United States

(Swan) Department of Preventive Medicine, Icahn School of Medicine at Mount Sinai, New York City, NY, United States

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687.

One-stage dorsal lingual mucosal graft urethroplasty for the treatment of failed hypospadias repair.

Li H.-B., Xu Y.-M., Fu Q., Sa Y.-L., Zhang J., Xie H.

Embase

Asian Journal of Andrology. 18 (3) (pp 467-470), 2016. Date of Publication: May 2016.

[Article]

AN: 610502865

The aim of this study was to retrospectively investigate the outcomes of patients who underwent one-stage onlay or inlay urethroplasty using a lingual mucosal graft (LMG) after failed hypospadias repairs. Inclusion criteria included a history of failed hypospadias repair, insufficiency of the local skin that made a reoperation with skin flaps difficult, and necessity of an oral mucosal graft urethroplasty. Patients were excluded if they had undergone a failed hypospadias repair using the foreskin or a multistage repair urethroplasty. Between January 2008 and December 2012, 110 patients with failed hypospadias repairs were treated in our center. Of these patients, 56 underwent a one-stage onlay or inlay urethroplasty using LMG. The median age was 21.8 years (range: 4-45 years). Of the 56 patients, one-stage onlay LMG urethroplasty was performed in 42 patients (group 1), and a modified Snodgrass technique using one-stage inlay LMG urethroplasty was performed in 14 (group 2). The median LMG urethroplasty length was 5.6 +/- 1.6 cm (range: 4-13 cm). The mean follow-up was 34.7 months (range: 10-58 months), and complications developed in 12 of 56 patients (21.4%), including urethrocuteaneous fistulas in 7 (6 in group 1, 1 in group 2) and neourethral strictures in 5 (4 in group 1, 1 in group 2). The total success rate was 78.6%. Our survey suggests that one-stage onlay or inlay urethroplasty with LMG may be an effective option to treat the patients with less available skin after failed hypospadias repairs; LMG harvesting is easy and safe, irrespective of the patient's age.

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Institution

(Li, Xu, Fu, Sa, Zhang, Xie) Department of Urology, Shanghai Jiao Tong University Affiliated Sixth People's Hospital, Shanghai Eastern Institute for Urologic Repair and Reconstruction, Shanghai, China

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Year of Publication

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688.

Foreskin morbidity in uncircumcised males.

Sneppen I., Thorup J.

Embase

Pediatrics. 137 (5) (no pagination), 2016. Article Number: e20154340. Date of Publication: May 2016.

[Article]

AN: 610484135

OBJECTIVE: As a consequence of the discussion on whether the health benefits of newborn male circumcision outweigh the risks and the discrepancies in reported figures of complications, we evaluated the incidence and morbidity of foreskin surgery due to medical indications in boys from the Capital Region of Denmark in 2014.

METHOD(S): Medical records from all boys operated on the foreskin due to medical reasons in the Capital Region in 2014 were reviewed. Patients with hypospadias, ritual circumcision, and redo-surgery because of complications to nontherapeutic circumcision were excluded.

RESULT(S): A total of 181 patients were included. The cumulative risk of undergoing foreskin operation before 18 years of age was 1.7%. Forty patients had histologic verified balanitis xerotica obliterans (BXO) corresponding to a total risk of 0.37% of developing BXO. Mean age at surgery was 10.1 years (range 1-17). Phimosis was the most frequently reported indication (95.0%). The remaining 5.0% underwent surgery because of frenulum breve causing problems during erection. Before surgery, 27.1% had foreskin-related voiding problems and 17.1% had at least 1 episode of balanitis. Circumcision was initially performed in 44 cases. The remaining 137 patients had a foreskin-preserving operation performed. Nine boys had secondary circumcision after initially having foreskin-preserving operation. Fifty patients initially had preputial histology performed. BXO was verified in 37 patients. Of the 9 patients with redo-surgery due to recurrent phimosis, a further 3 had histologically verified BXO.

CONCLUSION(S): Childhood foreskin-related problems in a region with no tradition of newborn male circumcision should not be neglected.

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Institution

(Sneppen, Thorup) Department of Pediatric Surgery, Surgical Clinic C Rigshospitalet, University Hospital of Copenhagen, 9 Blegdamsvej, Copenhagen DK-2100, Denmark (Thorup) Faculty of Health and Medical Sciences, University of Copenhagen, Copenhagen, Denmark

Publisher

American Academy of Pediatrics (141 Northwest Point Blvd, P.O. Box 927, Elk Grove Village IL 60007-1098, United States)

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689.

Comparison of tubularized incised plate urethroplasty combined with a meatus-based ventral dartos flap or dorsal dartos flap in hypospadias.

Jia W., Liu G.-C., Zhang L.-Y., Wen Y.-Q., Fu W., Hu J.-H., Wang Z., He Q.-M., Xia H.-M.

Embase

Pediatric Surgery International. 32 (4) (pp 411-415), 2016. Date of Publication: 01 Apr 2016.

[Article]

AN: 609071920

Purpose: Tubularized incised plate urethroplasty (TIPU) is the preferred surgical option for distal and mid-shaft hypospadias repair. Neourethra dartos flap coverage is routinely used as a protective layer with good results. We modified meatus-based ventral dartos flap (MBVDF) to TIPU by dissecting the proximal mid-ventral dartos attached urethra and leaving the subcutaneous fascia connecting the meatus, and retrospectively compared the outcomes of using MBVDF with single dorsal dartos flap (DDF) on the complication rates of TIPU.

Method(s): We present 2 surgeons' experiences with 356 patients with distal and mid-shaft hypospadias between January 2010 and December 2014. Patients were divided into two groups. Group DDF included 185 patients (mean age 29 months) underwent TIPU with DDF rotated laterally covering the suture lines of the neourethra. Group MBVDF included 171 patients (mean age 26 months) underwent TIPU with MBVDF covering the suture lines of the neourethra. Statistical analysis of patient basic information and complications was performed by two independent sample t test and Chi square test or Fisher's exact test.

Result(s): There were no statistical differences in age, type of hypospadias, and follow-up time between the two groups. The mean operative time in the group MBVDF (68.93 +/- 8.32 min) was

significantly shorter than in the group DDF (73.60 +/- 9.06 min). Ventral skin necrosis (2.7 %) and penile rotation (3.8 %) in group DDF was significantly higher than group MBVDF which did not occur. The differences in other complication rates including fistula rate (2.7 vs 2.9 %) between the groups were not statistically significant.

Conclusion(s): DDF and MBVDF with TIPU are similarly effective methods for decreasing fistula in hypospadias repair. MBVDF with TIPU may be an easier method and can avoid ventral skin necrosis and penile rotation.

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Status

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Author NameID

Xia, Hui-min; ORCID: <http://orcid.org/0000-0002-0103-1672>

Institution

(Jia, Xia) Southern Medical University, Guangzhou 510515, China (Jia, Liu, Zhang, Wen, Fu, Hu) Department of Pediatric Urology, Guangzhou Women and Children's Medical Center, Guangzhou 510623, China

(Wang, He, Xia) Department of Pediatric Surgery, Guangzhou Women and Children's Medical Center, Guangzhou 510623, China

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690.

Temporary re-catheterization as a treatment for early fistulas after hypospadias repair.

Chandrasekharam V.V.S.

Embase

Journal of Pediatric Urology. 12 (2) (pp 129-130), 2016. Date of Publication: 01 Apr 2016.

[Review]

AN: 608271661

Objective To present the results of temporary urethral re-catheterization in order to aid spontaneous closure of early fistulas after hypospadias repair. **Methods and technique** Children presenting with early fistulas (within 2 weeks of initial catheter removal) after hypospadias repair underwent urethral calibration and re-insertion of a urethral catheter under intravenous anesthesia. The catheter was removed after 2 weeks. **Results** Nine children (age 1-9 years) with early fistulas had re-catheterization: six (66%) had spontaneous healing of the fistula by 2 weeks, which remained closed at subsequent follow-up. **Conclusion** The simple technique of urethral re-catheterization may allow spontaneous healing of some early fistulas after hypospadias repair.

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Status

Embase

Institution

(Chandrasekharam) Pediatric Surgery, Pediatric Urology and MAS, Rainbow Children's Hospitals, Road No. 10 Banjara Hills, Hyderabad, Telangana, India

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691.

Ethnic patterns of hypospadias in New Zealand do not resemble those observed for cryptorchidism and testicular cancer: Evidence of differential aetiology?.

Gurney J.K., Stanley J., Shaw C., Sarfati D.

Embase

Andrology. 4 (1) (pp 82-86), 2016. Date of Publication: 01 Jan 2016.

[Article]

AN: 607874420

It has been proposed that hypospadias, cryptorchidism, poor semen quality and testicular cancer might share common prenatal causes. We have previously demonstrated similar ethnic patterns for the incidence of testicular cancer and cryptorchidism - a known risk factor for testicular cancer.

If the underlying exposure(s) that cause hypospadias, cryptorchidism and testicular cancer are shared, then we would expect the incidence relationship between ethnic groups to follow the same pattern across all three conditions. We followed a birth cohort of 318 345 eligible male neonates born in New Zealand between 2000-2010, and linked routinely collected maternity records with inpatient hospitalization and mortality records through to 2011. We searched hospitalization records for diagnoses of hypospadias, and used mortality records for censoring. We used Poisson regression methods to compare the relative risk of hypospadias between ethnic groups, adjusting for perinatal risk factors and total person time. We observed that European/Other children had the highest risk of hypospadias, with Maori, Pacific and Asian boys having around 40% lower risk of disease compared with this group (adjusted relative risk [RR]: Maori 0.62, 95% CI 0.55-0.70; Pacific 0.62, 95% CI 0.53-0.72; Asian 0.57, 95% CI 0.47-0.69). This contrasts substantially with our previous observations for cryptorchidism and testicular cancer, where Maori males have the greatest risk. Our observations suggest that - at least in New Zealand - the exposures that drive the development of hypospadias may differ to those that drive the development of cryptorchidism and/or testicular cancer.

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Institution

(Gurney, Stanley, Shaw, Sarfati) Department of Public Health, University of Otago, Wellington, New Zealand

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692.

TIP hypospadias repair: A pediatric urology indicator operation.

Snodgrass W., Bush N.

Embase

Journal of Pediatric Urology. 12 (1) (pp 11-18), 2016. Date of Publication: 01 Feb 2016.

[Review]

AN: 607358956

Objective We review development and evolution of TIP hypospadias repair, including technical changes made to improve its results. We also discuss general risk factors for hypospadias surgical complications. **Methods** We describe use of a database with prospective data entry to first identify our most common complications and their frequency, and then to monitor results of technical modifications made to reduce their occurrence. Multiple logistic regression of various factors recorded in the database was done to identify those predicting increased risk for urethroplasty complications. **Results** Fistula and glans dehiscence are the two most common complications we encountered after TIP repair. Changes in urethral plate tubularization and barrier layers covering the neourethra resulted in a significant reduction in fistulas after proximal TIP. Changes in glansplasty sutures and use of preoperative testosterone to increase glans size did not reduce likelihood for dehiscence, whereas increasing the extent of glans wings dissection did. Logistic regression analysis confirmed proximal meatal location and reoperation predicted increased complications, but also identified glans width ≤ 14 mm as an independent risk factor for hypospadias urethroplasty complications. **Conclusions** Systematic, prospective data collection facilitated identification of complications and their risk factors, and provided a means to assess results of modifications made to address them. Limiting the algorithm used for hypospadias repair increases expertise in those techniques used. Reported low surgical volumes for proximal hypospadias repair suggest subspecialization of these cases be carried out so that designated surgeons can achieve sufficient volume to analyze their results and make improvements.

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Status

Embase

Author NameID

Snodgrass, Warren; ORCID: <http://orcid.org/0000-0001-8715-9919>

Institution

(Snodgrass, Bush) PARC Urology, 5680 Frisco Square Boulevard, Frisco, TX 75034, United States

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693.

Staged segmental urethroplasty for scrotal/perineal hypospadias: a new concept.

Ishiyama A., Seo S., Murakami H., Ochi T., Okawada M., Doi T., Miyano G., Koga H., Lane G.J., Haruna H., Shimizu T., Yamataka A.

Embase

Pediatric Surgery International. 32 (4) (pp 403-409), 2016. Date of Publication: 01 Apr 2016.

[Article]

AN: 607223200

Purpose: We report the efficacy of staged segmental urethroplasty (SSUP) versus non-staged urethroplasty (NSUP) for treating scrotal/perineal hypospadias (SPH).

Method(s): Between 1997 and 2015, 29 SPH patients underwent UP (SSUP: n = 15; NSUP: n = 14). Incidences of urethrocutaneous fistula (UF), stenosis of the neourethra (SNU), diverticula formation, and residual chordee (RC) were compared. Differences were statistically significant if $p < 0.05$.

Result(s): The difference in mean age at NSUP (3.2 +/- 1.3 years) and at the final stage of SSUP (5.5 +/- 2.4 years) was significant ($p < 0.05$). Mean operative times for NSUP and SSUP (total for all stages) were not significantly different (231.5 +/- 117.5 versus 272.5 +/- 99.4 min); however, the incidence of postoperative complications was significantly less in SSUP (n = 1; UF) compared with NSUP (n = 6; 2 cases of UF, 3 cases of SNU, and 1 case of RC; ($p < 0.05$). Mean follow-up was significantly shorter in SSUP; 1.4 +/- 1.2 years versus 7.0 +/- 4.5 years in NSUP ($p < 0.05$).

Conclusion(s): SSUP would appear to be effective for treating SPH because of a significantly lower incidence of UF, SNU and RC during the first postoperative year, the period when complications have been reported to arise most frequently.

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Institution

(Ishiyama, Seo, Murakami, Ochi, Okawada, Doi, Miyano, Koga, Lane, Yamataka) Department of Pediatric General and Urogenital Surgery, Juntendo University School of Medicine, 2-1-1 Hongo,

Bunkyo-ku, Tokyo 113-8421, Japan (Haruna, Shimizu) Department of Pediatrics and Adolescent Medicine, Juntendo University School of Medicine, Tokyo, Japan

Publisher

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2016

694.

Dermal patch graft correction of severe chordee secondary to penile corporal body disproportion without urethral division in boys without hypospadias.

Zaontz M.R., Dean G.E.

Embase

Journal of Pediatric Urology. 12 (4) (pp 204), 2016. Date of Publication: 01 Aug 2016.

[Article]

AN: 613344000

Historically, significant ventral penile curvature secondary to corporal body disproportion has been corrected either by dorsal plication or division of the urethral plate. In the rare situations where there is severe chordee in the face of an intact urethra with an orthotopic meatus, division of the urethral plate is commonly performed at the time of grafting the ventral defect created by incising the tunica albuginea. Subsequently, a staged procedure is necessary to reconnect the urethra at a later date. Herein the authors present a novel technique that shows it is possible to perform successful dermal patch orthoplasty without division of the urethra in patients with a normal orthotopic meatus and urethra via urethral mobilization. Three patients over the past 3 years with severe ventral chordee, orthotopic meati and normal urethral anatomy presented for correction. Two patients were 18 years old and one was 10 years old. All three boys were circumcised. The two older boys insisted on dorsal plication as a first approach which worked only temporarily for about 6 months while the younger boy had no prior surgery performed. Each boy underwent a circumcising incision, degloving of the shaft skin, extensive urethral mobilization and dermal patch graft orthoplasty to correct chordee. All surgeries were performed in an outpatient setting. No urinary drainage was used in any patient and a simple bio-occlusive dressing was employed in each case. Follow-up ranged from 11 months to 2 years (mean 1.5 years). All three boys have strong straight erections, full well directed urinary streams and no

complications noted to date. Our conclusion based on this experience is that extensive urethral mobilization can allow for correction of severe ventral chordee without urethral division in a single operative setting in boys without hypospadias and a normal urethra. The accompanying movie herein describes the surgical technique.

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Status

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Author NameID

Zaontz M.R.; ORCID: <http://orcid.org/0000-0001-5636-0126>

Institution

(Zaontz) The Children's Hospital of Philadelphia, Philadelphia, PA, United States (Dean) Virtua Health System, Voorhees, NJ, United States

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695.

Long-Term Follow-Up after Treatment of Cryptorchidism.

Thorup J., Cortes D.

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European Journal of Pediatric Surgery. 26 (5) (pp 427-431), 2016. Date of Publication: 01 Oct 2016.

[Article]

AN: 612123764

When considering long-term prognosis and results in adult age following treatment of cryptorchidism in childhood there are three main issues to be discussed: cosmetics, fertility, and malignancy. In the present review, the most recent research on the topics related to summaries of well-known knowledge on the field is presented. To some extent a smaller testis in a higher scrotal position than normal must be accepted as a fair cosmetic result after orchidopexy in

childhood. The smaller testis size is related to the impaired fertility potential of the testis. In cases with atrophy, testicular prosthesis implantation is an option with good operative results. The risk of impaired fertility potential in adults treated in childhood for cryptorchidism is still significant and worst in bilateral disease. We need repetitive solid long-term follow-up data to show that orchidopexy performed within first year of life has markedly improved the fertility potential. Men previously having orchidopexy for cryptorchidism related to intra-abdominal testes, abnormal external genitalia and/or abnormal karyotype, and/or hypospadias are of special increased risk of developing testicular cancer. In these cases intratubular germ cell neoplasia may be diagnosed in prepubertal age. Early orchidopexy may lower the risk of developing testicular cancer.

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Institution

(Thorup) Department of Pediatric Surgery, Rigshospitalet, Copenhagen, Denmark (Thorup, Cortes) Institute for Clinical Medicine, Faculty of Health and Medical Sciences, University of Copenhagen, Copenhagen, Denmark

(Cortes) Department of Pediatrics, University Hospital, Hvidovre, Denmark

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696.

Surgery in disorders of sex development (DSD) with a gender issue: If (why), when, and how?.

Mouriquand P.D.E., Gorduza D.B., Gay C.-L., Meyer-Bahlburg H.F.L., Baker L., Baskin L.S., Bouvattier C., Braga L.H., Caldamone A.C., Duranteau L., El Ghoneimi A., Hensle T.W., Hoebeke P., Kaefer M., Kalfa N., Kolon T.F., Manzoni G., Mure P.-Y., Nordenskjold A., Pippi Salle J.L., Poppas D.P., Ransley P.G., Rink R.C., Rodrigo R., Sann L., Schober J., Sibai H., Wisniewski A., Wolffenbuttel K.P., Lee P.

Embase

Journal of Pediatric Urology. 12 (3) (pp 139-149), 2016. Date of Publication: 01 Jun 2016.

[Review]

AN: 610092104

Ten years after the consensus meeting on disorders of sex development (DSD), genital surgery continues to raise questions and criticisms concerning its indications, its technical aspects, timing and evaluation. This standpoint details each distinct situation and its possible management in 5 main groups of DSD patients with atypical genitalia: the 46,XX DSD group (congenital adrenal hyperplasia); the heterogeneous 46,XY DSD group (gonadal dysgenesis, disorders of steroidogenesis, target tissues impairments ...); gonosomic mosaicisms (45,X/46,XY patients); ovo-testicular DSD; and "non-hormonal/non chromosomal" DSD. Questions are summarized for each DSD group with the support of literature and the feed-back of several world experts. Given the complexity and heterogeneity of presentation there is no consensus regarding the indications, the timing, the procedure nor the evaluation of outcome of DSD surgery. There are, however, some issues on which most experts would agree: 1) The need for identifying centres of expertise with a multidisciplinary approach; 2) A conservative management of the gonads in complete androgen insensitivity syndrome at least until puberty although some studies expressed concerns about the heightened tumour risk in this group; 3) To avoid vaginal dilatation in children after surgical reconstruction; 4) To keep asymptomatic mullerian remnants during childhood; 5) To remove confirmed streak gonads when Y material is present; 6) It is likely that 46,XY cloacal exstrophy, aphallia and severe micropenis would do best raised as male although this is based on limited outcome data. There is general acknowledgement among experts that timing, the choice of the individual and irreversibility of surgical procedures are sources of concerns. There is, however, little evidence provided regarding the impact of non-treated DSD during childhood for the individual development, the parents, society and the risk of stigmatization. The low level of evidence should lead to design collaborative prospective studies involving all parties and using consensual protocols of evaluation.

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Status

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Author NameID

Gay, Claire-Lise; ORCID: <http://orcid.org/0000-0003-3355-8097> Hensle, Terry W.; ORCID: <http://orcid.org/0000-0002-8061-9094>

Bouvattier, Claire; ORCID: <http://orcid.org/0000-0001-7124-3016>

Institution

(Mouriquand, Gorduza, Gay, Mure) Department of Paediatric Urology/Paediatric Surgery, Universite Claude-Bernard, Hospices Civils de Lyon, Lyon, France (Mouriquand, Gorduza, Gay, Mure) Centre National de Reference Maladies Rares sur les Anomalies Congenitales du Developpement Genito-Sexuel, Lyon, France

(Meyer-Bahlburg) NYS Psychiatric Institute, New York, NY, United States

(Meyer-Bahlburg, Hensle) College of Physicians & Surgeons of Columbia University, New York City, NY, United States

(Baker) Children's Medical Center, University of Texas Southwestern Medical Center, Dallas, TX, United States

(Baskin) Pediatric Urology, UCSF Benioff Children's Hospital, San Francisco, CA, United States

(Bouvattier) Service d'Endocrinologie de l'enfant, GHU Paris-Sud, Hopital de Bicetre, Paris, France

(Bouvattier, Duranteau) Centre National de Reference Maladies Rares sur les Anomalies Congenitales du Developpement Genito-Sexuel, Paris, France

(Braga) Division of Urology, Department of Surgery, McMaster University, Toronto, Canada

(Caldamone) Pediatric Urology, Hasbro Children's Hospital, Providence, RI, United States

(Caldamone) Surgery (Urology) and Pediatrics, Brown University, Rhode Island Hospital, Providence, RI, United States

(Duranteau) Adolescent Gynaecology, Hopitaux Universitaires Paris Sud (Bicetre), Paris, France

(El Ghoneimi) Pediatric Surgery and Urology, University Hospital Robert Debre, APHP, University Paris Diderot, Sorbonne Paris Cite, Paris, France

(Hoebeke) Urology, Ghent University Hospital, Gent, Belgium

(Kaefer) Riley Children's Hospital, Indiana University School of Medicine, Indianapolis, IN, United States

(Kalfa, Rink) Service de Chirurgie Viscerale et Urologique Pediatrique, Hopital Lapeyronie, CHU de Montpellier, Universite de Montpellier, France

(Kolon) Pediatric Urology, Children's Hospital of Philadelphia, PA, United States

(Kolon) Perelman School of Medicine at University of Pennsylvania, PA, United States

(Manzoni) Pediatric Urology, Fondazione IRCCS CaGranda, Ospedale Maggiore Policlinico, Milan, Italy

(Nordenskjold) Department of Women's and Children's Health, Karolinska Institutet, Stockholm, Sweden

(Pippi Salle) Department of Surgery, Sidra Medical and Research Center, Doha, Qatar

(Poppas) Komansky Center for Children's Health, New York Presbyterian Hospital, Weill Cornell Medicine, New York, NY, United States

(Ransley) Great Ormond Street Hospital, Institute of Child Health, London, United Kingdom

(Rodrigo) Department of Surgery, Dalhousie University, IWK Health Centre, Halifax, Nova Scotia, Canada

(Rodrigo) Department of Urology, Dalhousie University, IWK Health Centre, Halifax, Nova Scotia, Canada

(Sann) Conseil d'ethique pediatrique, Centre Hospitalo-Universitaire de Lyon, France

(Schober) UPMC Hamot Medical Center, Erie, PA, United States

(Sibai) Paediatric Surgery, University of Casablanca, Morocco

(Wisniewski) Neuropsychology, University of Palo Alto, CA, United States

(Wolffenbuttel) Department of Urology and Pediatric Urology, Erasmus MC Sophia Children's Hospital, Rotterdam, Netherlands

(Lee) Penn State Hershey Pediatric Endocrinology, PA, United States

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697.

Fenestrated sheet split-thickness skin grafting for reconstruction of penile skin loss in pediatric population.

Chertin B., Kocherov S., Binenboym R., Gronovich Y., Tuchman I., Chertin L., Baskin L.

Embase

Journal of Pediatric Surgery. 51 (8) (pp 1362-1365), 2016. Date of Publication: 01 Aug 2016.

[Article]

AN: 608422637

Objectives We aimed to evaluate our experience with fenestrated sheet split-thickness skin grafts (STSGs) in the pediatric population. **Materials and methods** We retrospectively studied a cohort of 17 children 2-18 years old who underwent skin grafting owing to circumcision injuries (2 patients), traumatic penile injury (1) and after previous multiple hypospadias surgery (14). Fenestrated 0.012 in sheet STSGs from thigh area (15 patients) and buttock area (2) were fashioned to resurface the denuded penis following reconstruction. The median follow up was 13 years (range 1-19 years). **Results** There was 94% take of the grafts. One patient required additional grafting following first graft infection. Six patients underwent concomitant surgery at the

time of grafting (4 chordee repair and 2 meatoplasty). Two patients had slight chordee at 3 and 6 years postoperatively, and 2 with the history of preputial tubularized island flap hypospadias repair had developed a urethral stricture, which required staged repair with buccal mucosa 12 and 14 years following primary hypospadias repair. Six sexually active patients reported normal sexual intercourse and sensation following grafting. None of the patients demonstrated shrinkage of the STSGs over the follow up period. Conclusions Our data demonstrated that the use of fenestrated sheet STSGs in patients with penile skin loss yields satisfactory functional and cosmetic outcomes. The buttocks might be considered as a preferable donor site in terms of avoiding a visible scar.

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Status

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Author NameID

Chertin, Boris; ORCID: <http://orcid.org/0000-0002-8347-0969>

Institution

(Chertin, Kocherov, Chertin) Department of Pediatric Urology, Shaare Zedek Medical Center, Faculty of Medical Science, Hebrew University, Jerusalem, Israel (Binenboym, Gronovich, Tuchman) Department of Plastic Surgery, Shaare Zedek Medical Center, Faculty of Medical Science, Hebrew University, Jerusalem, Israel

(Baskin) Department of Pediatric Urology, UCSF, San Francisco, CA, United States

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698.

Morphological and functional hip long-term results after exstrophy repair.

Kertai M.A., Rosch W.H., Brandl R., Hirschfelder H., Zwink N., Ebert A.K.

Embase

European Journal of Pediatric Surgery. 26 (6) (pp 508-513), 2016. Date of Publication: 01 Dec 2016.

[Article]

AN: 606317953

Introduction Abnormalities of the bony pelvis in exstrophy-epispadias complex (EEC) and their possible relation to hip disease are well described. However, there is a lack of information about long-term orthopedic consequences and hip function in patients with EEC. Therefore, we investigated clinical and radiological results in an EEC patient cohort after long-term follow-up. **Patients and Methods** We conducted a cross-sectional study using standardized radiography, clinical investigation, and the Harris hip score. Seventeen postpuberty consecutive unselected EEC patients (3 female, 14 male; mean age 18.2 years) that presented to our clinic due to urological procedures or routine check-up from 2010 to 2011 were included. All had undergone symphysis approximation with a traction bandage without osteotomy in early childhood.

Radiological analysis was conducted offline by two independent investigators. **Results**

Radiological analysis showed a mean pubic diastasis of 5.1 cm (range 2.8-8.5 cm). Borderline hip dysplasia was present in four patients, one of them having had co-occurring developmental hip dysplasia in previous history. No severe dysplasia, subluxation, or luxation of the hip was found; however, one patient showed early hip arthrosis. Clinical examination revealed no relevant restriction of range of motion, although rotation and abduction were slightly altered in five patients. None of the EEC patients complained about pain or restriction in sports or daily activities. Harris hip score was perfect for all but one study participants. **Conclusion** Despite EEC-specific hip morphology, long-term hip function is not impaired in patients after symphyseal approximation without osteotomy in the newborn period. The symphysis diastasis after this procedure is comparable to available postosteotomy data. The large majority of EEC patients did not show dysplastic or degenerative hip disease. Functional hip score results confirmed reasonable age-related hip function in nearly all examined patients. However, postnatal ultrasound hip screening is recommended to prevent and adequately treat potential co-occurring developmental hip dysplasia.

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Status

Embase

Institution

(Kertai) Department of Pediatric Surgery, Klinik St. Hedwig, University Medical Center Regensburg, Regensburg, Germany (Rosch) Department of Pediatric Urology, Klinik St. Hedwig, University Medical Center Regensburg, Regensburg, Germany

(Brandl) Department of Radiology, Krankenhaus der Barmherzigen Bruder, Regensburg, Germany

(Hirschfelder) Department of Rehabilitation Medicine and Orthopaedics, Klinikum Nurnberg, Nurnberg, Germany

(Zwink) Division of Clinical Epidemiology and Aging Research, German Cancer and Research Center, Heidelberg, Germany

(Ebert) Department of Urology and Pediatric Urology, Ulm University, Prittwitzstr. 43, Ulm 89075, Germany

Publisher

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699.

Can Classic Bladder Exstrophy be Safely and Successfully Reconstructed at a Low Volume Center?.

Ben-Chaim J., Binyamini Y., Segev E., Sofer M., Bar-Yosef Y.

Embase

Journal of Urology. 195 (1) (pp 150-154), 2016. Date of Publication: 01 Jan 2016.

[Article]

AN: 607038270

Purpose Exstrophy reconstruction is challenging and requires expertise and experience. However, many patients are treated at low volume centers. We evaluated whether classic bladder exstrophy could be safely and successfully reconstructed at a low volume center. Materials and Methods A total of 31 patients with classic bladder exstrophy were primarily treated at our low volume center during a 17-year period. A total of 22 patients underwent primary closure within 5 days of birth and 9 underwent delayed closure with osteotomy. Of the patients 29 underwent planned modern staged repair and 2 underwent attempted complete primary repair. Results The bladder was successfully closed in all 31 children. All 22 newborns underwent primary bladder closure without osteotomy, including 4 with extremely small bladder plates. Bladder neck obstruction developed in 3 patients (10%), of whom 2 were treated successfully with transurethral dilation and 1 underwent open repair. Epispadias repair was successful in 12 of 15 patients

undergoing the Cantwell-Ransley technique and in 2 of 4 undergoing complete primary repair. A total of 16 patients underwent bladder neck reconstruction, of whom 9 are awaiting appropriate age or capacity, 4 were lost to followup, 1 is continent after bladder closure alone and 2 underwent continent diversion (1 after failed bladder neck reconstruction). Of the 15 patients with at least 1 year of followup after bladder neck reconstruction 9 are continent day and night, 2 are continent only during the daytime and 4 are incontinent, for a 73% post-bladder neck reconstruction continence rate (11 of 15 patients). Conclusions Successful exstrophy reconstruction is achievable at a low volume center, with results comparable to those of high volume centers.

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Institution

(Ben-Chaim, Binyamini, Segev, Sofer, Bar-Yosef) Pediatric Urology Unit, Dana-Dwek Children's Hospital, Tel Aviv Sourasky Medical Center, Sackler Faculty of Medicine, Tel Aviv University, Tel Aviv, Israel

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700.

Post-sialendoscopy ductoplasty by salivary duct stent placements.

Su C.-H., Lee K.-S., Tseng T.-M., Hung S.-H.

Embase

European Archives of Oto-Rhino-Laryngology. 273 (1) (pp 189-195), 2016. Date of Publication: 01 Jan 2016.

[Article]

AN: 602142338

With damage to a duct or papilla after sialendoscopy, a stent may be necessary to prevent re-stenosis and for maintaining the salivary duct open after complete sialendoscopy. However factors affecting outcomes and complications after stent placement remain unclear. This study

aimed to report preliminary experiences in salivary duct stent placement after sialendoscopy. Data from 35 procedures in 33 patients who received sialendoscopy with salivary duct stent placements at Mackay Memorial Hospital between October 2013 and June 2014 were recorded and compared for clinical data, as well as procedural techniques, findings, and outcomes. In the 35 stent placement procedures, the hypospadias silastic stent tubes were used in 27 and the Fr. 5 pediatric feeding tubes were used in the remaining eight. When the hypospadias silastic stent tubes were used for stenting, the stent obstruction and irritation rates were higher compared to those who used the Fr. 5 pediatric feeding tube (100 vs. 0 % and 67 vs. 33 %, respectively). None of the stents secured by a 5-0 nylon suture were complicated by dislocation but when the stents were secured by 6-0 nylon sutures, the dislocation rate went as high as 47.4 %. The duration needed for salivary duct stent placement might be potentially shortened to only 2 weeks. If a salivary duct stent is intended to be placed for a certain period before its scheduled removal, a suture strength equivalent or stronger than the 5-0 nylon suture should be considered for stent fixation.

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Status

Embase

Institution

(Su, Lee) Department of Otorhinolaryngology, Mackay Memorial Hospital, New Taipei City, Taiwan (Republic of China) (Su, Hung) Department of Otolaryngology, School of Medicine, Taipei Medical University, Taipei, Taiwan (Republic of China)

(Tseng, Hung) Department of Otolaryngology, Taipei Medical University Hospital, No. 252, Wu-Hsing Street, Taipei 110, Taiwan (Republic of China)

Publisher

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Impact of changing trends in technique and learning curve on outcome of hypospadias repair: An experience from tertiary care center.

Ansari M.S., Agarwal S., Sureka S.K., Mandhani A., Kapoor R., Srivastava A.

Embase

Indian Journal of Urology. 32 (3) (pp 216-220), 2016. Date of Publication: July-September 2016.

[Article]

AN: 611250556

Introduction: Apart from numerous clinical factors, surgical experience and technique are important determinants of hypospadias repair outcome. This study was aimed to evaluate the learning curve of hypospadias repair and the impact of changing trends in surgical techniques on the success of primary hypospadias repair.

Material(s) and Method(s): We retrospectively analyzed data of 324 patients who underwent primary repair of hypospadias between January 1997 and December 2013 at our center. During the initial 8 years, repairs were performed by multiple 5 different urologists. From 2005 onwards, all procedures were performed by a single urologist. The study cohorts were categorized into three groups; Group I, surgeries performed between 1997-2004 by multiple surgeons, Group II, between 2005-2006 during the initial learning curve of a single surgeon, and Group III, from 2007 onwards after completion of the learning curve of the single surgeon. The groups were compared in respect to surgical techniques, overall success and complications.

Result(s): Overall 296 patients fulfilled the inclusion criterion, 93 (31.4%), 50 (16.9%), and 153 (51.7%) in Group I, II, and III, respectively. Overall success was achieved in 60 (64.5%), 32 (64%), and 128 (83.7%) patients among the three groups respectively ($P < 0.01$). Nineteen (20.4%), 20 (40%), and 96 (62.7%) patients underwent tubularized incised plate repair in Group I, II, and III, with successful outcome in 12 (63.2%), 15 (75%), and 91 (94.8%) patients, respectively ($P < 0.01$). The most common complication among all groups was urethrocutaneous fistula, 20 (21.5%) in Group I, 11 (22%) in Group II, and 17 (11.1%) in Group III.

Conclusion(s): There is a learning curve for attaining surgical skills in hypospadias surgery.

Surgeons dedicated for this surgery provide better results. Tubularized incised plate urethroplasty appear promising in both distal and proximal type hypospadias.

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Embase

Institution

(Ansari, Agarwal, Sureka, Mandhani, Kapoor, Srivastava) Department of Urology, Sanjay Gandhi Post Graduate Institute of Medical Sciences, Lucknow, Uttar Pradesh 226 014, India

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702.

Results of distal hypospadias repair after pediatric urology fellowship training: A comparison of junior surgeons with their mentor.

Bush N.C., Barber T.D., Dajusta D., Prieto J.C., Ziada A., Snodgrass W.

Embase

Journal of Pediatric Urology. 12 (3) (pp 162.e1-162.e4), 2016. Date of Publication: 01 Jun 2016.

[Article]

AN: 611105516

Background Teaching and learning hypospadias repair is a major component of pediatric urology fellowship training. Educators must transfer skills to fellows, without increasing patient complications. Nevertheless, few studies report results of surgeons during their first years of independent practice. Purpose To review outcomes of distal hypospadias repairs performed during the same 2-year period by consecutive, recently matriculated, surgeons in independent practice, and to compare them to results by their mentor (with >20 years of experience). Materials Exposure to hypospadias surgery during fellowship was determined from case logs of five consecutive fellows completing training from 2007-2011. TIP was the only technique used to repair distal hypospadias. No fellow operated independently or performed complete repairs under supervision. Instead, the first 3 months were spent assisting their mentor, observing surgical methodology and decision-making. Then, each performed selected portions under direct supervision, including: degloving, penile straightening, developing glans wings, incising and tubularizing the urethral plate, creating a barrier layer, sewing the glansplasty, and skin closure. Overall fellow participation in each case was <50%. In 2011-2012, urethroplasty complications (fistula, glans dehiscence, meatal stenosis, urethral stricture, diverticulum) were recorded for consecutive patients undergoing primary distal repair by these recent graduates in their independent practices. The fellow graduating in 2011 provided 1 year of data. All patients undergoing repair during the study period were included in the analysis, except those lost to follow-up after catheter removal. Composite urethroplasty complications were compared between

junior surgeons, and between junior surgeons and their mentor, with Fisher's exact contingency test. Results Training logs indicated fellow participation ranged from 76-134 hypospadias repairs, including distal, proximal and reoperative surgeries. Post-graduation case volumes ranged from 25-68 by junior surgeons versus 136 by the mentor. With similar mean follow-up, urethroplasty complication rates were statistically the same between the former fellows, and between them versus the mentor, ranging from 5-13%. Nearly all were fistulas or glans dehiscence. Junior surgeons reported they performed TIP as learned during fellowship, with one exception who used 7-0 polydioxanone rather than polyglactin for urethroplasty. Discussion This is the first study directly comparing hypospadias surgical outcomes by recently graduated fellows in independent practice with those of their mentor. We found junior surgeons achieved similar results for distal TIP hypospadias repair. Although their participation during training largely comprised observation and surgical assistance, with discrete performance of key steps, skills sufficient to duplicate the mentor's results were transferred. These data suggest there should be no learning curve for distal hypospadias after training. This report raises several considerations for surgical educators. First, mentors should review their own results, to be certain that they are correctly performing and teaching procedures. Second, programs need to determine key steps for procedures they teach, and then emphasize their optimal performance. Finally, mentors should expect former fellows to report back their initial results of hypospadias repair to be certain lessons taught were learned. Otherwise, preventable complications resulting from technical errors will be multiplied in the children operated by their trainees as they enter independent practice.

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Status

Embase

Institution

(Bush, Snodgrass) PARC Urology, Frisco, TX, United States (Barber) Department of Pediatric Urology, Helen DeVos Children's Hospital, Grand Rapids, MI, United States

(Dajusta) Nationwide Children's Hospital, Columbus, OH, United States

(Prieto) Pediatric Urology, Children's Hospital of San Antonio, San Antonio, TX, United States

(Ziada) Department of Urology, University of Kentucky, Lexington, KY, United States

(Ziada) Department of Pediatrics, University of Kentucky, Lexington, KY, United States

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703.

Peyronie's disease: A literature review on epidemiology, genetics, pathophysiology, diagnosis and work-up.

Al-Thakafi S., Al-Hathal N.

Embase

Translational Andrology and Urology. 5 (3) (pp 280-289), 2016. Date of Publication: 01 Jun 2016.

[Review]

AN: 610767211

Peyronie's disease (PD), a fibromatous disorder of the tunica albuginea of the penile corpus cavernosum, named after the French physician Francois de la Peyronie, is characterized by pain, plaque formation, penile curvature, and plaque calcification. The epidemiological data on PD is inconsistent, with recent reports stating a prevalence of up to 9%, and the condition affecting men of all ages, from teenagers to septuagenarians. We are just beginning to elucidate the role of genetics as a causative factor for PD. Chromosomal abnormalities and single-nucleotide polymorphisms have been shown to be associated with fibrotic diatheses. Tunical mechanical stress and microvascular trauma are major contributory factors to the pathophysiology of PD. The diagnosis of PD can be made using a combination of clinical history, physical examination and, sometimes, imaging modalities. A better understanding of the molecular pathophysiology of this condition remains paramount for the development of newer and more effective disease-targeted interventions.

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Institution

(Al-Thakafi) Department of Urology, King Saud Medical City, Riyadh, Saudi Arabia (Al-Hathal) Department of Urology, King Faisal Specialist Hospital and Research Center, Riyadh 1211, Saudi Arabia

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704.

Tubularised incised-plate versus tubularisation of an intact and laterally augmented plate for hypospadias repair: A prospective randomised study.

Elbakry A., Hegazy M., Matar A., Zakaria A.

Embase

Arab Journal of Urology. 14 (2) (pp 163-170), 2016. Date of Publication: 01 Jun 2016.

[Article]

AN: 610571196

Objectives To compare the outcome of hypospadias repair using tubularised incised-plate (TIP) urethroplasty and tubularisation of an intact and laterally augmented urethral plate. **Patients and methods** This prospective randomised study included 370 patients with primary distal hypospadias. All had urethral plate widths of 8-10 mm and a glans of ≥ 15 mm. Exclusion criteria were previous repair, circumcision, a wide urethral plate of >10 mm or a narrow plate of <8 mm in diameter, a small glans of <15 mm in diameter, chordee of $>30^\circ$, and hormonal stimulation. Patients were randomised into two groups: Group 1 (185 patients) underwent TIP urethroplasty and Group 2 (185 patients) underwent tubularisation of the intact plate with lateral augmentation of the urethral plate using penile skin. The follow-up period was 12-28 months. **Results** There were 172 evaluable patients in Group 1 and 177 in Group 2. The urethroplasty was successful in 83.2% and 94.4% in Groups 1 and 2, respectively. Complications occurred in 16.8% in Group 1 and 5.6% in Group 2 ($P = 0.001$). Meatal stenosis occurred in 7% and 3.4% in Groups 1 and 2, respectively ($P = 0.130$). There were statistically significant differences in the wound dehiscence, fistula, and re-operation rates of Group 1 versus Group 2, at 6% versus 0%, 9.8% versus 2.8%, and 13.4% versus 5.6%, respectively. The presence of mild chordee did not affect the complication rate ($P = 0.242$). The mean (SD) operative time was 56.7 (8.9) min in Group 1 and 93.7 (8.3) min in Group 2 ($P < 0.001$). **Conclusion** The outcome of tubularised intact and laterally augmented plate is better than classical TIP urethroplasty of hypospadias. Further trials are mandatory to extend the indications of the technique.

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Status

Embase

Institution

(Elbakry, Hegazy, Matar, Zakaria) Department of Urology, Suez Canal University Hospital, Ismailia, Egypt

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Year of Publication

2016

705.

Fluconazole use and birth defects in the National Birth Defects Prevention Study.

Howley M.M., Carter T.C., Browne M.L., Romitti P.A., Cunniff C.M., Druschel C.M.

Embase

American Journal of Obstetrics and Gynecology. 214 (5) (pp 657e1-657e9), 2016. Date of Publication: 01 May 2016.

[Article]

AN: 610122157

Background Low-dose fluconazole is used commonly to treat vulvovaginal candidiasis, a condition occurring frequently during pregnancy. Conflicting information exists on the association between low-dose fluconazole use among pregnant women and the risk of major birth defects. Objective We used data from the National Birth Defects Prevention Study to examine this association. Study Design The National Birth Defects Prevention Study is a multisite, population-based, case-control study that includes pregnancies with estimated delivery dates from 1997 to 2011. Information on fluconazole use in early pregnancy was collected by self-report from 31,645 mothers of birth defect cases and 11,612 mothers of unaffected controls. Adjusted odds ratios and 95% confidence intervals were estimated for birth defects with 5 or more exposed cases; crude odds ratios and exact 95% confidence intervals were estimated for birth defects with 3-4 exposed cases. Results Of the 43,257 mothers analyzed, 44 case mothers and 6 control mothers reported using fluconazole. Six exposed infants had cleft lip with cleft palate, 4 had an atrial septal defect, and each of the following defects had 3 exposed cases: hypospadias, tetralogy of Fallot, d-transposition of the great arteries, and pulmonary valve stenosis. Fluconazole use was associated with cleft lip with cleft palate (odds ratio = 5.53; confidence interval = 1.68-18.24) and d-transposition of the great arteries (odds ratio = 7.56; confidence interval = 1.22-35.45). Conclusions The associations between fluconazole and both cleft lip with cleft palate and d-transposition of the great arteries are consistent with earlier published case reports but not recent epidemiologic studies. Despite the larger sample size of the National Birth Defects Prevention

Study, fluconazole use was rare. Further investigation is needed in large studies, with particular emphasis on oral clefts and conotruncal heart defects.

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Status

Embase

Institution

(Howley, Browne, Druschel) Congenital Malformations Registry, New York State Department of Health, Albany, NY, United States (Carter) Center for Human Genetics, Marshfield Clinic, Marshfield, WI, United States

(Browne, Druschel) Department of Epidemiology and Biostatistics, School of Public Health, University at Albany, Rensselaer, NY, United States

(Romitti) Department of Epidemiology, College of Public Health, University of Iowa, Iowa City, IA, United States

(Cunniff) Department of Pediatrics, Weill Cornell Medical College, New York, NY, United States

Publisher

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706.

Plastibell circumcision of 2,276 male infants: A multi-centre study.

Jimoh B.M., Odunayo I.S., Chinwe I., Akinfolarin O.O., Oluwafemi A., Olusanmi E.J.

Embase

Pan African Medical Journal. 23 (no pagination), 2016. Article Number: 35. Date of Publication: 09 Feb 2016.

[Article]

AN: 610073586

Introduction: The World Health Organization's manual on male circumcision listed Plastibell technique as a well-proven paediatric method with respect to the results and complications.

Although, literatures abound on its wide acceptability, there are few multi- centered reports from this environment. The objective was to evaluate the cases of infant circumcision by Plastibell device from two medical institutions.

Method(s): All consecutive infants who had Classical Plastibell Circumcision (PC) at the Federal Staff Medical Centre, Abuja and the Lagos State University Teaching Hospital, Ikeja between February 2011 and June 2015 were included in this cross-sectional study. The procedures were performed by surgical registrars and medical officers after ninety minutes of topical anesthesia to the penis. Data harvested from the standard proforma were analysed using Statistical Package for Social Science 20.0 for window.

Result(s): A total of 2,276 infants had classical PC within the study period. Their ages at circumcision ranged from 4 days to 3 months with a mean age of 17 days. Majority of the boys were circumcised at second week of life (n=1,394, 61.2%). All the cases were performed for religious (53%) and cultural (47%) reasons. The most common Plastibell size deployed was 1.3cm (n=1,040, 45.7%) while 1.6cm was the least commonly used ring (n=10, 0.4%). The mean time for device to fall-off was 6 days (range 4-12 days). There was no correlation between the age at circumcision and Plastibell size. We recorded an overall complication rate of 1.1% with postoperative bleeding leading the pack (n=12, 48%). No case of urethrocuteaneous fistula was seen. We detected 17 cases (0.7%) of distal hypospadias in whom circumcisions were postponed till the time of hypospadias repairs.

Conclusion(s): The main indication for infant circumcision in our environment was religious. The PC has good safety profile with few easily correctable early complications. Detailed attention to placement of ligature, selection of appropriate Plastibell size and adequate parental education are key to preventing post-procedure mishaps.

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Institution

(Jimoh) Department of Surgery, Federal Staff Medical Centre, Abuja, Nigeria (Odunayo, Akinfolarin, Olusanmi) Urology Unit, Department of Surgery, Lagos State University Teaching Hospital, Ikeja, Nigeria

(Chinwe, Oluwafemi) Department of Obstetrics and Gynecology, Federal Staff Medical Centre, Abuja, Nigeria

Publisher

African Field Epidemiology Network (E-mail: sec@afenet.net)

Year of Publication

2016

707.

Correlation of androgen receptor and SRD5A2 gene mutations with pediatric hypospadias in 46, XY DSD children.

Fu X.H., Zhang W.Q., Qu X.S.

Embase

Genetics and Molecular Research. 15 (1) (no pagination), 2016. Article Number: 15018232. Date of Publication: 31 Mar 2016.

[Article]

AN: 609902121

We performed an exploratory study by analyzing the correlation of 46, XY disorders of sex development (46, XY DSD) with androgen receptor (AR) and steroid 5alpha-reductase-2 (SRD5A2) gene mutations and a safety analysis of dihydrotestosterone (DHT) gel treatment for pediatric micropenis. We collected samples from 76 pediatric patients with 46, XY DSD and 50 healthy adult men with normal fertility as the control group. The pediatric patients were treated with DHT gel (0.1-0.3 mg/kg/day) for three to six months. The extended penis length, testicular volume, and multiple blood parameters were collected before treatment and one, three, and six months after treatment. Of the 76 cases with 46, XY DSD, 31.58% had hypospadias with micropenis and 6.58% had male pseudohermaphroditism. Through AR gene screening, it was found that 14 patients had AR point mutations and 22 patients had SRD5A2 mutations. After treatment with DHT, the penis length of the patients significantly improved after one, three, and six months of treatment, with longer treatment times resulting in greater improvement. Before treatment with DHT, the average serum DHT value of patients with 46, XY DSD was 24.29 pg/mL. After one, three, and six months of treatment, this value increased to 430.71, 328.9, and 323.6 pg/mL, respectively. We conclude that for pediatric patients who have male hermaphroditism or hypospadias with micropenis, AR and SRD5A2 gene mutation detection should be performed. Local application of DHT gel can promote penis growth effectively without systemic adverse reactions.

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Institution

(Fu, Zhang, Qu) Pediatric Surgery Department, People's Hospital of Binzhou, Binzhou, Shandong Province, China

Publisher

Fundacao de Pesquisas Cientificas de Ribeirao Preto (E-mail: funpecrp@uol.com.br)

Year of Publication
2016

708.

Use of human acellular dermal matrix during classic bladder exstrophy repair.

Bonitz R.P., Hanna M.K.

Embase

Journal of Pediatric Urology. 12 (2) (pp 114.e1-114.e5), 2016. Date of Publication: 01 Apr 2016.

[Article]

AN: 609069203

Introduction The extent of the abdominal wall defect in people with classic bladder exstrophy (CBE) varies, and can be extensive. In this study, human acellular dermis (HAD) was used to bridge the fascial gap, as an alternative to osteotomy, to support a fascial repair of the abdominal wall, and as a filler in selected cases of CBE. **Objective** To demonstrate the efficacy of the employed techniques of using HAD within the bladder exstrophy population. **Materials and methods** The medical records of six males, born with CBE, and who had abdominal wall defects were reviewed. Two children, aged 6 and 8 years old, were referred from overseas with unrepaired bladder exstrophy plates and large abdominal wall defects (8 cm and 12 cm wide). Both had their bladders reconstructed, placed within the pelvis, and HAD was used to replace the absent abdominal wall (bridged repair) without the use of pelvic osteotomy. In three other patients, HAD reinforced the native fascial repair (bolster repair). In three patients, HAD also served as a filler for the abdominal depression that was present following initial staged repair. Where HAD was used for bridged or bolster repair, the edges of the allograft were extended 2-3 cm circumferentially beyond the perimeter of the abdominal wall defect. **Results** All six patients healed well, without evidence of abdominal wall hernias at 1-3 years postoperatively. Functionally, each patient regained an appropriate level of abdominal wall strength. Two children successfully underwent a secondary procedure through the bridged allograft repair, as each required bladder neck reconstruction and bilateral ureteral reimplantation through the reconstructed abdominal wall. Continence was achieved in these two patients, with one child voiding at 2-hourly intervals and the second at 3-hourly intervals. One patient developed a urethral-cutaneous fistula, distant to location of the allograft. There were no associated wound complications. **Conclusions** In this series of patients born with classic bladder exstrophy, HAD

acted as a biologic scaffold and allowed native cellular ingrowth and tissue remodeling. It served as an alternative to pelvic osteotomy in older patients with unrepaired CBE. The material reinforced a weak or potentially suboptimal fascial repair and filled a tissue gap, resulting in improved aesthetics. Given its ease of preparation and the lack of significant morbidity associated with its use, combined with the functional and esthetic results in the present series, HAD may be considered during delayed reconstruction of abdominopelvic tissues in people born with CBE.

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Author NameID

Bonitz, Robert Paul; ORCID: <http://orcid.org/0000-0002-4607-7818>

Institution

(Bonitz, Hanna) Urology Division, Rutgers-New Jersey Medical School, 140 Bergen Street, Newark, NJ 07103, United States (Bonitz, Hanna) Children's Hospital of New Jersey, Saint Barnabas Health System, Livingston, NJ, United States

(Hanna) New York Presbyterian-Weill Cornell Medical Center, New York, NY, United States

Publisher

Elsevier Ltd

Year of Publication

2016

709.

A comparison between tourniquet application and epinephrine injection for hemostasis during hypospadias surgery: The effect on bleeding and postoperative outcome.

Alizadeh F., Fakoor A., Haghdani S.

Embase

Journal of Pediatric Urology. 12 (3) (pp 160.e1-160.e5), 2016. Date of Publication: 01 Jun 2016.

[Article]

AN: 608901822

Objective To compare tourniquet application and epinephrine injection for hemostasis during hypospadias surgery in terms of bleeding and complications. Methods Between April 2013 and September 2014, patients who were admitted for hypospadias repair were divided by random

allocation into either a diluted epinephrine injection (DE) group or tourniquet application (T) group for hemostasis during the procedure. In the T group, a rubber band was applied at the base of the penis after skin dissection, and in the DE group, epinephrine 1/100,000 was injected along the incision lines. The patients' ages, urethral defect lengths, pre-operative, intraoperative and postoperative variables were compared between the two groups. Results A total of 70 patients (35 in each group) were enrolled into the study. The mean ages and preoperative variables were not significantly different. Average blood loss was 23.51 +/- 15.36 cc in the tourniquet group and 15.99 +/- 10.00 cc in the epinephrine group, and was significantly higher in tourniquet group ($P = 0.022$); however, the mean operative time was not significantly different. Postoperative complications, described as Clavian classification, were reported in eight patients (23%) in the T group and 10 patients (30%) in the DE group, which was not significantly different. Conclusion Epinephrine injections minimize operative bleeding without significant harmful effects on postoperative outcomes; therefore, it could be considered to be a safe and effective method for preparation of a bloodless field during hypospadias surgery.

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Institution

(Alizadeh, Fakoore) Kidney Transplantation Research Center, Isfahan University of Medical Sciences (IUMS), Isfahan, Iran, Islamic Republic of (Haghdani) Hasheminejad Kidney Center (HKC), Iran University of Medical Sciences (IUMS), Tehran, Iran, Islamic Republic of

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710.

Scrotal-septal fasciocutaneous flap used as a multifunctional coverage for prior failed hypospadias repair.

Zhang S., Zhou C., Li F., Li S., Zhou Y., Li Q.

Embase

Urologia Internationalis. 96 (3) (pp 255-259), 2016. Date of Publication: 01 Apr 2016.

[Article]

AN: 608561208

Objectives: The study aims to report the method and outcome of using scrotal-septal fasciocutaneous flap as a multifunctional coverage for prior failed hypospadias repair.

Method(s): From January 2014 to June 2015, 18 hypospadias patients who have undergone repeated failed surgeries were enrolled. Their penile skin, urethral plate and dartos fascia are not enough to reconstruct the urethra, but have well-developed scrota. We performed urethroplasty by buccal mucosa free grafting and tubularized anastomosis 6 months after the urethroplasty. Then, scrotal-septal fasciocutaneous flap was used to be a multifunctional coverage on the surface of anastomotic urethra, which was a waterproof layer and cutaneous coverage.

Result(s): The skin flaps survived and the incisions healed in 18 patients. No fistula and stricture occurred. The scar of donor site seemed like a new scrotal raphe. The flap can slip slightly along with the preputial movement and retain the original sense of touch. All patients were followed up at 1 and 6 months by telephone or in person and gained good recoveries.

Conclusion(s): Scrotal-septal fasciocutaneous flap, including sufficient fascia tissue, reliable blood supply and skin coverage, is a good choice for the coverage of anastomotic urethra as both the waterproof layer and skin coverage, especially for hypospadias patients who have undergone several failed operations.

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Status

Embase

Institution

(Zhang, Zhou, Li, Li, Zhou, Li) 10th Department, Plastic Surgery Hospital, Chinese Academy of Medical Sciences, No. 33 Ba-Da-Chu Road, Shi Jing Shan District, Beijing 100144, China

Publisher

S. Karger AG

Year of Publication

2016

711.

Continence and quality of life with the modified Heitz-Boyer-Hovelacque rectal bladder for children with urinary incontinence following bladder exstrophy.

Messina M., Molinaro F., Ferrara F., Angotti R., Bulotta A.L., Di Maggio G., Bianchi A.

Embase

Journal of Pediatric Urology. 12 (3) (pp 174.e1-174.e6), 2016. Date of Publication: 01 Jun 2016.

[Article]

AN: 608421205

Background Bladder exstrophy and epispadias are severe congenital anomalies associated with an open bladder and urinary sphincter. Despite modern reconstruction, there is a significant incidence of residual or recurrent urinary incontinence that impacts on quality of life (QoL) and self-esteem, which in turn limits social interaction (Figure). The present study involved 14 patients, mainly from a Middle Eastern country, and reported the early findings with a modification of the Heitz-Boyer-Hovelacque rectal bladder technique for both urinary and faecal control. **Study design** Fourteen children, with a median age of 8.1 years, with poor quality of life and low self-esteem because of urinary incontinence and small polypoidal open bladders of 5-15 ml volume, mostly after bladder exstrophy surgery, were managed with a modification of the Heitz-Boyer-Hovelacque rectal bladder technique keeping an intact anal sphincter. The retrorectal pulled-through colon was anastomosed to the posterior wall of the rectum just above the external sphincter complex, thereby avoiding any possible injury to the anal sphincter. All patients had a normal colon and a competent anal sphincter without lumbosacral spinal or nerve anomalies. **Results** Ten children had a 5- to 10-year follow-up, one child had a 15-year follow-up, and three others, that were also continent, were excluded because of a <5-year follow-up. There were no postoperative complications, and all were dry and odour-free by day within 2-4 weeks of surgery. Two children still had minor urinary loss at night. There were no UTIs and renal function remained unimpaired. Eleven years after surgery, one child underwent excision of a pedunculated benign inflammatory polyp from the tip of the left ureter because of recurrent torsion and bleeding, there was no recurrence at the 2-year follow-up. None of the rectal or ureteric biopsies from any of the children showed metaplasia or neoplasia; however, in view of the potential long-term risks, all children were placed on a lifelong 'proctoscopy and biopsy' protocol. **Discussion** The ability to be dry and odour-free, and to wear normal clothing had a striking impact on QoL and psychological well-being of the children and their families. This was reflected in their positive overall approach, voluntary school attendance, and enthusiastic participation in communal events. All agreed that their improved genital appearance markedly contributed to their better body image and increased self-esteem. **Conclusion** These significant benefits, at a crucial time in the child's life, outweigh the potential risk of long-term neoplasia. Therefore, the Heitz-Boyer-Hovelacque rectal bladder technique is recommended with long-term proctoscopic follow-up.[Figure presented]

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Institution

(Messina, Molinaro, Ferrara, Angotti, Bulotta, Di Maggio) Division of Pediatric Surgery,
Department of Medical, Surgical and Neurological Sciences, University of Siena, Siena, Italy
(Bianchi) Neonatal and Paediatric Reconstructive Surgeon, Manchester, United Kingdom

Publisher

Elsevier Ltd

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2016

712.

Worldwide prevalence of hypospadias.

Springer A., van den Heijkant M., Baumann S.

Embase

Journal of Pediatric Urology. 12 (3) (pp 152.e1-152.e7), 2016. Date of Publication: 01 Jun 2016.

[Article]

AN: 607898124

Introduction Hypospadias is a common congenital malformation. Surgical repair and management of the long-term consequences require a substantial amount of socioeconomic resources. It is generally accepted that genetic and environmental factors play a major role in the etiology of hypospadias. There have been contradictory reports on rising hypospadias rates, and regional and ethnical differences. The exact prevalence of hypospadias is of major interest for healthcare providers, clinical medicine, and research. Objective To review the literature regarding the worldwide prevalence of hypospadias. Study design Pubmed, EMBASE and Google were systematically screened for: hypospadias, congenital malformation, anomaly, incidence, prevalence, and epidemiology. Exclusion criteria were surgical and risk-factor studies. To give an additional comprehensive overview, prevalence data were harvested from the Annual Report of the International Clearinghouse Centre for Birth Defects Surveillance and Research. Prevalence was reported as per 10,000 live births. Results Data were available from 1910 to 2013. The median study period was 9 years (range: 1-36 years). Approximately 90,255,200 births have been screened in all studies. The mean prevalence were: Europe 19.9 (range: 1-464), North America 34.2 (6-129.8), South America 5.2 (2.8-110), Asia 0.6-69, Africa 5.9 (1.9-110), and Australia 17.1-34.8. There were major geographical, regional, and ethnical differences, with an

extreme heterogeneity of published studies. Numerous studies showed an increasing prevalence; on the other hand, there were a lot of contradictory data on the prevalence of hypospadias. The summary table shows contradictory data from the five largest international studies available.

Discussion There was huge literature available on the prevalence of hypospadias. Most data derived from Europe and North America. Many methodological factors influenced the calculation of an accurate prevalence, and even more of the true changes in prevalence over time (no generally accepted and standardized definition of hypospadias, different monitoring systems, unclear efficiency of notification and data ascertainment, etc.). There was wide variation of prevalence according to countries and ethnicity, and there were conflicting data on the recent trends of prevalence. Moreover, there weren't any epidemiologic data available from many parts of the world. **Conclusion** True prevalence of hypospadias and trends were difficult to estimate. For the future, to be able to assess the true prevalence of hypospadias and changes in prevalence collaboration of national and international prospective registers is recommended.

[Table presented]

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Status

Embase

Author NameID

Springer A.; ORCID: <http://orcid.org/0000-0003-0300-6021>

Institution

(Springer, van den Heijkant) Interdisciplinary Center for Pediatric Urology, Medical University Vienna, Währinger Gurtel 18-20, Vienna 1090, Austria (Baumann) Department of Pediatric Surgery, Medical University Vienna, Währinger Gurtel 18-20, Vienna 1090, Austria

Publisher

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Year of Publication

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713.

Comparison of variables affecting the surgical outcomes of tubularized incised plate urethroplasty in adult and pediatric hypospadias.

Bhat A., Bhat M., Kumar V., Kumar R., Mittal R., Saksena G.

Embase

Journal of Pediatric Urology. 12 (2) (pp 108.e1-108.e7), 2016. Date of Publication: 01 Apr 2016.

[Article]

AN: 607756554

Introduction The American Academy of Pediatrics recommends operating on hypospadias between the ages of 6-12 months. Since most births in developed countries are conducted in a hospital, parents are likely to be well informed and counseled about the hypospadias. However, significant numbers of births in developing countries are still conducted at home, with illiteracy, poverty and ignorance often leading to late presentation at the hospital. Reported hypospadias-repair complication rates are higher in adults compared with those having surgery in childhood. The present study's objective was to evaluate the factors affecting surgical outcome in hypospadias patients undergoing tubularized and tubularized incised plate urethroplasty (TIPU) in adulthood compared with childhood. **Materials and methods** A prospective study of 60 adult patients >16 years, and 60 pediatric patients <5 years who underwent TIPU for primary hypospadias between May 2008 and May 2012. Patients were operated on by a single surgeon, under similar circumstances, and were pre-operatively examined to assess meatal location, chordee, and torsion; they were also examined intra-operatively for quality of spongiosum and urethral plate width. The outcomes were assessed by patient/parents for satisfaction regarding cosmesis, urinary stream and complications. **Results** The age of the patients varied from 16 to 27 years, with a mean of 20.8 years in adults, and 6 months to 5 years, with a mean of 2.1 years, in children. The type of hypospadias, degree of curvature, quality of spongiosum and urethral plate width were comparable in both groups, but complication rates were higher in adults (16.7%) than in the pediatric (6.7%) group (Figure 1A-D). Meatal stenosis responded well to dilatation, but fistulae required revision surgery and had a cure rate of 100%. The median follow-up was 37 months in adults, and 39 months in children. **Discussion** The higher complication rates in adults may be due to more frequent erections; increased susceptibility to infection along with relatively reduced vascularity lead to poor wound healing and increased complication rates. The limitation of the study was the small number of patients with mid and proximal hypospadias having lesser incidences in comparison with distal hypospadias. Adequate number of patients in these subgroups could have further strengthened the statistical correlation. Secondly, there was no objective criterion like uroflowmetry to assess urinary stream. **Conclusions** Complication rates were higher in adults undergoing TIPU compared with pediatric patients, which was also statistically significant in distal hypospadias. The important factors in surgical outcome were: severity of hypospadias, degree of curvature, quality of spongiosum, and urethral plate width. A. Bar diagram showing the effect of type of hypospadias on surgical outcome; B. the effect of chordee on surgical outcome; C. the effect of quality of spongiosum on surgical outcome; D. the effect of width of urethral plate on surgical outcome.

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Status

Embase

Institution

(Bhat, Bhat, Kumar, Kumar, Mittal, Saksena) Department of Urology, S.P. Medical College, C-15
Sadul Ganj, Bikaner, Rajasthan 334003, India

Publisher

Elsevier Ltd

Year of Publication

2016

714.

Continence in the cloacal exstrophy patient: What does it cost?.

Goldstein S.D., Inouye B.M., Reddy S., Lue K., Young E.E., Abdelwahab M., Grewal M.,
Wildonger S., Stec A.A., Gearhart J.P.

Embase

Journal of Pediatric Surgery. 51 (4) (pp 622-625), 2016. Date of Publication: 01 Apr 2016.

[Article]

AN: 607639976

Background Surgical advancements have made cloacal exstrophy (CE) a survivable condition, though management remains complex. Urologic, orthopedic, colorectal and gynecologic interventions are not standardized, and the cost of this care is high. While the importance of a successful primary closure in terms of outcomes is known, the economic consequences of failure remain uncharacterized. Methods A prospectively maintained institutional database of epispadias-exstrophy complex patients was reviewed for continent CE patients. Hospital charges for all inpatient admissions prior to achieving urinary continence were inflation-adjusted to year 2013 values using Consumer Price Index for medical care published by the United States Bureau of Labor Statistics. Records for which charge data were incomplete were completed by using single mean imputation, also inflation-adjusted. Descriptive data are presented as mean +/- standard deviation (SD). Results Of 102 CE patients, 35 had available hospital charge data: 15 who underwent successful primary closure at the authors' institution and 20 who presented after previously failed primary closures at referring institutions. The mean +/- SD hospital charges for

primary closure in the success group were \$136,201 +/- \$48,920. These patients then underwent subsequent additional surgeries that accrued charges of \$59,549 +/- \$25,189 in order to achieve continence. Overall, successful primary closures accumulated hospital charges of \$200,366 +/- \$40,071. In comparison, patients referred after prior failure required significantly more hospital admissions and additional charges of \$207,674 +/- \$65,820 were required to achieve continence ($p < 0.001$). Patients who failed primary closure are estimated to accumulate 70% more total health care charges compared to the group following successful primary closure. Conclusion The cost of CE management until urinary continence is high, averaging more than \$200,000 in inpatient hospital charges alone. Initial success is desirable from both an outcomes and economic perspective, as the cost of salvaging a failed primary closure at our institution is similar to the overall costs of a successful closure; this is in addition to the cost of any previous failed closures. Further studies will be required to determine the optimal timing of surgical management in terms of both patient outcomes and financial consequences.

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Institution

(Goldstein, Wildonger) Johns Hopkins University, School of Medicine, Division of Pediatric Surgery, 600 N. Wolfe Street, Baltimore, MD 21287-0005, United States (Inouye, Reddy, Lue, Young, Abdelwahab, Grewal, Stec, Gearhart) Johns Hopkins University, School of Medicine, James Buchanan Brady Urological Institute, Division of Pediatric Urology, 1800 Orleans St., Baltimore, MD 21287, United States

Publisher

W.B. Saunders

Year of Publication

2016

715.

Eccentric circummeatal based flap with limited urethral mobilization: An easy technique for distal hypospadias repair.

Ekinici S., Ciftci A.O., Karnak I., Senocak M.E.

Embase

[Article]

AN: 607625980

Background Hypospadias is a common congenital anomaly. Over 300 techniques have been described for repairing hypospadias. **Objective** Eccentric circummeatal based flap with combined limited urethral mobilization technique (ECMB-LUM) is a simple procedure to repair distal hypospadias with minimal complication rate. This study presents results of this technique, highlighting surgical pitfalls to achieve the best result. **Study design** Medical records of patients with distal hypospadias operated on using the same technique between 1998 and 2011 were reviewed retrospectively. Age at surgery, position of meatus preoperatively and postoperatively, duration of urethral catheterization and hospitalization, early and late complications, previous hypospadias repairs, and secondary surgical interventions were evaluated. In the surgical technique an eccentric circummeatal based flap is outlined. The proximal part of the flap is dissected from the underlying urethra and Buck's fascia. If the flap is not long enough, the distal urethra is mobilized a few millimeters (Figure). The eccentric flap is sutured to the tip of the glans. The glans wings are approximated in the midline. A urethral catheter of 6 Fr or 8 Fr is passed and left in the bulbous urethra or the urinary bladder. Diverged limbs of corpus spongiosum are approximated on the urethra, then, the glans and skin of the penile shaft are sutured. **Results** Of the 171 consecutive patients operated on using the ECMB-LUM technique; 115 had coronal, 47 had subcoronal, and nine had glanular meatus. The mean age at surgery was 4.5 (1-17) years. Patients were hospitalized for 2.2 +/- 0.7 days. Mean duration of urethral catheterization was 2.3 +/- 0.5 days. All but eight patients had ECMB-LUM as primary repair. There were no early complications such as bleeding, hematoma, and wound infection. All patients voided spontaneously after catheter removal. Late complications were meatal stenosis, urethrocutaneous fistula, meatal regression, and glandular dehiscence (Table). These patients were treated using dilatation, fistula repair, meatoplasty, and secondary repair with the same technique, respectively. Eventually all patients had a vertical slit-like meatus on the tip of a natural looking glans. **Discussion** The most commonly used distal hypospadias repair techniques are glanular approximation, meatal advancement and glanuloplasty, Koff, Mathieu, Thiersch-Duplay procedure, tubularized incised plate repairs, and modifications of these techniques. Cosmetic and functional results and complication rates of ECMB-LUM technique are comparable with those of the commonly used techniques. **Complications of ECMB-LUM repair.**

Complication	Number	Percentage
Meatal stenosis	4	(2%)
Fistula	4	(2%)
Meatal regression	4	(2%)
Glanular dehiscence	4	(2%)

 Eccentric circummeatal based flap and minimally mobilized urethra.

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Author NameID

Ekinci, Saniye; ORCID: <http://orcid.org/0000-0002-2961-7069>

Institution

(Ekinci, Ciftci, Karnak, Senocak) Hacettepe University Medical Faculty, Department of Pediatric Surgery, Shhiye, Ankara 06230, Turkey

Publisher

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Year of Publication

2016

716.

Can separation of the scrotal sac in proximal hypospadias reliably predict the need for urethral plate transection?.

Arnaud A., Ferdynus C., Harper L.

Embase

Journal of Pediatric Urology. 12 (2) (pp 121.e1-121.e5), 2016. Date of Publication: 01 Apr 2016. [Article]

AN: 607443520

Introduction One of the main challenges in proximal hypospadias repair is correcting curvature. The best technique to achieve this remains the object of debate. Indeed, some authors believe the urethral plate should be kept and used as often as possible. In some cases, however, even after extensive mobilization and dorsal plication, significant curvature remains and it is necessary to transect the urethral plate. Having a reliable pre-dissection marker of the need for urethral transection would be useful in choosing a technique. We wanted to determine if presence of marked separation of the scrotal sac (SSS), also referred to as bifid scrotum, could reliably predict the need for urethral plate transection. Study design We prospectively enrolled a series of boys with proximal hypospadias. We noted age, degree of hypospadias, meatal position, presence of cryptorchidism, and presence or absence of SSS. During surgery we fully degloved the penile shaft, freeing all ventral tissues, and radically dissected the more proximal bulbar urethra. We then performed an erection test. If there was residual curvature $<30^\circ$ we performed a dorsal plication, if it was $>30^\circ$ we transected the urethral plate. Results Twenty-nine patients were included, of whom 18 presented SSS. The average age was

comparable in both groups, as was type of hypospadias and meatal position. We estimated transection of the urethral plate to be necessary in 15 out of the 18 children with SSS, and 2 out of the 11 children without SSS. The relative risk for requiring urethral plate transection in case of SSS in this series was 4.58. Conclusion Techniques that commit to urethral plate transection are criticized because they preclude using the urethral plate. In our study presence of SSS was predictive for the need to transect the plate. Obviously one can decide to keep the urethral plate at all cost, and mobilize it more than we did, or accept more residual curvature, but in reality our aim was to determine a preoperative marker allowing us to define a patient category. We believe presence of SSS is a marker of severity, and that this "severity" translates into "a less usable urethra". As recent studies caution us about the evolution of the reconstructed native urethra and the possibility that it may not grow as well as the other penile tissues, we believe this extra information could influence the surgeon's decision as to the most appropriate technique for each patient. Results of bivariate analysis. Results

Total	No SSS	SSS	SpAge, years
4	23	44	70.68
Urethral transection	17 (58.6%)	2 (18.2%)	15 (83.3%)
	0.001		

A p-value <0.05 was considered statistically significant.

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Status

Embase

Institution

(Arnaud, Harper) Department of Pediatric Surgery, CHU F Guyon, Bellepierre 97405, Reunion

(Ferdynus) Methodology Unit (USM), CHU F Guyon, Bellepierre, Reunion

Publisher

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2016

717.

Prospective assessment of complications on a daily basis is essential to determine morbidity and mortality in routine pediatric surgery.

Sethi M.V.A., Zimmer J., Ure B., Lacher M.

Embase

Journal of Pediatric Surgery. 51 (4) (pp 630-633), 2016. Date of Publication: 01 Apr 2016.

[Article]

AN: 607215613

Aims of the study We aimed to assess postoperative complications prospectively on a daily basis and hypothesized that this would lead to an increase in the number of detected complications.

Methods Surgical complications were assessed prospectively during a period of 8 months.

Systematic documentation was carried out daily during a team meeting (period S). Data were compared to those of a preceding period of 8-months of nonsystematic assessment (period N) in which complications had been documented in a self-reporting fashion. Complications were classified according to the Clavien-Dindo classification. **Results** A total of 1291 patients (mean age: 6.6 years) were included. During period S complications were determined in 16% of 790 operations compared to 4% of 741 procedures in period N ($p < 0.01$). This difference was owing to an increased detection of minor complications (grade I-III), i.e. wound infection, dysuria after hypospadias repair or postoperative bleeding. In contrast, the incidence of severe complications (grade IV + V) was not significantly different between the time periods (1.3% in period S and 0.8% in period N). Most frequent major complications were cardiopulmonary arrest, enterocolitis, and death. Severe complications accounted for 8% of complications discussed during weekly morbidity and mortality conferences in period S versus 22% in period N ($p < 0.05$). **Conclusion** Our results indicate that a systematic documentation of complications on a daily basis reveals a more realistic picture of the incidence of pediatric surgical complications and should be the method of choice.

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Status

Embase

Author NameID

Sethi, Mohammad Valeed Ahmed; ORCID: <http://orcid.org/0000-0002-3426-2258>

Institution

(Sethi, Zimmer, Ure, Lacher) Center of Pediatric Surgery, Hannover Medical School, Carl-Neuberg-Str. 1, Hannover 30625, Germany (Lacher) Department of Pediatric Surgery, University of Leipzig, Liebigstrasse 20A, Leipzig 04103, Germany

Publisher

W.B. Saunders

Year of Publication

2016

718.

Role of routine cystoscopy and cystography in exstrophy-epispadias complex.

Bar-Yosef Y., Binyamini J., Sofer M., Ben-Chaim J.

Embase

Journal of Pediatric Urology. 12 (2) (pp 117.e1-117.e4), 2016. Date of Publication: 01 Apr 2016.

[Article]

AN: 607188345

Objective Our institutional protocol for the treatment of exstrophy-epispadias complex includes routine endoscopic and cystographic evaluation of the bladder with the child under general anesthesia. The protocols briefly described in the literature include a cystographic evaluation and the measurement of bladder capacity, but there are no reports on concurrent endoscopic findings and the value of examinations in this setting. Our objective is to evaluate the role and necessity of our management protocol by reviewing the findings in our patients' medical charts. **Study design** Cystoscopies are performed in children with exstrophy-epispadias complex during the second year of life and then every 18 months until a capacity of 90 mL has been reached and bladder neck reconstruction, the last stage of modern staged repair, is performed. Patients referred from other institutions are evaluated on presentation. The examinations are performed by means of a pediatric cystoscope with the child under general anesthesia. Cystography is performed under a pressure of 30 cm/H₂O. The contribution of the findings of these periodic evaluations vis-a-vis the surgical findings were studied. **Results** The medical charts of 49 patients who had been surgically treated for exstrophy-epispadias complex in our institution between 2000 and 2014 were reviewed. Thirty patients underwent at least one evaluation. Eighteen underwent serial examinations: four underwent two procedures, eleven underwent three procedures, and three underwent four procedures. The findings in eight cases were significant and they were treated on detection: bladder neck stricture (n = 5), bladder scar bridge (n = 2), and bladder stone (n = 1). Vesicoureteral reflux was present in all 30 patients, and high-grade reflux was present in 10 patients, of whom four had a bladder neck stricture. Fourteen of the 30 patients had a bladder capacity of 90 mL on the first evaluation, as did an additional eight children during later evaluations. Bladder capacity decreased below 90 mL in one child. Seven children did not reach the target capacity (Figure). **Conclusion** Cystoscopic and cystometrographic evaluation of an exstrophic bladder allows early diagnosis of treatable bladder pathologies and monitoring of bladder capacity. It is a valuable tool in the follow-up period preceding bladder neck reconstruction. (A), (B) Good bladder capacity (95 mL), bilateral low-grade VUR, smooth bladder contour. Scheduled for bladder neck reconstruction. (C), (D) Small bladder capacity (55 mL),

unilateral VUR, smooth bladder contour. (E), (F) High-grade bilateral VUR, bladder outlet obstruction, irregular bladder contour. A fibrotic bladder scar bridge was incised.

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Status

Embase

Institution

(Bar-Yosef, Binyamini, Sofer, Ben-Chaim) Pediatric Urology, Dana-Dwek Children's Hospital, Tel-Aviv Medical Center, Sackler Faculty of Medicine, Tel-Aviv University, 6 Weizmann St., Tel-Aviv 64239, Israel

Publisher

Elsevier Ltd

Year of Publication

2016

719.

Assessing caudal block concentrations of bupivacaine with and without the addition of intravenous fentanyl on postoperative outcomes in pediatric patients: A retrospective review.

Karkera M.M., Harrison D.R., Aunspaugh J.P., Martin T.W.

Embase

American Journal of Therapeutics. 23 (3) (pp e792-e798), 2016. Date of Publication: 2016.

[Article]

AN: 53136790

Caudal blocks are a significant and efficacious aspect of pediatric anesthesia, especially in urologic and many general surgery cases. This type of regional anesthesia is common because it has a high success rate and provides between 6 and 8 hours of postoperative pain control. The aim of this study was to determine whether the concentration of bupivacaine or the addition of intravascular (i.v.) fentanyl affected the postanesthesia care unit (PACU) discharge time. A retrospective cohort study comparing the outcomes in pediatric patients who have received varying caudal concentrations with and without the addition of i.v. fentanyl was performed. A total of 849 consecutive patients undergoing hypospadias repairs or circumcisions were reviewed and placed in one of the following 3 groups: 0.125% bupivacaine (group 1), 0.25% bupivacaine (group 2), or one of these concentrations of bupivacaine + i.v. fentanyl intraoperatively (group 3). Total

PACU time for each group was 46.1 minutes (group 1), 48.9 minutes (group 2), and 49.7 minutes (group 3). Our results revealed that there is no statistically significant difference between concentrations of bupivacaine administered in a caudal block with or without i.v. fentanyl with regard to the outcome of PACU duration (P 0.16). Overall, based on the retrospective cohort design, there is no difference in primary and secondary outcomes based on the concentration of bupivacaine, when administered at a volume of 1 mL/kg.

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Embase

Institution

(Karkera, Aunspaugh, Martin) Division of Pediatric Anesthesiology, University of Arkansas for Medical Sciences, Arkansas Children's Hospital, 1 Children's Way, Little Rock, AR 72202, United States (Harrison) Clinical Research Consultant, Little Rock, AR, United States

Publisher

Lippincott Williams and Wilkins (E-mail: kathiest.clai@apta.org)

Year of Publication

2016

720.

Reconstructive surgery for hypospadias: A systematic review of long-term patient satisfaction with cosmetic outcomes.

Adams J., Bracka A.

Embase

Indian Journal of Urology. 32 (2) (pp 93-102), 2016. Date of Publication: April-June 2016.

[Article]

AN: 609640966

Introduction: Research on long-term results of hypospadias has focused on surgical techniques and functional outcomes, and it is only recently that patient satisfaction with appearance and psychosocial outcomes have been considered. The aim of this study was to provide an evidence-based systematic review of adolescent and adult patient perceptions of cosmetic outcomes following childhood surgery for hypospadias.

Method(s): A systematic review was performed in accordance with the PRISMA and PICO guidelines, and studies assessed using the Oxford Centre for Evidence-Based Medicine system. MEDLINE, PsycInfo, EMBASE, and CINAHL databases were searched from 1974 to 2014 for clinical studies containing patient perceptions of appearance, deformity, and social embarrassment following hypospadias surgery.

Result(s): A total of 495 publications were retrieved, of which 28 met the inclusion criteria. Due to study design/outcome measure, heterogeneity data were synthesized narratively. Results indicate (i) patient perceptions of penile size do not differ greatly from the norm; (ii) perceptions of appearance findings are inconsistent, partially due to improving surgical techniques; (iii) patients who are approaching, or have reached, sexual maturity hold more negative perceptions and are more critical about the cosmetic outcomes of surgery than their prepubertal counterparts; (iv) patients report high levels of perceptions of deformity and social embarrassment; and (v) there is a lack of data using validated measurement tools assessing long-term patient perceptions of cosmetic outcomes, particularly with patients who have reached genital maturity.

Conclusion(s): Protocols for clinical postpuberty follow-up and methodologically sound studies, using validated assessment tools, are required for the accurate assessment of cosmetic and psychological outcomes of hypospadias surgery.

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Status

Embase

Institution

(Adams) Department of Applied Health, Faculty of Health, Edge Hill University, Ormskirk, Lancashire L39 4QP, United Kingdom (Bracka) Formerly at the Dudley Group of Hospitals, West Midlands, United Kingdom

Publisher

Medknow Publications (B9, Kanara Business Centre, off Link Road, Ghatkopar (E), Mumbai 400 075, India)

Year of Publication

2016

Evaluation of external genital anomalies and the underlying factors in male newborns.

Pakniyat A., Fallah M.R., Fakour Z., Moloudi F., Khezri S., Masoudi S.

Embase

Iranian Journal of Neonatology. 7 (1) (pp 52-57), 2016. Date of Publication: March 2016.

[Article]

AN: 609360499

Background: External genital anomalies are the most common congenital disorders in male infants with unknown etiology in the majority of cases. According to recent reports, incidence rate of these anomalies is rising in different countries. This study aimed to evaluate the prevalence of external genital anomalies and possible underlying factors in male newborns.

Method(s): This cross-sectional study was conducted on all the male neonates born in Motahari Hospital of Urmia during October 2009-June 2010. Neonatal screening results and demographic data were recorded separately in a questionnaire for each neonate. Data analysis was performed in SPSS V.20 using descriptive statistics (mean, frequency, percentage, and standard deviation) and Chi-square test to evaluate the correlations between quantitative variables.

Result(s): In this study, external genital anomalies were identified in 83 male neonates (8.3%). Moreover, 59 infants (5.9%) had cryptorchidism, 18 (1.8%) had hypospadias and 12 infants (1.2%) had microphallus. Moreover, a significant correlation was observed between the incidence of cryptorchidism and maternal comorbidities, low birth weight, prematurity, maternal gestational hypertension, and diabetes mellitus.

Conclusion(s): According to the results of this study, cryptorchidism is the most prevalent external genital anomaly in male newborns, followed by hypospadias. Occurrence of these anomalies is associated with several risk factors, such as prematurity, low birth weight, maternal diseases (e.g., diabetes and hypertension), and use of medications during pregnancy. In addition, cryptorchidism had a significant correlation with smoking habits, and further studies with larger sample sizes are required in this regard.

Status

Embase

Institution

(Pakniyat) Student Research Committee, Emergency Department, Arak University of Medical Science, Arak, Iran, Islamic Republic of (Fallah) Nephrology and Kidney Transplant Research Center, Urmia University of Medical Sciences, Urmia, Iran, Islamic Republic of

(Fakour) Department of Pediatrics, Urmia University of Medical Sciences, Urmia, Iran, Islamic Republic of

(Moloudi, Khezri, Masoudi) Urmia University of Medical Sciences, Urmia, Iran, Islamic Republic of

Publisher

Mashhad University of Medical Sciences (P.O. Box: 445, Daneshgah Avenue, Mashhad, Iran,
Islamic Republic of)

Year of Publication

2016

722.

Organic food consumption during pregnancy and hypospadias and cryptorchidism at birth: The Norwegian mother and child cohort study (MoBa).

Brantsaeter A.L., Torjusen H., Meltzer H.M., Papadopoulou E., Hoppin J.A., Alexander J.,
Lieblein G., Roos G., Holten J.M., Swartz J., Haugen M.

Embase

Environmental Health Perspectives. 124 (3) (pp 357-364), 2016. Date of Publication: March 2016.

[Article]

AN: 608769098

Background: The etiologies of the male urogenital anomalies hypospadias and cryptorchidism remain unclear. It has been suggested that maternal diet and environmental contaminants may affect the risk of these anomalies via placental or hormonal disturbances.

Objective(s): We examined associations between organic food consumption during pregnancy and prevalence of hypospadias and cryptorchidism at birth.

Method(s): Our study includes 35,107 women participating in the Norwegian Mother and Child Cohort Study (MoBa) who delivered a singleton male infant. Information about use of six groups of organically produced food (vegetables, fruit, bread/cereal, milk/dairy products, eggs, and meat) during pregnancy was collected by a food frequency questionnaire. Women who indicated that they sometimes, often, or mostly consumed organic foods in at least one of the six food groups were classified as organic food consumers in analyses. Hypospadias and cryptorchidism diagnoses were retrieved from the Medical Birth Registry of Norway. We estimated odds ratios (ORs) and 95% confidence intervals (CIs) using multiple logistic regression.

Result(s): Seventy-four male newborns were diagnosed with hypospadias (0.2%), and 151 with cryptorchidism (0.4%). Women who consumed any organic food during pregnancy were less likely to give birth to a boy with hypospadias (OR = 0.42; 95% CI: 0.25, 0.70, based on 21 exposed cases) than women who reported they never or seldom consumed organic food.

Associations with specific organic foods were strongest for vegetable (OR = 0.36; 95% CI: 0.15,

0.85; 10 exposed cases) and milk/dairy (OR = 0.43; 95% CI: 0.17, 1.07; 7 exposed cases) consumption. No substantial association was observed for consumption of organic food and cryptorchidism.

Conclusion(s): Consumption of organically produced foods during pregnancy was associated with a lower prevalence of hypospadias in our study population. These findings were based on small numbers of cases and require replication in other study populations.

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Status

Embase

Institution

(Brantsaeter, Torjusen, Meltzer, Papadopoulou, Haugen) Division of Environmental Medicine, Department of Exposure and Risk Assessment, Norwegian Institute of Public Health, Oslo, Norway (Torjusen, Roos) National Institute for Consumer Research (SIFO), Oslo, Norway (Hoppin) Center for Human Health and the Environment, Department of Biological Sciences, North Carolina State University, Raleigh, NC, United States

(Alexander) Office of the Director-General, Norwegian Institute of Public Health, Oslo, Norway

(Lieblein) Department of Plant Sciences, Norwegian University of Life Sciences, As, Norway

(Holten) Oikos-Organic Norway, Oslo, Norway

(Swartz) Department of Neuroscience Child and Adolescent Psychiatry, Uppsala University, Uppsala, Sweden

(Swartz) Vidarkliniken, Jarna, Sweden

Publisher

Public Health Services, US Dept of Health and Human Services

Year of Publication

2016

723.

Polymorphism of 3' UTR of MAMLD1 gene is also associated with increased risk of isolated hypospadias in Indian children: a preliminary report.

Ratan S.K., Sharma A., Kapoor S., Polipalli S.K., Dubey D., Mishra T.K., Sinha S.K., Agarwal S.K.

Embase

Pediatric Surgery International. 32 (5) (pp 515-524), 2016. Date of Publication: 01 May 2016.

[Article]

AN: 607966456

Objective: To study MAMLD1 gene polymorphisms, serum LH and testosterone levels amongst Indian children with isolated hypospadias (IH) and controls.

Material(s) and Method(s): Screening of the MAMLD1 gene was performed by PCR sequencing method in 100 Indian children aged 0-12 years presenting with IH and 100 controls. LH and testosterone hormone levels were also assessed (categorized in four age-wise groups).

Result(s): IH subjects had significantly higher incidence of MAMLD1 polymorphism as compared to controls (33 vs 15 %, $p = 0.01$). Of various genomic variants identified in this study, the noteworthy novel ones were missense mutation P299A and single nucleotide polymorphism c.2960C>T in 3' UTR of Exon 7. While p 299A was found to cause protein structural instability consequent to amino acid change, eighty percent subjects with c.2960C>T in 3' UTR of Exon 7 (corresponding to newly discovered currently non-validated exon 11) were found to have lower testosterone levels when compared with their age group mean. IH showed statistically higher incidence of c.2960C>T in comparison to controls (22 vs 10 %, p value 0.046) and about 2.5-folds higher risk of this anomaly.

Conclusion(s): Occurrence of MAMDL1 gene polymorphisms, specially of c.2960C>T in 3' UTR of its exon 7 is associated with a higher risk of IH in Indian children, probably by lowering androgenic levels.

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Status

Embase

Institution

(Ratan, Sinha, Agarwal) Department of Pediatric Surgery, Maulana Azad Medical College and Associated LokNayak Hospital, G-8/6, Second Floor, Malviya Nagar, New Delhi 110017, India

(Sharma, Polipalli, Mishra) Department of Biochemistry, Maulana Azad Medical College and Associated LokNayak Hospital, New Delhi, India

(Kapoor) Department of Pediatrics, Maulana Azad Medical College and Associated LokNayak Hospital, New Delhi, India

(Dubey) Department of Biophysics, All India Institute of Medical Sciences, New Delhi, India

Publisher

Springer Verlag (E-mail: service@springer.de)

Year of Publication

2016

724.

Penile appearance after hypospadias correction from a parent's point of view: Comparison of the hypospadias objective penile evaluation score and parents penile perception score.

Haid B., Becker T., Koen M., Berger C., Strasser C., Roesch J., Zniva C., Oswald J.

Embase

Journal of Pediatric Urology. 12 (1) (pp 33.e1-33.e7), 2016. Date of Publication: 01 Feb 2016.

[Article]

AN: 607400668

Introduction As there is only scarce information on the parents' view of the cosmetic outcome after hypospadias surgery we aimed to evaluate whether the results of the hypospadias objective penile evaluation (HOPE) score are transferable to parents satisfaction as measured by the pediatric penile perception score (PPPS). Patients 42 patients after hypospadias correction were included (2 (6.9%) glandular, 20 (68.9%) coronal, 6 (20.6%) penile and 1 (3.4%) scrotal hypospadias, median age 15.0 months). Two surgeons independently assessed HOPE score; the PPPS score as well as 4 questions specifically designed by a psychologist were completed by fathers and mothers. 29 (69.9%) full datasets were available for evaluation. Results Parents' assessment of the cosmetic results was worse than surgeons' assessment (81.13% [PPPS] vs. 92.81% [HOPE] of the respectively possible highest score, $P < 0.0001$). All 58 parents (100%) were convinced that surgery led to a better cosmetic aspect of their sons' genitalia although both, mothers and fathers, perceived the operation as a major encumbrance (fathers 3.62 vs. mothers 3.97 on a scale from 0-6, $P = 0.22$). Conclusion Parents can be encouraged preoperatively that a hypospadias operation, seen from their point of view will be a major amendment to the cosmetic appearance of their sons' genitalia even if the operation itself is perceived as a major psychological burden. In direct comparison of the highest possible score of either tool (HOPE or PPPS), the cosmetic results were judged significantly more optimistic by surgeons as compared to parents using validated tools. HOPE score results therefore may not be transferred uncritically to the parents view on the cosmetic results.

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Status

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Author NameID

Strasser, Christa; ORCID: <http://orcid.org/0000-0002-7735-3491>

Institution

(Haid, Becker, Koen, Berger, Strasser, Roesch, Oswald) Department of Pediatric Urology,
Hospital of the Sisters of Charity, Seilerstatte 4, Linz 4020, Austria (Zniva) Department of
Clinical Psychology, Hospital of the Sisters of Charity, Linz, Austria

Publisher

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2016

725.

Appraisal of adult genitalia after hypospadias repair: Do laypersons mind the difference?.

Ruppen-Greeff N.K., Landolt M.A., Gobet R., Weber D.M.

Embase

Journal of Pediatric Urology. 12 (1) (pp 32.e1-32.e8), 2016. Date of Publication: 01 Feb 2016.

[Article]

AN: 607325512

Introduction Men with corrected hypospadias often suffer from sexual inhibition and fear of being ridiculed by others because of their penile appearance. However, no investigation has thus far been made of the perception of hypospadias-affected surgically repaired genitals by laypersons unacquainted with hypospadias. Therefore, the aim of this study was to find out whether laypersons notice a difference between genitals of men with corrected hypospadias in comparison with circumcised genitals. Furthermore, the most relevant predictors of laypersons' perception of hypospadias-affected genitals were examined. Study design A cross-sectional study was performed in which a questionnaire with 10 standardized photographs of non-erect hypospadias-affected genitals and 10 circumcised genitals was presented to laypersons unacquainted with hypospadias to measure how they rated these genitals. Laypersons were 105 women and 70 men of three different age groups (age ranges 16-20, 25-30, and 40-45 years). Furthermore, laypersons were asked about demographic characteristics, their sexuality and their genital self-perception. Results The results showed that genitals with distal forms of hypospadias were rated similarly to circumcised genitals. In contrast, genitals with more proximal types were perceived as significantly less positive than circumcised genitals. However, the effect size was small. Higher age, being in an intimate relationship, higher socio-economic status, and a higher

sexual interest predicted a better layperson's perception of hypospadias-affected genitals.

Discussion These findings do not support the fear of some men with corrected hypospadias of being ridiculed by others because of their penile appearance. The results indicate that laypersons do not notice a difference between corrected distal types of hypospadias (which represent the majority of hypospadias) and circumcised genitals. Although the findings showed that laypersons perceive more proximal forms of hypospadias less positively than circumcised genitals, the difference does not appear to be clinically relevant as the effect size was small. A major strength of this study is its comprehensive study design. However, the low response rate of hypospadias patients and control individuals for photo documentation and of laypersons who rated these photosets is a limitation of the study. Therefore, generalization from the results must be made carefully.

Conclusions The results are relevant for patient counseling. Knowing that the penile appearance would not trouble laypersons may prevent the development of a negative genital self-perception and feelings of shame. The findings also suggest that hypospadias-affected genitals seem to be rated more positively when laypersons know more about the "normal" variation of penile appearance (e.g. with increasing sexual experience at a higher age).

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Status

Embase

Institution

(Ruppen-Greeff, Gobet, Weber) Division of Pediatric Urology, Department of Pediatric Surgery, University Children's Hospital Zurich, Steinwiesstrasse 75, Zurich CH-8032, Switzerland

(Ruppen-Greeff, Landolt, Gobet, Weber) Children's Research Center, University Children's Hospital Zurich, Zurich, Switzerland

(Landolt) Department of Psychosomatics and Psychiatry, University Children's Hospital Zurich, Zurich, Switzerland

(Landolt) Department of Psychology, University of Zurich, Zurich, Switzerland

Publisher

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2016

Anatomy of the cutaneous surface of the male genitalia, with elaboration upon the inner preputial coronal-shaft junction.

Naimer S.A., Zega K., Silverman W.F.

Embase

Journal of Pediatric Urology. 12 (1) (pp 53.e1-53.e5), 2016. Date of Publication: 01 Feb 2016.

[Article]

AN: 606698679

Introduction Descriptions of the penile prepuce in anatomical and clinical texts either omit details or contain a small, yet potentially serious, error with regard to the manner of its attachment to the penis. **Objective** This study sought to cast light on a ubiquitous but poorly understood and under-appreciated structure, while correcting a long-standing mistake in the medical literature. **Study design** The foreskins of five male stillborn babies were dissected and carefully examined. Tissue from the apposing surfaces of the various regions of the inner and outer prepuce surfaces and the transition zone itself were collected, embedded in paraffin, sectioned, stained, examined and photographed under microscopy. **Results** Contradicting the prevailing descriptions in the literature that the inner prepuce is a single, uniform sheath, this study's observations and histological findings demonstrated that it actually splits into separate laminae that connect distally to the shaft at the base of the corona and proximally with the shaft fascia, respectively (Figure). **Discussion** The penile prepuce is a discrete and deceptively complex part of the male anatomy, yet key details of its interposing surfaces are inaccurately described or entirely omitted in the literature. Understanding the normal anatomy of the prepuce is critically relevant, particularly for urologists and others involved in the performance of circumcision. For example, avoiding potentially catastrophic avulsion of the inner preputial remnant beyond the coronal sulcus during circumcision and accurate assessment of tissue positioning prior to penile reconstruction in cases of hypospadias. **Conclusion** The findings of this study correct a misunderstanding in the anatomy of the prepuce.

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Embase

Institution

(Naimer) Department of Family Medicine, Siaal Research Center for Family Medicine and Primary Care, Faculty of Health Sciences, Ben-Gurion University of the Negev, POB 653, Beer-Sheva 84105, Israel (Naimer) Elon Moreh Family Health Center, Shomron, Israel
(Zega, Silverman) Department of Physiology and Cell Biology, Faculty of Health Sciences, Ben-Gurion University of the Negev, Beer-Sheva, Israel

Publisher

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2016

727.

Newborn exstrophy closure without osteotomy: Is there a role?.

Inouye B.M., Lue K., Abdelwahab M., Di Carlo H.N., Young E.E., Turchi A., Grewal M., Hesh C., Sponseller P.D., Gearhart J.P.

Embase

Journal of Pediatric Urology. 12 (1) (pp 51.e1-51.e4), 2016. Date of Publication: 01 Feb 2016.

[Article]

AN: 606116931

Introduction Recent articles document successful classic bladder exstrophy (CBE) closure without osteotomy. Still, many patients require osteotomy if they have a large bladder template and pubic diastasis, or non-malleable pelvis. **Objective** To understand the indications and outcomes of bladder closure with and without pelvic osteotomy in patients younger than 1 month of age. **Methods** An institutional database of 1217 exstrophy-epispadias patients was reviewed for CBE patients closed at the authors' institution within the first month of life. Patient demographics, closure history, pubic diastasis distance, bladder capacity, and outcomes were recorded and compared using chi-square tests between osteotomy and non-osteotomy patients. Failure was defined as bladder dehiscence, prolapse, vesicocutaneous fistula, or bladder outlet obstruction requiring reoperation. Bladder capacity >100 mL was deemed sufficient for bladder neck reconstruction (BNR). **Results** One hundred CBE patients were included for analysis: 38 closed with osteotomy (26 male, 12 female), and 62 closed without osteotomy (42 male, 20 female). There were four failed closures in the osteotomy group (2 dehiscence, 2 prolapse) and four failed closures in the non-osteotomy group (2 dehiscence, 2 prolapse). This corresponded to statistically equivalent rates of failure between the osteotomy and non-osteotomy groups (10.5% vs. 6.5%, $p = 0.466$). There was no statistically significant difference between the groups' ability to achieve bladder capacity sufficient for BNR (82% vs. 71%, $p = 0.234$). **Discussion** A successful primary bladder closure, regardless of the use of osteotomy, has been shown to be the single most important predictor of eventual continence. Because of the complexity of exstrophy manifestations, a multidisciplinary team approach is of the utmost importance. Based on our institutional experience, closure without osteotomy is considered when patients are <72 h of life,

have a pubic diastasis <4 cm, malleable pelvis, and pubic apposition without difficulty. Rates of successful closure and attaining sufficient capacity for BNR were both statistically equivalent across groups. This retrospective study is limited by selection bias and the significant difference in follow-up time between groups. Nevertheless, as a high-volume exstrophy center this study draws from one of the largest cohorts available. Conclusions Regardless of the type of closure undertaken, there clearly is a role for newborn CBE closure without pelvic osteotomy in patients considered suitable for closure by both the pediatric urologist and orthopedic consultant. However, if there is any doubt concerning pubic diastasis width, pelvic malleability, or ease of pubic apposition, an osteotomy is highly recommended.

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Status

Embase

Author NameID

Inouye, Brian M.; ORCID: <http://orcid.org/0000-0001-9584-2435> Abdelwahab, Mahmoud;

ORCID: <http://orcid.org/0000-0003-1890-6647>

Young, Ezekiel E.; ORCID: <http://orcid.org/0000-0003-2988-1561>

Institution

(Inouye, Lue, Abdelwahab, Di Carlo, Young, Tourchi, Grewal, Hesh, Gearhart) Johns Hopkins University School of Medicine, James Buchanan Brady Urological Institute, Division of Pediatric Urology, Charlotte Bloomberg Children's Hospital, 1800 Orleans St., Baltimore, MD 21287, United States (Sponseller) Johns Hopkins University School of Medicine, Division of Pediatric Orthopaedics, Charlotte Bloomberg Children's Hospital, Baltimore, MD, United States

Publisher

Elsevier Ltd

Year of Publication

2016

728.

Management strategies for idiopathic urethritis.

Henderson L., Farrelly P., Dickson A.P., Goyal A.

Embase

Journal of Pediatric Urology. 12 (1) (pp 35.e1-35.e5), 2016. Date of Publication: 01 Feb 2016.

[Article]

AN: 605697217

Introduction Williams and Mikhael (1971) described idiopathic urethritis (IU) as a self-limiting condition that affects boys aged 5-15 years, with symptoms of urethrorrhagia, dysuria and haematuria. However, a proportion of boys will remain symptomatic for several years, and may develop urethral stricture (Poch et al., 2007; Palagiri et al., 2003). There is no universally effective treatment for IU, although various strategies have been employed. Objective To review the presentation and long-term outcomes of boys with IU, and present the efficacy of management strategies that have been utilised. Study design A retrospective review was performed of all boys with IU. It was based on clinical and cystoscopic findings for presentation, medical history, management and clinical progress. Results Fifty-four boys were included, with a median age of 11 years (range 5-15 years) at presentation. The median duration of symptoms was 18 months (range 2-132 months). The median follow-up was 18.5 months (range 1-120 months). Seven (13.0%) boys had early urethral stricture at initial cystourethroscopy, and one (1.9%) developed stricture during follow-up. Thirty-six boys (66.7%) had previous circumcision and four (7.4%) had meatal stenosis. Eight (14.8%) had previous hypospadias repair. Conclusion Whilst 50% of boys with IU do not require any specific treatment, those with severe/unremitting symptoms may benefit from a trial of urethral steroids or short-term urethral catheterisation. The mechanisms of benefit from these modalities are unclear and they require further evaluation.

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Status

Embase

Institution

(Henderson, Farrelly, Dickson, Goyal) Department of Paediatric Urology, Royal Manchester Children's Hospital, Manchester M13 9WL, United Kingdom

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2016

Proximal hypospadias: A persistent challenge. Single institution outcome analysis of three surgical techniques over a 10-year period.

Pippi Salle J.L., Sayed S., Salle A., Bagli D., Farhat W., Koyle M., Lorenzo A.J.

Embase

Journal of Pediatric Urology. 12 (1) (pp 28.e1-28.e7), 2016. Date of Publication: 01 Feb 2016.

[Article]

AN: 605632473

Introduction The optimal treatment of proximal hypospadias remains controversial. Several techniques have been described, but the best approach remains unsettled. **Objective** To evaluate and compare the complication rates of proximal hypospadias with and without ventral curvature (VC), according to three different surgical techniques: tubularized incised plate (TIP) urethroplasty, dorsal inlay graft TIP (DIG), and staged preputial repair (SR). It was hypothesized that SR performs better than TIP and DIG for proximal hypospadias. **Methods** Single-center, retrospective chart review of all patients with primary proximal hypospadias reconstructed between 2003 and 2013. The DIG was selectively employed in cases with narrow urethral plate (UP) and deficient spongiosum. Extensive urethral plate (UP) mobilization (UPM), dorsal plication (DP) and/or deep transverse incisions of tunica albuginea (DTITA) were selectively performed when attempting to spare transecting the UP. Division of UP and SR was favored in cases with severe VC (>50degree), which was often concurrently managed with DTITA if intrinsic curvature was present. For SR, tubularization of the graft was performed 6 months later. **Results** A total of 140 patients were included. Tubularized incised plate (TIP), DIG, and SR techniques were performed in 57, 23, and 60 patients, respectively. The TIP and DIG techniques achieved similar success rates, although DIG was performed in cases of narrow and spongiosum-deficient plates. Reoperation rates with TIP and DIG techniques was 52.6% and 52.1% (NS). Urethro-cutaneous fistulas were seen in 31.5% and 13% of TIP and DIG techniques, respectively. Staged repair accomplished better results than both TIP and DIG techniques, despite being performed in the most unfavorable cases (reoperation rate 28%). After technical modifications, the DIG technique achieved similar outcomes of SR. **Conclusions** Proximal hypospadias remains challenging, regardless of the technique utilized for its repair. Urethro-cutaneous fistulas were more commonly seen after long TIP repairs. Approximately half of the patients undergoing long TIP and DIG procedures needed re-intervention, although the percentage decreased significantly with late modifications in the DIG group. Recurrence of VC after TIP and DIG techniques seemed to be a significant and under-reported complication. Staged repairs, despite being performed for the most severe cases, resulted in overall better outcomes.

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Status

Embase

Institution

(Pippi Salle) Division of Urology, Sidra Medical and Research Center, PO Box 26999, Doha, Qatar (Sayed, Salle, Bagli, Farhat, Koyle, Lorenzo) Division of Urology, Hospital for Sick Children, Department of Surgery, University of Toronto, Toronto, Canada

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730.

Tubularized incised plate urethroplasty for distal penile hypospadias repair; our experience in prince Rashid bin Alhassan Hospital, Amman, Jordan.

Dajaa M., Alibrahim A., Khasawneh G., Gora A.A., Al-Mefleh W.

Embase

Rawal Medical Journal. 41 (1) (pp 43-45), 2016. Date of Publication: 2016.

[Article]

AN: 608428475

Objective: To report our success rate, complications and cosmetic results with distal penile hypospadias treated by tabularized incised plate urethroplasty. Methodology: This retrospective study was done at Prince Rashid bin Alhassan Hospital from June 2011 to April 2013 and included 83 patients with distal penile hypospadias who underwent tabularized incised plate urethroplasty for the first time. All patients were followed for up to 24 months after surgery.

Result(s): Patients age ranged from 2-8 years; mean age was 4 years. 69 (83%) patients had excellent functional and cosmetic results without any complications. Complications were seen in 14 patients. Urethrocutaneous fistula was seen in 8 (10%) patients, meatal stenosis in 2 (2.4%), complete disruption of the glans in 1 (1.2%) and penile rotation was seen in 3 (3.6%) patients.

Conclusion(s): Tabularized incised plate urethroplasty for distal penile hypospadias repair resulted in excellent functional neourethra, excellent glans and meatus shape and low complications rate.

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Status

Embase

Institution

(Dajaa, Alibrahim, Khasawneh, Gora, Al-Mefleh) Department of Pediatric Surgery, Jordanian Royal Medical Services, Amman, Jordan

Publisher

Pakistan Medical Association

Year of Publication

2016

731.

Bisphenol A and other phenols in human placenta from children with cryptorchidism or hypospadias.

Fernandez M.F., Arrebola J.P., Jimenez-Diaz I., Saenz J.M., Molina-Molina J.M., Ballesteros O., Kortenkamp A., Olea N.

Embase

Reproductive Toxicology. 59 (pp 89-95), 2016. Date of Publication: January 01, 2016.

[Article]

AN: 608149157

Embryo-foetal exposure to low doses of endocrine disrupting chemicals (EDCs) has been related to reproductive tract diseases in experimental animals but not convincingly in human populations. The aim of this case-control study was to explore the relationship between exposure to non-persistent EDCs during pregnancy and male genital development. Exposure to bisphenol-A (BPA), benzophenones (BPs) [BP-1, BP-2, BP-3, BP-6, BP-8 and 4-hydroxybenzophenone (4-OH-BP),] and parabens (PBs) [methyl-, ethyl-, propyl- and butyl-PB] was analyzed by means of ultra-high performance liquid chromatography-tandem mass spectrometry in placenta samples from a subsample of 28 cases and 51 healthy controls nested in a cohort of newborns recruited between 2000 and 2002. The multivariable regression analyses indicated a statistically significant association between exposure to BPA and propyl-PB and the risk of malformations [adjusted odd ratio (95% CIs) in the third tertile of exposure: 7.2 (1.5-35.5) and 6.4 (1.2-35.5) for BPA and propyl-PB, respectively].

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Institution

(Fernandez, Arrebola, Jimenez-Diaz, Saenz, Molina-Molina, Olea) Instituto de Investigacion Biosanitaria (ibs.GRANADA), University of Granada, San Cecilio University Hospital, Granada 18071, Spain (Fernandez, Arrebola, Molina-Molina, Olea) CIBER en Epidemiologia y Salud Publica (CIBERESP), Madrid, Spain

(Jimenez-Diaz, Ballesteros) Research Group of Analytical Chemistry and Life Sciences, Department of Analytical Chemistry, University of Granada, Granada, Spain

(Kortenkamp) Institute of the Environment, Health and Societies, Brunel University, Kingston Lane, Uxbridge UB8 3PH, United Kingdom

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Year of Publication

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732.

Efficacy of dexamethasone on postoperative analgesia in children undergoing hypospadias repair.

Shirazi M., Mahmoudi H., Nasihatkon B., Ghaffaripour S., Eslahi A.

Embase

Pakistan Journal of Medical Sciences. 32 (1) (pp 125-129), 2016. Date of Publication: 2016.

[Article]

AN: 608040851

Background and Objective: Management of post operative pain in children undergoing hypospadias repair, accounts for optimized surgery outcomes and improved patients' satisfaction. Thus, various studies have widely investigated the best approaches for the pain management. In this study our aim was to determine the effect of dexamethasone in combination with penile nerve block on the postoperative pain and complications in the children undergoing hypospadias surgery.

Method(s): In this randomized double-blind placebo controlled trial, after obtaining informed consent from parents or legal guardians, 42 children undergoing surgical treatment of hypospadias were randomized in two groups to receive either IV dexamethasone 0.5 mg/kg (n=23) or placebo (normal saline) (n=19) during the operation. Penile block was performed in

both groups using Bupivacaine 0.5% (1mg/kg) at the end of the procedure. By the end of the operation, FLACC (Face, Leg, Activity, Cry, Consolability) pain score was assessed as the primary outcome of the study. Secondary outcomes includes timing and episodes of rescue medication consumption, post operative nausea \vomiting and bleeding. All the outcomes were assessed in the recovery room and after 2, 6, 12, and 24 hours.

Result(s): The median of FLACC pain scores at the recovery room and 2, 6, 12, and 24 hours post operation was 2, 1, 1, 1, and 2 for the dexamethasone group and 8, 8, 7, 7, and 8 for the placebo group respectively. This were significantly different ($P<0.000$). The median time of first rescue medication consumption was 8 hours post operation for the dexamethasone group and three hours for the placebo group which was significantly different ($z= 4.57$, $p<0.000$). The maximum episode of post operative rescue medication consumption in dexamethasone group was 4 episodes in only one patient and the minimum was one episode in 11 patients. In comparison numbers in placebo group were five episodes in seven patients and three episodes in four patients. The result indicated that there was statistically significant difference between two groups in terms of episodes of rescue medication consumption ($\text{Chi}^2= 31.4$, $p<0.000$).

Conclusion(s): Single dose of intravenous dexamethasone (0.5 mg/kg) in combination with penile block decreased the post operative pain measures, and total post operative analgesic requirement. It also increased the onset of the first analgesic requirement compared to penile block alone.

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Embase

Institution

(Shirazi, Nasihatkon, Eslahi) Department of Urology, Shiraz University of Medical Sciences, Shiraz, Iran, Islamic Republic of (Mahmoudi, Ghaffaripour) Department of Anesthesiology, Anesthesiology and Critical Care Research Center, Shiraz University of Medical Sciences, Shiraz, Iran, Islamic Republic of

Publisher

Professional Medical Publications (Raja Ghazanfar Ali Road, Saddar, Karachi, Pakistan)

Year of Publication

2016

How to improve outcome in surgery for proximal hypospadias?.

Qureshi A.H., Zaidi S.Z.

Embase

Journal of the Pakistan Medical Association. 66 (2) (pp 207-209), 2016. Date of Publication: February 2016.

[Article]

AN: 607824990

Objective: To evaluate the role of subdartos fascial tissue as watertight layer in improving outcome for 2-stage proximal hypospadias surgery.

Method(s): The experimental study was conducted at the Department of Urology, Indus Hospital, Karachi, and comprised an audit of patients with proximal hypospadias who underwent surgery from July 1, 2007, to December 31, 2011. The initial two-stage repair of proximal hypospadias led to a high rate of urethrocutaneous fistula formation (Group A), and, thus, a modification was introduced and subdartos facial double layer was applied over the urethral suture line (Group B). The results were compared regarding age, type of hypospadias, graft failure and urethrocutaneous fistula in these patients.

Result(s): There were 27 patients in Group A and 16(59.3%) of them ended up having urethrocutaneous fistula. Group B had 25 patients and only 2(8%) had fistula formation.

Conclusion(s): The application of dartos facial flap waterproofing layer reduced fistula rate.

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Status

Embase

Institution

(Qureshi, Zaidi) Department of Urology, Indus Hospital Karachi, Pakistan

Publisher

Pakistan Medical Association

Year of Publication

2016

Perfluorooctane sulfonate concentrations in amniotic fluid, biomarkers of fetal leydig cell function, and cryptorchidism and hypospadias in Danish boys (1980-1996).

Toft G., Jonsson B.A., Bonde J.P., Norgaard-Pedersen B., Hougaard D.M., Cohen A., Lindh C.H., Ivell R., Anand-Ive R., Lindhard M.S.

Embase

Environmental Health Perspectives. 124 (1) (pp 151-156), 2016. Date of Publication: January 2016.

[Article]

AN: 607485772

Background: Exposure to perfluorooctane sulfonate (PFOS) may potentially disturb fetal Leydig cell hormone production and male genital development.

Objective(s): We aimed to study the associations between levels of amniotic fluid PFOS, fetal steroid hormone, and insulin-like factor 3 (INSL3) and the prevalence of cryptorchidism and hypospadias.

Method(s): Using the Danish National Patient Registry, we selected 270 cryptorchidism cases, 75 hypospadias cases, and 300 controls with stored maternal amniotic fluid samples available in a Danish pregnancy-screening biobank (1980-1996). We used mass spectrometry to measure PFOS in amniotic fluid from 645 persons and steroid hormones in samples from 545 persons. INSL3 was measured by immunoassay from 475 persons. Associations between PFOS concentration in amniotic fluid, hormone levels, and genital malformations were assessed by confounder-adjusted linear and logistic regression.

Result(s): The highest tertile of PFOS exposure (> 1.4 ng/mL) in amniotic fluid was associated with a 40% (95% CI: -69, -11%) lower INSL3 level and an 18% (95% CI: 7, 29%) higher testosterone level compared with the lowest tertile (< 0.8 ng/mL). Amniotic fluid PFOS concentration was not associated with cryptorchidism or hypospadias.

Conclusion(s): Environmental PFOS exposure was associated with steroid hormone and INSL3 concentrations in amniotic fluid, but was not associated with cryptorchidism or hypospadias in our study population. Additional studies are needed to determine whether associations with fetal hormone levels may have long-term implications for reproductive health.

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Status

Embase

Institution

(Toft) Department of Occupational Medicine, Aarhus University Hospital, Aarhus, Denmark

(Toft) Department of Clinical Epidemiology, Aarhus University Hospital, Aarhus, Denmark

(Jonsson, Lindh) Division of Occupational and Environmental Medicine, Lund University, Lund, Sweden

(Bonde) Department of Occupational and Environmental Medicine, Copenhagen University Hospital, Bispebjerg, Copenhagen, Denmark

(Norgaard-Pedersen, Hougaard, Cohen) Danish Center for Neonatal Screening, Department of Clinical Biochemistry and Immunology, Statens Serum Institute, Copenhagen, Denmark

(Ivell, Anand-Ive) School of Biosciences, University of Nottingham, Nottingham, United Kingdom

(Lindhard) Department of Pediatrics, Regional Hospital of Randers, Randers, Denmark

(Lindhard) Perinatal Epidemiology Research Unit, Department of Pediatrics, Aarhus University Hospital, Skejby, Denmark

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Public Health Services, US Dept of Health and Human Services

Year of Publication

2016

735.

A novel technique for repair of mid-penile hypospadias using a preputial skin flap: results of 110 patients.

Elmoghazy H., Hussein M.M., Mohamed E., Badawy A., Alsagheer G., Abd Elhamed A.M.

Embase

International Urology and Nephrology. 48 (12) (pp 1943-1949), 2016. Date of Publication: 01 Dec 2016.

[Article]

AN: 612179065

Background: Several techniques have been used to repair mid-penile hypospadias; however, high failure rates and major complications have been reported. In this study, we describe a novel technique using a well-vascularized flap of the inner and outer preputial skin.

Method(s): A total of 110 male children with hypospadias underwent repair by our technique between 2008 and 2015. The inclusion criteria were children with mid-penile or slightly more proximal hypospadias, with or without ventral chordae, and an intact prepuce of the cobra eyes variety. Recurrent cases, patients with other preputial types, and circumcised children were excluded from this study. The prepared flap was sutured in its natural longitudinal orientation to

the created urethral plate strip to form a neo-urethra over a urethral catheter. Outcome measures included surgical success without the formation of a urethra-cutaneous fistula, no ischaemia of the flaps, glans dehiscence or infection and functional outcome and cosmetic appearance.

Result(s): The median follow-up duration was 3.3 years. There were 63 cases of mid-penile hypospadias (57.3 %), and in 47 cases (42.7 %), the meatus was slightly more proximal. The age of the patients ranged from 1.1 to 8.0 years, with a mean age of 4.6 +/- 1.2 years. Surgery was successful in 106 (96.4 %) cases. Minor complications occurred in 11 patients (10 %) and included oedema of glans in ten patients and bluish discoloration on the ventral aspect of the glans close to the suture line in three patients. All patients improved within 2 weeks after surgery. Long-term follow-up revealed a properly functioning urethra with a forward, projectile, single, compact, and rifled urinary stream of adequate calibre and cosmetically acceptable repair. No cases of meatal retraction, meatal stenosis, urethral stricture, or acquired urethral diverticulum occurred.

Discussion(s): Our technique is different from the split prepuce in situ technique. We create a narrow strip of the urethral plate that facilitates glanular closure, and we use the inner and adjacent outer skin in a vertical manner to preserve excess skin for penile coverage. Prepuce is split at midline to preserve more preputial skin with favourable dartos tissue for penile skin coverage. The glans is closed using a stitch-by-stitch method that has not been described previously.

Conclusion(s): This study presents a novel technique for mid-penile hypospadias repair using a preputial skin flap with excellent results in terms of short- and long-term outcomes.

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Status

Embase

Institution

(Elmoghazy, Hussein, Mohamed, Badawy, Abd Elhamed) Department of Urology, Sohag University Hospital, Alshark District, Sohag, Sohag Governorate, Egypt (Alsagheer) Department of Urology, South Valley University Hospital, Qena, Egypt

Publisher

Springer Netherlands

Year of Publication

2016

736.

Congenital renal anomalies in cloacal exstrophy: Is there a difference?.

Suson K.D., Inouye B., Carl A., Gearhart J.P.

Embase

Journal of Pediatric Urology. 12 (4) (pp 207.e1-207.e5), 2016. Date of Publication: 01 Aug 2016.

[Article]

AN: 611163458

Introduction Cloacal exstrophy (CE) is the most severe manifestation of the epispadias-exstrophy spectrum. Previous studies have indicated an increased rate of renal anomalies in children with classic bladder exstrophy (CBE). Given the increased severity of the CE defect, it was hypothesized that there would be an even greater incidence among these children. **Objective** The primary objective was to characterize renal anatomy in CE patients. Two secondary objectives were to compare these renal anatomic findings in male and female patients, and female patients with and without Mullerian anomalies. **Study design** An Institutional Review Board-approved retrospective review of 75 patients from an institutional exstrophy database. Data points included: age at analysis, sex, and renal and Mullerian anatomy. Abnormal renal anatomy was defined as a solitary kidney, malrotation, renal ectopia, congenital cysts, duplication, and/or proven obstruction. Abnormal Mullerian anatomy was defined as uterine or vaginal duplication, obstruction, and/or absence. **Results** The Summary Table presents demographic data and renal anomalies. Males were more likely to have renal anomalies. Mullerian anomalies were present in 65.7% of female patients. Girls with abnormal Mullerian anatomy were 10 times more likely to have renal anomalies than those with normal Mullerian anatomy (95% CI 1.1-91.4, $P = 0.027$). **Discussion** Patients with CE had a much higher rate of renal anomalies than that reported for CBE. Males and females with Mullerian anomalies were at greater risk than females with normal uterine structures. Mesonephric and Mullerian duct interaction is required for uterine structures to develop normally. It has been proposed that women with both Mullerian and renal anomalies be classified separately from other uterine malformations on an embryonic basis. In these patients, an absent or dysfunctional mesonephric duct has been implicated as potentially causal. This provided an embryonic explanation for uterine anomalies in female CE patients. There were also clinical implications. Women with renal agenesis and uterine anomalies were more likely to have endometriosis than those with isolated uterine anomalies, but were also more likely to have successful pregnancies. Males may have had an analogous condition with renal agenesis and seminal vesicle cysts. Future research into long-term kidney function in this population, uterine function, and possible male sexual duct malformation is warranted. **Conclusion** Congenital renal

anomalies occurred frequently in children with CE. They were more common in boys than in girls. Girls with abnormal Mullerian anatomy were more likely to have anomalous renal development. Mesonephric duct dysfunction may be embryologically responsible for both renal and Mullerian maldevelopment. [Table presented]

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Status

Embase

Institution

(Suson) Children's Hospital of Michigan, Pediatric Urology, 3901 Beaubien, Detroit, MI 48201, United States (Inouye, Carl, Gearhart) Johns Hopkins University School of Medicine, Jeffs Division of Pediatric Urology, Brady Urological Institute, 1800 Orleans Street Suite 7302, Baltimore, MD 21287, United States

Publisher

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Year of Publication

2016

737.

Testicular prostheses in children: Is earlier better?.

Peycelon M., Rossignol G., Muller C.O., Carricaburu E., Philippe-Chomette P., Paye-Jaouen A., El Ghoneimi A.

Embase

Journal of Pediatric Urology. 12 (4) (pp 237.e1-237.e6), 2016. Date of Publication: 01 Aug 2016.

[Article]

AN: 610626344

Introduction The absence of a testis occurs for various reasons in children, but testicular prosthesis implantation in children is uncommon. The optimal time for prosthesis placement is still unclear, and its complication rate has been poorly studied in children. Objective The aim of this study was to determine the risk factors of complications in cases of testicular prosthesis implantation in children. Study design A monocentric, retrospective review was performed of

children implanted with a testicular prosthesis between 2008 and 2014. All implantations were performed through an inguinal incision with a standardized procedure. Children were divided into two groups depending on the interval after orchiectomy: (A) early implantation (delay between surgeries <1 year); and (B) delayed surgeries (delay ≥1 year). Statistical analysis was performed with Student and Fisher tests. Results Twenty-six patients (A, 15; B, 11) had a total of 38 testicular prostheses placements. Mean follow-up was 36.2 months. First surgery was performed at the mean age of 11.8 years (range 0-17.9) (A, 14.1; B, 8.1; $P = 0.01$) and testicular prosthesis implantation at the mean age of 14.7 years (range 9-18) (A, 14.3; B, 14.6) with a mean delay of 36.1 months (A, 1.3; B, 80.3). Indications were mainly spermatic cord torsion (27%), bilateral anorchia (27%), and testicular atrophy after cryptorchidism surgery (19.2%). Complications (10.5%) included two cases of extrusion, one infection and one migration. Patient 1 had a history of acute lymphoblastic leukemia with testicle relapse 2 years after induction therapy. High-dose chemotherapy, total body irradiation and bilateral orchiectomies were performed, and bilateral prostheses were implanted 12 years after the end of chemotherapy. Complications happened 85 days after surgery. Patient 2 was followed-up for a proximal hypospadias. The tunica vaginalis flap, which was used during a redo urethroplasty, lead to testicular atrophy. Thirteen years after the last penile surgery, a testicular prosthesis was placed through an inguinal incision, and extrusion occurred 203 days after surgery. Bacterial cultures of the prostheses were sterile and histological review showed no sign of granuloma or graft rejection. The complication rate was significantly higher if the delay between the two surgeries exceeded 1 year ($P = 0.01$). Indications of orchiectomy, prior scrotal incision, and prosthesis size were not risk factors. Conclusions Testicular prosthesis implantation was relatively safe in a pediatric cohort. The complication rate was significantly higher if the delay between the orchiectomy and the prosthetic placement exceeded 1 year. These results suggest that reducing the delay between orchiectomy and prosthesis implantation may lead to fewer complications.

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Status

Embase

Institution

(Peycelon, Rossignol, Muller, Carricaburu, Philippe-Chomette, Paye-Jaouen, El Ghoneimi)

Department of Pediatric Surgery and Urology, Robert-Debre University Hospital, Assistance-Publique Hopitaux de Paris, Universite Paris Diderot, Sorbonne Paris Cite, Paris, France

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Year of Publication

2016

738.

Hypospadias anatomy: Elastosonographic evaluation of the normal and hypospadiac penis.

Camoglio F.S., Bruno C., Zambaldo S., Zampieri N.

Embase

Journal of Pediatric Urology. 12 (4) (pp 199.e1-199.e5), 2016. Date of Publication: 01 Aug 2016.

[Article]

AN: 608829466

Background Hypospadias is one of the most common congenital anomalies in childhood. The aim of this study is to apply elastosonography on normal and hypospadiac penis to verify the structural differences in tissues composition and stiffness. Materials and methods We analyzed medical chart of patients treated at our Institution for hypospadias during the period December 2005 and December 2014 (group 1). Other two groups were enrolled for this study: group 2- patients with hypospadias waiting for surgery and group 3-patients without hypospadias. Inclusion and exclusion criteria were created; all patients underwent penile ultrasound and elastosonography. Elastographic index of elasticity was defined as soft, medium-hard or hard. We assigned the value 1 to soft tissue, 2 and 3 to medium-hard and hard respectively. Results During the study period 294 patients were treated for hypospadias. After reviewing medical chart 115 patients were considered for analysis (group 1). 22 patients were enrolled in group 2 and 38 patients were enrolled in group 3. Group 1: 7 proximal hypospadias, 29 penile hypospadias, 79 distal hypospadias. Patients with hypospadias had malformation also at corpus spongiosum and cavernosum respect to controls. Elastography showed a corpus spongiosum stiffness defined as medium-hard or hard in all cases of the pathologic group and soft in all the subjects of the control group ($p < 0.05$). Conclusions Elastosonography showed how the hypospadias anatomy is deeply altered, even in an anatomical area far from meatal abnormality: corpus spongiosum in hypospadiac penis seems to be globally stiffer and less elastic and cavernous corpora are less developed.[Figure presented]

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Status

Embase

Institution

(Camoglio, Zambaldo, Zampieri) Department of Surgery, Pediatric Surgical Unit, Policlinico G.B.

Rossi, University of Verona, Verona, Italy (Bruno) Department of Radiology, Policlinico G.B.

Rossi, University of Verona, Verona, Italy

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Year of Publication

2016

739.

Long-term outcome after urinary diversion using the ileocecal segment in children and adolescents: Complications of the efferent segment.

Deuker M., Roos F.C., Grossmann A., Fae P., Thuroff J.W., Stein R.

Embase

Journal of Pediatric Urology. 12 (4) (pp 247.e1-247.e7), 2016. Date of Publication: 01 Aug 2016.

[Article]

AN: 610632380

Introduction Long-term outcomes are of special concern in children after urinary diversion. In a single institution study we evaluated retrospectively the long-term outcomes of urinary diversion in children, in whom the ileocecal segment had been used, in respect to complications of the efferent segment. **Methods** The Mainz pouch was used in 107 children for continent urinary diversion. Indications were neurogenic bladder (53%, 57/107), exstrophy-epispadias complex (25%, 27/107), malignancy (13%, 14/107), and others (9%, 9/107). Continent cutaneous diversion was performed in 95 patients, and 12 patients received bladder augmentation/substitution with a continent cutaneous stoma. As efferent segment, we used the in situ submucosally embedded appendix in 55/107, and an intussuscepted ileal nipple valve in 45/107 patients, other in seven patients. Complication rates and degree of satisfaction with urinary diversion were evaluated by assessing medical records and using follow-up questionnaires. **Results and discussion** The median follow-up time was 15.9 years (0.4-27.6 years). There was a significant difference between the stenosis rates of appendical stoma and the intussuscepted ileal nipple valve. In general, 38% of patients (41/107) with a continent cutaneous stoma developed a stoma stenosis

at the skin level, with 41% (17/41) of those having recurrent stenoses (2-6x). Of the patients with an appendix stoma, 49% (27/55) developed a stenosis, compared with 20% (9/45) of those with an intussuscepted ileal nipple valve ($p < 0.001$). In an attempt to reduce the incidence of stenosis, we implemented use of the ACE stopper during the last 2 years, but whether this is successful is subject to further investigations. For surgical treatment of stoma stenosis, reanastomosis of the efferent segment to the skin ($n = 37$, re-stenosis rate 37%, [14/37]) was superior to endoscopic scar incision ($n = 44$, re-stenosis rate 59% [26/44], OR 2.4). The overall continence rate was 85% (91/107). At the latest follow-up, 89% (49/55) of patients with an appendix stoma and 82% (37/45) of those with an intussuscepted ileal nipple valve were completely continent. Conclusion The in situ submucosally embedded appendix and the intussuscepted ileal nipple valve are reliable continence mechanisms in the long-term median follow-up of nearly 16 years. The stomal stenosis rate is approximately double for the submucosal appendix, likely because of its smaller diameter in comparison with the ileal valve. Although we encountered a high overall rate of complications, the satisfaction rate with the diversion was high (99% [88/89] satisfied or very satisfied patients).

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Status

Embase

Institution

(Deuker, Roos, Thuroff) Johannes Gutenberg University, School of Medicine, Department of

Urology, Mainz, Germany (Grosman, Fae) Johannes Gutenberg University, School of

Medicine, Division of Pediatric Urology, Mainz, Germany

(Stein) University Medical Center Mannheim, Department of Pediatric and Adolescent Urology,

Germany

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2016

Long-term functional outcomes after penoscrotal hypospadias repair: A retrospective comparative study of proximal TIP, Onlay, and Duckett.

Hueber P.-A., Salgado Diaz M., Chaussy Y., Franc-Guimond J., Barrieras D., Houle A.-M.

Embase

Journal of Pediatric Urology. 12 (4) (pp 198.e1-198.e6), 2016. Date of Publication: 01 Aug 2016.

[Article]

AN: 613344722

Introduction A variety of techniques are available for proximal hypospadias repair. Onlay, proximal tubularized incised plate (TIP), and Duckett are among the popular choices because they can be performed as a one-step procedure. However, the decision to select a procedure often comes down to the surgeon's preference rather than that supported by evidence-based data. In particular, there is a paucity of literature on the long-term urinary outcomes after proximal hypospadias repair. **Objective** The aim of this study was to evaluate the evolution of long-term uroflowmetry parameters after proximal hypospadias surgery over a long-term follow-up including the adolescent period. **Study design** Files from patients who underwent primary proximal hypospadias repair at our institution between 1997 and 2001 were reviewed. Only patients with documented serial postoperative uroflowmetry profiles at follow-up visits were included.

Comparison between surgeries (pTIP vs. Onlay vs. Duckett) was performed according to the following postoperative time interval endpoints: 0-1 years, 1-2, >2-4, >4-6, >6-10, >10-12, and >12 years. Maximal urinary flow rate (Qmax) in relation to voiding volume (VV) adjusted for age or body surface area (BSA) were also evaluated in comparison to normal children using established Miskolc nomograms and compared between surgery techniques. **Results** Fifty-two patients met the inclusion criteria with a median follow-up of 10 years: 25 (59.6%) TIP, 18 (34/6%) Onlay, and nine (17.3%) Duckett. Overall, Qmax increased progressively according to time and age in particular during the period covering adolescence. At follow-up 12 years postoperatively, median Qmax values were 18.5 mL/second, 13.8 mL/second and 16.6 mL/second for TIP, Onlay, and Duckett, respectively, with no significant difference detected between groups ($p = 0.5$) (see figure). Compared with normal children when adjusted for voiding volume and BSA, the proportion of obstructive uroflowmetry patterns defined as $Q_{max} < 25^{\text{th}}$ percentile of nomograms was more prevalent in patients aged 3-7 years old at 83.8% but decreased to less than 21.2% in patients aged >13 years for all procedures (see figure).

Discussion These results are consistent with previous work showing frequently reduced Qmax after hypospadias surgery with great potential for improvement at puberty. **Conclusion** These results suggest that the obstructive urinary flow pattern observed in patients early on is a frequent finding after proximal hypospadias surgery. However, because of the remarkable improvement observed at puberty a watchful waiting approach is proposed in order to avoid unnecessary intervention. [Figure presented]

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Author NameID

Salgado Diaz, Monica; ORCID: <http://orcid.org/0000-0002-6377-1238> Chaussy, Yann; ORCID: <http://orcid.org/0000-0002-7175-1341>

Institution

(Hueber, Salgado Diaz, Chaussy, Franc-Guimond, Barrieras, Houle) Department of Surgery, Centre Hospitalier Universitaire Sainte-Justine, University of Montreal, Montreal, Quebec, Canada

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741.

Is glans penis width a risk factor for complications after hypospadias repair?.

Faasse M.A., Johnson E.K., Bowen D.K., Lindgren B.W., Maizels M., Marcus C.R., Jovanovic B.D., Yerkes E.B.

Embase

Journal of Pediatric Urology. 12 (4) (pp 202.e1-202.e5), 2016. Date of Publication: 01 Aug 2016. [Article]

AN: 613343370

Introduction Recent studies have suggested that a smaller glans penis size may be associated with a higher likelihood of complications after hypospadias repair. Accurate identification of risk factors other than the well-understood variable of meatal location would allow development of better prognostic models and individualized risk stratification. **Objective** To test the hypothesis that a smaller width of the glans penis predicts adverse outcomes after hypospadias surgery. **Methods** Prospectively recorded clinical data were reviewed from a single-institution registry of primary hypospadias repairs performed between 2011 and 2014. Follow-up records were

examined for occurrence of complications. Urethroplasty complications were defined to include meatal stenosis, dehiscence, urethrocuteaneous fistula, urethral stricture, and/or urethral diverticulum. The subset of meatal stenosis and dehiscence were regarded as glanular complications. Regression analyses were performed to determine association between glans width and occurrence of complications. Because pre-operative androgen stimulation is known to increase glans penis size, separate subgroup analyses were included of patients with and without pre-operative use of testosterone cream. Results A total of 159 patients met criteria for inclusion in the study cohort: 140 patients underwent a single-stage repair, while 19 patients had a two-stage repair. The median glans penis width was 15 mm (range 10-22). Eighty-four patients (53%) received testosterone cream pre-operatively and had a significantly wider glans penis than the 75 patients who did not (median 15.5 vs 14 mm; $P < 0.001$). Median clinical follow-up was 7 months (IQR 1-12), with a minimum time elapsed since surgery of 10 months at the time of chart review. Twenty-four patients (15%) had one or more urethroplasty complications, including 11 (7%) with glanular complications. Overall, there was no statistically significant association between glans width and urethroplasty complications ($P = 0.26$) or glanular complications ($P = 0.90$) (Summary Table). Subgroup analyses of patients with and without pre-operative testosterone also revealed no significant associations between glans width and complications. Conclusions Glans penis width was not a risk factor for complications after hypospadias repair. This finding differs from the results of other recent studies and encourages further research into the value of measuring penile parameters in patients undergoing hypospadias repair. [Table presented]

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Author NameID

Lindgren B.W.; ORCID: <http://orcid.org/0000-0002-1863-9887> Faasse M.A.; ORCID:

<http://orcid.org/0000-0002-1524-7239>

Institution

(Faasse, Johnson, Bowen, Lindgren, Maizels, Marcus, Yerkes) Division of Urology, Ann & Robert H. Lurie Children's Hospital of Chicago, Northwestern University Feinberg School of Medicine, 225 E. Chicago Ave, Chicago, IL 60611, United States (Faasse) Advocate Children's Hospital, 4400 W. 95th St., Chicago, IL 60453, United States

(Jovanovic) Department of Preventive Medicine-Biostatistics, Northwestern University Feinberg School of Medicine, 680 N. Lake Shore Dr., Chicago, IL 60611, United States

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2016

742.

Hypospadias: Are we as good as we think when we correct proximal hypospadias?.

Long C.J., Canning D.A.

Embase

Journal of Pediatric Urology. 12 (4) (pp 196.e1-196.e5), 2016. Date of Publication: 01 Aug 2016.

[Review]

AN: 613343342

Objective Hypospadias surgery is a humbling art form. Although outcomes with distal hypospadias are favorable, recent publications have suggested that the complication rates are much higher than previously anticipated for proximal hypospadias. The present review examined the literature concerning proximal hypospadias, to explore some of the inadequacies and identify some of the reasons behind these shortfalls in the reported data. Methods A systematic review of the published literature was conducted using keywords relevant to proximal hypospadias and long-term outcomes. Results The literature for hypospadias was reviewed, and outcomes for distal vs proximal variants were compared. The quality of the literature for proximal hypospadias was examined, and the shortcomings that led to underreporting of the surgical outcomes were identified. Special focus was on the lack of standardized documentation, the subsequent inability to objectify the severity of the phenotype, and the underestimation of complications due to lack of long-term follow-up. There was also a great deal of variability in the utilized techniques, and the literature was filled with small case series from single institutions. To enable scientific assessment of outcomes, it is proposed that the following be implemented: acceptance and incorporation of standardized phenotype assessment scores in the pre-operative period, objective intraoperative hypospadias characterization, and postoperative score assessment. Conclusions Treatment of proximal hypospadias is much less successful than the distal variant. A specialty wide commitment to standardize the hypospadias language is required to make advancement in surgical outcomes. Boys need to be followed through puberty into adulthood, and honest reporting of outcomes must be discussed so that surgical techniques for this complicated disease process can be advanced.

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Status

Embase

Author NameID

Long C.J.; ORCID: <http://orcid.org/0000-0002-2783-7754>

Institution

(Long, Canning) Division of Urology, The Children's Hospital of Philadelphia, Perelman School of Medicine at the University of Pennsylvania, 3rd Floor Wood Center, 3401 Civic Center Boulevard, Philadelphia, PA 19104, United States

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743.

Kelly procedure for male primary epispadias.

Mishra P., Rajendran S., Asimakidou M., Mushtaq I.

Embase

Journal of Pediatric Urology. 12 (4) (pp 212.e1-212.e2), 2016. Date of Publication: 01 Aug 2016.

[Article]

AN: 613343333

Introduction Primary epispadias is a rare congenital malformation involving the urogenital system. In patients with epispadias, bladder closure enhances bladder growth and continence. **Methods** Several steps were carried out to bring the bladder neck to the midline and allow tension-free bladder neck reconstruction and recreation of the natural angulation of urethra. The urethral plate and penile shaft were dissected and the corpora cavernosa separated, and then the bladder neck repair was performed. The urethral plate was tubularised and brought ventrally. The separated corpora were reapposed, avoiding torsion. The skin was reoriented to provide cover to the penis. **Results** The Kelly procedure improves cosmesis and continence by reconstructing a tension-free bladder neck repair and lengthening the penis. **Conclusion** The video demonstrates the Kelly procedure for primary epispadias in a male child.

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Status

Embase

Institution

(Mishra, Rajendran, Asimakidou, Mushtaq) Great Ormond Street Hospital for Children, Great Ormond Street, London WC1N 3JH, United Kingdom

Publisher

Elsevier Ltd

Year of Publication

2016

744.

Biometry of the hypospadiac penis after hormone therapy (testosterone and estrogen): A randomized, double-blind controlled trial.

Paiva K.C.D.C., Bastos A.N., Miana L.P., Barros E.D.S., Ramos P.S., Miranda L.M., Faria N.M., Avarese de Figueiredo A., de Bessa J., Netto J.M.B.

Embase

Journal of Pediatric Urology. 12 (4) (pp 200.e1-200.e6), 2016. Date of Publication: 01 Aug 2016.

[Article]

AN: 613342973

Introduction The use of preoperative hormonal stimulation before hypospadias surgery aims to increase penile size and achieve better surgical results; however, the rules are not clear in the literature. We evaluated the effects of topical testosterone and estrogen in the hypospadiac penis regarding biometric measures, side effects, and serum hormonal levels. To our knowledge, this is the first study using estradiol prior to hypospadias surgery. **Patients and methods** Sixty-nine children with hypospadias were randomly divided into three study groups: a control group (CG) of 17 children given placebo cream; a testosterone group (TG) of 28 children who used 1% testosterone propionate; and an estradiol group (EG) of 24 children using 0.01% estradiol. All subjects applied the topical ointment on the entire penis, twice daily for 30 days before surgical correction. Biometric evaluation of the penis included penile length and diameter, glans diameter,

distance from the urethral meatus to the tip, and the width of the urethral plate. These measurements as well as serum hormone level, and side effects were evaluated prior to hormone use, and 30 and 90 days after. Results After 30 days an increase in penile diameter and length and diameter of the glans were observed in TG ($p < 0.05$). The width of the urethral plate and distance from meatus to the tip increased in TG, although not significantly. The most frequent side effects were appearance of pubic hair and darkening of the genital skin, mainly in TG, but these were transient and disappeared after 90 days of treatment. No significant variations were seen in serum hormonal levels (Table). Conclusion As in previous studies, an increase in penile length and diameter, and glans diameter was observed with the use of testosterone. Proximal urethral plate width and distance from the meatus to the tip of the penis had a tendency to increase also in TG. Estradiol did not change biometric measure of the penis. Few side effects occurred after both hormones, and any that did improved after 90 days follow-up and did not change hormone serum levels. Conclusion Preoperative use of topical testosterone increases penile size, diameter of the penis and glans. The use of estradiol does not change penile biometry. Side effects occur mainly with the use of testosterone and are transient. No significant and persistent hormonal changes were observed. [Table presented]

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Status

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Author NameID

Netto, Jose Murillo B.; ORCID: <http://orcid.org/0000-0002-9959-6160> de Bessa, Jose Jr.;

ORCID: <http://orcid.org/0000-0003-4833-4889>

Institution

(Paiva) Division of Pediatric Surgery, Department of Pediatrics, Federal University of Juiz de Fora (UFJF), MG, Brazil (Paiva) Division of Pediatric Surgery, Department of Pediatrics, Hospital and Maternidade Therezinha de Jesus of School of Medical Sciences and Health of Juiz de Fora (HMTJ/SUPREMA), MG, Brazil

(Miana, Barros, Netto) Division of Urology, Department of Surgery, Hospital and Maternidade Therezinha de Jesus of School of Medical Sciences and Health of Juiz de Fora (HMTJ/SUPREMA), MG, Brazil

(Ramos) School of Medical Sciences and Health of Juiz de Fora (HMTJ/SUPREMA), MG, Brazil

(Miranda, Faria, Avarese de Figueiredo, Netto) Division of Urology, Department of Surgery, Federal University of Juiz de Fora (UFJF), MG, Brazil

(de Bessa) Division of Urology, Department of Surgery, State University of Feira de Santana (UEFS), BA, Brazil

(Bastos) Department of Morphology, Federal University of Juiz de Fora (UFJF), MG, Brazil

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745.

Incidence of hip dysplasia associated with bladder exstrophy.

Mundy A., Kushare I., Jayanthi V.R., Samora W.P., Klingele K.E.

Embase

Journal of Pediatric Orthopaedics. 36 (8) (pp 860-864), 2016. Date of Publication: 2016.

[Article]

AN: 613402501

Background: Exstrophy of the bladder is a rare congenital defect seen in 2.15 children out of every 100,000 live births, with the most severe variant, cloacal exstrophy (CE), only occurring in 1 in 200,000. Developmental dysplasia of the hip (DDH) describes a spectrum of disease ranging from mild hip instability to frank dislocation. Underlying malformations, such as myelomeningocele and arthrogryposis, are often associated with the most severe variant of hip dysplasia, teratologic hip dislocation. The varying degrees of severity in DDH have been encountered in classic bladder exstrophy (CBE) patients, but the exact incidence is unknown. We sought to determine the incidence of DDH in CBE and CE patients.

Method(s): We performed a retrospective review of all children with CBE or CE presenting to a single pediatric center between 1994 and 2014. Each chart was reviewed for correct diagnosis of CBE or CE, patient age and demographics, associated medical conditions, pertinent surgeries performed, and the age at operation. Patient imaging was reviewed to determine whether bilateral hip imaging was available.

Result(s): In a 20-year retrospective review, we identified 66 patients who were diagnosed with either CBE or CE and had available hip imaging (38 males and 28 females). Of these, 11 patients were found to have radiographic evidence of DDH, for an incidence of 16.7% (11/66). Five of these patients had CE, whereas 6 presented with CBE. The first radiographic evidence of DDH was noted at a mean age of 5.75 years (range, birth to 22 y).

Conclusion(s): We advocate the use of routine hip screening ultrasound in all infants born with either CBE or CE. Early identification of DDH in these patients may allow additional treatment options to coincide with frequently used osteotomy and orthopaedic interventions.

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Status

Embase

Institution

(Mundy, Kushare, Samora, Klingele) Department of Orthopedic Surgery, Nationwide Children's Hospital, 700 Children's Drive, Columbus, OH 43205-2696, United States (Jayanthi) Department of Urology, Nationwide Children's Hospital, Columbus, OH, United States

Publisher

Lippincott Williams and Wilkins (E-mail: kathiest.clai@apta.org)

Year of Publication

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746.

Correction of penile ventral curvature in patients with minor or no hypospadias: a single surgeon's experience of 43 cases.

Seo S., Ochi T., Yazaki Y., Murakami H., Okawada M., Doi T., Miyano G., Koga H., Lane G.J., Yamataka A.

Embase

Pediatric Surgery International. 32 (10) (pp 975-979), 2016. Date of Publication: 01 Oct 2016.

[Article]

AN: 611563657

Purpose: To report our experience of correcting penile ventral curvature associated with minor or no hypospadias.

Method(s): We reviewed 43 penile ventral curvature patients treated by a single surgeon from 1997 to 2015. Of these, 23 had minor hypospadias. Curvature was corrected using degloving, chordectomy, dorsal plication (DP), tunica albuginea incision (TAI), or a combination of these.

Outcome was confirmed by induced artificial erection and post-operative appearance.

Result(s): Mean age at curvature correction was 3.2 +/- 2.6 years. 17/43 had degloving and chordectomy (DC), 16/43 had DP after DC, and 10/43 had TAI after DC, because of ventral shortening and severe curvature caused by a short hypoplastic urethra. Other procedures required were primary meatoplasty (n = 4) and urethroplasty (UP; n = 1) at the time of curvature correction, and UP after correction of curvature (n = 11). Complications included recurrence of curvature after DP (n = 3/16; 18.8 %) and urethral stenosis after UP with tubed peritoneum (n = 1/10; 10 %). There were no recurrences of curvature in TAI cases. Parents reported penile cosmesis as good (n = 38; 88.4 %), acceptable (n = 4; 9.3 %), or poor (n = 1; 2.3 %).

Conclusion(s): We recommend TAI followed by UP for correcting penile ventral curvature with short hypoplastic urethra. Tubed peritoneum is not recommended for UP.

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Status

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Institution

(Seo, Ochi, Yazaki, Murakami, Okawada, Doi, Miyano, Koga, Lane, Yamataka) Department of Pediatric General and Urogenital Surgery, Juntendo University School of Medicine, 2-1-1 Hongo, Bunkyo-ku, Tokyo 113-8421, Japan

Publisher

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Year of Publication

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747.

The cost of failure: The economic impact of failed primary closure in classic bladder exstrophy.

Hesh C.A., Young E., Intihar P., Gearhart J.P.

Embase

Journal of Pediatric Surgery. 51 (8) (pp 1312-1316), 2016. Date of Publication: 01 Aug 2016.

[Article]

AN: 610951162

Purpose Failure of primary closure in classic bladder exstrophy (CBE) is a significant cause of morbidity, and yet its relative economic impact has not been well characterized. The authors aim to determine whether CBE patients who underwent failed primary closure incur greater economic burden in the year following their successful closure than those patients who underwent a successful primary closure. Materials and Methods After institutional review board approval CBE patients who were successfully closed between 1993 and 2013 were identified in an institutional exstrophy-epispadias database. Patients who were never closed at the study institution and those who had no documented successful closure were excluded. Inpatient hospital charges, hospital costs, and professional fees were collected for the year following successful closure. Results 162 patients met the inclusion and exclusion criteria and accounted for 312 inpatient admissions in the year following and including their respective successful bladder closures. 62 of the patients failed their primary closure and the remaining 100 succeeded. Adjusting for covariates, patients who underwent successful primary closure experienced a reduction in inpatient hospital charges of \$8497, hospital costs of \$9046 and professional fees of \$11,180 in the year following their successful closure compared to those patients who failed their primary closure. Conclusion Apart from the self-evident financial advantages of a successful primary closure, namely the avoidance of reclosure, there appears to be a lasting negative financial impact of failed primary closure even after these patients undergo successful reclosure at the study institution.

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Author NameID

Hesh, Christopher A.; ORCID: <http://orcid.org/0000-0003-2559-7758>

Institution

(Hesh, Young, Gearhart) The Johns Hopkins School of Medicine, Department of Urology, Division of Pediatric Urology, Baltimore, MD, United States (Intihar) The Johns Hopkins Medical Institutions, Financial Analysis Unit, Baltimore, MD, United States

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W.B. Saunders

Year of Publication

2016

748.

Complexities of Mullerian Anatomy in 46XX Cloacal Exstrophy Patients.

Suson K.D., Preece J., Di Carlo H.N., Baradaran N., Gearhart J.P.

Embase

Journal of Pediatric and Adolescent Gynecology. 29 (5) (pp 424-428), 2016. Date of Publication: 01 Oct 2016.

[Article]

AN: 611366922

Study Objective To characterize Mullerian anatomy in 46,XX cloacal exstrophy patients. **Design** Retrospective review of prospectively maintained, institutionally approved exstrophy-epispadias-cloacal exstrophy database. **Setting** Tertiary care, high-volume exstrophy center (Division of Pediatric Urology, The Johns Hopkins Hospital, Baltimore, Maryland). **Participants** We included 31 patients who were genetically female with cloacal exstrophy for whom records included detailed evaluation of Mullerian anatomy. **Interventions** None. **Main Outcome Measures** Mullerian structures, method of evaluation, management, and sexual activity. **Results** Of our patients, 12.9% (3/31) had no identified abnormalities. Vaginal anatomy was described for 30 patients; 3/30 had vaginal agenesis, 14/30 had a single vagina, and 13/30 had vaginal duplication. Of 14 patients with 1 vagina, 5 had atresia/hypoplasia, and 1 had a lateral displacement. One patient with 2 vaginas also had distal atresia. Of the cervixes evaluated, 9/14 were duplicated (2/9 with a solitary vagina), and 19/27 of the uteri were duplicated (6/22 with 1 vagina, 1/22 with no vagina). Five patients required imaging to fully characterize their anatomy, and 7 patients had studies that failed to identify Mullerian structures seen in the operating room or on physical examination. **Common reconstructive surgeries** included vaginoplasties, incisions of vaginal septa, colporrhaphies, and hysterectomies. **Sexual activity** was confirmed for 3 patients, 2 of whom had conceived. **Conclusion** Most female cloacal exstrophy patients exhibit abnormalities of the Mullerian system. Axial imaging and ultrasound are helpful diagnostic adjuncts but do not replace careful physical examination and assessment in the operating room. Further studies of sexual activity and fertility are warranted.

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Status

Embase

Institution

(Suson, Di Carlo, Baradaran, Gearhart) Division of Pediatric Urology, The Johns Hopkins Hospital, Baltimore, Maryland, United States (Suson) Urology, Children's Hospital of Michigan, Detroit, Michigan, United States
(Preece) Division of Urology, University of Maryland Medical Center, Baltimore, Maryland, United States
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749.

Improved outcomes after technical modifications in tubularized incised plate urethroplasty for mid-shaft and proximal hypospadias.

Tam Y.H., Pang K.K.Y., Wong Y.S., Tsui S.Y., Wong H.Y., Mou J.W.C., Chan K.W., Lee K.H.

Embase

Pediatric Surgery International. 32 (11) (pp 1087-1092), 2016. Date of Publication: 01 Nov 2016.

[Article]

AN: 611534325

Purpose: To investigate and compare the outcomes after tubularized incised plate (TIP) urethroplasty in mid-shaft and proximal hypospadias using a standard and a modified technique.

Method(s): We conducted a retrospective study in 104 consecutive children who underwent mid-shaft or proximal TIP repairs from Jan 2007 to Sept 2015. Patients in Cohort One had dorsal dartos (DD) neourethral coverage while patients in Cohort Two had either de-epithelialized split preputial (DESP) or tunica vaginalis (TV) flap coverage. TV flap was used only when DESP flap was not sufficient to cover the neourethra.

Result(s): There were 52 patients each in Cohort One (DD, n = 52) and Cohort Two (DESP, n = 38; TV, n = 14) with no difference in ratio of mid-shaft/proximal between the two cohorts. At a median follow-up of 28 months, 36 patients (34.6 %) developed 47 complications including fistula (n = 19; 18.3 %) and neourethral dehiscence (n = 4; 3.8 %). Cohort One patients had significantly more fistula (28.8 vs 7.7 %; p = 0.005) and neourethral dehiscence (7.7 vs 0 %; p = 0.04) than Cohort Two. There was no difference between the two cohorts in the complication rates of meatal stenosis, recurrent ventral curvature and neourethral stricture.

Conclusion(s): Both DESP and TV flap appear to be superior to DD in preventing fistula and neourethral dehiscence in non-distal TIP repairs.

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Institution

(Tam, Pang, Wong, Tsui, Wong, Mou, Chan, Lee) Division of Paediatric Surgery and Paediatric Urology, Department of Surgery, Prince of Wales Hospital, The Chinese University of Hong Kong, Shatin, NT, Hong Kong

Publisher

Springer Verlag (E-mail: service@springer.de)

Year of Publication

2016

750.

Pudendal versus caudal block in children undergoing hypospadias surgery a randomized controlled trial.

Kendigelen P., Tutuncu A.C., Emre S., Altindas F., Kaya G.

Embase

Regional Anesthesia and Pain Medicine. 41 (5) (pp 610-615), 2016. Date of Publication: 23 Aug 2016.

[Article]

AN: 611933304

Background and Objectives: Postoperative pain management after hypospadias surgery is often challenging. Caudal block is used for analgesia but has limitations. This study compares the analgesic efficiency of pudendal block with that of caudal block in pediatric patients undergoing hypospadias repair surgery.

Method(s): This prospective, double-blind, randomized, controlled study enrolled 84 patients receiving pudendal block or caudal block before hypospadias surgery. In the pudendal group, the pudendal nerve was identified using a nerve stimulator, and the block consisted of 0.25%

bupivacaine 0.5 mL/kg. In the caudal group, the caudal block used 0.2% bupivacaine 1 mL/kg. Our primary outcome was pain intensity within 24 hours postoperatively. The trial was registered at ClinicalTrials.gov (number: NCT02390388).

Result(s): For the primary outcome, patients in the pudendal group had lower postoperative pain intensity when compared with the caudal group ($P < 0.001$). Three patients in the pudendal group and all of the patients in the caudal group needed additional analgesia within 24 hours after the surgery ($P < 0.001$). The family satisfaction rate was significantly higher in the pudendal group ($P < 0.001$).

Conclusion(s): For the pudendal group, the pain scores for the first 24 hours after the surgery were significantly lower and the duration of analgesia was longer.

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Status

Embase

Institution

(Kendigelen, Tutuncu, Altindas, Kaya) Departments of Anesthesiology and Intensive Care, Cerrahpasa Medical Faculty, Istanbul University, St. No. 53 Fatih, Istanbul 34098, Turkey (Emre) Departments of Pediatric Surgery, Cerrahpasa Medical Faculty, Istanbul University, Istanbul, Turkey

Publisher

Lippincott Williams and Wilkins (E-mail: kathiest.clai@apta.org)

Year of Publication

2016

751.

Circumcision-related tragedies seen in children at the Komfo Anokye Teaching Hospital, Kumasi, Ghana Pediatric Urology.

Appiah K.A.A., Gyasi-Sarpong C.K., Azorliade R., Aboah K., Laryea D.O., Otu-Boateng K., Baah-Nyamekye K., Maison P.O.M., Arthur D., Antwi I.O., Frimpong-Twumasi B., Yenli E.M., Togbe S.K., Amoah G.

Embase

BMC Urology. 16 (1) (no pagination), 2016. Article Number: 65. Date of Publication: 08 Nov 2016.
[Article]

AN: 613123729

Background: Circumcision is a common minor surgical procedure and it is performed to a varying extent across countries and religions. Despite being a minor surgical procedure, major complications may result from it. In Ghana, although commonly practiced, circumcision-related injuries have not been well documented. This study is to describe the scope of circumcision-related injuries seen at the Komfo Anokye Teaching Hospital in Kumasi, Ghana.

Method(s): The study was conducted at the Urology Unit of the Komfo Anokye Teaching Hospital in Kumasi. Consecutive cases of circumcision-related injuries seen at the unit over an 18 month period were identified and included in the study. Data was collected using a structured questionnaire. Data was entered and analysed using SPSS version 16. Charts and tables were generated using Microsoft Excel.

Result(s): A total of 72 cases of circumcision-related injuries were recorded during the 18 month period. Urethrocuteaneous fistula was the commonest injury recorded, accounting for 77.8 % of cases. Other injuries recorded were glans amputations (6.9 %); iatrogenic hypospadias (5.6 %), and epidermal inclusion cysts (2.8 %). The majority of children were circumcised in health facilities (75 %) and nurses were the leading providers (77.8 %). The majority of circumcisions were conducted in the neonatal period (94.7 %).

Conclusion(s): Circumcision-related injuries commonly occurred in the neonatal period. Most of the injuries happened in health facilities. The most common injury recorded was urethrocuteaneous fistula but the most tragic was penile amputation. There is the need for education and training of providers to minimise circumcision-related injuries in Ghana.

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Status

Embase

Author NameID

Appiah, Kwaku Addai Arhin; ORCID: <http://orcid.org/0000-0002-7583-8938>

Institution

(Appiah, Azorliade, Otu-Boateng, Baah-Nyamekye, Maison, Arthur, Antwi, Frimpong-Twumasi, Togbe, Amoah) Department of Surgery, Komfo Anokye Teaching Hospital, Kumasi, Ghana
(Gyasi-Sarpong, Aboah) Department of Surgery, School of Medical Sciences-KNUST, Kumasi, Ghana

(Laryea) Public Health Unit, Komfo Anokye Teaching Hospital, Kumasi, Ghana

(Yenli) Department of Surgery, Tamale Teaching Hospital, Kumasi, Ghana

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Year of Publication

2016

752.

Isolated Male Epispadias: Anatomic Functional Restoration Is the Primary Goal.

Spinoit A.-F., Claeys T., Bruneel E., Ploumidis A., Van Laecke E., Hoebeke P.

Embase

BioMed Research International. 2016 (no pagination), 2016. Article Number: 6983109. Date of Publication: 2016.

[Article]

AN: 612579418

Background. Isolated male epispadias (IME) is a rare congenital penile malformation, as often part of bladder-exstrophy-epispadias complex (BEEC). In its isolated presentation, it consists in a defect of the dorsal aspect of the penis, leaving the urethral plate open. Occurrence of urinary incontinence is related to the degree of dorsal displacement of the meatus and the underlying underdevelopment of the urethral sphincter. The technique for primary IME reconstruction, based on anatomic restoration of the urethra and bladder neck, is here illustrated. Patients and **Methods.** A retrospective database was created with patients who underwent primary IME repair between June 1998 and February 2014. Intraoperative variables, postoperative complications, and outcomes were assessed. A descriptive statistical analysis was performed. **Results and Limitations.** Eight patients underwent primary repair, with penopubic epispadias (PPE) in 3, penile epispadias (PE) in 2, and glandular epispadias (GE) in 3. Median age at surgery was 13.0 months [7 -47]; median follow-up was 52 months [9 -120]. Complications requiring further surgery were reported in two patients, while further esthetic surgeries were required in 4 patients. **Conclusion.** Anatomical restoration in primary IME is safe and effective, with acceptable results given the initial pathology.

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PMID

27722172 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=27722172>]

Status

Embase

Author NameID

Spinoit, Anne-Francoise; ORCID: <http://orcid.org/0000-0002-9045-9127> Claeys, Tom; ORCID: <http://orcid.org/0000-0001-6677-6846>

Institution

(Spinoit, Claeys, Bruneel, Ploumidis, Van Laecke, Hoebeke) Department of Urology, Ghent University Hospital, Ghent, Belgium

Publisher

Hindawi Publishing Corporation (410 Park Avenue, 15th Floor, 287 pmb, New York NY 10022, United States)

Year of Publication

2016

753.

Reinforcing the ventral penile shaft with pedicled fat/connective tissues before urethroplasty lowers the risk for post-urethroplasty complications in hypospadias.

Sueyoshi R., Seo S., Ochi T., Murakami H., Yazaki Y., Takeda M., Nakamura H., Lane G.J., Yamataka A.

Embase

Journal of Pediatric Surgery. 51 (12) (pp 2021-2024), 2016. Date of Publication: 01 Dec 2016.

[Article]

AN: 613312154

Purpose CHARGE is our technique for reinforcing the ventral penile shaft with pedicled pericordal/scrotal fat, pedicled perimeatal connective tissue, or a combination of these at the time of initial hypospadias surgery. Such pedicled grafts "charge" poorly developed urethral plates and thin ventral foreskin prior to urethroplasty to improve compromised vascular perfusion that could prevent post-urethroplasty complications (post-UPC). Methods We reviewed post-UPC in 179 staged hypospadias repair patients (1997-2015). CHARGE, adopted routinely in 2010 was used in 39 patients (C-group), not indicated in 7 because ventral connective tissue was thick, and not used in 133 (NC-group). Initial hypospadias surgery included foreskin degloving with or without chordectomy, dorsal plication, tunica albuginea incision, or a combination of these. Results Subject demographics were similar. NC had significantly more post-UPC than C (25 versus 0; $p <$

.01) comprising stenosis (n = 14), fistula (n = 7), diverticulum (n = 2), and wound infection (n = 2) that developed after a mean of 0.7 +/- 0.2 years (range: 1 day-2.8 years). Extra time taken for CHARGE was less than 15 minutes in all cases. Mean follow-up after urethroplasty (years) was significantly shorter in C (1.5 +/- 1.0 versus 5.7 +/- 3.8) (p < .01), but almost double the time taken to develop post-UPC. Conclusion CHARGE would appear to prevent post-UPC. Level of evidence Retrospective Comparative Study - Level III

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PMID

27686484 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=27686484>]

Status

Embase

Institution

(Sueyoshi, Seo, Ochi, Murakami, Yazaki, Takeda, Nakamura, Lane, Yamataka) Department of Pediatric General and Urogenital Surgery, Juntendo University School of Medicine, Tokyo, Japan

Publisher

W.B. Saunders

Year of Publication

2016

754.

Diethylstilbestrol (DES): Also harms the third generation.

Anonymous

Embase

Prescrire International. 25 (177) (pp 294-298), 2016. Date of Publication: December 2016.

[Review]

AN: 613765885

* Diethylstilbestrol (DES) is a synthetic nonsteroidal oestrogen and endocrine disruptor that was used in the 1950s-1970s to prevent spontaneous abortion, despite its lack of proven efficacy. Millions of women worldwide took DES during pregnancy. * In France, between 1951 and 1981, about 160 000 children were exposed to DES during the first trimester of their intrauterine life, and in some cases almost throughout the entire pregnancy. They are referred to as "DES daughters" and "DES sons". In 2010, in France, about 25 000 DES daughters were aged 33 to 40

years: pregnancies among these women are foreseeable until about 2020. * In utero exposure to DES can have harmful effects. In particular, DES daughters have an increased risk of cancer and structural abnormalities of the uterus that can adversely affect their pregnancies. * What are the consequences of taking DES during pregnancy for the third generation, i.e. the children of DES children? To answer this question, we reviewed the available data in mid- 2016 using the standard Prescrire methodology. * According to a retrospective study conducted in France by Réseau DES France, published in 2016, which included 4409 DES grandchildren (2228 girls and 2181 boys) and about 6000 controls, about one-quarter of DES grandchildren are born prematurely. * Preterm delivery exposes neonates to serious neonatal complications, including neurosensory disorders, disabilities and increased neonatal mortality. The more premature the baby, the greater the risk of complications. In the Réseau DES France study, cerebral palsy was more frequent in the DES grandchildren group: 59/10 000, versus 6/10 000 in the control group. * A study conducted in the United States in about 4500 DES daughters found that preterm delivery occurred at a frequency of about 26%, much higher than that reported in controls. Neonatal mortality was 8 times higher among DES grandchildren, and the risk of stillbirth was twice as high. * Other smaller studies have also shown an increased risk of preterm birth. * A cohort study conducted in about 5000 DES grandchildren found that the risk of malformations of any type was higher than in the unexposed control group. * Epidemiological studies, conducted in several countries, found an increased frequency of hypospadias in DES grandsons. The relative risk was about 5 in the largest study. Other, less robust studies found no statistically significant difference. * Several studies in several countries have shown a twofold increase in the risk of oesophageal atresia or tracheo-oesophageal fistula in DES grandchildren. * The data on congenital heart defects or musculoskeletal malformations are limited and uninformative. * Epidemiological studies have not identified a significant increase in the risk of gynaecological anomalies or cancers in DES granddaughters. * Limited data are available on the risk of malformations in the children of DES sons. * The data obtained in rodents exposed to DES (and other endocrine disruptors) make it entirely plausible that in utero exposure to DES, in humans too, provokes epigenetic effects that are passed on to future generations not directly exposed to DES. * In practice, these data should be discussed with DES daughters, their partners and healthcare teams so that appropriate monitoring, clinical management and follow-up can be arranged for both mother and baby. The harms of taking DES during pregnancy last for decades and affect future generations.

Status

Embase

Publisher

Association Mieux Prescrire (E-mail: international@prescrire.org)

Year of Publication

2016

755.

Parental Decisional Regret after Primary Distal Hypospadias Repair: Family and Surgery Variables, and Repair Outcomes.

Ghidini F., Sekulovic S., Castagnetti M.

Embase

Journal of Urology. 195 (3) (pp 720-724), 2016. Date of Publication: 2016.

[Article]

AN: 608302513

Purpose Decisional regret is defined as distress after making a health care choice and can be an issue for parents electing distal hypospadias repair for their sons. We assessed the influence on decisional regret of variables related to the family, surgery and outcomes. Materials and Methods Charts for 372 patients undergoing primary distal hypospadias repair between 2005 and 2012 were reviewed, and validated questionnaires, including the Decisional Regret Scale, Pediatric Penile Perception Score and Dysfunctional Voiding and Incontinence Scoring System, were administered to parents. Results Data were available for 172 of 372 families (response rate 46.2%). Of 323 parents 128 (39.6%) presented with moderately strong decisional regret, with good agreement within couples. Predictors of decisional regret included intermediate parental educational level (OR 3.19, 95% CI 1.52-6.69), patient not being the first born (OR 2.01, 95% CI 1.07-3.78), family history of hypospadias (OR 4.42, 95% CI 1.96-9.97), initial desire to avoid surgery (OR 2.07, 95% CI 1.04-4.12), younger age at followup (OR 0.81, 95% CI 0.72-0.91), presence of lower urinary tract symptoms (OR 4.92, 95% CI 1.53-15.81) and lower Pediatric Penile Perception Score (OR 0.86, 95% CI 0.75-0.99). Decisional regret was unrelated to parental desire to avoid circumcision, surgical variables, development of complications and duration of followup. Conclusions Decisional regret is a problem in a significant proportion of parents electing distal hypospadias repair for their sons. In our experience family variables seemed to be predictors of decisional regret, while surgical variables did not. Predictors of decisional regret included worse parental perception of penile appearance and the presence of lower urinary tract symptoms. However, the latter could be unrelated to surgery. Irrespective of the duration of followup, decisional regret seems decreased in parents of older patients.

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PMID

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Status

Embase

Institution

(Ghidini, Sekulovic, Castagnetti) Section of Pediatric Urology, Urology Unit, University Hospital of Padova, Padua, Italy

Publisher

Elsevier Inc. (E-mail: usjcs@elsevier.com)

Year of Publication

2016

756.

Grafted tubularised incised-plate urethroplasty: An objective assessment of outcome with lessons learnt from surgical experience with 263 cases.

Gupta V., Yadav S.K., Alanzi T., Amer I., Salah M., Ahmed M.

Embase

Arab Journal of Urology. 14 (4) (pp 299-304), 2016. Date of Publication: 01 Dec 2016.

[Article]

AN: 613549959

Objective Snodgrass urethroplasty remains the preferred technique in primary distal hypospadias but development of meatal stenosis often limits distal extension of the midline incision of the urethral plate (MIUP), which remains a limiting factor in reconstructing an apical neomeatus (NM). We here-in assess the cosmetic and functional outcome with distal extension of the MIUP in grafted tubularised incised-plate urethroplasty (G-TIP) repair. Patients and methods This prospective study included the surgical experience of 263 cases of primary hypospadias operated upon between 2012 and 2015. The G-TIP technique included standard steps of Snodgrass urethroplasty, including degloving and harvesting of glans wings, followed by MIUP that was extended distally beyond the margins of the urethral plate (UP) into the glans. The incised bed was grafted with a free preputial skin graft and fixed to the bed with polydioxanone 7-0 suture. The UP was tubularised and the suture line reinforced with a Dartos flap. The urethral catheter was removed at 7-10 days after the repair and the outcome was assessed at follow-up using the Hypospadias Objective Scoring Evaluation (HOSE) system. Results An apical NM was achieved

in 96% of the patients with a 3.7% incidence of urethrocutaneous fistula. The presence of suture tracks and graft at the margins of the NM were seen in the initial 4% and 5% of cases, respectively. Acceptable cosmetic results, with objective HOSE scores of >14, were achieved in 96% of cases. Conclusion The G-TIP repair is a straightforward and feasible technique facilitating reconstruction of an apical NM, with an optimum outcome based on HOSE scoring. However, multicentre data are needed for undertaking comparative analysis and to assess the universal applicability of this technique in primary hypospadias.

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Institution

(Gupta, Yadav, Alanzi, Amer, Salah, Ahmed) Department of Pediatric Surgery and Urology, Ibn Sina Hospital, Kuwait City, Kuwait

Publisher

Arab Association of Urology (E-mail: araburo@yahoo.com)

Year of Publication

2016

757.

Patient Reported Outcomes in Preoperative and Postoperative Patients with Hypospadias.

Keays M.A., Starke N., Lee S.C., Bernstein I., Snodgrass W.T., Bush N.C.

Embase

Journal of Urology. Part 2. 195 (4) (pp 1215-1220), 2016. Date of Publication: April 2016.

[Article]

AN: 608674070

Purpose Current outcome tools for hypospadias have limited focus on the caregiver or patient perspective of important patient centered outcomes. In this study we collaborated with patients, caregivers, and lay and medical experts to develop and pilot a patient reported outcome measure for hypospadias. Materials and Methods We developed a patient reported outcome measure based on systematic review of the literature and focus group input. The patient reported outcome measure was piloted in caregivers for boys younger than 8 years and in patients older than 8 years who presented for urology consultation before meeting with the surgeon. Patients were

classified with uncorrected hypospadias, successful repair or failed repair based on the presence or absence of complications (fistula, diverticulum, meatal stenosis/stricture, greater than 30-degree recurrent curvature, glans dehiscence and/or skin reoperation). Results A patient reported outcome measure was developed and administered to 347 patients and/or caregivers-proxies, including 105 uncorrected cases, 162 successful repair cases and 80 failed cases. Satisfaction with appearance was highest in those with successful hypospadias repair compared to failed repair and uncorrected hypospadias (93% vs 77% and 67%, respectively). Voiding symptoms such as spraying or a deviated stream were highest in failed and uncorrected cases (39% and 37%, respectively). Overall dissatisfaction with voiding was highest for uncorrected hypospadias and failed repair compared to successful cases (54% and 47%, respectively, vs 15%).

Conclusions The evaluation of patient and caregiver-proxy reported outcomes in preoperative and postoperative patients with hypospadias allows for the quantification of benefits derived from hypospadias repair and may ultimately represent the gold standard outcome measure for hypospadias. This pilot study identified preliminary patient centered themes and demonstrated the feasibility of administering hypospadias patient reported outcome measures in clinical practice.

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26926541 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=26926541>]

Status

Embase

Institution

(Keays) Children's Hospital of Eastern Ontario, University of Ottawa, Ottawa, Ontario, Canada

(Starke, Lee, Bernstein) University of Texas Southwestern Medical Center, Dallas, Texas, United States

(Snodgrass, Bush) PARC Urology, Frisco, Texas, United States

Publisher

Elsevier Inc. (E-mail: usjcs@elsevier.com)

Year of Publication

2016

The incidence of apparent congenital urogenital anomalies in North Indian newborns: A study of 20,432 pregnancies.

Bhat A., Kumar V., Bhat M., Kumar R., Patni M., Mittal R.

Embase

African Journal of Urology. 22 (3) (pp 183-188), 2016. Date of Publication: 01 Sep 2016.

[Article]

AN: 613244818

Introduction and objectives Over the last few decades, congenital anomalies of the urogenital system have increased globally as a consequence of advanced maternal age at pregnancy and developments in assisted reproductive techniques. The aim of this study was to determine the incidence of apparent congenital urogenital anomalies in North Indian newborns and the causative factors. **Subjects and methods** The data of all newborns delivered at our institute between September 2012 and August 2014 were collected for this prospective study. The predetermined data format included the newborns' birth weight and gestational age, the maternal age, parity and infertility treatment, if any. Newborns weighing less than 1000 g or born before 32 weeks of gestation were excluded from the study. **Results** During the study period, 20,432 deliveries were recorded (10,952 male and 9480 female babies). Apparent urogenital congenital anomalies were diagnosed in 799, with an incidence of 39.1 per 1000 newborns. The most common anomaly was cryptorchidism found in 678 newborns, while hypospadias was noted in 61, ambiguous genitalia in 34, congenital hernia/hydrocele in 20 and an exstrophy-epispadias complex in 5 children. Prune belly syndrome was seen in 1 newborn. Newborns weighing less than 2500 g had a higher proportion of anomalies (9.64%) in comparison to those weighing over 2500 g (1.99%) ($p = 0.0001$). A maternal age >30 years, parity >2 and infertility treatment were recorded in 5.40%, 4.93% and 9.80%, respectively, and all were independently associated with an increased risk of urogenital anomalies ($p = 0.0001$). **Conclusions** The incidence of apparent congenital urogenital anomalies was 3.91%. Infertility treatment, parity >2 and a maternal age >30 years were independently associated with an increased risk of congenital urogenital anomalies.

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Author NameID

Bhat A.; ORCID: <http://orcid.org/0000-0002-2141-4726>

Institution

(Bhat) Department of urology, Dr. S.N. Medical College, Jodhpur, India (Bhat, Kumar, Bhat, Kumar, Mittal) Department of Urology, S.P. Medical College, Bikaner, Rajasthan 334003, India

(Patni) Department of Obstetric and Gynecology, S.P. Medical College, Bikaner, Rajasthan
334003, India

Publisher

Pan African Urological Surgeons Association(PAUSA) (E-mail: sunnydoodu@yahoo.com)

Year of Publication

2016

759.

A simple method for closure of urethrocutaneous fistula after tubularized incised plate repair:

Preliminary results.

Shirazi M., Ariafar A., Babaei A.H., Ashrafzadeh A., Adib A.

Embase

Nephro-Urology Monthly. 8 (6) (no pagination), 2016. Article Number: e40371. Date of

Publication: November 2016.

[Article]

AN: 613239424

Background: Urethrocutaneous fistula (UCF) is the most prevalent complication after hypospadias repair surgery. Many methods have been developed for UCF correction, and the best technique for UCF repair is determined based on the size, location, and number of fistulas, as well as the status of the surrounding skin.

Objective(s): In this study, we introduced and evaluated a simple method for UCF correction after tubularized incised plate (TIP) repair.

Method(s): This clinical study was conducted on children with UCFs ≤ 4 mm that developed after TIP surgery for hypospadias repair. The skin was incised around the fistula and the tract was released from the surrounding tissues and the dartos fascia, then ligated with 5-0 polydioxanone (PDS) sutures. The dartos fascia, as the second layer, was covered on the fistula tract with PDS thread (gauge 5-0) by the continuous suture method. The skin was closed with 6-0 Vicryl sutures. After six months of follow-up, surgical outcomes were evaluated based on fistula relapse and other complications.

Result(s): After six months, relapse occurred in only one patient, a six-year-old boy with a single 4-mm distal opening, who had undergone no previous fistula repairs. Therefore, in 97.5% of the cases, relapse was non-existent. Other complications, such as urethral stenosis, intraurethral

obstruction, and epidermal inclusion cysts, were not seen in the other patients during the six-month followup period.

Conclusion(s): This repair method, which is simple, rapid, and easily learned, is highly applicable, with a high success rate for the closure of UCFs measuring up to 4 mm in any location.

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Embase

Institution

(Shirazi, Ashrafzadeh) Department of Urology, School of Medicine, Shiraz University of Medical Sciences, Shiraz, Iran, Islamic Republic of (Ariaifar) Urology Oncology Research Center, Shiraz University of Medical Sciences, Shiraz, Iran, Islamic Republic of

(Babaei, Adib) Student Research Committee, Shiraz University of Medical Sciences, Shiraz, Iran, Islamic Republic of

Publisher

Kowsar Corp (E-mail: info@anesthpain.com)

Year of Publication

2016

760.

Surgical pathologies in children presenting for non-medical. Tibbi endikasyon disi sunnet icin basvuran cocuklarda cerrahi patolojiler <Tibbi endikasyon disi sunnet icin basvuran cocuklarda cerrahi patolojiler.>

Thomas D.T.

Embase

Marmara Medical Journal. 29 (2) (pp 84-87), 2016. Date of Publication: 2016.

[Article]

AN: 611771138

Objectives: Circumcision is the most commonly performed surgical procedure in the world. In this study, we determined the frequency of additional surgical pathologies in patients presenting for non-medical circumcision.

Material(s) and Method(s): Children presenting to a secondary healthcare facility between June 2013 - June 2014 for non-medical circumcision were included in this study. Patients' ages, presenting at outpatient clinics were noted together with the results of physical examinations.

Result(s): A total of 2088 children presented for non-medical circumcision. Their average age was 5.2 years. 56.3% of patients presented to the Pediatric Surgery outpatient clinic, 25.3% presented to the Urology outpatient clinic and 18.3% presented to the General Surgery outpatient clinic. Additional surgical pathologies were noted in 3.9% of patients and these were: phimosis (n=36), inguinal hernia/hydrocele (n=12), buried penis (n=10), undescended testis (n=9), retractile testis (n=5), hypospadias (n=3), megameatus (n=2), umbilical hernia (n=2) and varicocele (n=2). The surgical plans for 37 (1.8%) patients changed due to the findings at examination.

Conclusion(s): Changes in surgical plans were required for 1.8% of patients. It is therefore important that all patients, including those presenting with a request for circumcision, have a detailed physical examination.

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Status

Embase

Institution

(Thomas) Department of Pediatric Surgery, Faculty of Medicine, Maltepe University, Ataturk Cad. Cam Sok. No 3, Maltepe, Istanbul, Turkey

Publisher

Marmara University

Year of Publication

2016

761.

Fertility potential in adult hypospadias.

Kumar S., Tomar V., Yadav S.S., Priyadarshi S., Vyas N., Agarwal N.

Embase

Journal of Clinical and Diagnostic Research. 10 (8) (pp PC01-PC05), 2016. Date of Publication: 01 Aug 2016.

[Article]

AN: 611501622

Introduction: Hypospadias is a congenital anomaly of the urogenital tract characterized by abnormal location of the external urethral meatus over ventral aspect of penis. The ideal time to correct primary hypospadias is when aged 6-12 months. However, in some developing countries, such as ours, this anomaly may be left untreated until adulthood. There are multiple risk factors responsible for development of this anomaly, out of which paternal sub fertility is one of them. As the child grows into adulthood, fertility status becomes an important issue, frequently raised by patients.

Aim(s): To evaluate the fertility potential of adult hypospadiac patients and to rule out the effect of age of surgical correction over fertility potential.

Material(s) and Method(s): Seventy three adult patients of hypospadias were prospectively evaluated, out of which 43 were operated during adulthood. All patients underwent measurement of penile length and circumference in both flaccid and erect positions along with testicular volume, serum LH, FSH and testosterone and semen analysis. A set of questionnaire was given to all the patients which included assessment of erectile function (IIEF-5), ejaculatory function, strength of libido and level of satisfaction after sexual intercourse. Results were compared with an age matched control group of 70 healthy person.

Result(s): Out of 73 patients with a mean age of 23.73 years, 14(19.17%) had proximal and rest 59(80.82%) had distal type of hypospadias. Mean penile length in both type of hypospadiac patients under both flaccid and erect conditions (7.92 ± 1.33 and 9.62 ± 1.31 cm) were significantly shorter than those of control (10.78 ± 0.94 and 13.15 ± 1.15 cm) ($p < 0.001$). In spite of short penile length, the level of satisfaction of all patients and their partner after penetrating intercourse were comparable to control ($p > 0.05$). The strength of libido ($p > 0.05$) was comparable with control in both type of hypospadiac patients; however IIEF-5 scores was poor in the proximal type of hypospadias. Semen volume (ml), sperm concentration (mill/ml), active sperm motility (%), and normal sperm morphology were comparable between distal hypospadiac group and control ($p > 0.05$), however these parameters were poor in proximal type. The serum FSH and LH levels were significantly higher and serum testosterone level was significantly lower in hypospadiac patients than control ($p < 0.001$). However, no difference in testicular size was found. The patients who were operated during childhood had shorter penile length compared to patients operated during adulthood.

Conclusion(s): Fertility potential parameters in distal type of hypospadias were comparable with control. However the proximal type had poor erectile function and semen quality. Patients operated in either childhood or in adulthood, there was no significant difference in fertility potential.

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Status

Embase

Institution

(Kumar, Tomar, Yadav, Priyadarshi, Vyas, Agarwal) Department of Urology, S.M.S Medical College, Jaipur, Rajasthan, India

Publisher

Journal of Clinical and Diagnostic Research (No 3, 1/9 Roop Nagar, G T Road, Delhi 110007, India)

Year of Publication

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762.

This Month in Pediatric Urology.

Cain M.P.

Embase

Journal of Urology. 196 (2) (pp 297-298), 2016. Date of Publication: 01 Aug 2016.

[Article]

AN: 611160727

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Embase

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Year of Publication

2016

763.

Modified Penile Augmentation by Dermal-Fat Graft in Post-Hypospadias Adults.

Xu L., Zhao M., Yang Z., Chen W., Li Y., Ma N., Wang W., Feng J., Liu Q., Ma T.

Embase

Aesthetic plastic surgery. 40 (1) (pp 120-129), 2016. Date of Publication: 01 Feb 2016.

[Article]

AN: 621449179

BACKGROUND: Although a considerable part of patients desire further improvement in penile size after hypospadias repairs, penile augmentation is only considered in patients with congenital penile hypoplasia or acquired penile retraction. Modified penile augmentation by free dermal-fat graft is introduced to satisfy these patients and improve surgical safety. **METHODS:** From April 2012 to December 2014, a total of 15 male adults after hypospadias repairs, aged 18-24 years, underwent modified penile augmentation which involved girth enhancement by a free dermal-fat graft and penile elongation (suprapubic skin advancement-ligamentolysis). A specially designed tunneling instrument was introduced to facilitate pericavernous thickening without degloving. Outcome evaluation was mainly based on objective penile measurements and results of the Male Genital Image Scale.

RESULTS: With 6 months' follow-up, all patients had achieved excellent cosmetic results, with a significant average dimensional increase of 1.53 ± 0.23 cm in flaccid girth and 1.67 ± 0.24 cm in flaccid length. No erection deficiencies or urinary fistula occurred. Patients' perception of male genitalia improved postoperatively, with the average MGIS score rising from 31.73 ± 4.86 to 40.20 ± 4.54 .

CONCLUSIONS: This modified technique is safe and effective in enlarging penile size. The use of the specially designed tunneling instrument simplifies penile girth enhancement, avoiding unnecessary trauma to the neo-urethra and neurovascular bundle. It is confirmed that physical dimensional enhancement does contribute to improving their underestimation of penile size.

LEVEL OF EVIDENCE IV: This journal requires that authors assign a level of evidence to each article. For a full description of these Evidence-Based Medicine ratings, please refer to the Table of Contents or the online Instructions to Authors www.springer.com/00266.

PMID

26705273 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=26705273>]

Institution

(Xu, Zhao, Yang, Chen, Li, Ma, Wang, Feng, Liu, Ma) The 2nd Department of Plastic Surgery, Plastic Surgery Hospital, Chinese Academy of Medical Sciences & Peking Union Medical College, No. 33, Ba-da-chu Road, Shijingshan District, Beijing, 100144, China

Year of Publication

2016

764.

Consultation for Small-Sized Penis in the Egyptian Males: A Case Control Study.

Salama N.

Embase

American journal of men's health. 10 (3) (pp 220-227), 2016. Date of Publication: 01 May 2016.

[Article]

AN: 619787375

This study aimed to report penile dimensions in adult Egyptian males consulting for small-sized penis (SSP) and describe their demographics and andrological profile. A case control study was designed through retrospective data analysis of patients (n = 239) seeking advice for SSP and a control group (n = 59). This included sociodemographics, other andrological complaints, and penile dimensions (pendulous length [PL], penopubic or total length [TL], and circumference [CF]) at flaccid and erect states and the size of the prepubic fatty pad. The results reported that most patients were single, students, and smokers and had not completed a university education. Several patients reported falsely premature ejaculation (PE), penile curvature (PC), and small-sized testes. Most penile dimensions of the patients (mean, cm) were significantly lower than those of the controls, whether in flaccid (PL: 7.4 vs. 8.05, p = .008; CF: 8.7 vs. 8.98, p = .026) or erect state (PL: 11.8 vs. 13, p = .000; TL: 14.2 vs. 15, p = .000; CF: 11.3 vs. 11.8, p = .003). However, no patient presented with a pendulous penile length <4 cm in flaccid or <7 cm in erect state. In conclusion, Egyptian men consulting for SSP did not have true small organs, but their penile dimensions were slightly smaller than those of men without such complaint. The proper sexual education program is highly advisable in these situations to avoid the myths and misconceptions about sexuality.

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Institution

(Salama) Alexandria Faculty of Medicine, Alexandria, Egypt

Year of Publication

2016

765.

Copy Number Variations of the Azoospermia Factor Region and SRY Are Not Associated with the Risk of Hypospadias.

Kon M., Saito K., Mitsui T., Miyado M., Igarashi M., Moriya K., Nonomura K., Shinohara N., Ogata T., Fukami M.

Embase

Sexual development : genetics, molecular biology, evolution, endocrinology, embryology, and pathology of sex determination and differentiation. 10 (1) (pp 12-15), 2016. Date of Publication: 2016.

[Article]

AN: 619690390

We investigated the frequency of copy number variations (CNVs) in the Y chromosome of Japanese children with hypospadias. We analyzed the copy number of the azoospermia factor (AZF) region and SRY, using multiplex ligation-dependent probe amplification. Four AZF-linked CNVs, including one novel simple duplication, were identified in 39 of 89 patients, at a frequency comparable to that of those in unaffected individuals. SRY-linked CNVs were absent in our patients. The results imply that CNVs in the AZF region and SRY are not associated with the risk of hypospadias in the Japanese population, although the pathogenicity of the AZF-linked simple duplication remains to be elucidated.

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PMID

27023068 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=27023068>]

Institution

(Kon) Department of Molecular Endocrinology, National Research Institute for Child Health and Development, Tokyo, Japan

Year of Publication

2016

766.

NR5A1 Loss-of-Function Mutations Lead to 46,XY Partial Gonadal Dysgenesis Phenotype: Report of Three Novel Mutations.

Fabbri H.C., Ribeiro de Andrade J.G., Maciel-Guerra A.T., Guerra-Junior G., de Mello M.P.

Embase

Sexual development : genetics, molecular biology, evolution, endocrinology, embryology, and pathology of sex determination and differentiation. 10 (4) (pp 191-199), 2016. Date of Publication: 2016.

[Article]

AN: 619648471

Mutations in the NR5A1 gene, which encodes the steroidogenic factor 1 (SF1), are responsible for different phenotypes of disorders of sex development (DSD), such as bilateral anorchia and hypospadias. Furthermore, they can be associated with primary amenorrhea, premature ovarian failure, male infertility, adrenal tumors, and endometriosis. Direct sequencing of the 7 NR5A1 exons including ~1,000 bp of the 5'-upstream and 3'-downstream regions and all intron-exon boundaries was performed in patients with DSD. Three different in silico tools were used to assess the consequences of a splice site mutation. As a result, 3 novel NR5A1 mutations were identified in 3 patients with 46,XY partial gonadal dysgenesis: p.Lys38* and p.Leu80Trpfs*8 lead to premature translation termination codons within the SF1 DNA-binding domain, and the intronic nucleotide substitution c.1138+1G>T at the intron 6 donor splice site is considered to modify correct splicing. We assume that the anomalous mRNA produced as a result of p.Lys38* and p.Leu80Trpfs*8 will be degraded by nonsense-mediated mRNA decay even before translation, leading to SF1 haploinsufficiency. The c.1138+1G>T mutation is expected to produce a truncated protein. Heterozygous SF1 loss-of-function mutations in these cases resulted in mild DSD manifestations, such as dysgenetic testes, spontaneous puberty, and preserved adrenal function.

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PMID

27463801 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=27463801>]

Institution

(Fabbri) Center for Molecular Biology and Genetic Engineering - CBMEG, Campinas, Brazil

Year of Publication

2016

Surgical Repair of Mid-shaft Hypospadias Using a Transverse Preputial Island Flap and Pedicled Dartos Flap Around Urethral Orifice.

Liang W., Ji C., Chen Y., Zhang G., Yao Y., Zhang J.

Embase

Aesthetic plastic surgery. 40 (4) (pp 535-539), 2016. Date of Publication: 01 Aug 2016.

[Article]

AN: 618409729

OBJECT: To evaluate the effects, particularly the incidence of anastomotic fistula, of a pedicled dartos flap around the urethral orifice in the treatment of urethroplasty of mid-shaft hypospadias.

METHODS: A total of 46 cases of congenital mid-shaft hypospadias were included in this study.

The patients ranged in age from 0.7 to 25.4 years and the average was 5.8 years. The patients received penis chordee correction. A transverse preputial island flap was developed for urethral reconstruction. The proximal dartos of the urethral orifice was used to develop a pedicled dartos flap, which was transposed to cover and strengthen neourethral anastomosis. The ventral penile skin defect was repaired by another flap.

RESULTS: The 46 patients were examined during follow-up visits for 6 months to 3 years. An anastomotic fistula was observed in one case (2.2 %). Scar healing without fistula was observed in another patient due to poor blood supply to part of the ventral penile skin. No other incidences of fistula, urethral rupture, flap necrosis, wound infections, urinary tract (meatal) stenosis, or urethral diverticulum were observed in the patients.

CONCLUSION: A pedicled dartos flap around the urethral orifice can take advantage of well-vascularized local tissue to add a protective layer to the proximal aspect of the neourethral anastomosis for reducing the incidence of anastomotic fistula in mid-shaft hypospadias repair using a transverse preputial island flap.

LEVEL OF EVIDENCE V: This journal requires that authors assign a level of evidence to each article. For a full description of these Evidence-Based Medicine ratings, please refer to the Table of Contents or the online Instructions to Authors www.springer.com/00266 .

PMID

27286850 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=27286850>]

Institution

(Liang, Ji, Chen, Zhang, Yao, Zhang) Department of Plastic Surgery, Sun Yat-sen Memorial Hospital, Sun Yat-sen University, Guangzhou, 510120, Guangdong Province, China (Zhang) Department of Plastic Surgery, Huizhou Central Hospital, Huizhou, 516000, Guangdong Province, China

Year of Publication

2016

768.

Our experience of proximal hypospadias repair using the Cloutier-Bracka technique at the Gynaeco-Obstetric and Paediatric Hospital, Yaounde-Cameroon.

Faustin Felicien M.T., Nwaha Makon A.S., Kamadjou C., Fossi G., Le Coultre C., Andze O.G., Sosso M.A., Mure P.Y.

Embase

African journal of paediatric surgery : AJPS. 13 (4) (pp 193-195), 2016. Date of Publication: 01 Oct 2016.

[Article]

AN: 618115491

BACKGROUND: In parts of Africa, routine circumcision is practised and sometimes even on children with hypospadias. The lack of preputial foreskin renders urethroplasty more difficult and often requires to use of a mucosal graft as described by Bracka. OBJECTIVE: The authors describe their experience of hypospadias repair using Bracka's technique.

MATERIALS AND METHODS: Over a period of 5 years, 100 cases of proximal hypospadias were operated in our institution. All patients aged 0-18 years who had already been circumcised were included in this study.

RESULTS: The outcome of the 12 cases operated according to Bracka's technique was analysed. The mean age was 11.5 years. The ectopic meatus was penoscrotal in three cases, scrotal in one case and perineal in eight cases. After reconstruction, the new meatus was sutured at the top of the glans in one case, at the prepuce in seven cases and at the penile midshaft in one case. The main complications noted were surgical site infection, wound dehiscence, residual chordee and urethrocutaneous fistula. No neourethral stenosis nor urethrocele was recorded.

DISCUSSION: The buccal mucosal graft urethroplasty as described by Bracka is associated with a lower risk of meatal strictures compared to other free mucosal grafts. The buccal mucosa is easier to harvest and causes less scarring than bladder mucosa.

CONCLUSION: Repair of severe hypospadias remains a challenge for paediatric surgeons. The functional and cosmetic outcomes depend on the choice of the donor site for the graft and objective assessment of successful reconstruction criteria during follow-up.

PMID

28051050 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=28051050>]

Institution

(Faustin Felicien, Nwaha Makon, Kamadjou, Fossi, Andze) Yaounde Gynaeco-obstetric and Paediatric Hospital, Cameroon (Le Coultre) University of Geneva Teaching Hospital, Switzerland

(Sosso) Department of Surgery and Subspecialties, Faculty of Medicine and Biomedical Sciences, University of Yaounde I, Cameroon

(Mure) Lyon Teaching Hospital, France

Year of Publication

2016

769.

Penile curvature: an update for management from 20 years experience in a high volume centre.

Sasso F., Vittori M., D'Addessi A., Bassi P.F.

Embase

Urologia. 83 (3) (pp 130-138), 2016. Date of Publication: 26 Sep 2016.

[Review]

AN: 617858109

Our aim was to review the literature and discuss about penile curvature in order to have an update for management after 20 years experience in the field. Penile curvature may be congenital or acquired. Congenital penile curvature is a relatively uncommon condition that may present in late adolescent or early adult life. The incidence is estimated to be 0.6 %. On the other side, acquired penile curvature has an overall prevalence of 0.5-13%. Three main factors seem to increase the risk of developing an acquired penile curvature, often related to Peyronie's disease: penile traumatism, genetic and familiar conditions and a history of diseases of the genital tract. In treating Peyronie's disease, no medical therapy is fully effective, and surgery remains the gold standard in cases of severe deformity and/or erectile dysfunction. Peyronie's disease is associated with significant psychological stress for patients and their partners. Appropriate treatment should be individualized and tailored to the patient's goals and expectations. There is not the 'best' surgical technique and outcomes are satisfactory when proper treatment decisions are made.

PMID

27103093 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=27103093>]

Institution

(Sasso, Vittori, D'Addessi, Bassi) Agostino Gemelli Teaching Hospital Foundation, Catholic University School of Medicine, Rome - Italy

Year of Publication

2016

770.

Modification of the Koyanagi Technique for the Single-Stage Repair of Proximal Hypospadias.

Chen Y., Zhang J., Ji C., Liang W., Pan S., Wu B.

Embase

Annals of plastic surgery. 76 (6) (pp 693-696), 2016. Date of Publication: 01 Jun 2016.

[Article]

AN: 617855112

We describe a modification of the Koyanagi technique for hypospadias in an attempt to reduce complications and improve results. Between January 2005 and July 2012, 21 patients underwent treatment of proximal hypospadias using a modified hypospadias repair. The procedure involved making a U-incision similar to that in a Thiersch-Duplay repair to preserve the blood supply to the tubular neourethra. The neourethra was reconstructed by island penile skin. An M-incision was made in the glans, and the meatus was attached to tip of glans. Flaps were harvested from both sides of the penis preventing postoperative penile turn. Cosmetic and functional, long-term (mean, 12 months) recovery was excellent. Complications consisted of 3 cases of urethrocutaneous fistula (14.2%), which were corrected surgically, and 2 cases of penile skin necrosis. There were no instances of meatal stenosis, diverticulum, or urethral stricture. Bifid scrotum was present in 6 patients, and associated penoscrotal transposition was present in 15 patients. Both of these associated defects were corrected at the time of surgery. The modified technique permits 1-stage repair of proximal hypospadias with a low complication rate, the satisfactory recovery.

PMID

27176562 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=27176562>]

Institution

(Chen) From the Department of Plastic and Reconstruction Surgery, Sun Yat-sen Memorial Hospital, Sun Yat-sen University, Guangzhou, China

Year of Publication

2016

771.

A wet dressing for male genital surgery: A phase II clinical trial.

Vilar F.O., Pinto F.C., Albuquerque A.V., Martins A.G., Araujo L.A., Aguiar J.L., Lima S.V.

Embase

International braz j urol : official journal of the Brazilian Society of Urology. 42 (6) (pp 1220-1227), 2016. Date of Publication: 01 Nov 2016.

[Article]

AN: 617651105

PURPOSE: This study was to confirm the safety and efficacy of BC dressing when used in surgical male wound healing at the urogenital area. METHODS: Open, non-controlled clinical study of phase II. A total of 141 patients, among those children, adolescents and adults with hypospadias (112), epispadias (04), phimosis (13) and Peyronie's disease (12) that had a BC dressing applied over the operated area after surgery. A written informed consent was obtained from all participants. Study exclusion criteria were patients with other alternative treatment indications due to the severity, extent of the injury or the underlying disease. The outcomes evaluated were efficacy, safe and complete healing. The costs were discussed.

RESULTS: In 68% patients, the BC dressing fell off spontaneously. The BC was removed without complications in 13% of patients at the outpatient clinic during the follow-up visit and 17% not reported the time of removal. In 3% of the cases, the dressing fell off early. Complete healing was observed between 8th and 10th days after surgery. The BC dressings have shown a good tolerance by all the patients and there were no reports of serious adverse events.

CONCLUSION: The bacterial cellulose dressings have shown efficacy, safety and that can be considered as a satisfactory alternative for postoperative wound healing in urogenital area and with low cost.

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PMID

27649111 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=27649111>]

Institution

(Vilar, Lima) Servico de Urologia do Hospital das Clinicas, Departamento de Cirurgia do Centro de Ciencias da Saude da Universidade Federal de Pernambuco, UFPE, Brasil (Pinto,

Albuquerque, Aguiar) Departamento de Cirurgia do Centro de Ciencias da Saude da
Universidade Federal de Pernambuco, UFPE, Brasil
(Martins, Araujo) Servico de Cirurgia Pediatrica do Hospital das Clinicas, Departamento de
Cirurgia do Centro de Ciencias da Saude da Universidade Federal de Pernambuco, UFPE, Brasil
Year of Publication
2016

772.

Craniofacial anomalies associated with hypospadias. Description of a hospital based population
in South America.

Fernandez N., Escobar R., Zarante I.

Embase

International braz j urol : official journal of the Brazilian Society of Urology. 42 (4) (pp 793-797),
2016. Date of Publication: 01 Jul 2016.

[Article]

AN: 617370339

INTRODUCTION: Hypospadias is a congenital abnormality of the penis, in which there is incomplete development of the distal urethra. There are numerous reports showing an increase of prevalence of hypospadias. Association of craniofacial malformations in patients diagnosed with hypospadias is rare. The aim of this study is to describe the association between hypospadias and craniofacial congenital anomalies. MATERIALS AND METHODS: A retrospective review of the Latin-American collaborative study of congenital malformations (ECLAMC) data was performed between January 1982 and December 2011. We included children diagnosed with associated hypospadias and among them we selected those that were associated with any craniofacial congenital anomaly.

RESULTS: Global prevalence was 11.3 per 10.000 newborns. In this population a total of 809 patients with 1117 associated anomalies were identified. On average there were 1.7 anomalies per patient. Facial anomalies were present in 13.2%. The most commonly major facial anomaly associated to hypospadias was cleft lip/palate with 52 cases. We identified that 18% have an association with other anomalies, and found an association between craniofacial anomalies and hypospadias in 0.59 cases/10.000 newborns.

DISCUSSION: Hypospadias is the most common congenital anomaly affecting the genitals. Its association with other anomalies is rare. It has been reported that other malformations occur in 29.3% of the cases with hypospadias. The more proximal the meatus, the higher the risk for having another associated anomaly.

CONCLUSION: Associated hypospadias are rare, and it is important to identify the concurrent occurrence of craniofacial anomalies to better treat patients that might need a multidisciplinary approach.

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PMID

27564292 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=27564292>]

Institution

(Fernandez) Hospital Universitario San Ignacio Pontificia Universidad Javeriana - Urologia - Genetica Bogota, Colombia (Escobar) Universidad del Rosario - Epidemiologia, Bogota, Colombia

(Zarante) Hospital Universitario San Ignacio Pontificia Universidad Javeriana - Genetica, Bogota, Colombia

Year of Publication

2016

773.

Mixed Gonadal Dysgenesis with an unusual "inverted" Y chromosome.

Makhija D., Shah H., Tiwari C., Jayaswal S., Desale J.

Embase

Developmental period medicine. 20 (3) (pp 178-180), 2016. Date of Publication: 01 Jan 2016.

[Article]

AN: 616855863

Mixed gonadal dysgenesis is a rare disorder of sex development associated with sex chromosome aneuploidy and mosaicism of the Y chromosome. It is characterized by a unilateral non-palpable (usually intra-abdominal) testis, a contralateral streak gonad and persistent mullerian structures. The clinical presentation can vary from a typical male to female phenotype including all degrees of cryptorchidism, labial fusion, clitoromegaly, epispadias and hypospadias.

It is the second most common cause of ambiguous genitalia in the neonatal period. We report a case of Mixed Gonadal Dysgenesis with an inverted Y chromosome.

PMID

27941186 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=27941186>]

Institution

(Makhija) Dept of Paediatric Surgery, TNMC & BYL Nair Hospital, Mumbai Central, Mumbai, Maharashtra. India, E-Mail ID: deepee.sweety@gmail.com (Shah, Tiwari, Jayaswal, Desale)
Dept of Paediatric Surgery, TNMC & BYL Nair Hospital, Mumbai Central, Mumbai, Maharashtra.
India

Year of Publication

2016

774.

Clinical Study of 23 Male Patients with Congenital Ventral Penile Angulation without Hypospadias.

Patoulas I., Patoulas D., Farmakis K., Kalogirou M.

Embase

Acta medica (Hradec Kralove). 59 (4) (pp 113-116), 2016. Date of Publication: 01 Jan 2016.

[Article]

AN: 616705376

Congenital ventral penile angulation without hypospadias is a rare disease and causes great anxiety to the parents. The aim of our study is the presentation of this disease, especially the indications of surgical treatment and the protocol applied in our clinic. We retrospectively studied 23 male patients aged 2.5 to 7 years old (av 5.2 y) with important penile angulation (over 45degree) without hypospadias, treated during the past 15 years in our department. In 9 patients the cause was the skin chordee (fibrosis of the ventral part of the prepuce), in 4 the fibrotic fascia (incomplete development of dartos and Buck's fascia) and in 10 the disproportion of the corpora cavernosa. No case of congenital short urethra was reported. In our opinion, the appliance of the algorithm suggested by Donnahoo KK et al. in uncomplicated cases, along with the experience of the surgical team, results in satisfactory treatment and avoidance of complications.

PMID

28440213 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=28440213>]

Institution

(Patoulas, Patoulas, Farmakis, Kalogirou) 1st Department of Pediatric Surgery, Aristotle University of Thessaloniki, GH G. Gennimatas, Thessaloniki, Greece

Year of Publication

2016

775.

Results of a Two-stage Technique for Treatment of Proximal Hypospadias with Severe Curvature: Creation of a Urethral Plate Using a Vascularized Preputial Island Flap.

Ozcan R., Emre S., Kendigelen P., Elicevik M., Emir H., Soylet Y., Buyukunal S.N.

Embase

Urology journal. 13 (2) (pp 2629-2634), 2016. Date of Publication: 16 Apr 2016.

[Article]

AN: 616599529

MATERIALS AND METHODS: The medical records of children with proximal hypospadias and severe curvature were retrospectively analyzed. A 2-stage procedure was performed in 30 children. In the first stage, the release of chordee was performed, and a well-vascularized preputial island flap was created. The vascularized island flap was brought anteriorly and sutured over the ventral surface of the glans and degloved penile shaft. The second stage was performed 6-8 months later. A neourethra was reconstructed by the tubularization of the preputial-urethral plate utilizing the principles of Duplay technique. All surgical procedures were performed between 2005 and 2011. **RESULTS:** The mean age of the patients was 4.4 years (1-17 years). The mean duration of urethral catheterization was 6 days after the first stage and 10 days following the second stage. The flaps were viable in all of the children. There was no residual chordee. Following the second stage (n = 30), complications developed in 11 children (36%), namely, a fistula in 7, a pinpoint fistula in 3, and a diverticulum formation in 1. The cosmetic outcome was satisfactory. Uroflowmetry measurements were evaluated, and only one patient had a diverticulum formation at the late follow-up.

CONCLUSION: Vascularized preputial island flap is an alternative to free grafts for the reconstruction of the urethra. The main advantage of this flap technique is the creation of a thick, healthy and well-vascularized urethral plate. The advantages of this technique include better

aesthetic appearance, an acceptable complication rate, and a very low rate of diverticula formation.

PURPOSE: To present the results of a two-stage technique used for the treatment of proximal hypospadias with severe curvature.

PMID

27085564 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=27085564>]

Institution

(Ozcan, Emre, Elicevik, Emir, Soylet, Buyukunal) Department of Pediatric Surgery, Division of Pediatric Urology, Istanbul University, Cerrahpasa Medical Faculty, Istanbul, 34098, Turkey

(Kendigelen) Department of Pediatric Anesthesiology, Istanbul University, Cerrahpasa Medical Faculty, Istanbul, 34098, Turkey

Year of Publication

2016

776.

Technical refinements to improve outcomes following distal hypospadias repair.

Lacy J.M., Hendrix L.N., Bole R., Habib E., Wootton C.W., Ziada A.M.

Embase

The Canadian journal of urology. 23 (1) (pp 8184-8187), 2016. Date of Publication: 01 Feb 2016.

[Article]

AN: 616095605

INTRODUCTION: Hypospadias complications, most notably meatal stenosis, are commonly reported to occur after tubularized incised plate (TIP) hypospadias repair. We focus on a point of technique in TIP repair and its effect on outcome of this possible complication, as well as other commonly reported complications. Meatal stenosis after TIP can be avoided if the urethra and overlying glans are dissected and sutured separately with no attempt at cross suturing whether the urethra ends below, behind, or above the glans sutures. This hypothesis was evaluated by a prospective data collection before and after implementation to evaluate the effect of a technical refinement on rates of meatal stenosis in TIP hypospadias repair. **MATERIALS AND**

METHODS: All cases of coronal to midpenile hypospadias repair during two periods were included in our study. Group 1 included 140 consecutive patients over a 30 month period. Group 2 included 122 consecutive patients over a 36 month period during which the above mentioned

technical changes were implemented by all participating pediatric urologists. Rates of complications between the two groups were compared with special emphasis on meatal stenosis.

RESULTS: Median follow up for both groups was > 1 year. Overall complication rate in Group 1 was 31.5% compared to 9.8% in Group 2. Meatal stenosis was significantly reduced from 13 patients (9.3%) in Group 1 to 2 patients (1.6%) in Group 2, $p = 0.008$.

CONCLUSION: The technical refinements described resulted in reduction of complication rates and a decrease in incidence of meatal stenosis.

PMID

26892064 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=26892064>]

Institution

(Lacy) Department of Urology, University of Kentucky Medical Center, Lexington, Kentucky, USA

Year of Publication

2016

777.

Safety of Tioguanine During Pregnancy in Inflammatory Bowel Disease.

van den Berg S.A., de Boer M., van der Meulen-de Jong A.E., Jansen J.M., Hoentjen F., Russel M.G., Mahmmod N., van Bodegraven A.A., van der Woude C.J., Mulder C.J., de Boer N.K.

Embase

Journal of Crohn's & colitis. 10 (2) (pp 159-165), 2016. Date of Publication: 01 Feb 2016.

[Article]

AN: 615867227

BACKGROUND AND AIMS: Conventional thiopurine [azathioprine and mercaptopurine] treatment during pregnancy in patients with inflammatory bowel disease [IBD] is considered to be safe; however data on the safety and teratogenicity of the non-conventional thiopurine tioguanine [TG] in pregnant IBD patients are lacking. We aim to describe the safety and teratogenicity of TG treatment during pregnancy in IBD patients. METHODS: This was a retrospective, multicentre descriptive case series of female IBD patients using TG during pregnancy. Data on disease and medication history, pregnancy complications, pregnancy outcome, mode of delivery, preterm birth, birthweight, congenital abnormalities, laboratory signs of myelosuppression or hepatotoxicity, and 6-thioguaninenucleotide [6-TGN] concentrations in mother and neonate were collected.

RESULTS: In all, 13 patients [77% Crohn's disease, 23% ulcerative colitis] used TG [median dose 18 g/day] during pregnancy; 19 pregnancies, including 1 twin pregnancy, were included. Spontaneous abortion occurred in three pregnancies. In 7 of the 16 ongoing pregnancies a caesarean section was performed. One neonate had a mild congenital abnormality [distal shaft hypospadias]. In the singleton pregnancies, the median birthweight was 3410 g at a median of gestational age of 39 weeks. No preterm birth [< 37 weeks] or low birthweight [< 2500 g] was observed in the singleton newborns. In the twin pregnancy an induction of labour was performed at 35 + 1 weeks of gestation because of pre-eclampsia. Both neonates had a low birthweight.

CONCLUSIONS: This relatively small case series supports safe use of TG in pregnant IBD patients. Still, consideration should be given to the indication and continuation of TG during pregnancy.

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PMID

26503525 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=26503525>]

Institution

(van den Berg, Mulder) Department of Gastroenterology and Hepatology, VU University Medical Center, Amsterdam, The Netherlands (de Boer) Department of Obstetrics and Gynaecology, VU University Medical Center, Amsterdam, The Netherlands

(van der Meulen-de Jong) Department of Gastroenterology and Hepatology, Leiden University Medical Center, Leiden, The Netherlands

(Jansen) Department of Gastroenterology and Hepatology, Onze Lieve Vrouwe Gasthuis Amsterdam, Amsterdam, The Netherlands

(Hoentjen) Department of Gastroenterology and Hepatology, Radboud University Medical Center, Nijmegen, The Netherlands

(Russel) Department of Gastroenterology and Hepatology, Medical Spectrum Twente, Enschede, The Netherlands

(Mahmmod) Department of Gastroenterology and Hepatology, St Antonius Hospital, Nieuwegein, The Netherlands

(van Bodegraven) Department of Gastroenterology and Hepatology, VU University Medical Center, Amsterdam, The Netherlands Department of Internal Medicine, Geriatrics and Gastroenterology, Orbis Medical Center, Sittard-Geleen, The Netherlands

(van der Woude) Department of Gastroenterology and Hepatology, Erasmus Medical Center, Rotterdam, The Netherlands

(de Boer) Department of Gastroenterology and Hepatology, VU University Medical Center, Amsterdam, The Netherlands khn.deboer@vumc.nl

Year of Publication
2016

778.

The Ideal Use of Catheters in Hypospadias Repair: An Experimental Study.

Polat H., Gulacti U.

Embase

Urology journal. 13 (5) (pp 2856-2859), 2016. Date of Publication: 10 Oct 2016.

[Article]

AN: 615596827

PURPOSE: To find answers to some catheter-related questions in hypospadias repair such as which type of catheter should be used, with which catheter balloon inflation volume, and when should the catheter be removed from the urethra? As catheter use and post-op retention time varies among surgeons in hypospadias repair. **MATERIAL AND METHODS:** Fifty-four 10 French all-silicone- and 54 latex Foley catheters were prepared and assigned to groups as senary. The catheter's balloons were inflated with 2, 3 and 5 mL of sterile water. The catheters were submerged in artificial human urine and then removed from the solution at 24, 72, and 168 h after submersion. The catheter balloon volume losses, increases in the transverse diameter of the catheters, and angulation of the catheter tips were measured to determine catheter degradation. **RESULTS:** The minimum balloon volume loss was 0.4 mL in the group of all-silicone catheters that were inflated with 2mL and deflated after 24h (2mL 24h). According to balloon volume and deflation time, there were no increases in transverse diameter of the four groups of all-silicone catheters; 2mL 24h, 3mL 24h, 5mL 24h, and 2mL 72h. With 1 mm expansion, the lowest increase on transverse diameter of the latex catheters occurred in five groups; 2mL 24h, 3mL 24h, 5mL 24h, 2mL 72h, and 2mL 168h.

CONCLUSION: An all-silicone catheter inflated with 2mL and removed from the urethra within 24-72 hours may be the ideal catheter use in hypospadias repair.

PMID

27734429 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=27734429>]

Institution

(Polat) Adiyaman University Medical Faculty, Department of Urology, Adiyaman, Turkey.

(Gulacti) Adiyaman University Medical Faculty, Department of Emergency, Adiyaman, Turkey

Year of Publication
2016

779.

Factors affecting post-pubertal penile size in patients with hypospadias.

Moriya K., Nakamura M., Nishimura Y., Kitta T., Kanno Y., Chiba H., Kon M., Shinohara N.

Embase

World journal of urology. 34 (9) (pp 1317-1321), 2016. Date of Publication: 01 Sep 2016.

[Article]

AN: 615595342

OBJECTIVES: To evaluate actual post-pubertal penile size and factors affecting it in hypospadias patients, we retrospectively reviewed medical charts. PATIENTS AND METHODS: Hypospadias patients whose external genitalia were categorized into Tanner stage 5, and whose stretched penile length was evaluated at 15 years old or older from April 2008 to April 2015, were enrolled in the present study. Stretched penile length was measured by a single examiner. Actual post-pubertal stretched penile length and factors affecting the post-pubertal stretched penile length were estimated. Statistical analysis was performed using Mann-Whitney U test and univariate and multivariate linear regression models for the determination of independent factors.

RESULTS: Thirty patients met the inclusion criteria. Median age at evaluation was 17.2 years. Thirteen and 17 had mild and severe hypospadias, respectively. Endocrinological abnormality was identified in 5. Multivariate analysis showed that the severity of hypospadias and endocrinological abnormality were significant factors affecting stretched penile length. Stretched penile length in 25 patients without endocrinological abnormality was significantly longer than that in those with endocrinological abnormality ($p = 0.036$). Among patients without endocrinological abnormality, stretched penile length in 13 with severe hypospadias was significantly shorter than that in 12 with mild hypospadias ($p = 0.004$).

CONCLUSIONS: While the severity of hypospadias and endocrinological abnormality at post-pubertal evaluation were factors affecting post-pubertal penile size, stretched penile length in patients with severe hypospadias was shorter even in cases without endocrinological abnormality. These results suggest that severe hypospadias is not only a disorder of urethral development, but also a disorder of penile development.

PMID

26792579 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=26792579>]

Institution

(Moriya, Nakamura, Nishimura, Kitta, Kanno, Chiba, Kon, Shinohara) Department of Renal and Genitourinary Surgery, Hokkaido University Graduate School of Medicine, North-15, West-7, Kita-ku, Sapporo, 060-0824, Japan

Year of Publication

2016

780.

Transverse preputial onlay island flap urethroplasty for single-stage correction of proximal hypospadias.

Singal A.K., Dubey M., Jain V.

Embase

World journal of urology. 34 (7) (pp 1019-1024), 2016. Date of Publication: 01 Jul 2016.

[Article]

AN: 615394558

AIMS AND OBJECTIVES: Transverse preputial onlay island flap urethroplasty (TPOIF) was described initially for distal hypospadias, but has seen extended application for proximal hypospadias. We describe a set of modifications in the technique and results in a large series of proximal hypospadias. **MATERIALS AND METHODS:** All children who underwent TPOIF repair for proximal hypospadias (proximal penile, penoscrotal and scrotal) from June 2006 to June 2013 by a single surgeon were prospectively followed till June, 2014. A standard technique and postoperative protocol were followed. Salient points to be emphasized in the technique: (1) dissection of the dartos pedicle till penopubic junction to prevent penile torsion, (2) incorporation of the spongiosum in the urethroplasty, (3) midline urethral plate incision in glans (hinging the plate), (4) Dartos blanket cover on whole urethroplasty.

RESULTS: Out of 136 children with proximal hypospadias, 92 children who underwent TPOIF formed the study group. Out of 92 children, 48 (52 %) children required a tunica albuginea plication for chordee correction. In total, 16 (17 %) patients developed 24 complications and 11 children (12 %) required second surgeries: fistula closure in 7 (with meatoplasty in 5), glansplasty for glans dehiscence in 2 and excision of diverticulum in 2. Two children required a third surgery.

Only 5 children had a noticeable penile torsion (less than 30 degree), and 7 had a patulous meatus.

CONCLUSIONS: Transverse preputial onlay island flap urethroplasty can deliver reliable cosmetic and functional outcomes in proximal hypospadias.

PMID

26394625 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=26394625>]

Institution

(Singal) MITR Hospital, Kharghar, Navi Mumbai, India. arbinders@gmail.com (Singal)

Hypospadias Foundation, Kharghar, Navi Mumbai, India. arbinders@gmail.com

(Singal) Division of Pediatric Urology, MGM Institute of Health Sciences, Kamothe, Maharashtra, India. arbinders@gmail.com

(Singal) One Vision Health and Research Foundation, Navi Mumbai, India. arbinders@gmail.com

(Dubey) MITR Hospital, Kharghar, Navi Mumbai, India

(Dubey, Jain) Hypospadias Foundation, Kharghar, Navi Mumbai, India

(Jain) One Vision Health and Research Foundation, Navi Mumbai, India

(Jain) Case Western Reserve (MetroHealth) Program, Department of Pediatrics, Case Western Reserve University, Cleveland, OH, USA

Year of Publication

2016

781.

Outcomes of Patients with Glanular Hypospadias or Dorsal Hood Deformity with Mild Chordee Treated by Modified Firlit's Technique.

Alizadeh F., Shirani S.

Embase

Urology journal. 13 (6) (pp 2908-2910), 2016. Date of Publication: 08 Dec 2016.

[Article]

AN: 615393907

PURPOSE: To assess the success and complication rates of glanular hypospadias or dorsal hood deformity surgery, using a modified Firlit's technique without glanuloplasty. MATERIALS AND METHODS: Between May 2013 and December 2015, 41 patients with glanular hypospadias or dorsal hood deformity without hypospadias and mild ventral chordee underwent surgery. Thirty-

eight who completed the 1 week and 6-month follow-up were retrospectively evaluated. Exclusion criteria were complete absence of corpus spongiosum resulting in very thin distal urethra, moderate to severe ventral chordee or deep urethral plate that seemed to be better served by tubularization techniques. Modified Firlit's technique (a submeatal inverted V incision in addition to the classic technique) was applied to all patients.

RESULTS: The mean age was 20.4 $\pm$ 13 months (range: 6-52 months). The only complication was a narrow-band ventral skin necrosis in 1 patient (2.6%) that was replaced by new skin growth without need for further intervention. No other complications including unresolved chordee, urethrocutaneous fistula, meatal stenosis, hematoma, infection or post-operative bleeding was observed. Defining the satisfactory result as the glanular position of the meatus and the absence of residual chordee, all patients had satisfactory outcome.

CONCLUSION: Modified Firlit's technique is a simple method with excellent cosmetic results and low complication rate that avoids unnecessary glans dissections. When prepuceplasty is not desired, this technique can be considered as a viable option.

PMID

27928812 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=27928812>]

Institution

(Alizadeh, Shirani) Isfahan Kidney Transplantation Research Center. Isfahan University of Medical Sciences. Department of Urology. Al-Zahra Hospital. Isfahan, Iran

Year of Publication

2016

782.

THE TREATMENT OF INTERSEX AND THE PROBLEM OF DELAY: THE AUSTRALIAN SENATE INQUIRY INTO INTERSEX SURGERY AND CONFLICTING HUMAN RIGHTS FOR CHILDREN.

O'Connor M.

Embase

Journal of law and medicine. 23 (3) (pp 531-543), 2016. Date of Publication: 01 Mar 2016.

[Article]

AN: 613677967

When a child is born with indeterminate genitalia (so-called intersex or disordered sex development), it becomes very difficult to balance the child's right to determine their own sexual future against the problems of living as a child with an indeterminate gender. Moreover, the initial assignment of gender may prove to be inappropriate and major psychological disturbances in the recipient can arise during adolescence and adult life. The problems of these children were explained to the Australian Senate Committee during its inquiry into intersex surgery in 2013. As a result, the Committee made a number of recommendations, including a proposal that all surgery be deferred until the child is able to consent to treatment. The author argues that the Committee's proposal to delay all modifications of indeterminate genitalia is impractical. The inclusion in the definition of intersex of common conditions (such as hypospadias in genetic male infants) means that necessary and uncontroversial surgery will be delayed until after puberty. This delay may be harmful and adverse to some children's best interests.

PMID

27323632 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=27323632>]

Year of Publication

2016

783.

Correcting Glanular Dislocation in Hypospadias With Up-to-Tip Technique and Double Wing Flaps.

Ji C., Liang W., Pan S., Chen Y., Zhang J., Yao Y.

Embase

Annals of plastic surgery. 76 (1) (pp 102-107), 2016. Date of Publication: 01 Jan 2016.

[Article]

AN: 613498573

BACKGROUND: The surgical treatment of hypospadias contains urethral reconstruction and correction of penile curvature. In some severe type of hypospadias, it also needs correction of penoscrotal transposition and cleft of scrotum. However, there are few literatures that refer to the correcting of deformity of glanular dislocation. OBJECTIVE: This study aimed to correct the deformity of glanular dislocation in patients with hypospadias with up-to-tip technique and double wing flaps.

METHODS: From May of 2011 to September of 2012, 49 patients who underwent hypospadias with glanular dislocation repairing received up-to-tip technique to correct the deformity of glanular dislocation in our department by a single surgeon (J.Z.). An X-shaped incision was made in the ventral median of glans, and the glans was dissected from the penis cavernosus, then 2 wing-shaped flaps appeared. The 2 sides of flaps were rotated and lifted with new urethra inside. The divergent glans flaps were closed with vertical mattress suture. Follow-up was from 12 to 30 months.

RESULTS: All patients had excellent cosmetic results immediately after the operation. The glans was straight and was upturned obviously. The wing flaps of glans were partially dehiscent in 2 (4.08%) cases after the operation. The other patients had good wound healing. No urethral stricture in distal urethra was observed.

CONCLUSIONS: Up-to-tip technique and double wing flap can effectively correct the deformity of glanular dislocation in the patients with hypospadias. After the operation, the glans configuration was improved.

PMID

25003422 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=25003422>]

Institution

(Ji, Zhang, Liang, Pan, Chen, Zhang, Yao) From the Department of Plastic Surgery, Sun Yat-sen Memorial Hospital, Sun Yat-sen University, Guangzhou City, Guangdong Province, China

Year of Publication

2016

784.

Human anogenital distance: An update on fetal smoke-exposure and integration of the perinatal literature on sex differences.

Fowler P.A., Filis P., Bhattacharya S., Le Bizec B., Antignac J.-P., Morvan M.-L., Drake A.J., Soffientini U., O'Shaughnessy P.J.

Embase

Human Reproduction. 31 (2) (pp 463-472), 2016. Date of Publication: 24 May 2016.

[Article]

AN: 608914350

STUDY QUESTION Do sex and maternal smoking effects on human fetal anogenital distance (AGD) persist in a larger study and how do these data integrate with the wider literature on perinatal human AGD, especially with respect to sex differences? **SUMMARY ANSWER** Second trimester sex differences in AGD are broadly consistent with neonatal and infant measures of AGD and maternal cigarette smoking is associated with a temporary increase in male AGD in the absence of changes in circulating testosterone. **WHAT IS KNOWN ALREADY** AGD is a biomarker of fetal androgen exposure, a reduced AGD in males being associated with cryptorchidism, hypospadias and reduced penile length. Normative fetal AGD data remain partial and windows of sensitivity of human fetal AGD to disruption are not known. **STUDY DESIGN, SIZE, DURATION** The effects of fetal sex and maternal cigarette smoking on the second trimester (11-21 weeks of gestation) human fetal AGD were studied, along with measurement of testosterone and testicular transcripts associated with apoptosis and proliferation. **PARTICIPANTS/MATERIALS, SETTING METHODS** AGD, measured from the centre of the anus to the posterior/caudal root of penis/clitoris (AGDapp) was determined in 56 female and 70 male morphologically normal fetuses. These data were integrated with current literature on perinatal AGD in humans. **MAIN RESULTS AND THE ROLE OF CHANCE** At 11-13 weeks of gestation male fetal AGDapp was 61% ($P < 0.001$) longer than in females, increasing to 70% at 17-21 weeks. This sexual dimorphism was independent of growth characteristics (fetal weight, length, gonad weight). We confirmed that at 14-16 weeks of gestation male fetal AGDapp was increased 28% ($P < 0.05$) by in utero cigarette smoke exposure. Testosterone levels were not affected by smoking. To develop normative data, our findings have been integrated with available data from in vivo ultrasound scans and neonatal studies. Inter-study variations in male/female AGD differences lead to the conclusion that normalization and standardization approaches should be developed to enable confidence in comparing data from different perinatal AGD studies. **LIMITATIONS, REASONS FOR CAUTION** Sex differences, and a smoking-dependent increase in male fetal AGD at 14-16 weeks, identified in a preliminary study, were confirmed with a larger number of fetuses. However, human fetal AGD should be re-assessed once much larger numbers of fetuses have been studied and this should be integrated with more detailed analysis of maternal lifestyle. Direct study of human fetal genital tissues is required for further mechanistic insights. **WIDER IMPLICATIONS OF THE FINDINGS** Fetal exposure to cigarette smoke chemicals is known to lead to reduced fertility in men and women. Integration of our data into the perinatal human AGD literature shows that more work needs to be done to enable reliable inter-study comparisons. **STUDY FUNDING/COMPETING INTEREST(S)** The study was supported by grants from the Chief Scientist Office (Scottish Executive, CZG/1/109 & CZG/4/742), NHS Grampian Endowments (08/02), the European Community's Seventh Framework Programme (FP7/2007-2013) under grant agreement no 212885 and the Medical Research Council, UK

(MR/L010011/1). The authors declare they have no competing interests, be it financial, personal or professional.

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PMID

26732622 [<http://www.ncbi.nlm.nih.gov/pubmed/?term=26732622>]

Status

Embase

Institution

(Fowler, Filis) Institute of Medical Sciences, University of Aberdeen, Foresterhill, Aberdeen AB25 2ZD, United Kingdom (Bhattacharya) Institute of Applied Health Sciences, University of Aberdeen, Foresterhill, Aberdeen AB25 2ZD, United Kingdom

(Le Bizec, Antignac, Morvan) USC INRA 1329, Laboratoire d'Etude des Residus et Contaminants dans les Aliments, LUNAM Universite, Oniris, Nantes F-44307, France

(Drake) Endocrinology Unit, Queen's Medical Research Institute, University/BHF Centre for Cardiovascular Science, University of Edinburgh, 47 Little France Crescent, Edinburgh EH16 4TJ, United Kingdom

(Soffientini, O'Shaughnessy) Institute of Biodiversity, Animal Health and Comparative Medicine (IBAHCM), College of Medical, Veterinary and Life Sciences, University of Glasgow, Bearsden Rd, Glasgow G61 1QH, United Kingdom

Publisher

Oxford University Press

Year of Publication

2016

785.

Progestogen for preventing miscarriage in women with recurrent miscarriage of unclear etiology
Haas, David M. Hathaway, Taylor J. Ramsey, Patrick S. Institution David MHaas. TI Progestogen for preventing miscarriage in women with recurrent miscarriage of unclear etiology.

EBM Reviews - Cochrane Database of Systematic Reviews

Cochrane Database of Systematic Reviews. 11, 2019.

[Systematic Review]

AN: 00075320-100000000-02431

Background

Progesterone, a female sex hormone, is known to induce secretory changes in the lining of the uterus essential for successful implantation of a fertilized egg. It has been suggested that a causative factor in many cases of miscarriage may be inadequate secretion of progesterone. Therefore, clinicians use progestogens (drugs that interact with the progesterone receptors), beginning in the first trimester of pregnancy, in an attempt to prevent spontaneous miscarriage. This is an update of a review, last published in 2013.

Since publication of the 2018 update of this review, we have been advised that the Ismail 2017 study is currently the subject of an investigation by the Journal of Maternal-Fetal & Neonatal Medicine. We have now moved this study from 'included studies' to 'Characteristics of studies awaiting classification' until the outcome of the investigation is known.

Objectives

To assess the efficacy and safety of progestogens as a preventative therapy against recurrent miscarriage.

Search methods

For this update, we searched Cochrane Pregnancy and Childbirth's Trials Register, the WHO International Clinical Trials Registry Platform () (6 July 2017) and reference lists from relevant articles, attempting to contact trial authors where necessary, and contacted experts in the field for unpublished works.

Selection criteria

Randomized or quasi-randomized controlled trials comparing progestogens with placebo or no treatment given in an effort to prevent miscarriage.

Data collection and analysis

Two review authors independently assessed trials for inclusion and risk of bias, extracted data and checked them for accuracy. Two reviewers assessed the quality of the evidence using the GRADE approach.

Main results

Twelve trials (1,856 women) met the inclusion criteria. Eight of the included trials compared treatment with placebo and the remaining four trials compared progestogen administration with no treatment. The trials were a mix of multicenter and single-center trials, conducted in India, Jordan, UK and USA. In five trials women had had three or more consecutive miscarriages and in seven trials women had suffered two or more consecutive miscarriages. Routes, dosage and duration of progestogen treatment varied across the trials. The majority of trials were at low risk of bias for most domains. Ten trials (1684 women) contributed data to the analyses.

The meta-analysis of all women, suggests that there may be a reduction in the number of miscarriages for women given progestogen supplementation compared to placebo/controls

(average risk ratio (RR) 0.73, 95% confidence interval (CI) 0.54 to 1.00, 10 trials, 1684 women, moderate-quality evidence). A subgroup analysis comparing placebo-controlled versus non-placebo-controlled trials, trials of women with three or more prior miscarriages compared to women with two or more miscarriages and different routes of administration showed no clear differences between subgroups for miscarriage.

None of the trials reported on any secondary maternal outcomes, including severity of morning sickness, thromboembolic events, depression, admission to a special care unit, or subsequent fertility.

There was probably a slight benefit for women receiving progestogen seen in the outcome of live birth rate (RR 1.07, 95% CI 1.00 to 1.13, 6 trials, 1411 women, moderate-quality evidence). We are uncertain about the effect on the rate of preterm birth because the evidence is very low-quality (RR 1.13, 95% CI 0.53 to 2.41, 4 trials, 256 women, very low-quality evidence). No clear differences were seen for women receiving progestogen for the other secondary outcomes including neonatal death, fetal genital abnormalities or stillbirth. There may be little or no difference in the rate of low birthweight and trials did not report on the secondary child outcomes of teratogenic effects or admission to a special care unit.

Authors' conclusions

For women with unexplained recurrent miscarriages, supplementation with progestogen therapy may reduce the rate of miscarriage in subsequent pregnancies.

786.

Probiotics for the prevention of pediatric antibiotic-associated diarrhea

Guo, Qin. Goldenberg, Joshua Z. Humphrey, Claire. El Dib, Regina. Johnston, Bradley C. Institution Bradley C Johnston. TI Probiotics for the prevention of pediatric antibiotic-associated diarrhea.

EBM Reviews - Cochrane Database of Systematic Reviews

Cochrane Database of Systematic Reviews. 5, 2019.

[Systematic Review]

AN: 00075320-100000000-03901

Background

Antibiotics alter the microbial balance commonly resulting in antibiotic-associated diarrhea (AAD). Probiotics may prevent AAD via providing gut barrier, restoration of the gut microflora, and other potential mechanisms of action.

Objectives

The primary objectives were to assess the efficacy and safety of probiotics (any specified strain or dose) used for the prevention of AAD in children.

Search methods

MEDLINE, Embase, CENTRAL, CINAHL, and the Web of Science (inception to 28 May 2018) were searched along with registers including the ISRCTN and Clinicaltrials.gov. We also searched the NICE Evidence Services database as well as reference lists from relevant articles.

Selection criteria

Randomized, parallel, controlled trials in children (0 to 18 years) receiving antibiotics, that compare probiotics to placebo, active alternative prophylaxis, or no treatment and measure the incidence of diarrhea secondary to antibiotic use were considered for inclusion.

Data collection and analysis

Study selection, data extraction, and risk of bias assessment were conducted independently by two authors. Dichotomous data (incidence of AAD, adverse events) were combined using a pooled risk ratio (RR) or risk difference (RD), and continuous data (mean duration of diarrhea) as mean difference (MD), along with corresponding 95% confidence interval (95% CI). We calculated the number needed to treat for an additional beneficial outcome (NNTB) where appropriate. For studies reporting on microbiome characteristics using heterogeneous outcomes, we describe the results narratively. The certainty of the evidence was evaluated using GRADE.

Main results

Thirty-three studies (6352 participants) were included. Probiotics assessed included *Bacillus* spp., *Bifidobacterium* spp., *Clostridium butyricum*, *Lactobacilli* spp., *Lactococcus* spp., *Leuconostoc cremoris*, *Saccharomyces* spp., or *Streptococcus* spp., alone or in combination. The risk of bias was determined to be high in 20 studies and low in 13 studies. Complete case (patients who did not complete the studies were not included in the analysis) results from 33 trials reporting on the incidence of diarrhea show a precise benefit from probiotics compared to active, placebo or no treatment control.

After 5 days to 12 weeks of follow-up, the incidence of AAD in the probiotic group was 8% (259/3232) compared to 19% (598/3120) in the control group (RR 0.45, 95% CI 0.36 to 0.56; $I^2 = 57\%$, 6352 participants; NNTB 9, 95% CI 7 to 13; moderate certainty evidence). Nineteen studies had loss to follow-up ranging from 1% to 46%. After making assumptions for those lost, the observed benefit was still statistically significant using an extreme plausible intention-to-treat (ITT) analysis, wherein the incidence of AAD in the probiotic group was 12% (436/3551) compared to 19% (664/3468) in the control group (7019 participants; RR 0.61;

95% CI 0.49 to 0.77; $P < 0.00001$; $I^2 = 70\%$). An a priori available case subgroup analysis exploring heterogeneity indicated that high dose (≥ 5 billion CFUs per day) is more effective than low probiotic dose (< 5 billion CFUs per day), interaction P value = 0.01. For the high dose studies the incidence of AAD in the probiotic group was 8% (162/2029) compared to 23% (462/2009) in the control group (4038 participants; RR 0.37; 95% CI 0.30 to 0.46; $P = 0.06$; moderate certainty evidence). For the low dose studies the incidence of AAD in the probiotic group was 8% (97/1155) compared to 13% (133/1059) in the control group (2214 participants; RR 0.68; 95% CI 0.46 to 1.01; $P = 0.02$). Again, assumptions for loss to follow-up using an extreme plausible ITT analysis was statistically significant. For high dose studies the incidence of AAD in the probiotic group was 13% (278/2218) compared to 23% (503/2207) in control group (4425 participants; RR 0.54; 95% CI 0.42 to 0.70; $P < 0.00001$; $I^2 = 68\%$; moderate certainty evidence).

None of the 24 trials (4415 participants) that reported on adverse events reported any serious adverse events attributable to probiotics. Adverse event rates were low. After 5 days to 4 weeks follow-up, 4% (86/2229) of probiotics participants had an adverse event compared to 6% (121/2186) of control participants (RD 0.00; 95% CI -0.01 to 0.01; $P < 0.00001$; $I^2 = 75\%$; low certainty evidence). Common adverse events included rash, nausea, gas, flatulence, abdominal bloating, and constipation.

After 10 days to 12 weeks of follow-up, eight studies recorded data on our secondary outcome, the mean duration of diarrhea; with probiotics reducing diarrhea duration by almost one day (MD - 0.91; 95% CI -1.38 to -0.44; $P < 0.00001$; low certainty evidence). One study reported on microbiome characteristics, reporting no difference in changes with concurrent antibiotic and probiotic use.

Authors' conclusions

The overall evidence suggests a moderate protective effect of probiotics for preventing AAD (NNTB 9, 95% CI 7 to 13). Using five criteria to evaluate the credibility of the subgroup analysis on probiotic dose, the results indicate the subgroup effect based on high dose probiotics (≥ 5 billion CFUs per day) was credible. Based on high-dose probiotics, the NNTB to prevent one case of diarrhea is 6 (95% CI 5 to 9). The overall certainty of the evidence for the primary endpoint, incidence of AAD, based on high dose probiotics was moderate due to the minor issues with risk of bias and inconsistency related to a diversity of probiotic agents used. Evidence also suggests that probiotics may moderately reduce the duration of diarrhea, a reduction by almost one day. The benefit of high dose probiotics (e.g. *Lactobacillus rhamnosus* or *Saccharomyces boulardii*) needs to be confirmed by a large well-designed multi-centered randomized trial. It is premature to draw firm conclusions about the efficacy and safety of 'other' probiotic agents as an adjunct to antibiotics in children. Adverse event rates were low and no serious adverse events were attributable to probiotics. Although no serious adverse events were observed among inpatient

and outpatient children, including small studies conducted in the intensive care unit and in the neonatal unit, observational studies not included in this review have reported serious adverse events in severely debilitated or immuno-compromised children with underlying risk factors including central venous catheter use and disorders associated with bacterial/fungal translocation.

787.

Statins for Smith-Lemli-Opitz syndrome

Ballout, Rami A. Bianconi, Simona. Livinski, Alicia. Fu, Yi-Ping. Remaley, Alan T. Porter, Forbes D. Institution Rami A Ballout. TI Statins for Smith-Lemli-Opitz syndrome.

EBM Reviews - Cochrane Database of Systematic Reviews

Cochrane Database of Systematic Reviews. 1, 2020.

[Protocol]

AN: 00075320-100000000-11921

This is a protocol for a Cochrane Review (Intervention). The objectives are as follows:

n/a

To evaluate the efficacy of statin therapy in reducing the frequency or severity of the neurobehavioral abnormalities seen in people with SLOS (e.g. aggression, anxiety, irritability, self-mutilation, autistic behaviors, sleep disturbances, etc.) ().

To evaluate the potential effects of statin therapy on survival.

788.

One-step techniques for primary distal hypospadias in children and adolescents

Gomes, Adriano L. da Silva, MK Edina. Atallah, Alvaro N. Carnevale, Jose. Porfirio, JM Gustavo. Baptista-Silva, CC Jose. Institution Adriano L Gomes . TI One-step techniques for primary distal hypospadias in children and adolescents.

EBM Reviews - Cochrane Database of Systematic Reviews

Cochrane Database of Systematic Reviews. 3, 2019.

[Protocol]

AN: 00075320-100000000-08743

789.

Progestogen for treating threatened miscarriage

Wahabi, Hayfaa A. Fayed, Amel A. Esmail, Samia A. Bahkali, Hassan Khawater. Institution

Hayfaa A Wahabi .TI Progestogen for treating threatened miscarriage.

EBM Reviews - Cochrane Database of Systematic Reviews

Cochrane Database of Systematic Reviews. 9, 2018.

[Systematic Review]

AN: 00075320-100000000-04728

Background

Miscarriage is a common complication encountered during pregnancy. It is defined as spontaneous pregnancy loss before 20 weeks' gestation. Progesterone's physiological role is to prepare the uterus for the implantation of the embryo, enhance uterine quiescence and suppress uterine contractions, hence, it may play a role in preventing rejection of the embryo. Inadequate secretion of progesterone in early pregnancy has been linked to the aetiology of miscarriage and progesterone supplementation has been used as a treatment for threatened miscarriage to prevent spontaneous pregnancy loss. This update of the Cochrane Review first published in 2007, and previously updated in 2011, investigates the evidence base for this practice.

Objectives

To determine the efficacy and the safety of progestogens in the treatment of threatened miscarriage.

Search methods

We searched Cochrane Pregnancy and Childbirth's Trials Register, and the WHO International Clinical Trials Registry Platform () (8 August 2017) and reference lists of retrieved trials.

Selection criteria

Randomised, quasi-randomised or cluster-randomised controlled trials, that compared progestogen with placebo, no treatment or any other treatment for the treatment of threatened miscarriage in women carrying singleton pregnancy.

Data collection and analysis

At least two review authors assessed the trials for inclusion in the review, assessed trial quality and extracted the data and graded the body of evidence.

Main results

We included seven trials (involving 696 participants) in this update of the review. The included trials were conducted in different countries, covering the full spectrum of the World Bank's economic classification, which enhances the applicability of evidence drawn from this review. Two trials were conducted in Germany and Italy which are high-income countries, while four trials were conducted in upper-middle income countries; two in Iran, one in Malaysia and the fourth in Turkey, and the seventh trial was conducted in Jordan, which is a lower-middle income country. In six trials all the participants met the inclusion criteria and in the seventh study, we included in the meta-analysis only the subgroup of participants who met the inclusion criteria. We assessed the body of evidence for the main outcomes using the GRADE tool and the quality of the evidence ranged from very low to moderate. Downgrading of evidence was based on the high risk of bias in six of the seven included trials and a small number of events and wide confidence intervals for some outcomes.

Authors' conclusions

The results of this Cochrane Review suggest that progestogens are probably effective in the treatment of threatened miscarriage but may have little or no effect in the rate of preterm birth. The evidence on congenital abnormalities is uncertain, because the quality of the evidence for this outcome was based on only two small trials with very few events and was found to be of very low quality.