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Guidelines

Congenital Lower Urinary Tract Obstruction: Update and Summary of the European Association of Urology and European Society for Paediatric Urology Guidelines

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Abstract

Background and objective: The literature on congenital lower urinary tract obstruction (CLUTO) is still limited, resulting in a generally low level of evidence. These guidelines aim to provide a practical approach based on a consensus from the European Association of Urology (EAU)/European Society for Paediatric Urology (ESPU) Paediatric Urology Guidelines Panel. The primary aim of this update was to revise and expand the 2024 EAU/ESPU paediatric urology guidelines, focusing on the comprehensive management of CLUTO.

Methods: A structured literature review was performed for all relevant publications published from the last update until March 21, 2023.

Key findings and limitations: Antenatal management should be considered based on ultrasound findings, foetal urine biochemistry, amniotic fluid levels, and chromosomal status. In newborns with a suspected diagnosis of infravesical obstruction, bladder drainage should be performed and antibiotic prophylaxis initiated. Voiding cystography should be conducted as soon as possible in cases where posterior urethral valves (PUVs) are suspected. A serum creatinine nadir of above 0.85 mg/dl is associated with a poor prognosis. Despite optimal treatment, 20% of patients will progress to end-stage renal disease. Lifelong monitoring and management of both bladder and renal

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function are essential. Neonatal circumcision, as an adjunct to antibiotic prophylaxis in PUV patients, significantly reduces the risk of febrile urinary tract infections during the first 2 yr of life.

Conclusions: This paper is a summary of the updated 2024 EAU/ESPU guidelines, and it provides practical considerations for patients with CLUTO.

Patient summary: In this summary and update of the European Association of Urology/European Society for Paediatric Urology guidelines, we provide practical considerations for the management of children with congenital lower urinary tract obstruction.

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1. Introduction

These guidelines aim to provide a practical approach to the management of congenital lower urinary tract obstruction (CLUTO), based on risk assessment and specific indications for both diagnostic evaluations and interventions.

The acronym CLUTO refers to intrauterine dilatation of the bladder and/or upper urinary tract, typically detected during pregnancy via ultrasound (US). While isolated upper urinary tract dilatation is not a defining feature of CLUTO, it may in some cases serve as an early indicator of underlying posterior urethral valves (PUVs), especially in bilateral presentations. A wide range of conditions can lead to such dilatation, with PUVs being the most common cause. Postnatally, CLUTO encompasses various anatomical and functional disorders, including anterior urethral valves (AUVs), urethral atresia/stenosis, prune belly syndrome, dilating vesicoureteral reflux (VUR), cloacal malformation, prolapsing ureterocele, and syndromes such as megacystis-microcolon-intestinal hypoperistalsis or megacystis-megaureter syndrome. Owing to the complexity and rarity of CLUTO, referral to a tertiary centre with multidisciplinary expertise in both prenatal and postnatal management is strongly recommended [1].

This publication summarises the 2024 updates to the CLUTO chapter in the European Association of Urology (EAU) and European Society for Paediatric Urology (ESPU) guidelines.

To our knowledge, no other guidelines in the current literature consolidate and present, in a single document, the comprehensive knowledge necessary for managing children with CLUTO. These guidelines are created to provide clinicians with the extensive knowledge and practical tools required for diagnosis and treatment [2].

Unfortunately, the available scientific literature is still limited, and the lack of robust, prospective randomised controlled trials weakens the evidence supporting the current guidelines. Most studies are retrospective, include heterogeneous groups of children, and have limited quality stratification.

2. Evidence acquisition

Chapters of the EAU/ESPU guidelines on paediatric urology are updated at regular intervals (every 4 yr). For the update of this chapter, a structured literature search was conducted by a research information specialist and reviewed by at

least two panel members for all relevant publications published since the previous update (January 2020) until March 21, 2023 [3]. This was followed by a critical appraisal of the full texts and a discussion with the complete guideline panel in a face-to-face meeting. The literature search was performed using the following databases: MEDLINE, Embase, Cochrane CENTRAL, and Cochrane CDSR (via Ovid). Keywords related to CLUTO were searched using Boolean operators. Controlled vocabularies as well as free text words were searched; variations of root words were searched. There was no restriction on study design. Conference abstracts were excluded. Only English publications were included. In total, 601 citations were retrieved for screening. The search strategy is provided as a supplement to the guidelines chapter (Supplementary material).

3. Evidence synthesis

Diagnosis of CLUTO can occur either prenatally or postnatally, with a broad range of presentations [4]. The aetiopathogenesis of foetal CLUTO involves bladder outflow obstruction during urinary tract development, leading to progressive bladder dilation, thickening of the bladder wall, and potential secondary complications such as hydroureteronephrosis (HUN) and compression of the renal parenchyma. In severe cases, this obstruction can result in oligohydramnios or anhydramnios, which may cause lung hypoplasia and lead to foetal or perinatal mortality [5].

The most common cause of CLUTO is a PUV (57%) [6]. Less common causes are AUVs, anterior urethral diverticula (AUDs), syringoceles, Cobb's collar, urethral atresia/hypoplasia, and posterior urethral polyps (PUPs). The remaining cases are associated with genetic syndromes, and developmental or chromosomal abnormalities, including anorectal malformations or undefined pathologies (36.5%) [7].

3.1. Prenatal megacystis

In the 1st trimester, foetal megacystis is defined as a bladder with a longitudinal (craniocaudal) diameter of ≥ 7 mm. A longitudinal diameter between 7 and 12 mm is usually transient, resolving in about 90% of cases by the 2nd trimester. However, a measurement of >15 mm suggests CLUTO and is unlikely to resolve spontaneously [1]. In the 2nd and 3rd trimesters, megacystis is defined by an enlarged bladder that fails to empty during an extended

US examination lasting for at least 40 min. The prognosis of the foetus depends on the underlying pathology, timing of diagnosis, presence of oligohydramnios or anhydramnios, and bladder volume [1,6,7].

3.2. Posterior urethral valves

3.2.1. Epidemiology and pathophysiology

PUVs are one of the few life-threatening congenital anomalies of the urinary tract identified during the neonatal period. The risk of developing chronic kidney disease (CKD) is estimated to be 65%, with up to 20% progressing to end-stage renal disease (ESRD) [8]. PUVs account for up to 17% of paediatric ESRD cases, and the incidence is estimated to be 1 in 7000–8000 live births [9].

An obstruction at the level of the urethra, as occurs with PUVs, affects the entire urinary tract to varying degrees:

1. The prostatic urethra becomes distended, and the ejaculatory ducts may dilate due to VUR.
2. The bladder neck becomes hypertrophied and rigid.
3. The hypertrophied bladder wall may occasionally develop multiple diverticula.
4. Nearly all patients with a PUV exhibit upper urinary tract dilatation, which may result from the valve itself and increased bladder pressure, or from obstruction of the ureterovesical junction caused by the hypertrophied bladder.
5. In cases of secondary VUR, the affected kidney typically exhibits poor function.

Secondary VUR is observed in at least 50% of patients with a PUV [10]. It is widely accepted that unilateral high-grade VUR, associated with ipsilateral renal dysplasia, acts as a “pressure pop-off valve”, potentially protecting the contralateral kidney and leading to a more favourable prognosis [11]. Other types of *pop-off mechanisms* include large bladder diverticulum, urinary extravasation (with or without urinary ascites), and a patent urachus [12]. However, the renoprotective effects of *pop-off phenomena* remain equivocal, as long-term outcomes from various studies have been inconsistent. Some studies have demonstrated a protective effect, while others have shown no significant effect. A possible explanation for these discrepancies may lie in differences in defining the exact nature of what constitutes a pop-off mechanism [13,14].

3.2.2. Classification systems of the urethral valves

Up until today, the original classification by Young et al [15] is the most commonly used classification, which described three categories: type I, type II, and type III. However, today, only types I and III are considered to be obstructive.

3.2.3. Diagnostic evaluation

Most cases of PUVs are suspected prenatally (Fig. 1). During prenatal US screening, HUN and a distended bladder are suspicious signs of a PUV. A thick-walled bladder seems to better predict a PUV than a dilated posterior urethra (“key-hole” sign) [16]. However, differentiation between obstructive and nonobstructive aetiologies on prenatal US is challenging, as both have a similar sonographic appearance

[17]. In the presence of increased echogenicity of the kidney, dilatation of the urinary tract, and oligohydramnios, diagnosis of a PUV should strongly be considered. Prenatal US is adequate in most of the cases (90%) [18]. However, in certain circumstances, such as when US images are limited due to technical factors such as challenging foetal positioning or maternal obesity, foetal magnetic resonance imaging (MRI) may provide additional information, especially in complex cases where further clarity is needed [19].

Postnatally, creatinine, blood urea nitrogen, and electrolytes should be monitored closely during the first few days. Initial management includes a multidisciplinary team involving a paediatric nephrologist. The clinician must be aware of a noteworthy association between PUVs and undescended testicles (UDTs) and/or inguinal hernia and patent urachus. UDTs occurred in 12–17% of PUVs, which is consistent with a ten-fold increase [20].

A voiding cystourethrogram (VCUG; including lateral views of the urethra during the voiding phase without a catheter in situ) is recommended to assess the presence of a PUV. This study is essential whenever there is a suspicion of infravesical obstruction, as the urethral anatomy is well outlined during voiding. The extent of posterior urethral deformity, as expressed by the posterior urethral height: width ratio, appears to correlate positively with nadir creatinine at 12 mo of age [21]. Nuclear renography, using techniques such as dimercaptosuccinic acid or mercaptoacetyl triglycine, is important for assessing both split renal function and potential renal scarring, providing valuable information on the individual contribution of each kidney to overall renal function.

3.2.4. Management

3.2.4.1. Prenatal treatment. The main challenges for prenatal intervention are the timely and accurate diagnosis of conditions that may lead to renal damage. Since renal dysplasia is irreversible, it is essential to identify foetuses with preserved renal function early enough to ensure timely treatment.

The primary goal of prenatal vesicoamniotic shunting varies with the timing of the procedure: in late interventions, the aim is to restore amniotic fluid volume and reduce the risk of pulmonary hypoplasia, whereas early interventions, although aimed at preserving renal function, have not yet been definitively shown to offer clear benefits to long-term renal outcomes [22]. The decision to intervene prenatally can be guided by a staging system based on renal US findings, amniotic fluid volume, and foetal urine biochemistry [23]. Early intervention (before 16 wk of gestation) may benefit renal function, but accurate diagnosis and detection of other severe comorbidities are extremely challenging at this stage [24]. Later interventions primarily benefit lung development rather than renal function [25]. Emerging reports of interventions as early as the end of the 1st trimester suggest potential long-term renal preservation. However, these results are still preliminary, and the techniques are complex, with a higher risk of foetal demise at this fragile stage of development [26–28].

Foetal urine samples obtained before 23 wk of gestation, particularly measurements of β 2-microglobulin, sodium,

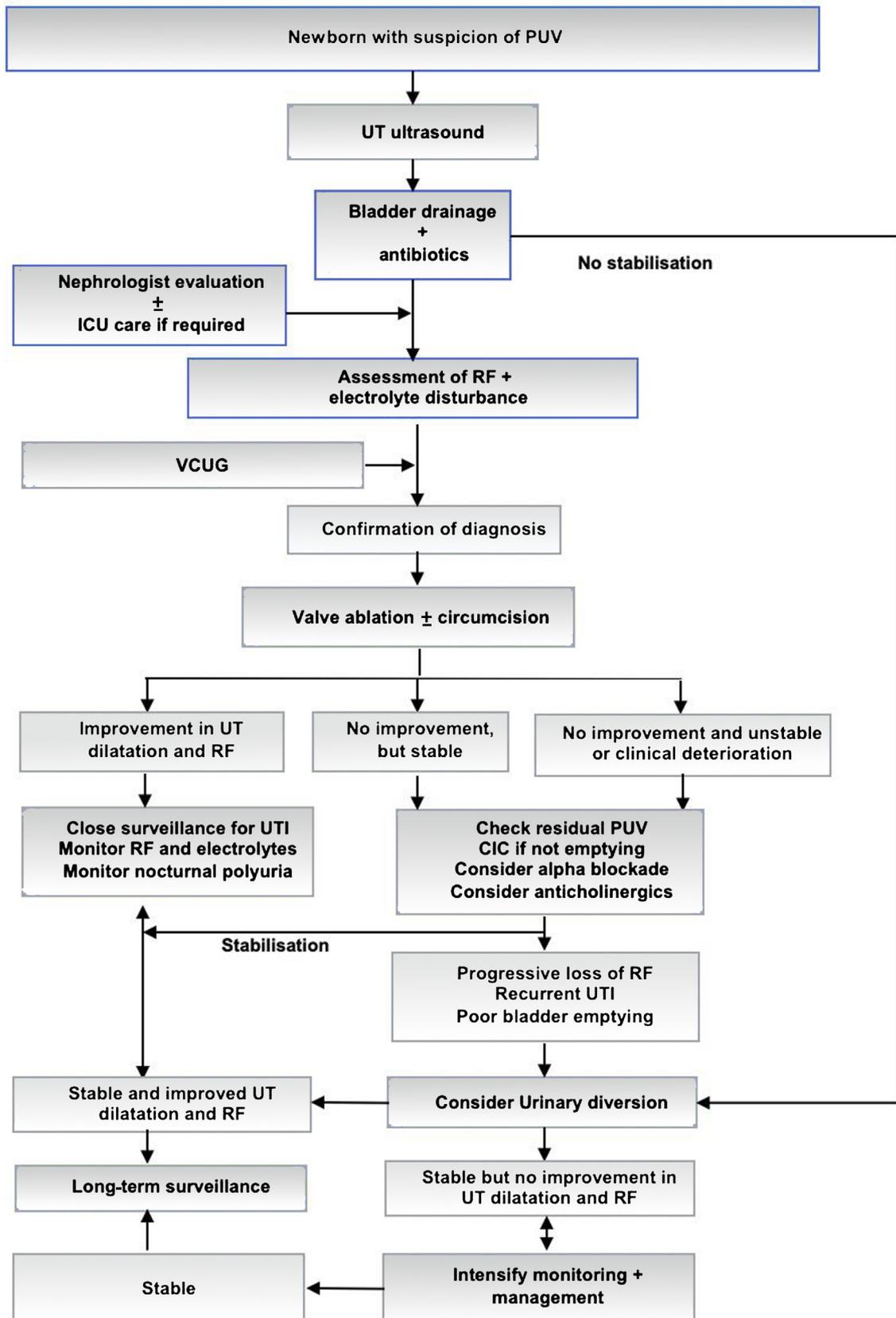


Fig. 1 – An algorithm on the assessment, management, and follow-up of newborns with a possible PUV. CIC = clean intermittent catheterisation; ICU = intensive care unit; PUV = posterior urethral valve; RF = renal function; UT = urinary tract; UTI = urinary tract infection; VCUg = voiding cystourethrogram.

chloride, and calcium, may help distinguish between those who could benefit from intrauterine therapy and those with a more compromised prognosis [29]. Normal biochemistry values (sodium <100 mmol/l, chloride <90 mmol/l, calcium <8 mg/dl, and β 2-microglobulin <6 mg/l) in the first foetal urine sample, or improvement in these values between two sequential samples, were associated with higher foetal survival and normal renal function at 5 yr in a small study [30]. Amniotic fluid status, renal appearance, and foetal urine biochemistry can aid in counselling. A proteomic analysis of foetal urine, using a 12-peptide signature found in foetuses that develop ESRD by age 2, may hold promise for assessing CLUTO, although this remains experimental.

The placement of a vesicoamniotic shunt (VAS) is a prenatal intervention designed to restore amniotic fluid circulation. Reported complication rates range from 21% to 59%, with shunt dislocation being the most common issue [22]. The PLUTO trial (a randomised study) failed to demonstrate a long-term benefit on renal function from VAS placement [31]. A meta-analysis of interventions for CLUTO reported that VAS resulted in a higher perinatal survival rate than conservative management (57.1% vs 38.8%), with no significant differences in survival at 6–12 mo, 2 yr, or postnatal renal function [32].

Foetal cystoscopy with laser ablation carries a high complication rate and lacks strong evidence supporting its effectiveness [33]. To avoid the severe complications associated with laser ablation, balloon dilation has been attempted [34]. However, the small number of patients and study designs are insufficient to support any formal recommendations.

Parental counselling is crucial. The natural history of CLUTO, including postnatal outcomes with or without prenatal treatment, as well as the uncertainties and controversies surrounding the diagnosis and management of CLUTO, should be discussed thoroughly [22].

3.2.4.2. Postnatal treatment.

3.2.4.2.1. *Bladder drainage.* Following delivery, the bladder should be drained transurethrally or suprapubically. This catheter drainage tube can then be used to perform a VCUG to confirm the diagnosis.

3.2.4.2.2. *Valve ablation.* Once the medical condition of the neonate has stabilised, endoscopic valve ablation can be performed, provided that the urethra is accessible with available equipment. If the urethra is too small, urinary drainage should be maintained until valve ablation can be accomplished. Small paediatric cystoscopes and resectoscopes are available for incising, ablating, or resecting the valve at the 5, 7, and the 12 o'clock positions. It is crucial to avoid extensive electrocoagulation, as stricture formation is the most common complication of such procedures. Two studies have shown a lower rate of urethral strictures when using a cold knife than when using diathermy [35,36]. Currently, there is no strong evidence to support the use of laser ablation for PUVs; however, preliminary studies on holmium:YAG and thulium lasers indicate that laser fulguration is safe and effective [37].

3.2.4.2.3. *Vesicostomy.* A vesicostomy is indicated if the child is too small to undergo endoscopic surgery, has

experienced failure of endoscopic valve ablation, or has shown no clinical or biochemical improvement following valve ablation. This procedure serves as an alternative to prolonged catheter drainage, and has been shown to stabilise or improve the upper urinary tracts in up to 90% of cases [38]. The most commonly performed vesicostomy procedure in children was described by Blocksom and later modified by Duckett [39]. Common complications following a vesicostomy include stomal stenosis, mucosal prolapse, peristomal dermatitis, and bladder calculi. The risk of prolapse is typically associated with extensive mobilisation of the bladder and the placement of the stoma too inferiorly on the abdominal wall, which can allow the posterior bladder wall to evert through the stoma.

3.2.4.2.4. *High diversion.* In cases where bladder drainage is insufficient to prevent recurrent infections of the upper urinary tract, improve renal function, or reduce upper tract dilatation, high urinary diversion should be considered. This recommendation reflects the consensus of the panel (as supported by the urinary tract infection (UTI) chapter of the EAU/ESPU guidelines). The choice of the diversion method, such as high loop ureterostomy, ring ureterostomy, end ureterostomy, or pyelostomy, should be based on individual clinical scenarios, with careful evaluation of the advantages and disadvantages of each technique. Urinary diversion can also play a role in delaying progression to end-stage renal failure [40].

3.2.4.2.5. *Bladder neck incision.* Bladder neck incision has been proposed as a method for managing bladder neck obstruction [41]. However, there is currently no evidence to demonstrate that bladder neck incision prevents the need for reintervention or reduces rehospitalisation rates. Therefore, it cannot be recommended as a routine management option.

3.2.4.2.6. *Medical treatment.* Early administration of oxybutynin may improve bladder function and enhance the resolution of hydronephrosis (HN) and VUR; however, it has no discernible effect on renal function or the risk of UTIs [42,43]. In patients with poor bladder emptying, α -blockers can be used to reduce the postvoid residual [44].

3.2.4.2.7. *Night bladder drainage and clean intermittent catheterisation.* Deterioration of renal function without anatomical obstruction, coupled with higher urine output (polyuria), may lead to bladder overdistension during the night. Therefore, bladder drainage during the night via catheterisation may benefit both HN and renal function [45].

Patients with high daytime postvoid residual urine volumes may benefit from clean intermittent catheterisation (CIC) [46]. For those who are unable or unwilling to perform CIC via the urethra, the placement of a catheterisable channel is a viable alternative [47]. CIC has been shown to delay the onset of dialysis in patients with CKD progressing to ESRD. Additionally, it has resulted in significantly better 10-yr graft survival rates in transplanted patients [48].

3.2.4.2.8. *Additional measures to prevent UTIs.* VUR is highly prevalent in patients with a PUV, occurring in up to 72% of cases, with bilateral involvement seen in up to 32%. A recent prospective observational study identified high-grade VUR as an independent risk factor for the development of febrile

UTIs, particularly within the first 9 mo of life. Antibiotic prophylaxis should be considered for these patients [49,50]. High-grade VUR is typically associated with poorly functioning kidneys; however, early removal of a nonfunctioning renal unit in an asymptomatic patient appears unnecessary.

Circumcision may be discussed as a strategy to further reduce the risk of UTIs. In the aforementioned multicentre randomised controlled trial, after 2 yr of follow-up, the study demonstrated a statistically significant effect of circumcision as an adjunct to antibiotic prophylaxis in preventing febrile UTIs. The hazard ratio for developing a febrile UTI in the group receiving antibiotics alone was 10.3 (95% confidence interval: 1.3–82.5) compared with the combined group receiving both prophylactic antibiotics and circumcision [51].

3.2.5. Follow-up

Within 3 mo following the initial treatment, the effectiveness of the intervention should be assessed through clinical improvement (evaluated via US and renal function), a control VCUG, or a repeat cystoscopy, depending on the clinical course [52].

Several prognostic factors have been identified for predicting future renal function in patients with a PUV. The literature reports varying serum nadir creatinine levels, ranging from 0.85 to 1.2 mg/dl ($\mu\text{mol/l}$) [53,54]. Additionally, renal parenchymal quantity (total renal parenchymal area) and quality (corticomedullary differentiation and renal echogenicity) observed on initial postnatal US also possess a prognostic value [55].

Life-long monitoring of these patients is mandatory, as bladder dysfunction (“valve bladder”) is common and the delay in day- and night-time continence is a significant problem [56]. Urodynamic studies play an important role in the management of patients with valve bladder, especially in those with a suspicion of bladder dysfunction; however, there is no consensus regarding the optimal timing or frequency of such studies [57].

CKD develops in up to 65% of PUV patients, with approximately 20% progressing to ESRD [58]. Renal transplantation in these patients can be performed safely and effectively. Deterioration of graft function is primarily associated with lower urinary tract dysfunction [59].

Data regarding sexual function and fertility in PUV patients are limited. Long-term studies have indicated normal erectile function and fertility potential. However, it is important to consider the potential negative impact on individual fertility, as these patients have a higher risk for bilateral cryptorchidism, recurrent epididymitis, and ESRD [60].

3.3. Anterior urethral valve

An AUV is a semilunar or iris-like band of tissue on the ventral aspect of the urethra. It can be isolated or seen in association with an AUD. The aetiology of isolated an AUV is speculated to be secondary to congenital urethral obstruction, malunion of glandular and penile urethra, congenital cystic dilatation of periurethral glands, or ruptured distal lip of a syringocele. It can be present in the bulbous urethra,

penoscrotal junction, and penile urethra. Patients may present with a poor urinary stream, penile ballooning, UTIs, or haematuria. The diagnosis is based on a VCUG with possible findings of a dilated or elongated posterior urethra, dilatation of the anterior urethra, thickened trabeculated bladder, hypertrophied bladder neck, VUR, and urethral diverticula. In doubtful cases, retrograde urethrography may be helpful showing a linear filling defect along the ventral wall, or it may show a dilated urethra ending in a smooth bulge or an abrupt change in the calibre of the dilated urethra on a VCUG.

Treatment is mainly by endoscopic valve ablation. In selected patients, temporary diversion may be considered until the child is big enough for endoscopy. Open surgery is reserved for patients with a very large diverticulum and defective spongiosum. Renal failure may develop in up to 22% of patients, with the highest risk observed in those with pretreatment azotaemia, VUR, and UTIs [61].

3.4. Anterior urethral diverticulum

The common postnatal presenting features of an AUD include compressible ventral penile swelling, postmicturition dribble, voiding difficulties, poor urinary stream, and recurrent UTIs. Diagnosis is established through a VCUG with or without a retrograde urethrogram.

For small AUDs, endoscopic techniques such as cutting or deroofting of the distal lip of the diverticulum may suffice for treatment. Larger diverticula typically require excision followed by a two-layered urethroplasty, or alternatively, marsupialisation with staged urethroplasty. Anatomically, AUDs are often associated with abnormalities in the development of the corpus spongiosum, which may contribute to their formation. These anomalies underline the variable presentation of an AUD and its relationship with other urological conditions, necessitating individualised diagnostic and therapeutic approaches [62].

3.5. Syringocele

Cowper’s glands, also known as bulbourethral glands, are two glands located within the urogenital diaphragm, opening into the urethra approximately 1–2 cm distal to the sphincter. Syringocele refers to the cystic dilatation of these glands, which can be classified as congenital or acquired, the latter often resulting from trauma or infection. Syringoceles are categorised into four types: simple, imperforate, perforate, and ruptured. A simplified classification merges simple, perforate, and ruptured types into “open syringoceles”, while the imperforate type is termed “closed syringoceles”.

Closed syringoceles typically cause obstructive symptoms, whereas open syringoceles function as diverticula, leading to postvoid dribbling and, in some cases, obstruction. Depending on the type of syringocele, patients may present with a variety of symptoms, including postvoid dribbling, urethral discharge, UTIs, perineal pain, haematuria, obstructive voiding symptoms, dysuria, or urinary retention.

Diagnosis is primarily based on an antegrade and/or a retrograde urethrogram, which reveals a cystic defect distal to the prostate. If imaging studies are inconclusive or

Table 1 – Summary of evidence for the management of posterior urethral valves

Summary of evidence
Posterior urethral valves are one of the few life-threatening congenital anomalies of the urinary tract.
Antenatal therapy could be considered based on ultrasound findings, foetal urine biochemistry, amniotic fluid level, and chromosomal status.
Serum creatinine nadir above 0.85 mg/dl is correlated with a poor prognosis.
Neonatal circumcision as an adjunct to antibiotic prophylaxis in PUV patients significantly decreases the risk of developing febrile UTIs during the first 2 yr of life.
Early pharmacological management with oxybutynin may improve bladder function.
Despite optimal treatment, 20% of patients will develop ESRD.
Renal transplantation in these patients is safe and effective, if the bladder function is managed.
ESRD = end-stage renal disease; PUV = posterior urethral valve; UTI = urinary tract infection.

Table 2 – Summary of recommendations for the management of posterior urethral valves

Recommendations	Strength rating
Drain the bladder in newborns with a suspected diagnosis of infravesical obstruction and place on antibiotic prophylaxis.	Strong
Perform a VCUG in patients in whom a diagnosis of PUV is suspected.	Strong
Attempt endoscopic valve ablation after bladder drainage and stabilisation of the child.	Strong
Consider neonatal circumcision as an adjunct to antibiotic prophylaxis to decrease the risk of a UTI in those with a PUV, especially in the presence of high-grade VUR.	Strong
Offer prolonged urinary diversion (suprapubic/transurethral) for bladder drainage if the child is too small for valve ablation.	Strong
Use serum creatinine nadir as a prognostic marker.	Strong
Assess split renal function by a dimercaptosuccinic acid scan or mercaptoacetyltriglycine clearance.	Strong
Consider high urinary diversion if bladder drainage is insufficient to drain the upper urinary tract, or in the absence of a clinicobiochemical improvement.	Strong
Monitor and manage bladder and renal function lifelong.	Strong
PUV = posterior urethral valve; UTI = urinary tract infection; VCUG = voiding cystourethrogram; VUR = vesicoureteral reflux.	

additional clarification is needed, US and/or MRI may be utilised. Asymptomatic syringoceles can be managed conservatively.

Endoscopic deroofing using various energy sources, such as cold knife, electrocautery, or holmium laser, is an effective technique in both obstructing and nonobstructing syringoceles. In cases where the endoscopic approach is not feasible, open correction may be considered [63].

3.6. Cobb's collar

Cobb's collar is a congenital membranous stricture of the bulbar urethra, distinct from congenital obstructive posterior urethral membrane. This condition is independent of the veru montanum and external sphincter, and is thought to result from the persistence of a portion of the urogenital membrane. Diagnosis is typically made by a VCUG, which reveals narrowing in the proximal bulbar urethra with folds extending proximally, along with dilation of the posterior urethra, a prominent bladder neck, and other signs indicative of infravesical obstruction. However, delayed diagnosis is common and can lead to significant complications, including renal dysfunction and persistent bladder issues. Early recognition and treatment are crucial to prevent these outcomes. Clinicians should maintain a high index of

suspicion when encountering recurrent urinary tract symptoms or bladder outlet obstruction in paediatric patients. Treatment with an endoscopic cold-knife incision has been shown to result in lower recurrence rates than electrocautery [64,65].

3.7. Urethral atresia/hypoplasia

Male urethral atresia is a congenital condition characterised by a complete obstruction of the urethra, typically caused by a membrane located at the distal end of the prostatic urethra. The segment of the urethra distal to this obstruction is usually hypoplastic, presumably due to a lack of foetal voiding. This condition is often associated with bladder distention, VUR, HN, and renal dysplasia. Many reported cases exhibit phenotypic characteristics resembling those of prune belly syndrome.

Antenatal intervention may improve foetal survival rates. Although the Progressive Augmentation by Dilating the Urethra Anterior (PADUA) procedure has been described as a treatment option, most cases ultimately necessitate some form of vesical diversion [66].

3.8. Posterior urethral polyps

PUPs are polypoid, pedunculated, fibroepithelial tumours that arise in the posterior urethra, proximal to the veru montanum. Although PUPs do not cause antenatal HN, these can lead to obstruction later in life. The polyp typically lies on the floor of the urethra, with its tip extending into the bladder neck. An obstruction occurs due to the distal displacement of the polyp during micturition. Patients commonly present with dysuria, haematuria, and obstructive symptoms, such as a poor urinary stream and episodes of intermittent retention. Diagnosis can be suspected through a VCUG and/or US, but is confirmed via cystourethroscopy. Treatment is typically performed through endoscopic resection of the polyp. The course of the disease is generally benign, with no recurrences reported in the literature [67].

4. Conclusion/summary update

Conclusions are summarised in two tables (summary of evidence [Table 1] and recommendations [Table 2]).

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Acquisition of data: Gnech, Rawashdeh.

Analysis and interpretation of data: Gnech, Rawashdeh.

Drafting of the manuscript: Gnech, Rawashdeh.

Critical revision of the manuscript for important intellectual content: Gnech, 't Hoen, Bogaert, Castagnetti, O'Kelly, Quaedackers, Silay, Kennedy, Skott, van Uitert, Yuan, Radmayr, Burgu, Rawashdeh.

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Appendix A. Supplementary material

Supplementary data to this article can be found online at <https://doi.org/10.1016/j.euf.2025.01.012>.

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