EAU GUIDELINES ON PRIAPISM

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Introduction

Priapism is a pathological condition representing a true disorder of penile erection that persists more than 4 hours and is beyond or unrelated to sexual interest or stimulation. Erections lasting up to 4 hours are defined by consensus as 'prolonged'. Priapism may occur at all ages.

Classification

Ischaemic priapism is a persistent erection marked by rigidity of the corpora cavernosa and by little or no cavernous arterial inflow, although often proximally there is a compensated high velocity picture with little flow distally. The patient typically complains of penile pain and examination reveals a rigid erection.

Arterial priapism is a persistent erection caused by unregulated cavernous arterial inflow. The patient typically reports an erection that is not fully rigid and is not associated with pain although fully rigid erections may occur with sexual stimulation.

Stuttering (recurrent or intermittent) priapism is a distinct condition that is characterised by repetitive and painful episodes of prolonged erections. Erections are self-limited with intervening periods of detumescence. These are analogous

to repeated episodes of low flow (or ischaemic) priapism. The duration of the erectile episodes is generally shorter than in ischaemic priapism. The frequency and/or duration of these episodes is variable and a single episode can sometimes progress into a major ischaemic priapic episode.

Ischaemic (Low-Flow or Veno-Occlusive) Priapism

Diagnostic evaluation

Table 1: Key points in taking the history of priapism

• Duration of erection
• Presence and degree of pain
• Previous episodes of priapism and method of treatment
• Current erectile function, especially the use of any erectogenic therapies prescription or nutritional supplements
• Medications and recreational drugs
• Sickle cell disease, haemoglobinopathies, hypercoagulable states
• Trauma to the pelvis, perineum, or penis

Table 2: Key findings in priapism

	Ischaemic priapism	Arterial priapism
Corpora cavernosa fully rigid	Usually	Seldom
Penile pain	Usually	Seldom
Abnormal penile blood gas	Usually	Seldom
Haematological abnormalities	Usually	Seldom
Recent intracorporeal injection	Sometimes	Sometimes
Perineal trauma	Seldom	Usually

Table 3: Typical blood gas values

Source	pO_2 (mmHg)	pCO_2 (mmHg)	pH
Normal arterial blood (room air) [similar values are found in arterial priapism]	> 90	< 40	7.40
Normal mixed venous blood (room air)	40	50	7.35
Ischaemic priapism (first corporal aspirate)	< 30	> 60	< 7.25

Recommendations for the diagnosis of ischaemic priapism	GR
Take a comprehensive history for diagnosis which can help to determine the underlying type of priapism.	B
Include physical examination of the genitalia, the perineum and the abdomen in the diagnostic evaluation which may help to determine the underlying type of priapism.	B
For laboratory testing, include complete blood count, white blood count with blood cell differential, platelet count and coagulation profile. Direct further laboratory testing by the history and clinical and laboratory findings. In children with priapism, perform a complete evaluation of all possible causes.	B
Analyse blood gas of blood aspirated from the penis for the differentiation between ischaemic and arterial priapism.	B

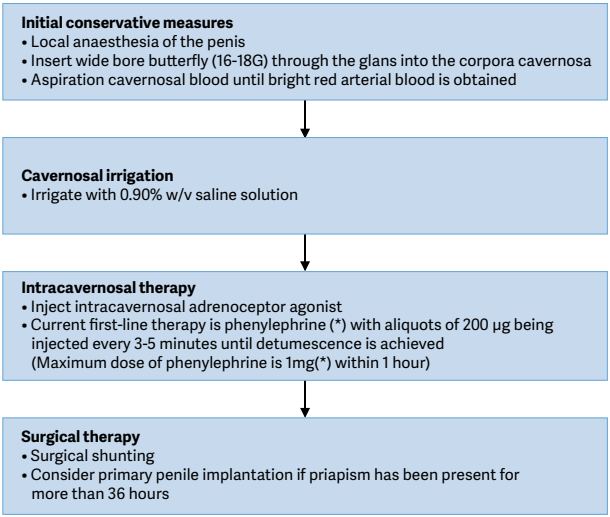
Perform colour duplex ultrasound of the penis and perineum for the differentiation between ischaemic and arterial priapism as an alternative or adjunct to blood gas analysis.	B
In cases of prolonged ischaemic priapism, use magnetic resonance imaging of the penis to predict smooth muscle viability and confirm erectile function restoration.	B
Perform selected pudendal arteriogram when embolisation is planned for the management of arterial priapism.	B

US = ultrasound.

Disease management

The treatment is sequential and the physician should move on to the next stage if the treatment fails.

Figure 1: Treatment of ischaemic priapism



(*) The dose of phenylephrine should be reduced in children. It can result in significant hypertension and should be used with caution in men with cardiovascular disease and monitoring of pulse, blood pressure and ECG is advisable in all patients during administration and for 60 minutes afterwards. Its use is contraindicated in men with a history of cerebro-vascular disease and significant hypertension.

Table 4: Medical treatment of ischaemic priapism

Drug	Dosage/Instructions for use
Phenylephrine	- Intracavernous injection of 200 µg every three to five minutes.
	- Maximum dosage is 1 mg within 1 hour.
	- The lower doses are recommended in children and patients with severe cardiovascular disease.
Etilephrine	- Intracavernosal injection at a concentration of 2.5 mg in 1-2 mL normal saline.
Methylene blue	- Intracavernous injection of 50-100 mg, left for five minutes. It is then aspirated and the penis compressed for an additional five minutes.
Adrenaline	- Intracavernous injection of 2 mL of 1/100,000 adrenaline solution up to five times over a 20-minute period.
Terbutaline	- Oral administration of 5 mg for prolonged erections lasting more than 2.5 hours that have arisen following intracavernosal injection of vasoactive agents.

Recommendations for the treatment of ischaemic priapism	GR
Start management of ischaemic priapism as early as possible (within four to six hours) and follow a step-wise approach.	B
First, decompress the corpora cavernosa by penile aspiration until fresh red blood is obtained.	C
In priapism secondary to intracavernous injections of vasoactive agents, replace blood aspiration with intra-cavernous injection of a sympathomimetic drug as the first step.	C
In priapism that persists despite aspiration, proceed to the next step, which is intracavernous injection of a sympathomimetic drug.	B
In cases that persist despite aspiration and intracav-ernous injection of a sympathomimetic drug, repeat these steps several times before considering surgical intervention.	C
Treat ischaemic priapism due to sickle cell anaemia in the same fashion as idiopathic ischaemic priapism. Provide other supportive measures (intravenous hydration, oxygen administration with alkalisation with bicarbonates, blood exchange transfusions), but do not delay initial treatment to the penis.	B
Proceed to surgical treatment only when blood aspira-tion and intracavernous injection of sympathomimetic drugs have failed or for priapism events lasting $\leq$ 72 hours.	C
Perform distal shunt surgical procedures first followed by proximal procedures in case of failure.	C
Discuss the immediate implantation of a penile pros-thesis with the patient in cases of priapism presenting > 36 hours after onset, or in cases for which all other interventions have failed.	B

Arterial (High-Flow or Non-Ischaemic) Priapism

Diagnostic evaluation

History

A comprehensive history is also mandatory in arterial priapism diagnosis and follows the same principles as described in Table 1.

Recommendations for the diagnosis of arterial priapism

The same recommendations as for ischaemic priapism apply.

Disease management

Recommendations for the treatment of arterial priapism	GR
Because high-flow priapism is not an emergency, perform definitive management at the discretion of the treating physician.	B
Manage conservatively with the use of ice applied to the perineum or site-specific perineal compression as the first step, especially in children. Use androgen deprivation therapy only in adults.	C
Perform selective artery embolisation, using temporary or permanent substances.	B
Repeat the procedure for the recurrence of arterial priapism following selective artery embolisation.	B
Reserve selective surgical ligation of the fistula as a final treatment option when embolisation has failed.	C

Stuttering (Recurrent or Intermittent) Priapism

Diagnostic evaluation

History

A comprehensive history is mandatory and follows the same principles as described in Table 1.

Disease management

Recommendations for the treatment of stuttering priapism	GR
Manage each acute episode similar to that for ischaemic priapism.	B
Use hormonal therapies (mainly GnRH agonists or antagonists) and/or antiandrogens for the prevention of future episodes in patients with frequent relapses. Do not use them before sexual maturation is reached.	C
Initiate treatment with PDE5Is only when the penis is in its flaccid state.	C
Use digoxin, α-adrenergic agonists, baclofen, gabapentin or terbutaline) only in patients with very frequent and uncontrolled relapses.	C
Use intracavernosal self-injections at home of sympathomimetic drugs for the treatment of acute episodes on an interim basis until ischaemic priapism has been alleviated.	C

GnRH = gonadotropin-receptor hormone
PDE5Is = phosphodiesterase type 5 inhibitors

This short booklet text is based on the more comprehensive EAU Guidelines (ISBN 978-90-79754-98-4), available to all members of the European Association of Urology at their website, http://www.uroweb.org.