EAU GUIDELINES ON PRIAPISM

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Introduction

Priapism is a pathological condition representing a true disorder of penile erection that persists for more than four hours and is beyond or unrelated to sexual interest or stimulation. Erections lasting up to four 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 blood flow distally. The patient typically complains of penile pain and clinical examination reveals a rigid erection.

Non-ischaemic 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 still 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 often

self-limited with intervening periods of detumescence. These are analogous to repeated episodes of ischaemic (or low flow) 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 full-blown ischaemic priapism.

Ischaemic (Low-Flow or Veno-Occlusive) Priapism

Diagnostic Evaluation

Table 1: Key points when taking the history of priapism

Duration of erection
Presence and severity 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 drug use
Sickle cell disease, haemoglobinopathies, hypercoagulable states
Trauma to the pelvis, perineum, or penis

Table 2: Key findings in priapism

	Ischaemic priapism	Non-ischaemic priapism
Corpora cavernosa fully rigid	Usually	Seldom
Penile pain	Usually	Seldom
Abnormal penile blood gas	Usually	Seldom
Haematological abnormalities	Sometimes	Seldom
Recent intracavernosal injection	Sometimes	Sometimes
Perineal trauma	Seldom	Usually

Table 3: Typical blood gas values

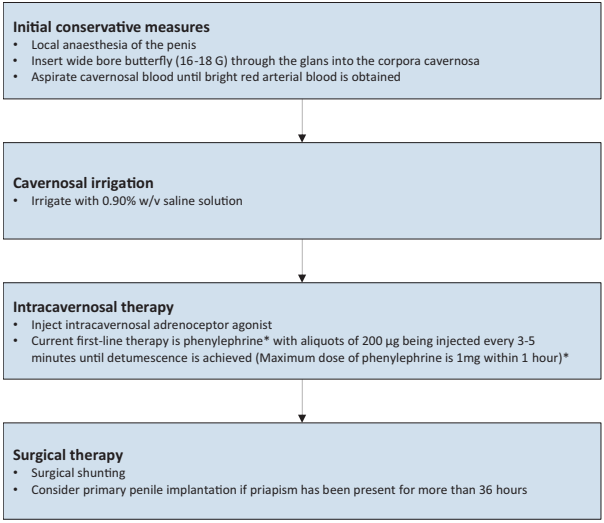
Source	pO_2 (mmHg)	pCO_2 (mmHg)	pH
Normal arterial blood (room air) [similar values are found in arterial priapism]	> 90	< 40	7.40
Normal mixed venous blood (room air)	40	50	7.35
Ischaemic priapism (first corporal aspirate)	< 30	> 60	< 7.25

Recommendations for the diagnosis of ischaemic priapism	Strength rating
Take a comprehensive history to establish the diagnosis which can help to determine the priapism subtype.	Strong
Include a physical examination of the genitalia, the perineum and the abdomen in the diagnostic evaluation.	Strong
For laboratory testing, include complete blood count, white blood count with blood cell differential, platelet count and coagulation profile. Direct further laboratory testing based on history, clinical and laboratory findings. In children with priapism, perform a complete evaluation of all possible causes.	Strong
Analyse the blood gas parameters from blood aspirated from the penis to differentiate between ischaemic and non-ischaemic priapism.	Strong
Perform colour duplex ultrasound of the penis and perineum for the differentiation between ischaemic and non-ischaemic priapism as an alternative or adjunct to blood gas analysis.	Strong
In cases of prolonged ischaemic priapism, use magnetic resonance imaging of the penis to predict smooth muscle viability and confirm erectile function restoration.	Strong
Perform selected pudendal arteriogram when embolisation is planned for the management of non-ischaemic priapism.	Strong

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. Monitoring of pulse, blood pressure and electrocardiogram (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 one hour. • 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 twenty minute period.
Terbutaline	• Oral administration of 5 mg for prolonged erections lasting more than 2.5 hours, after intracavernous injection of vasoactive agents.

Recommendations for the treatment of ischaemic priapism	Strength rating
Start management of ischaemic priapism as early as possible (within four to six hours) and follow a stepwise approach.	Strong

First, decompress the corpora cavernosa by penile aspiration until fresh red blood is obtained.	Weak
In priapism secondary to intracavernous injections of vasoactive agents, replace blood aspiration with intracavernous injection of a sympathomimetic drug as the first step.	Strong
In priapism that persists despite aspiration, proceed to the next step, which is intra-cavernous injection of a sympathomimetic drug.	Strong
In cases that persist despite aspiration and intracavernous injection of a sympathomi-metic drug, repeat these steps several times before considering surgical intervention.	Strong
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.	Strong
Proceed to surgical treatment only when blood aspiration and intracavernous injection of sympathomimetic drugs have failed or for priapism events lasting < 72 hours.	Strong
Perform distal shunt surgical procedures first followed by proximal procedures in case of failure.	Strong
Consider insertion of a penile prosthesis if priapism episode is > 36 hours after onset, or in cases for which all other interventions have failed.	Strong

Non-ischaemic (High-Flow or Arterial) Priapism

Diagnostic Evaluation

History

A comprehensive history is also mandatory in the diagnosis of non-ischaemic priapism and follows the same principles as described in Table 1.

Recommendations for the diagnosis of non-ischaemic priapism

The same recommendations as for ischaemic priapism apply.

Disease Management

Recommendations for the treatment of non-ischaemic priapism	Strength rating
Because non-ischaemic priapism is not an emergency, perform definitive management at the discretion of the treating physician.	Weak
Manage conservatively with the use of site specific perineal compression as the first step, especially in children. Consider androgen deprivation therapy only in adults.	Weak
Perform superselective arterial embolisation, using temporary material.	Strong
Repeat the procedure with temporary or permanent material for recurrent non-ischaemic priapism following selective arterial embolisation.	Weak
Reserve selective surgical ligation of a fistula as a final treatment option when embolisation has failed.	Weak

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	Strength rating
Manage each acute episode similar to that for ischaemic priapism.	Weak
Use hormonal therapies (mainly gonadotropin-receptor hormone agonists or antagonists) and/or anti-androgens for the prevention of future episodes in patients with frequent relapses. Do not use them before sexual maturation is reached.	Weak
Initiate treatment with phosphodiesterase type 5 inhibitors only when the penis is in its flaccid state.	Strong
Use digoxin, α-adrenergic agonists, baclofen, gabapentin or terbutaline only in patients with very frequent and uncontrolled relapses.	Weak
Use intracavernous self-injections at home of sympathomimetic drugs for the treatment of acute episodes on an interim basis until ischaemic priapism has been alleviated.	Weak

This short booklet text is based on the more comprehensive EAU Guidelines (ISBN 978-94-92671-01-1), available to all members of the European Association of Urology at their website, http://www.uroweb.org/guidelines.