GUIDELINES ON PRIAPISM

(Text update March 2015)

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Eur Urol 2014 Feb;65(2):480-9

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. 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
A comprehensive history is key for diagnosis and can help to determine the underlying type of priapism.	B
Physical examination of the genitalia, the perineum and the abdomen must be included in the diagnostic evaluation and may help to determine the underlying type of priapism.	B
Laboratory testing should include complete blood count, white blood count with blood cell differential, platelet count and coagulation profile. Further laboratory testing should be directed by the history and clinical and laboratory findings. Priapism in children requires a complete evaluation of all possible causes.	B
Blood gas analysis of blood aspirated from the penis is recommended for the differentiation between ischaemic and arterial priapism.	B

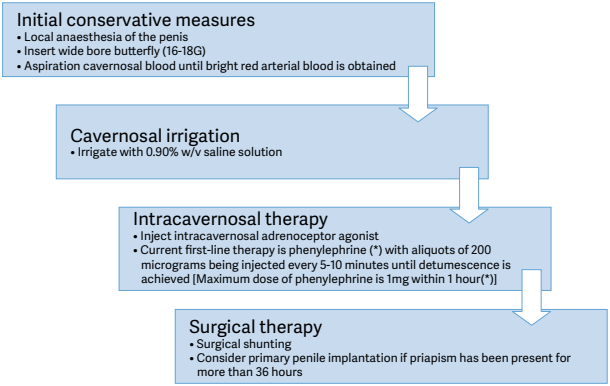
Colour duplex US of the penis and perineum is recommended for the differentiation between ischaemic and arterial priapism as an alternative or adjunct to blood gas analysis. It can also be helpful in localisation of the site and extend of fistula in arterial priapism as well as in the determination of successful resolution of ischaemic priapism.	B
Magnetic resonance imaging of the penis can predict smooth muscle viability and erectile function restoration.	B
Selected pudendal arteriogram should be reserved for the management of arterial priapism when embolisation is undertaken.	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 3-5 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. 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 5 minutes. It is then aspirated and the penis compressed for an additional 5 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
Ischaemic priapism is an emergency condition and rapid intervention is compulsory.	B
The specific aim is to restore painless penile flaccidity, in order to prevent chronic damage to the corpora cavernosa.	C
Management of ischaemic priapism should start as early as possible (within 4-6 hours) and should follow a stepwise approach. Erectile function preservation is directly related to the duration of priapism.	B
Initial management is decompression of the corpora cavernosa by penile aspiration until fresh red blood is obtained.	C
In priapism secondary to intracavernous injections of vasoactive agents blood aspiration can be replaced by intracavernous injection of a sympathomimetic drug as the first step.	C
In priapism that persists despite aspiration, the next step is intracavernous injection of a sympathomimetic drug. Phenylephrine is the recommended drug due to its favourable safety profile on the cardiovascular system compared to other drugs. Phenylephrine is usually diluted in normal saline with a concentration of 100-500 µg/mL and given in 1 mL doses every 3-5 minutes directly into the corpus cavernosum, up to a maximum dosage of 1 mg for no more than 1 hour. Patients at high cardiovascular risk should be given lower doses. Patient monitoring is highly recommended.	B
In cases that persist despite aspiration and intracavernous injection of a sympathomimetic drug, these steps should be repeated several times before considering surgical intervention.	C

Ischaemic priapism due to sickle cell anaemia is treated in the same fashion as idiopathic ischaemic priapism. Other supportive measures are recommended (intravenous hydration, oxygen administration with alkalisation with bicarbonates, blood exchange transfusions) but these should not delay initial treatment to the penis.	B
Surgical treatment is recommended only when blood aspiration and intracavernous injection of sympathomimetic drugs have failed or for priapism events lasting ≤ 72 hours.	C
Distal shunt surgical procedures should be performed first followed by proximal procedures in case of failure. The efficacy of these procedures is questionable and cavernous biopsy may be considered to diagnose muscle necrosis. No clear recommendation on one type of shunt over another can be given.	C
In cases of priapism presenting > 36 hours after onset, or in cases for which all interventions have failed, erectile dysfunction is inevitable and the immediate implantation of a penile prosthesis should be discussed with the patient. Implantation of penile prosthesis at a later stage can be difficult due to severe corporal fibrosis.	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
The management of high-flow priapism is not an emergency and definitive management can therefore be considered.	B
Conservative management includes the use of ice applied to the perineum or site-specific perineal compression. It may be successful particularly in children. Androgen deprivation therapy may enable closure of the fistula reducing spontaneous and sleep-related erections.	C
Selective artery embolisation, using temporary or permanent substances, is the suggested treatment modality and has high success rates. No definitive statement can be made on the best substance for embolisation in terms of sexual function preservation.	B
The recurrence of arterial priapism following selective artery embolisation requires the procedure to be repeated.	B
The preservation rate of sexual function is about 80%.	C
Selective surgical ligation of the fistula should be reserved as a last 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
The primary goal in the management of patients with stuttering priapism is the prevention of future episodes, which can generally be achieved pharmacologically.	B
The management of each acute episode is similar to that for ischaemic priapism.	B
Hormonal therapies (mainly gonadotropin-receptor hormone agonists or antagonists) and/or antiandrogens may be used for the prevention of future episodes. They should not be used before sexual maturation is reached.	C
Phosphodiesterase type 5 inhibitors (PDE5Is) have a paradoxical effect in alleviating and preventing stuttering priapism, mainly in patients with idiopathic and sickle cell disease associated priapism. Treatment should be initiated only when the penis is in its flaccid state.	C
Other systemic drugs (digoxin, alpha-adrenergic agonists, baclofen, gabapentin, terbutaline) can be considered, but data are even more limited.	C

Intracavernosal self-injections at home of sympathomimetic drugs can be considered for the treatment of acute episodes on an interim basis until ischaemic priapism has been alleviated.	C

This short booklet text is based on the more comprehensive EAU Guidelines (ISBN 978-90-79754-80-9), available to all members of the European Association of Urology at their website, http://www.uroweb.org.