Guidelines on
Priapism

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1. BACKGROUND

1.1 Introduction

Priapism is a pathological condition representing a true disorder of penile erection that persists beyond or is unrelated to sexual interest or stimulation (1) (LE: 4). Overall, erections lasting up to 4 hours are by consensus defined as 'prolonged' (LE: 4).

Priapism may occur at all ages. Current data show that the incidence of priapism in the general population is low (0.5-0.9 cases per 100,000 person-years) (2,3). In patients with sickle cell disease, which is an inherited disease that causes chronic haemolytic anaemia, the prevalence of priapism is up to 3.6% in patients < 18 years of age (4) increasing up to 42% in patients ≥ 18 years of age (5-7).

The Guidelines Office of the European Association of Urology (EAU) has appointed an Expert Panel to provide the first EAU Guidelines for Priapism.

1.2 Methodology

The EAU Guidelines on Priapism are based on a systemic literature search performed by the Expert Panel members. The MedLine database was searched using the major Medical Subject Headings term 'priapism' with search cut-off date of January 2013. This search yielded 1,199 articles (125 review articles, 404 original articles and 670 case reports). The Panel also identified critical problems and knowledge gaps, enabling priorities to be established for future clinical research.

1.3 Level of evidence and grade of recommendation

References in the text have been assessed according to their level of scientific evidence (LE) and guideline recommendations (GR) have been graded follow the listings in Tables 1 and 2, which are based on the Oxford Centre for Evidence-based Medicine Levels of Evidence (8). Grading aims to provide transparency between the underlying evidence and the recommendation given.

Table 1: Level of evidence*

Level	Type of evidence
1a	Evidence obtained from meta-analysis of randomized trials
1b	Evidence obtained from at least one randomized trial
2a	Evidence obtained from one well-designed controlled study without randomization
2b	Evidence obtained from at least one other type of well-designed quasi-experimental study
3	Evidence obtained from well-designed non-experimental studies, such as comparative studies, correlation studies and case reports
4	Evidence obtained from expert committee reports or opinions or clinical experience of respected authorities

Modified from (8).

It should be noted that when recommendations are graded, the link between the level of evidence (LE) and grade of recommendation (GR) is not directly linear. The availability of randomized controlled trials (RCTs) may not necessarily translate into a grade A recommendation where there are methodological limitations or a disparity in published results.

However, the absence of high level of evidence does not necessarily preclude a grade A recommendation, provided there is overwhelming clinical experience and consensus. There may be exceptional situations where corroborating studies cannot be performed, perhaps for ethical or other reasons, and in this case unequivocal recommendations are considered helpful. Whenever this occurs, it is indicated in the text as 'upgraded based on panel consensus'. The quality of the underlying scientific evidence, although this is a very important factor, has to be balanced against benefits and burdens, values and preferences, and costs when a grade is assigned to a recommendation.

The EAU Guidelines Office does not perform structured cost assessments, nor can they address local/national preferences in a systematic fashion. However, whenever these data are available, the Expert Panel will include the information.

Table 2: Grade of recommendation*

Grade	Nature of recommendations
A	Based on clinical studies of good quality and consistency that addressed the specific recommendations, including at least one randomized trial
B	Based on well-conducted clinical studies, but without randomized clinical trials
C	Made despite the absence of directly applicable clinical studies of good quality

Modified from (8).

1.4 Publication history

The EAU Guidelines on priapism are a new publication by the EAU Male Sexual Dysfunction Guidelines Panel. The same Panel also provided the 'EAU Guidelines on Male Sexual Dysfunction: Erectile Dysfunction and Premature Ejaculation' and the 'EAU Guidelines on Penile Curvature' (see the relevant sections for more information).

Alongside a scientific publication (9), a quick reference document (pocket guidelines) is available, presenting key findings of the Priapism Guidelines. These reference documents follow the updating cycle of the underlying large texts. All available material can be viewed and downloaded for personal use at the EAU website. The EAU website also includes a selection of translations and republications produced by national urological associations: http://www.uroweb.org/guidelines/online-guidelines/.

This document was peer-reviewed prior to publication.

1.5 Potential conflict of interest statement

The Expert Panel has submitted potential conflict of interest statements, which can be viewed on the EAU website: http://www.uroweb.org/guidelines/online-guidelines/.

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2. CLASSIFICATION

2.1 Ischaemic (low-flow or veno-occlusive) priapism

Ischaemic priapism is a persistent erection marked by rigidity of the corpora cavernosa and by little or no cavernous arterial inflow (1). The patient typically complains of penile pain and the examination reveals a rigid erection. Resolution of ischaemic priapism is characterized by the penis returning to a flaccid non-painful state. However, in many cases, persistent penile oedema, ecchymosis and partial erections can occur and it may mimic unresolved priapism. When left untreated, resolution may take days and erectile dysfunction invariably results.

2.2 Arterial (high flow or non-ischaemic) priapism

Arterial priapism is a persistent erection caused by unregulated cavernous arterial inflow (1). The patient typically reports an erection that is not fully rigid and is not associated with pain. Fully rigid erections under sexual stimulation may occur, before returning to the previous state of penile tumescence. In this case, it is not associated with erectile dysfunction.

2.3 Stuttering (recurrent or intermittent) priapism

Stuttering priapism, also termed intermittent or recurrent priapism, is a distinct condition that is characterized by repetitive and painful episodes of prolonged erections. Erections are self-limited with intervening periods of detumescence (2,3). The duration of the erectile episodes in stuttering priapism is generally shorter than in the low-flow ischaemic type (4). The frequency and/or duration of these distressing priapic episodes may increase and a single episode can sometimes develop into a major period of ischaemic priapic episodes.

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3. EPIDEMIOLOGY AND PATHOPHYSIOLOGY

3.1 Ischaemic (low flow or veno-occlusive) priapism

Ischaemic priapism is the most common form of priapism, accounting for more than 95% of all priapism episodes (1,2). It is usually painful, with a rigid erection characterized clinically by absent or reduced intracavernous arterial inflow. In ischaemic priapism, there are time-dependent modifications in the corporal metabolic environment, progressively leading to hypoxia, hypercapnia, and acidosis.

Although not all forms of priapism require immediate intervention, ischaemic priapism beyond 4 hours is considered a compartment syndrome, characterized by pressure within the closed space of the corpora cavernosa, which severely compromises circulation in the cavernous tissues. A compartment syndrome requires emergency medical intervention to minimize potential irreversible consequences, such as corporal fibrosis and permanent erectile dysfunction (3,4). The duration of priapism represents the most significant predictor of maintenance of premorbid erectile function; in this context, interventions beyond 48-72 hours since onset may eventually help to relieve erection and pain, but have little benefit in preserving erectile functioning. Histologically, by 12 hours, corporal specimens show interstitial oedema, progressing to destruction of sinusoidal endothelium, exposure of the basement membrane and thrombocyte adherence at 24 hours. At 48 hours, thrombi can be found in the sinusoidal spaces and smooth muscle necrosis with fibroblast-like cell transformation is evident (4).

In terms of pathophysiology (Table 3), ischaemic priapism has been identified as idiopathic in the majority of cases since no specific cause could be identified (2,5). Moreover, ischaemic priapism has been associated with sickle cell anaemia, haematological dyscrasias, neoplastic syndromes, and the use of several different medications. Ischaemic priapism may occur (0.4-35%) after intracavernous injections of papaverine, phentolamine and/or prostaglandin E1 (2,3,6-8) (Table 3). However, most of the these cases were treated with papaverine-based combinations while the prevalence of priapism is < 1% in the case of prostaglandin E1 (7). Since their introduction on the market, a few cases of priapism have been described in men who have taken phosphodiesterase type 5 inhibitors (PDE5is) (2). Most of these men had histories of increased risk for priapism, including sickle cell disease, spinal cord injury, combined administration of PDE5is and intracavernosal injection of vasoactive agents, a history of penile trauma, abuse of narcotics or taking psychotropic medication or who had used PDE5is for recreational purposes without any medical reasons (2).

Sickle cell disease is the most common aetiology of ischaemic priapism in childhood, accounting for 63% of the cases. It is the primary aetiology in 23% of adult cases of priapism (9), with a lifetime probability of developing ischaemic priapism of 29-42% in men with sickle cell disease (2,9,10) (LE: 4). Mechanisms of sickle cell disease associated priapism in the human penis may involve dysfunctional nitric oxide synthase and ROCK signaling, and increased oxidative stress associated with NADPH oxidase mediated signaling (11).

Priapism resulting from metastatic or regional infiltration is not widely studied or reported. The cases in the literature seem to indicate that this is an infiltrative and not a haemodynamic process like ischaemic or high flow priapism (12). As such the recommendations for pharmacological treatment likely will not work and certainly all of these men should be imaged with magnetic resonance imaging (MRI) and offered supportive care for their primary cancer.

Table 3: Potential causative factors for ischaemic priapism

•	Idiopathic
•	Haematological dyscrasias (sickle cell disease, thalassemia, leukaemia; multiple myeloma, Hb Olmsted variant, fat emboli during hyperalimentation, haemodialysis, glucose-6-phosphate dehydrogenase deficiency, Factor V Leiden mutation)
•	Infections (toxin-mediated) (i.e. scorpion sting, spider bite, rabies, malaria)
•	Metabolic disorders (i.e. amyloidosis, Fabry's disease, gout)
•	Neurogenic disorders (i.e. syphilis, spinal cord injury, cauda equina syndrome, autonomic neuropathy, lumbar disc herniation, spinal stenosis, cerebrovascular accident, brain tumour, spinal anaesthesia)
•	Neoplasms (metastatic or regional infiltration) (i.e. prostate, urethra, testis, bladder, rectal, lung, kidney)
•	Medications
o	Vasoactive erectile agents (i.e. papaverine, phentolamine, prostaglandin E1/alprostadil, combination of intracavernous therapies)
o	Alpha-adrenergic receptor antagonists (i.e. prazosin, terazosin, doxazosin, tamsulosin)

o	Antianxiety agents (hydroxyzine)
o	Anticoagulants (heparin, warfarin)
o	Antidepressants and antipsychotics (i.e. trazodone, bupropion, fluoxetine, sertraline, lithium, clozapine, risperidone, olanzapine, chlorpromazine, thiorizadine, phenothiazines)
o	Antihypertensives (i.e. hydralazine, guanethidine, propranolol)
o	Hormones (i.e. gonadotropin-releasing hormone, testosterone)
o	Recreational drugs (i.e. alcohol, marijuana, cocaine [intranasal and topical], crack, cocaine)

3.2   Arterial (high flow or non-ischaemic) priapism

Epidemiological data on arterial priapism are almost exclusively derived from small case series (2,13-16). The usual cause of high flow priapism is blunt perineal trauma (17). The injury results in a laceration in the cavernosal artery leading to a high-flow fistula between the artery and the lacunar spaces of the sinusoidal tissue (15). This unregulated flow results in a persistent erection, probably via a mechanism that involves stimulation of endothelial nitric oxide synthase by the turbulent blood flow (18). Partial erections are enhanced after sexual stimulation, as the trabecular smooth muscle fully relaxes, activating the corporal veno-occlusive mechanism (15,19).

There is often a delay between the injury and the development of the priapism that may be up to 2-3 weeks (19). This reflects either spasm or ischaemic necrosis of the injured artery with the fistula only developing as the spasm resolves or when the ischaemic segment blows out.

Occasional cases are associated with metastatic malignancy to the penis (20,21), with acute spinal cord injury (22) and occasionally following intracavernosal injections or aspiration (23,24). Under these circumstances, it may complicate low-flow priapism. It has also been reported to occur following internal urethrotomy (25) and a Nesbit procedure (26). Although sickle cell disease is usually associated with low-flow priapism, occasional cases of high-flow priapism have been reported (27). High-flow priapism may be a consequence of repeated invasive procedures performed in attempt to reverse ischaemia.

3.3   Stuttering (recurrent or intermittent) priapism

Epidemiological studies of stuttering priapism are lacking. Our current understanding of this stressful entity has been derived from observations in men with sickle cell disease in which the incidence of priapism is high (28,29). Recurrent priapism episodes occur in men with sickle cell disease in between 42 and 64% (30,31). In a multicentre study that involved 98 boys (adolescents and young men with sickle cell disease) ranging in age from 5 to 20 years, the incidence of priapism was 35%, of whom 72% had a history of stuttering priapism (28).

The aetiology of stuttering priapism is similar to that of ischaemic priapism. Sickle cell disease is the most common cause of stuttering priapism. The cause can also be idiopathic and rarely due to a neurological disorder. Moreover, men who have suffered from an acute ischaemic priapic event, especially one which has been prolonged (more than 4 hours) may be at risk for developing stuttering priapism (32). The underlining mechanism is similar to that of other types of ischaemic priapism: a deficiency of endothelial nitric oxide in the penis causes down-regulation of its specific downstream effectors, a cyclic guanosine monophosphate (cGMP)-dependent protein kinase including phsosphodiesterase type 5 dysregulation (33,34). Under this condition, the control system of corporal smooth muscle tone is functioning at a low point. Hence, the response to any sexual or non-sexual stimulus, such as that which can occur during rapid eye movement sleep, will induce a prolonged erectile episode.

Recently, several studies have emerged, proposing novel mechanisms for the occurrence of this entity, specifically in patients with sickle cell disease. These studies postulate that factors involved in pathways affecting inflammation, cellular adhesion, nitric oxide metabolism, vascular reactivity and coagulation may all play a role in the pathophysiology of this entity (2,11,35-37).

3.4 Conclusions on the epidemiology and pathophysiology of priapism

	LE
Ischaemic priapism is most common, accounting for more than 95% of all cases	1b
Ischaemic priapism is identified as idiopathic in the vast majority of patients, while sickle cell anaemia is the most common cause in childhood	1b
Ischaemic priapism occurs relatively often (up to 35%) after intracavernous injections of papaverine-based combinations while it is rare (< 1%) after prostaglandin E1 monotherapy	2a
Priapism is rare in men who have taken phosphodiesterase type 5 inhibitors (PDE5is) with only sporadic cases reported	1a
Arterial priapism usually occurs after blunt perineal trauma	2
Stuttering priapism has the same aetiology as the ischaemic type, with sickle cell disease being the most common cause. But the cause can also be idiopathic and in rare cases may be due to a neurological disorder	3

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4. DIAGNOSTIC EVALUATION OF PRIAPISM

4.1 History

A comprehensive history is the mainstay in priapism diagnosis (1,2). The medical history must include a history of sickle cell disease or other haematological abnormality (3,4) and a history of pelvic, genital or perineal trauma. The sexual history must include complete details of the duration of erection, the presence and degree of pain, prior medical drug use, any previous history of priapism and erectile function prior to the last priapism episode (Table 4).

The history can help to determine the underlying type of priapism (Table 5). Ischaemic priapism is associated with progressive penile pain and the erection is rigid. With most cases of ischaemic priapism of idiopathic origin, the patient history may reveal one of the causes presented in Table 3.

Arterial priapism is suspected when there is no pain and erections are not fully rigid. It can be associated with full erections under sexual stimulation and there is a history of coital trauma or blunt trauma to the penis. The onset of post-traumatic high-flow priapism in adults and children may be delayed by hours to days following the initial injury. Sexual intercourse is usually not compromised.

The history of stuttering priapism is characterized by recurrent episodes of prolonged erections, usually non-resolving morning erections. The onset of the priapic episodes usually occurs during sleep and detumescence does not occur upon waking. Generally, these priapic episodes are not painful and only cause the patient to seek medical help when the discomfort interferes with daily life.

The patient usually presents following several recurring priapic episodes. Upon investigation, there is no obvious underlying aetiology (such as a blood dyscrasia or medications), which has been noticed by the patient to precipitate the event. Stuttering priapism can also affect the general well-being of the patient who may become worried when he engages in sexual activity. Furthermore, some of these men may develop a prolonged episode of full-blown, low-flow, priapism, which could potentially require emergency medical intervention (5).

Table 4: Key points in taking the history of priapism (adapted from Broderick et al [1])

•	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

4.2 Physical examination

Physical examination of the genitalia, the perineum and the abdomen must be included in the diagnostic evaluation of priapism (1,2). In ischaemic priapism, the corpora are fully rigid and tender but the glans penis is soft. In arterial priapism, the corpora are tumescent but not fully rigid (Table 5). Abdominal and perineal examination may reveal evidence of trauma or malignancy.

4.3 Laboratory testing

Laboratory testing should include a complete blood count, white blood count with blood cell differential, platelet count and coagulation profile to assess anaemia, detect haematological abnormalities and to make sure that the patient can safely tolerate any necessary surgical interventions. (1,2). Blood aspiration from the corpora cavernosa shows bright red arterial blood in arterial priapism, while blood is dark in ischaemic priapism (Table 5) (LE: 2b). Blood gas analysis is essential to differentiate between arterial and ischaemic priapism (Table 6). Further laboratory testing should be directed by history, clinical and laboratory findings. These may include specific tests for the diagnosis of sickle cell anaemia or other haemoglobinopathies (e.g. haemoglobin electrophoresis) or urine and plasma toxicological studies when there is suspected use of recreational psychoactive drugs.

4.4 Penile imaging

Colour duplex ultrasound (US) of the penis and perineum is recommended in the evaluation of arterial priapism because it can identify approximately 70% of cases and can differentiate arterial from ischaemic priapism as an alternative or adjunct to blood gas analysis (6-8) (LE: 2b).

Ultrasound should be performed in the lithotomy position and examination of the entire penile shaft and perineum is recommended. In arterial priapism US will show turbulent flow at the fistula, which helps to localize the site of trauma since patients with arterial priapism have normal to high blood velocities in the cavernous arteries, while patients with ischaemic priapism will have no blood flow in the cavernous arteries. The return of the cavernous artery waveform will accompany successful detumescence (1,6,9). Colour duplex US of the penis should be performed before aspiration in ischaemic priapism. After aspiration, a reactive hyperaemia may develop with a high arterial flow that may mislead the diagnosis as arterial priapism.

A pudendal arteriogram in selected patients can reveal a characteristic blush at the site of the injury to the cavernosal artery in patients with arterial priapism (10,11). However, due to its invasiveness and the lack of availability of colour duplex US, it should be reserved for the management of arterial priapism, when embolization is undertaken (1,2) (LE: 3).

The role of MRI in the diagnostic evaluation of priapism is still controversial. In arterial priapism, its role is limited since the small penile vessels and arteriovenous fistulae cannot be easily demonstrated (12). On the contrary, it may helpful in cases of ischaemic priapism to assess the viability of the corpora cavernosa and the presence of penile fibrosis (13). In a prospective study in 38 patients with ischaemic priapism, the sensitivity of MRI in predicting non-viable smooth muscle was 100%, as confirmed by corporal biopsy (14). In this study, all patients with viable smooth muscle on MRI maintained erectile function on clinical follow-up (LE: 3).

Table 5: Key findings in priapism (adapted from Broderick et al [1])

	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 6: Typical blood gas values (adapted from Broderick et al [1])

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

4.5 Recommendations for the diagnosis of priapism

Recommendation	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)	B
Colour duplex US of the penis and perineum is recommended for the differentiation between ischaemic and arterial priapism and for localization of the site of fistula in arterial 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 embolization is undertaken	B

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5.   MANAGEMENT OF PRIAPISM

5.1   Management of ischaemic priapism

Acute ischaemic priapism is an emergency condition and rapid intervention is compulsory (LE: 4), and should follow a stepwise approach. The aim of any treatment is to restore penile flaccidity, without pain, in order to prevent eventual chronic damage to the corpora cavernosa. In many cases, penile oedema may persist, with ecchymosis and partial erection, which could eventually mimic unresolved priapism.

5.1.1   *First-line treatments*

First-line treatments in ischaemic priapism of more than 4 hours duration are highly recommended before any surgical treatment (LE: 4). Conversely, first-line treatments initiated beyond 72 hours may benefit in relieving the unwanted erection and associated pain, but have little documented benefit in terms of potency preservation (LE: 4).

Several first line treatments have been described historically including exercise, ejaculation, ice packs, cold baths, and cold water enemas (1). However, there is lack of evidence on the efficacy of such measures. The so-called 'simpler cases' of drug-induced priapism are typically caused by a single intracavernosal administration of a drug, such as alprostadil. The first step in treatment of this case can be the direct injection of a sympathomimetic agent (most often, phenylephrine or etilephrine), using a 30G needle, without prior aspiration of blood from the corpora cavernosa (LE: 4). The outcome of the intracavernosal injection may be improved by massaging the corpora cavernosa in a 'milking' manoeuvre in order to aid distribution of the sympathomimetic agent (LE: 4). However, these 'simpler cases' can be successfully treated with blood aspiration alone (LE: 4).

5.1.1.1   *Penile anaesthesia/systemic analgesia (as indicated)*

It is possible to perform blood aspiration and intracavernosal injection of a sympathomimetic agent without any anaesthesia. However, anaesthesia may be necessary when there is severe penile pain. The treatment options for penile anaesthesia/systemic analgesia include:
•   dorsal nerve block; however, this is not able to reduce a painful sensation caused by high intracavernous pressure;
•   circumferential penile block;
•   subcutaneous local penile shaft block;
•   oral conscious sedation (for paediatric patients).

5.1.1.2   *Aspiration ± irrigation with normal saline solution*

The first intervention for an episode of priapism lasting more than 4 hours consists of corporal aspiration (LE: 4) to drain stagnant blood from the corporal bodies, thus making it possible to relieve the compartment syndrome-like condition of the penis.

Decompression of the corpora cavernosa usually promotes the recovery of intracorporal blood circulation, which should result in the relief of penile pain and counteract local acidotic and anoxic metabolic derangements caused by the priapism itself. Blood aspiration may be performed with intracorporeal access through the glans or with a percutaneous needle access on either lateral aspect of the proximal penile shaft, using either a 16G or 18G angiocatheter or butterfly needle. The needle must penetrate the skin, the subcutaneous tissue and the tunica albuginea to eventually drain the priapic corpus cavernosum (LE: 4). Some clinicians use two angiocatheters or butterfly needles at the same time to accelerate drainage, as well as aspirating and irrigating simultaneously with a saline solution (2) (LE: 4). Overall, aspiration must be continued until fresh red, oxygenated, blood is aspired (LE: 4).

The aspiration of corporal blood, with or without saline irrigation, has up to a 30% chance of promoting penile detumescence and thus terminating priapism. Overall, there are insufficient data to conclude that aspiration followed by saline intracorporeal irrigation is more effective than aspiration alone (LE: 4).

5.1.1.3 Aspiration ± irrigation with normal saline solution in combination with intracavernosal injection of pharmacological agents

This combination is currently considered the standard of care in the treatment of ischaemic priapism (1,3,4) (LE: 4). Pharmacological agents include sympathomimetic drugs or alpha-adrenergic agonists with a resolution rate of up to 80%.

Options for intracavernosal sympathomimetic agents include phenylephrine, etilephrine, ephedrine, epinephrine, norepinephrine and metaraminol (1,4-11) (LE: 2b). The use of intracavernosal adrenalin injection alone has also been sporadically reported (12). Overall, the specific agent used may depend upon pharmaceutical availability, according to geographical diversity.

5.1.1.3.1 Phenylephrine

Phenylephrine has been suggested as the drug of choice due to its high selectivity for the alpha-1-adrenergic receptor, without concomitant beta-mediated ionotropic and chronotropic cardiac effects (5-7). In this context, although there have been no comparative trials of sympathomimetic agents in the management of priapism, phenylephrine is widely considered to be the agent of choice (LE: 4).

A chart for the extemporaneous preparation of dilutions of alpha-adrenergic agonists for intermittent injection or irrigation has been proposed (13). 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 (LE: 4). A lower concentration or volume is applicable for children and patients with severe cardiovascular disease (LE: 4).

Phenylephrine use is limited due to the potential systemic cardiovascular side effects (1,4-8) and it is therefore recommended that vital signs (blood pressure and pulse) are measured before and after injection, and monitored every 15 minutes (9). This is particularly important in older men with existing cardiovascular diseases. After injection, the puncture site may be compressed and the corpora cavernosa massaged to facilitate drug distribution.

Ischaemic priapism patients may not respond properly to conventional doses of phenylephrine, potentially due to the attenuated, contractile response, which is a consequence of hypoxia and acidosis (14-16), so that higher doses may be required to achieve penile detumescence. Preclinical data (6,7) report widespread apoptosis of the cavernosal smooth muscle preventing further contraction (7). Clinical benefit is therefore from repeated doses at various time intervals or high-dose phenylephrine administration in men, i.e. up to a total cumulative dose of 50,000 µg (6), especially in younger men without any cardiovascular risk factors (LE: 3).

The potential treatment-related side effects of intracavernous phenylephrine (and other sympathomimetic agents) include headache, dizziness, hypertension, reflex bradycardia, tachycardia and palpitations, irregular cardiac rhythms (men with a high cardiovascular risk should be more accurately monitored with an electrocardiogram) and sporadic subarachnoid haemorrhage (8). Overall, the administration of intracavernosal sympathomimetic agents is contraindicated in patients suffering from malignant or poorly controlled hypertension and in those who are concurrently taking monoamine oxidase inhibitors (LE: 4).

5.1.1.3.2 Etilephrine

Etilefrine is the second most widely used sympathomimetic agent, administered by intracavernosal injection at a concentration of 2.5 mg in 1-2 L normal saline (1,3,4,17-19) (LE: 3).

5.1.1.3.3 Methylene blue

Methylene blue is a guanylate cyclase inhibitor, which may be a potential inhibitor of endothelial-mediated cavernous relaxation. It has therefore been suggested for treating short-term pharmacologically induced priapism (20,21) (LE: 3). Methylene blue, 5 mL or 100 mg (21), should be injected intracavernously and left for 5 minutes. It is then aspirated and the penis compressed for an additional 5 minutes (20). Treatment-related side effects include a transient burning sensation and blue discolouration of the penis following injection of methylene blue.

5.1.1.3.4 Adrenaline

Intracavernosal adrenaline alone (dosage of 2 mL of 1/100,000 adrenalin solution up to five times throughout a 20-minute period [12]), has been used as first-line treatment in patients with ischaemic priapism, which was mainly due to an intracavernosal injection of vasoactive agents. Success rate of over 50% after a single injection, with an overall success rate of 95% with repeated injections is achieved. A combined alpha- and

beta-adrenergic effect on the venous system was assumed to be the underlying mechanism of action (LE: 3). There are no reports of major side effects or erectile dysfunction.

5.1.1.3.5 Oral terbutaline
Oral terbutaline is a beta-2-agonist with minor beta-1 effects and some alpha-agonistic activity. A dosage of 5 mg has been suggested to treat ischaemic priapism lasting more than 2.5 hours after intracavernosal injection of vasoactive agents, with a mechanism of action not adequately elucidated (22-24) (LE: 1b). In men with sickle cell disease, the vascular relaxation may allow oxygenated arterial blood to enter the corpora cavernosa, which then washes out the stagnant sickle cells. However, the mechanism of action is not clear. Terbutaline should be given cautiously in patients with coronary artery disease, increased intravascular fluid volume, oedema and hypokalaemia (24).

5.1.1.4   Management of sickle cell disease related priapism
Rapid intervention is compulsory (LE: 4). This approach is similar to the previously described in other cases of ischaemic priapism (25-27) (LE: 4). As with other haematological disorders, other therapeutic practices may also need to be implemented (26-28).

Specific measures for sickle cell disease related priapism include the administration of intravenous hydration and parental narcotic analgesia while preparing the patient for aspiration and irrigation. In addition, supplemental oxygen administration is required and alkalinization with bicarbonate (25,29). Exchange blood transfusion has been also proposed, with the aim of increasing the tissue delivery of oxygen.
Once it has been decided to transfuse blood, the transfused blood should be Hb S negative, Rh and Kell antigen matched (30). However, the evidence is not sufficiently robust to conclude that exchange transfusion itself promotes resolution of the state of priapism, as defined by an acceleration of time to detumescence, in men with sickle-cell disease. It should also be noted that several reports suggest that this treatment may result in serious neurological sequelae (31). Because of these considerations, the routine use of this therapy cannot be recommended by the Expert Panel (LE: 4).

5.1.2   Second-line treatments
Second-line intervention typically refers to surgical intervention in the form of penile shunt surgery. In an acute situation, surgery for ischaemic priapism should be considered only when conservative management options fail, with the specific purpose of relieving penile ischaemia and to lessen any pathological sequelae in highly difficult presentations of major ischaemic priapism (LE: 4).
However, there is no experimental evidence detailing the amount of time allowed for first-line treatment before moving on to a second-line therapy. Overall, the consensus recommendations suggest a course of first-line treatment of at least 1 hour prior to moving to surgery (LE: 4). However, this time interval could be longer in cases with partial success to first-line treatments.
A number of clinical indicators may suggest failure of first-line treatment and the persistence of the priapism; continuing corporal rigidity, acidosis and anoxia by cavernous blood gas testing, absence of cavernosal artery inflow by penile colour duplex US, or elevated intracorporal pressures by pressure monitoring (LE: 4).

5.1.2.1   Penile shunt surgery
Penile shunt surgery aims to restore an exit for blood from the corpora cavernosa and at the same time to re-establish blood circulation within these structures. For this purpose, any shunt creates an opening in the tunica albuginea of the corpora cavernosa, which may eventually communicate with the glans, the corpus spongiosum, or a vein for blood drainage (1,4,32). In 2009, the International Society for Sexual Medicine (ISSM) Standards Committee stated that shunting should be considered for priapism events lasting 72 hours (LE: 4) (1).

In general, the type of shunt procedure chosen is suggested by the surgeon's preference and procedure familiarity (LE: 4). It is preferable for distal shunt procedures to be tried before proximal shunting is considered (LE: 4). However, the efficacy of this treatment strategy is questionable and cavernous biopsy may be considered to diagnose muscle necrosis. Moreover, considering the type of surgery to treat refractory ischaemic priapism, which has failed any medical, less-invasive approach, it is compulsory to consider:
•   the clinical feature of the disease (aetiology and duration)
•   types and number of previous/failed treatments
•   success rates of the proposed intervention
•   potential risk of complications
•   technical ease of the suggested procedure and the surgeon's familiarity with it (LE: 4).

In every day practice, it is important to immediately assess the success of any type of surgical correction of ischaemic priapism. This is done either by direct observation (i.e. penile detumescence or oxygenated red blood discharging from the corpora cavernosa) or assessment using different techniques (i.e. cavernous blood gas testing, penile colour duplex US, intracoporal pressure monitoring) or by using the penile compression manoeuvre (squeeze and release) (LE: 4) (1,4).

The postoperative recovery rates of erectile function in men submitted to shunt surgery for prolonged erections are very low (33,34). Priapism events prolonged for more than 36 hours appear to impair irreversibly erectile tissue both structurally and functionally (34). Overall, it has been considered that in patients suffering from major ischaemic priapism (lasting continuously for a prolonged time ≥ 36 hours), any shunt procedure may only serve to limit pain sensations, without adequately preserving erectile functioning (LE: 4).

Four categories of shunt procedures have been reported (1,3,32). The limited available data preclude a recommendation of a greater efficacy for one procedure over another based on accurate outcome estimates (LE: 4).

5.1.2.1.1 Percutaneous distal (corporoglanular) shunts
Winter's procedure: This procedure uses a biopsy to create a fistula between the glans penis and each corpora cavernosa body (1,3,35-37) (LE: 3). Postoperative sequelae are uncommon (38). Winter's shunt is relatively easy to perform, but has been reported as the least successful operation to create a distal shunt (33).

Recently, a modification of Winter's shunt has been proposed in paediatric patients with refractory ischaemic priapism. Multiple punctures are made in both the corporal bodies by partial withdrawal of the needle and changing the direction of its tip. After removal of the needle, the puncture wound in the glans is closed (39). No major complications have been reported with this modification (LE: 3).

Ebbehoj's technique: This technique involves the execution of multiple tunical incision windows between the glans and each tip of the corpus cavernosum by means of a number size 11 blade scalpel passed several times percutaneously (1,3,37,40,41) (LE: 3).

Lue's procedure: This technique involves performing either a unilateral or bilateral T-shunt procedure using a number size 10 blade scalpel placed vertically through the glans until fully within the corpus cavernosum. The blade is then rotated 90 degrees away from the urethra and pulled out (1,3,37,42) (LE: 3). The whole tunnelling procedure could be performed using ultrasonographic guidance, mainly in order to avoid urethral injury (42).

Relative contraindications are:
•   bleeding diathesis
•   phimosis (since a dorsal slit will be required to expose the glans)
•   narrow penis with a corporal diameter that will not accommodate a number size 10 blade (42).

5.1.2.1.2 Open distal (corporoglanular) shunts
Al-Ghorab's procedure: This procedure consists of an open excision of circular cone segments of distal tunica albuginea, along with a subsequent skin closure by means of a running suture with absorbable material (1,3,37,43,44) (LE: 3).

Burnett's technique: In cases of highly refractory ischaemic priapism, even after less invasive distal penile shunt procedures, a modification of the Al-Ghorab corporoglanular shunt surgery involves the retrograde insertion of a 7/8 Hegar dilator into the distal end of each corpus cavernosum through the original Al-Ghorab glanular excision.
        After removal of the dilator from the corpus cavernosum, blood evacuation is facilitated by manual compression of the penis sequentially from a proximal to distal direction. After detumescence, the glans penis skin is closed as in the Al-Ghorab procedure (1,3,37,45,46) (LE: 3). Reported complications included wound infection, penile skin necrosis and an urethrocutaneous fistula (46). Erectile function was not preserved in all patients (45-47), but this is mostly thought to be due to the priapism duration rather than the treatment.

Recently, a further modification of these two open distal (corporoglanular) shunts has been suggested, using the Al-Ghorab distal shunt combined with cavernous tunnelling with blunt cavernosotomy (with a Pean forceps) to create a large blood drainage route by removing the necrotic or fibrous cavernous tissues (48).

5.1.2.1.3 Open proximal (corporospongiosal) shunts
Quackles's technique: Through a trans-scrotal or perineal approach, a proximal open shunt technique creates a communication between the corpus cavenosum and the corpus spongiosum. The most frequent complications include an unwanted urethra-cavernous fistula, a urethral stricture or the development of cavernositis (1,3,32,49) (LE: 3).

Sacher's technique: This comprises a bilateral performance of the Quackles procedure, together with staggered corpora cavernosa-corpus spongiosum shunts to reduce the risk of urethral stricture adjacent to the shunts (1,3,32,50) (LE: 3).

5.1.2.1.4 Vein anastomoses/shunts
Grayhack's procedure: This mobilizes the saphenous vein below the junction of the femoral vein and anastomoses, the vein end-to-side in the corpus cavernosum. Venous shunts may be complicated by saphenofemoral thrombus formation and pulmonary embolism (1,3,51-53) (LE: 3).

Barry's procedure: A venous bypass is created between the corpus cavernosum and either the deep or the superficial dorsal vein through a small surgical field, without requiring saphenous vein mobilization (1,3,54) (LE: 3).

5.1.2.2 Immediate surgical prosthesis implantation
Intractable, therapy-resistant, acute ischaemic priapism or episodes lasting more than 48-72 hours usually result in complete erectile function impairment, along with possible major penile deformity. In these cases, immediate penile prosthesis surgery has been suggested (55-58) (LE: 3).

The immediate insertion of a penile prosthesis has been recommended to avoid the difficulty of surgery and the risk of complications, e.g. urethral injury, tunical erosions, infection and/or penile shortening, which may occur whenever surgery is performed some time after long-term corporal fibrosis has already developed. Potential complications that could compromise immediate penile prosthesis implantation include distal erosion and cavernositis (56,58), along with a mild rate of revision surgery (56).

However, there are no clear indications for immediately implanting a penile prosthesis in a man with acute ischaemic priapism (4). In this context, penile prosthesis (either malleable or a three-piece inflatable prosthesis) at the time of presentation could be taken into consideration if (1) (LE: 4):
•   ischaemia has been presented for more than 36 hours (mainly in sickle cell disease patients) (55);
•   aspiration and sympathomimetic intracavernous injections have failed;
•   distal and proximal shunting have failed.
Overall, an MRI prior to surgery or corporal biopsy at implant time is highly recommended to document corporal smooth muscle necrosis (1,56) (LE: 4).

5.1.2.3 Surgery for non-acute sequelae after ischaemic priapism
Structural changes may occur after ischaemic priapism: They include penile scarring, megalophallic deformities, penile shortening, and possible penile loss, which result after cavernosal tissue necrosis and fibrosis (32,56,59,60). Erectile function impairment is also often observed after ischaemic priapism (1,61). Unfortunately, these outcomes may still occur despite the successful resolution of priapism following an effective first-line or second-line treatment.

Penile prosthesis surgery: Prosthesis implantation is indicated for patients who are unable to perform sexual intercourse due to severe erectile dysfunction. In particular in sickle cell patients, since other therapeutic options to promote erectile functioning (e.g. PDE5is, intracavernosal injections and vacuum erection devices) are avoided in case they may provoke a further priapism event (1,4).

In severe corporal fibrosis, inserting semi-rigid prosthetic devices is preferable to an inflatable implant (56,62) (LE: 3). Overall, due to the challenges of corporal fibrosis, early implantation (6-18 months after ischaemic priapism) has been promoted mainly in men with sickle cell disease (32,63,64). This reduces the risks of procedural complications, e.g. urethral injury, tunical erosions, infection.

Penile reconstructive surgery: Specialized surgical techniques may be required following severe priapism that has resulted in penile destruction with complicated deformities or even loss of penile tissue. In these circumstances, penile reconstruction and concomitant prosthesis implant may be considered (63) (LE: 3).

5.1.3   Recommendations for the treatment of ischaemic priapism

Recommendations	GR
Ischaemic priapism is an emergency condition and rapid intervention is compulsory	B
The specific aim of any emergent treatment is to retrieve penile flaccidity, without pain, in order to prevent eventual 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
The first step in the management of ischaemic priapism is decompression of the corpora cavernosa by penile aspiration until fresh red blood is obtained. In drug-induced priapism after intracavernous injections of vasoactive agents for the treatment of erectile dysfunction, blood aspiration can be replaced by intracavernous injection of a sympathomimetic drug as the first step	C
In priapism recurrence after 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 case of priapism recurrence after aspiration and intracavernous injection of a sympathomimetic drug, these steps should be repeated several times before considering surgical intervention. No clear recommendation for the highest phenylephrine dose to be administered can be given	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 alkalization with bicarbonates, blood exchange transfusions) but these should not delay initial treatment	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 is recommended. Implantation of penile prosthesis at a later stage can be difficult due to severe corporal fibrosis	B

5.2   Management of arterial priapism

The management of high-flow priapism is not an emergency because the penis is not ischaemic. Definitive management can therefore be considered and should be discussed with the patient so that they understand the risks and complications of treatment (1,65) (LE: 3).

5.2.1   Conservative management

This may include applying ice to the perineum or site-specific perineal compression (66-69). It is an option in all cases, particularly children (70) (LE: 3). The fistula occasionally closes spontaneously. Even in those cases when it does not, the response to a sexual stimulus does allow for intercourse.

Blood aspiration is not an option for the treatment of arterial priapism and the use of alpha-adrenergic antagonists is not recommended due to potential severe adverse effects, e.g. transfer of the drug into the systemic circulation.

5.2.2 Selective arterial embolization

Selective arterial embolization can be performed using either an autologous clot (71-73), gel foam or sponge (72,74), or more permanent substances, such as coils (72,74-76) or acrylic glue (77) (LE: 3). Success rates of up to 89% have been reported (78) in relatively small, non-randomized studies. There are no robust data to demonstrate the relative merits of the different substances. At least, theoretically, the use of an autologous clot has some attractions. It temporarily seals the fistula, but when the clot is lysed, the arterial damage has usually resolved and the blood flow of the penis can therefore return to normal. The use of a permanent device, such as a coil, would permanently block an artery and may lead to adverse effects upon spontaneous sexual function. Other potential complications include penile gangrene, gluteal ischaemia, cavernositis and perineal abscess (1,79).

Following percutaneous embolization, a follow-up is appropriate within 1-2 weeks. Assessment by clinic examination and by colour duplex US can determine whether the embolization has been successful (80). If there is doubt, a repeat arteriogram is required. Recurrence rates of 7-27% after a single treatment of embolization have been reported (72,73,81) (LE: 3). In a few cases, repeat embolization is necessary. Sexual function following embolization can be adversely affected although there is full restoration of potency in around 80% of men (81,82) (LE: 3).

Embolization in children, although reportedly successful, is technically challenging and requires treatment within a specialist paediatric vascular radiology department (83,84).

5.2.3 Surgical management

Surgical treatment consists of selective ligation of the fistula through a transcorporeal approach under the guidance of colour duplex ultrasound (3,85,86). Although surgery has been successful in treating arterial priapism, it is technically challenging and may pose significant risks, mainly erectile dysfunction due to accidental ligation of the cavernous artery instead of the fistula. Nowadays, it is rarely performed and only in cases that have pseudocapsule formation around the fistula (which makes it easier to identify the fistula). It may also be considered when there are contraindications for selective embolization, no availability of the technique or embolization failure (LE: 4).

5.2.4 Recommendations for the treatment of arterial priapism

Recommendation	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	C
Selective artery embolization, using temporary or permanent substances, is the suggested treatment modality and has high success rates	B
The recurrence of arterial priapism following selective artery embolization requires the procedure to be repeated	B
The preservation rate of sexual function is about 80%. No definitive statement can be made on the best substance for embolization in terms of sexual function preservation	C
Selective surgical ligation of the fistula should be reserved as a last treatment option when embolization has failed	C

5.3 Management of stuttering priapism

The primary goal in the management of patients with stuttering priapism is the prevention of future episodes, which can usually be achieved pharmacologically. The management of each acute episode is similar to that for ischaemic priapism; aspiration/irrigation in combination with intracavernous injections of alpha-adrenergic agonists. Unfortunately, the efficacy and safety of the various treatment modalities reported in the medical literature are poorly characterized. Specifically, most reports are from small case series and the Expert Panel is not aware of any published, well-designed, controlled studies on the efficacy and safety of these treatments (25,29,87).

5.3.1 Hormonal manipulations of circulating testosterone

The aim of hormonal manipulation is to down-regulate circulating testosterone levels to suppress the action of androgens on penile erection (25,29,88). This can be done through the use of gonadotropin-releasing hormone

(GnRH) agonists or antagonists (89) (LE: 4). Potential side effects may include hot flushes, gynaecomastia, impaired erectile function, loss of libido and asthenia. Antiandrogens (i.e. flutamide, bicalutamide) (90,91) and oestrogens (92,93) are used to reduce circulating testosterone levels and have a similar efficacy profile to GnRH agonists or antagonists (LE: 4). However, the potential cardiovascular toxicity of oestrogens limits their clinical use.

5-alpha-reductase inhibitors (finasteride, dutasteride) block the conversion of testosterone to dihydrotestosterone. In a non-controlled study of 35 patients with sickle cell disease, finasteride (3 or 5 mg daily for 120 days) produced a significant decrease in the number of recurrent priapic episodes (94) (LE: 3). Ketoconazole, an antifungal agent that reduces adrenal and testicular androgen production, may also be a potential treatment for priapism (88,95) (LE: 4).

The duration of hormonal treatment for effective suppression of recurrent priapic events is still problematic. The length of treatment varies from weeks to years and depends on the agent type and investigator suggestions. Since this information has been derived from small case series in men with idiopathic stuttering priapism and patients with sickle cell disease, it is not possible to make any conclusions on the efficacy, dose and the duration of treatment. Moreover, hormonal agents have a contraceptive effect and interfere with normal sexual maturation. Caution is therefore strongly advised when prescribing hormonal treatments to prepubertal boys, adolescents or those men who are trying for their female partner to conceive. The side effects of these medications often result in castrate levels of testosterone, which have a contraceptive effect, interfere with growth, and significantly affect sexual function.

Of the hormonal agents suggested for preventing priapism, GnRH agonists and anti-androgens appear to be the most efficacious and safe. They are recommended as primary treatments for the management of stuttering priapism in adult men.

5.3.2 *Alpha-adrenergic agonists*
Studies of oral alpha-adrenergic agonists to treat stuttering priapism have suggested the use of limited daily dosing of these agents as effective prevention (96). Side effects of drug therapy (usually prescribed at bedtime) include tachycardia and palpitations.

Pseudoephedrine, widely used as an oral decongestant, can also be used as a first-line treatment (22). However, its effect on corporal smooth muscle is not fully understood. Etilefrine is an alpha-adrenergic agonist used successfully to prevent stuttering priapism due to sickle cell anaemia. It is taken orally at doses of 50-100 mg daily, resulting in response rates of up to 72% (97,98).

5.3.3 *Digoxin*
Digoxin (a cardiac glycoside and a positive inotrope) is used to treat patients with congestive heart failure. Digoxin regulates smooth muscle tone through a number of different pathways leading to penile detumescence (25,29,99). The use of maintenance digoxin doses (0.25-0.5 mg daily) in idiopathic stuttering priapism has been proven to reduce the number of hospital visits and to improve quality of life (25).

A small, clinical, double-blind, placebo-controlled study, using digoxin (0.25-0.5 mg daily) produced a decrease in sexual desire and excitement with a concomitant reduction in penile rigidity, regardless of any significant change in plasma levels of testosterone, oestrogens and luteinizing hormone (99) (LE: 2b). Common side effects may include a decreased libido, anorexia, nausea, vomiting, confusion, blurred vision, headache, gynaecomastia, rash and arrhythmia.

5.3.4 *Terbutaline*
Terbutaline, a beta-agonist that causes vasodilation, resulting in smooth muscle relaxation of the vasculature (25,29). Oral terbutaline prevents stuttering priapism with detumescence rates of 36% in patients with alprostadil-induced priapism (22) (LE: 3). The only randomized, placebo-controlled study (n = 68) in patients with pharmacologically-induced priapism, showed detumescence in 42% of the terbutaline-treated group compared to only 15% in the placebo-treated group (24) (LE: 1b). Common side effects include nervousness, shakiness, drowsiness, heart palpitations, headache, dizziness, hot flashes, nausea and weakness.

5.3.5 *Gabapentin*
Gabapentin has anticonvulsant, antinociceptive and anxiolytic properties and is widely used as an analgesic and antiepileptic agent. Its proposed mechanism of action is to inhibit voltage-gated calcium channels, which attenuates synaptic transmission (88), and reduces testosterone- and follicle-stimulating hormone levels (100).

It is given at a dose of 400 mg, four times a day, up to 2400 mg daily, until complete penile detumescence occurs, with subsequent maintenance administration of gabapentin, 300 mg daily (101) (LE: 4). Common side effects may include anorgasmia and impaired erectile function.

5.3.6 Baclofen

Baclofen is a gamma-aminobutyric acid (GABA) derivative that acts as a muscle relaxant and antimuscle spasm agent. It can inhibit penile erection and ejaculation through GABA activity and prevents recurrent reflexogenic erections or prolonged erections from neurological diseases (29). Oral baclofen has little efficacy and it is not usually used in stuttering priapism but intrathecal baclofen dosing is more effective (25,102-104) (LE: 4). Common side effects include drowsiness, confusion, dizziness, weakness, fatigue, headache, hypotension and nausea.

5.3.7 Hydroxyurea

Hydroxyurea blocks the synthesis of DNA by inhibiting ribonucleotide reductase, which has the effect of arresting cells in the S-phase (88,105). It is an established treatment for ameliorating sickle cell disease in most patients and improving their life expectancy (27,106). For patients with sickle cell disease and recurrent priapism, there is limited evidence to suggest a medical prophylactic role for hydroxyurea (LE: 3) (88,105,107). Potential side effects are oligospermia and leg ulcers.

5.3.8 Phosphodiesterase type 5 inhibitors (PDE5is)

Phosphodiesterase type 5 inhibitors act by increasing PDE5 function, i.e. by increasing the concentration of cGMP in the smooth muscle in a nitric oxide dysfunctional state. This state occurs in priapism in association with the underlying disease. It may result in a change in the nitric oxide pathway, producing downregulation of PDE5 in the penis and therefore preventing the complete degradation of cGMP in the corpora cavernosa (25,29,108,109).

Low doses of PDE5is (sildenafil, 25 mg daily, or tadalafil, 5 mg three times weekly) have a paradoxical effect in alleviating and preventing stuttering priapism, mainly in patients with idiopathic and sickle cell disease associated priapism (25,29,108-112) (LE: 3). When using PDE5is to treat priapism, it is important to remember that therapy should be started only when the penis is in its flaccid state and not during an acute episode of priapism. There is a delay of one week after starting systemic PDE5is dosing before treatment is effective. There are no reported impairments in male sexual function (LE: 3).

5.3.9 Intracavernosal injections

Some patients with stuttering priapism, who have been started on systemic treatments to prevent recurrence of unwanted erections, may not see therapeutic effects immediately. They may temporarily require intracavernous self-injections at home with sympathomimetic agents, until ischaemic priapism has been alleviated (25,29).

The most commonly used drugs are phenylephrine and etilephrine (as described above in the treatment of ischaemic priapism) (1,3,17,98). Metaraminol (a long-acting potent alpha-1- beta-l-receptor agonist with vasoconstrictive properties) has been also suggested for treatment of stuttering priapism episodes (113) (LE: 3).

Common side effects may include hypertension, coronary ischaemia and cardiac arrhythmias. Tissue plasminogen activator (TPA) is a secreted serine protease that converts the proenzyme plasminogen to plasmin, which acts as a fibrinolytic enzyme. Limited clinical data have suggested that a single intracavernosal injection of TPA can successfully treat patients with recalcitrant priapism (88,114) (LE: 3). Mild bleeding is the most commonly observed side effect.

5.3.10 Recommendations for the treatment of stuttering priapism

Recommendation	GR
The primary goal in the management of patients with stuttering priapism is the prevention of future episodes, which can generally be achieved pharmacologically. 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

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6.   ABBREVIATIONS USED IN THE TEXT

This list is not comprehensive for the most common abbreviations.

cGMP   cyclic guanosine monophosphate
EAU   European Association of Urology
GABA   gamma-aminobutyric acid
GnRH   gonadotropin-releasing hormone
GR   grade of recommendation
LE   level of evidence
MRI   magnetic resonance imaging
PDE5is   phosphodiesterase type 5 inhibitors
RCT   randomized controlled trial
TPA   tissue plasminogen activator
US   ultrasound