Journal of Pediatric Urology (2014) 10, 11—25

Journal of Pediatric urology

REVIEW ARTICLE

Priapism in children: a comprehensive review and clinical guideline

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Received 8 May 2013; accepted 30 July 2013
Available online 8 September 2013

KEYWORDS
Priapism;
Prolonged erections;
Leukaemia;
Sickle cell disease;
Trauma

Abstract *Objective:* We review the English literature between 1980 and 2013 and summarize the clinical classification, aetiology, physiology, and pathophysiology of paediatric priapism. We propose a clinical guideline for the management of priapism in children.
Patients: Male patients aged ≤18 years.
Results: Priapism, a prolonged penile erection lasting >4 h, is a rare condition in childhood. There are 3 widely accepted types of priapism: 1) ischaemic priapism, the commonest type seen in children; 2) stuttering priapism, recurrent, self-limiting prolonged erections; and 3) non-ischaemic priapism, rare in children, usually due to trauma. Neonatal priapism has also been described.

Ischaemic priapism is a urological emergency causing fibrosis of the corpora cavernosa, subsequent erectile dysfunction and penile disfigurement. The commonest causes of priapism in children are sickle cell disease (65%), leukaemia (10%), trauma (10%), idiopathic (10%), and pharmacologically induced (5%).
Conclusions: Priapism in children must be assessed urgently. Rapid resolution of ischaemic priapism prevents permanent cavernosal structural damage and is associated with improved prognosis for potency later in life. Stuttering priapism requires careful counselling for episodic management. Chronic prophylaxis may be obtained using α-adrenergic sympathomimetics, phosphodiesterase type 5 inhibitors and, in sickle cell disease, hydroxyurea. Non-ischaemic and neonatal priapism may generally be treated less urgently.

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1477-5131/$36
http://dx.doi.org/10.1016/j.jpurol.2013.07.024

Nomenclature

Priapism a prolonged erection of the penis unrelated to sexual stimuli lasting four or more hours.
Ischaemic priapism a prolonged erection of the penis lasting four or more hours associated with ischaemia of the corpora cavernosa.
Non-ischaemic priapism a prolonged erection of the penis lasting four or more hours not associated with ischaemia.
Stuttering priapism recurrent, self-limiting prolonged erections, which may precede an episode of ischaemic priapism.
Neonatal priapism a prolonged erection of the penis lasting four or more hours in a new-born infant.

Introduction

Priapism (International Classification of Diseases N48.3) [1] is a prolonged full or partial penile erection lasting ≥4 h unrelated to sexual stimulus [2]. Priapism is a urological emergency. Its management aims to prevent penile disfigurement/shortening, erectile dysfunction (ED) and psychological sequelae [3]. There are currently no widely accepted guidelines on the management of priapism in children.

The term priapism is derived from the Greek and Roman mythological figure Priapus, son of Aphrodite and God of fertility who was depicted with a giant erect phallus and was a symbol for male generative power [4]. References to priapism are made in Ebers' ancient Egyptian papyrus, and priapism was first described in the modern literature in 1616 by Petraens [4]. However, it was not until 260 years later that priapism was first reported in a child [5]. Priapism of the clitoris ("clitorism") has also been described in a child but is beyond the scope of this article.

In this article we comprehensively review the literature and describe the clinical classification, aetiology, physiology, pathophysiology and current management regimens of priapism in children. We suggest an algorithm for the clinical assessment and treatment of priapism in children.

Materials and methods

The electronic resources PubMed, Embase, Cochrane Database, and the Database of Abstracts and Reviews of Effects (DARE) were searched (1980—2013) using the medical subject heading (MeSH) "priapism". Limitations included "all children (birth-18 years)" and English-language. All articles' abstracts were read to select significant articles for full text. All case series and significant case reports were identified and comprehensively reviewed. Where the relevance of a paper was unclear a review of the full paper was undertaken.

The salient literature on priapism in adults was also reviewed including a PubMed search for the MeSH term "priapism" (2003—2013) for article types: clinical trial, guideline, meta-analysis, multicentre study, practice guideline, randomized controlled trial, review, and systematic review. The limitations adult (≥19 years) and English language were also applied. The references of selected papers were reviewed to identify potentially significant articles not included in the initial searches.

Results

A total of 337 abstracts were identified: 293 articles on priapism in children and 44 on adults. One randomized control trial (RCT) was identified but was not included as this studied only 11 adults with stuttering priapism.

Articles selected for inclusion were reviewed, interpreted, and discussed by the authors to propose recommendations for clinical practice based on Level 4—5 evidence and therefore makes Grade C—D recommendations. The reported findings are summarized herein.

Clinical classification

There are three widely accepted types of priapism: ischaemic (low-flow, veno-occlusive), stuttering (intermittent, recurrent ischaemic), and non-ischaemic (high-flow, arterial). A fourth, neonatal priapism is also described.

Ischaemic priapism is the commonest type seen in children and is typically painful. Sexual activity (including masturbation) and nocturnal erections are common precipitators. Marked rigidity of the corpora cavernosa with a flaccid glans and spongiosum is typical. Elevated interstitial pressures ensue, causing microvascular compromise and ischaemia: a compartment syndrome within the tunica albuginea.

Stuttering priapism describes recurrent "unwanted and painful erections" which are often self-limiting but may precede an "unrelenting" ischaemic priapism [6]. Nocturnal erections are the most common trigger. It was first described by Emond [7] in 1980 in Jamaican sickle cell disease (SCD) patients, in whom they often start in childhood. Stuttering priapism has a significant effect on quality of life: recurrent visits to healthcare providers (often at night), sleep deprivation, embarrassment, and sexual performance anxiety [8].

Non-ischaemic priapism is a partial erection due to unregulated cavernous arterial flow which is usually painless. Piesis sign (perineal compression resulting in penile detumescence which recurs after removal of perineal pressure) strongly suggests non-ischaemic priapism in children (less common in adults) [4,9].

Neonatal priapism is a prolonged (≥4 h) erection during the first 28 days of life; 18 cases have been reported [5,10]. It usually occurs in the first few days of life and persists for 2—12 days (average 5 days).

Incidence and age distribution

The incidence of priapism in males of any age is estimated at 0.3—1.5 per 100,000 per annum, most frequently

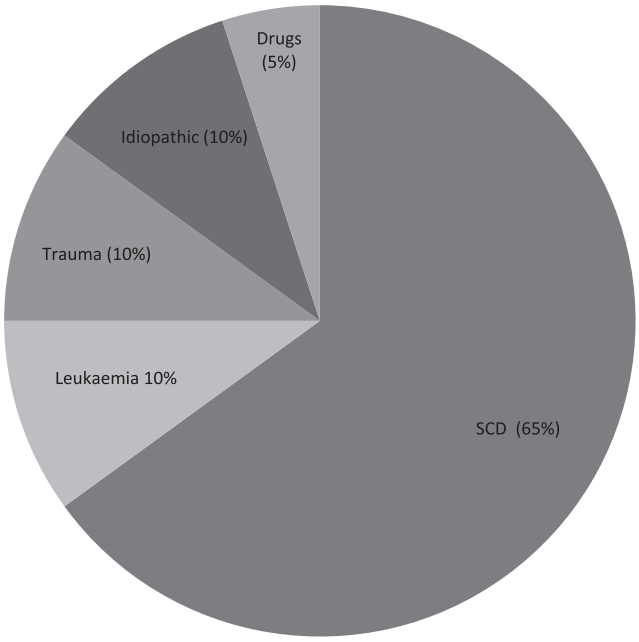

Figure 1 Major causes of priapism and their frequencies. Data represent estimations from the literature search; adapted from Ref. [13]. SCD: sickle cell disease; Drugs: pharmacologically induced.

affecting men in their fifth decade [11,12]. There are no generalizable data on the prevalence of priapism in all children, which is considered rare. However, paediatric priapism may be under-reported as embarrassment might prevent boys or parents seeking medical attention. Variables affecting the incidence and reporting of childhood priapism include population ethnicity (priapism's prevalence varies proportionately to SCD incidence) and the definition of priapism [13].

SCD is the commonest cause of priapism in children (Fig. 1). Adeyoju et al. [14] conducted a detailed international multicentre survey of 130 patients with SCD aged 4—66 years (mean 25 years); 35% had experienced priapism; 72% of cases were stuttering. Mean age at first episode was 15 years. Perhaps surprisingly, 25% of children with SCD-related priapism are pre-pubertal [3]. One study concluded that 90% of men with SCD experience priapism before their 20th birthday [15].

Non-ischaemic priapism is uncommon in both adults and children: our understanding is based upon small case series. An observational study estimated neonatal priapism in 15 per 100,000 live births, suggesting it may also be under-reported [16].

Physiology

Recent advances in ED have revealed the molecular basis for tumescence. Erections may be initiated by genital stimulation (reflexogenic), central stimulation (psychogenic: stimuli may be audiovisual, fantasy, or memory) or "central origination" (nocturnal: mediated by androgens during rapid eye movement sleep in adolescent boys) [17].

Penile flaccidity is maintained by high resting arterial and cavernosal smooth muscle tone, limiting cavernosal arterial inflow. Resting tone may be further increased in cold conditions or after sympathomimetic intracorporeal injection (ICI) [17]. Tumescence is initiated by relaxation of cavernosal arteries and sinusoidal smooth muscle; increasing arterial inflow and capacitance (latent phase). Sinusoids trap blood and tumescence ensues, compressing subtunical venular plexi (tumescent phase). This stretches the tunica albuginea, which occludes emissary veins (full erection phase) [18].

Contraction of the ischiocavernous muscles further increases cavernosal pressure; which usually exceeds systolic blood pressure (rigid erection phase) for short periods [17]. The corpus spongiosum and glans are subject to similar increases in arterial inflow but lack a tunical covering so act largely as an arteriovenous shunt during erections.

Parasympathetic and somatic neurons (arising from the second to fourth sacral segments) stimulate neuronal and endothelial (acetylcholine mediated) nitric oxide synthase (NOS) catalysing nitric oxide (NO) production from L-arginine and oxygen. NO acts on both cavernosal artery and sinusoidal smooth muscle, activating guanylate cylase and thus elevating cyclic guanosine monophosphate (cGMP). This in turn activates cGMP-dependent protein kinases, which reduce intracellular calcium levels and promotes smooth muscle relaxation.

Sympathetic neurons (arising from the 12th thoracic to second lumbar segments) release noradenaline (norepinephrine) to increase cellular inositol triphosphate and diacylglycerol levels causing a rise in calcium [18]. Type 5 phosphodiesterase (PDE-5) reduces cGMP, rendering it inactive. This heralds smooth muscle contraction which is propagated by the RhoA and Rho-kinase system as inhibition of myosin light chain kinases, and thus myosin-actin cross-bridge formation ensues [17].

Androgens increase endothelial and neuronal NOS expression and activity (stimulating tumescence) but also enhance expression and activity of PDE-5 (promoting flaccidity). The antithetic actions of androgens are postulated to mediate the libido's effects on erections [19].

Reflexogenic erections are physiological, even in neonates and pre-pubertal boys. Detumesence should occur after removal of the stimulus. Erections are commonly observed during bathing, diaper changing, urethral catheterization, and with a full bladder [2].

Aetiology and pathophysiology

Ischaemic priapism

Hinman's classic theory that "congestion and slowing of the blood stream" increases blood viscosity and subsequent ischaemia cause ischaemic priapism is supported by the presence of dark, deoxygenated blood when aspirating the corpora [2]. The molecular basis of a "final common pathway" in ischaemic priapism is outlined in Fig. 2. Cavernosal smooth muscle necrosis leads to cavernosal fibrosis, which causes ED and penile distortion/shortening.

SCD typically causes ischaemic or stuttering priapism, but has also been associated with non-ischaemic priapism. Deoxygenated haemoglobin S (HbS) causes sickling

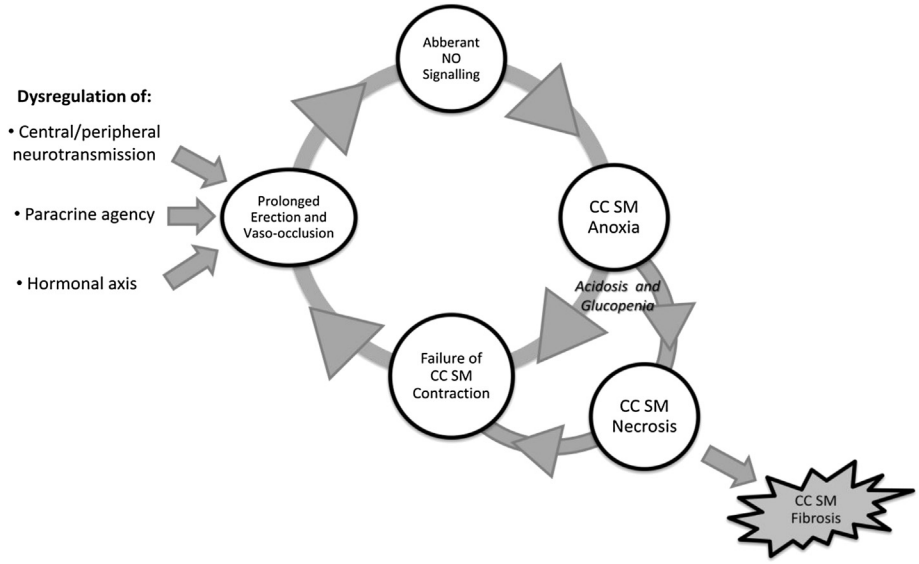

Figure 2 The molecular pathophysiology of ischaemic priapism. A prolonged erection may be initiated by a variety of factors which affect central and peripheral neurotransmission and paracrine agencies within the corporal tissue or the hormonal axis. Prolonged veno-occlusion leads to aberrant nitric oxide production and disruption of the molecular signalling (including guanylate cyclise, cGMP, PDE-5, and thus calcium). This results in prolonged cavernosal smooth muscle hypoxia leading to acidosis and glucopenia and hence impaired smooth muscle contraction. This in turn prolongs the erection and hence hypoxia; ultimately resulting in smooth muscle necrosis which heralds cavernosal fibrosis. Adapted from Ref. [51]. CC: corpora cavernosa; NO: nitric oxide; SM: smooth muscle.

(adhesive interactions between erythrocytes, endothelial cells, and leucocytes) and thus microvascular obstruction. Vaso-occlusive episodes (painful sickle crises, osteonecrosis, and acute chest syndrome), seen in homogenous genotypes (SS), are no longer thought to explain priapism [20].

Sickling also stimulates haemolysis and thus increases serum free haemoglobin levels, which deactivate NO. Sickled erythrocytes also remove L-arginine (the substrate for NO synthesis) from plasma. NO is thus deactivated and depleted, known as haemolysis-associated endothelial dysfunction [20]. Decreased NO availability also decreases PDE-5, RhoA, and Rho-kinase and disrupts adenosine signalling. These mechanism cause priapism, pulmonary hypertension, strokes, and leg ulcers.

Thirteen cases of ischaemic priapism (including 1 child) have been associated with sickle cell trait (heterozygous genotype: AS); however, an alternative cause (e.g. pharmacological) is identifiable in the majority of these cases [21].

Nocturnal erections, sexual activity, dehydration, fever, and exposure to cold are the most common precipitants of priapism in children with SCD [3]. Anaemia and raised serum haemolysis markers (reticulocytes, indirect bilirubin, lactate dehydrogenase, and aspartate aminotransferase) are seen in SCD priapism [20].

Childhood leukaemias may cause priapism: hyperleucocytosis and thrombocytosis are usually present [22]. In hyperleucocytosis (white cell count $\geq$50—100 $\times$ 10^9/L), direct interaction between leukaemic blasts and endothelial cells causes a loss of vascular integrity, activating prothrombotic mechanisms, hence an increased risk of pulmonary or cerebral leucostasis. Cavernosal leucostasis may similarly lead to thrombus formation and venous outflow obstruction, activating the common pathway and hence ischaemic priapism [22].

Other causes of priapism in children are listed in Table 1. Despite recent advances in our understanding of the pathophysiology of priapism, it remains idiopathic in 10% of children.

Stuttering priapism

SCD is the commonest cause of stuttering priapism in children, suggesting a similar aetiology to ischaemic priapism, if less pronounced [3]. However, its exact pathophysiology is poorly understood. It is postulated that cavernosal endothelial NO deficiency causes downregulation of protein kinase G (PKG), PDE-5, and Rho A/Rho-kinase [24]. Tonic cavernosal smooth muscle tone is reduced without PDE-5 and other mechanisms to regulate cGMP levels, producing over-responses to sexual or androgenic stimulation and hence recurrent prolonged erections [20].

Other postulated mechanisms include impairment of adrenoceptors, upregulation of transforming growth factor (TGF)-β, scarring of intracavernous venules, and abnormal central neurological control mechanisms [2,8].

Non-ischaemic priapism

Penile, perineal, or pelvic trauma (typically straddle or coital injuries) are the commonest cause of non-ischaemic priapism. Laceration of cavernous arterioles, usually in the crura or corporal bodies, may cause an arteriolar—sinusoidal fistula. Rarely, a cavernous or internal

Table 1 Causes of priapism in children (in approximate order of frequency compiled from the literature search).

Ischaemic	**Pharmacologically induced**
Haemaglobinopathy (SCD, thalassaemia, G6PD, CDA)	PDE-5 inhibitors
Leukaemia (ALL, CML)	Hormones (e.g. testosterone)
Tumour (rhabdomyosarcoma, testicular)	Anti-psychotics, anti-depressants
Infection	Anti-hypertensives
Neurogenic (spinal cord injury, CES)	(inc. α-blockers)
Toxins (malaria, scorpion, spider)	Erythropoietin
Henoch–Schönlein purpura	Anaesthetic (inc. spinal)
Haemodialysis	Recreational (alcohol, cocaine, marijuana)
Parental nutrition	
Non-ischaemic	**Neonatal**
Trauma	Polycythaemia
Haematological (SCD, leukaemia)	Infection (syphilis, pyocavernositis)
Fabry's disease[a]	Cranial birth trauma (forceps)
Iatrogenic (aspiration/surgery)	Respiratory distress syndrome
	Umbilical artery catheterization

ALL: acute lymphoblastic leukaemia; CDA: congenital dyserythropoietic anaemia; CES: cauda equina syndrome; CML: chronic myeloid leukaemia; G6PD: glucose-6-phosphate dehydrogenase deficiency; PDE5: phosphodiesterase type 5; SCD: Sickle cell disease.

[a] Fabry's disease is a rare congenital glycosphingolipid metabolism disorder.

pudendal artery laceration can produce a fistula. The fistula causes a high in-flow state and pooling of blood in sinusoidal spaces. This causes mechanical stimulation of endothelial NOS, increasing NO/cGMP and thus smooth muscle relaxation [9]. Non-ischaemic priapism is therefore self-propagating.

The onset of priapism is typically delayed by a few days following trauma (3 h–7 days) [9]. It is postulated that penile haemodynamic changes caused by a subsequent erection (typically nocturnal) rupture the clot formed following the initial injury and hence fistula formation. Non-ischaemic priapism without a fistula or a high-flow haemodynamic state of the cavernous arteries may also occur, the aetiology of which remains unclear. Non-ischaemic priapism has also been described following medical or surgical management of ischaemic priapism in adults [2].

Neonatal priapism

Most neonatal priapism is idiopathic: subclinical perineal birth canal trauma is hypothesized to cause most cases [2]. Other reported associations are cited in Table 1. SCD is not associated with neonatal priapism due to the presence of foetal haemoglobin (HbF). Ischaemic priapism has not been reported in a neonate and no cases are thought to have caused discomfort/agitation. Colour Doppler ultrasonography (CDU) in two newborns and cavernosal blood gas analysis in one support non-ischaemic priapism. Full functional recovery is reported in all cases, although follow-up is limited to $\leq$8 years [5]. This suggests a favourable natural history and benign pathophysiology.

Pharmacologically induced priapism

Priapism is associated with a wide range of drugs in children (Table 1) administered therapeutically or following an overdose (intentional or accidental). PDE-5 inhibitors are used in pulmonary hypertension but have a 1% incidence of priapism at therapeutic doses [25]. Testosterone may cause stuttering, ischaemic, or rarely non-ischaemic priapism, usually 5–8 days after injection. Following testosterone deprivation, prolonged testosterone exposure is postulated to augment NOS expression more profoundly than PDE-5 [19].

Anti-psychotics are the most frequent cause of pharmacologically induced priapism in adults (due to α-1 adrenergic antagonism); however, this is rare in children, typically only seen with polypharmacy. Illicit drug use, notably cocaine, is a common cause of priapism in adults.

Management

Priapism must be assessed and treated by experienced clinicians in a timely manner as ischaemic priapism may cause future ED, anxiety, attenuated sexual aversion behaviour, and intimacy avoidance due to a fear of recurrent priapic pain.

The management of priapism in neonates

Initial evaluation of prolonged erections in newborns should include clinical examination and/or ultrasonography, full blood count, and C-reactive protein [5]. Urgent CDU should be performed. If ischaemic priapism cannot be excluded corporal aspiration and/or ketamine anaesthesia should be considered [10]. However, corporal injection or surgical intervention have never been required in neonatal priapism.

Red cell volume reduction (venesection) should be performed for polycythaemia [10]. Careful observation alone is appropriate in most idiopathic neonatal priapism: the majority resolve spontaneously without sequelae [5]. We advocate fortnightly CDU and clinical review in idiopathic neonatal priapism.

Initial management in older children

Initial management aims to assess the type of priapism and achieve detumescence (Fig. 3). Opiate analgesia is usually required in ischaemic priapism, which may inhibit tumescence [26]. "First-aid" measures include physical exercise (e.g. running up stairs), urination, a cold bath, ejaculation, and fluids [27].

Cold packs are analgesic, may have a cytoprotective effect (limiting ischaemic damage) and may cause

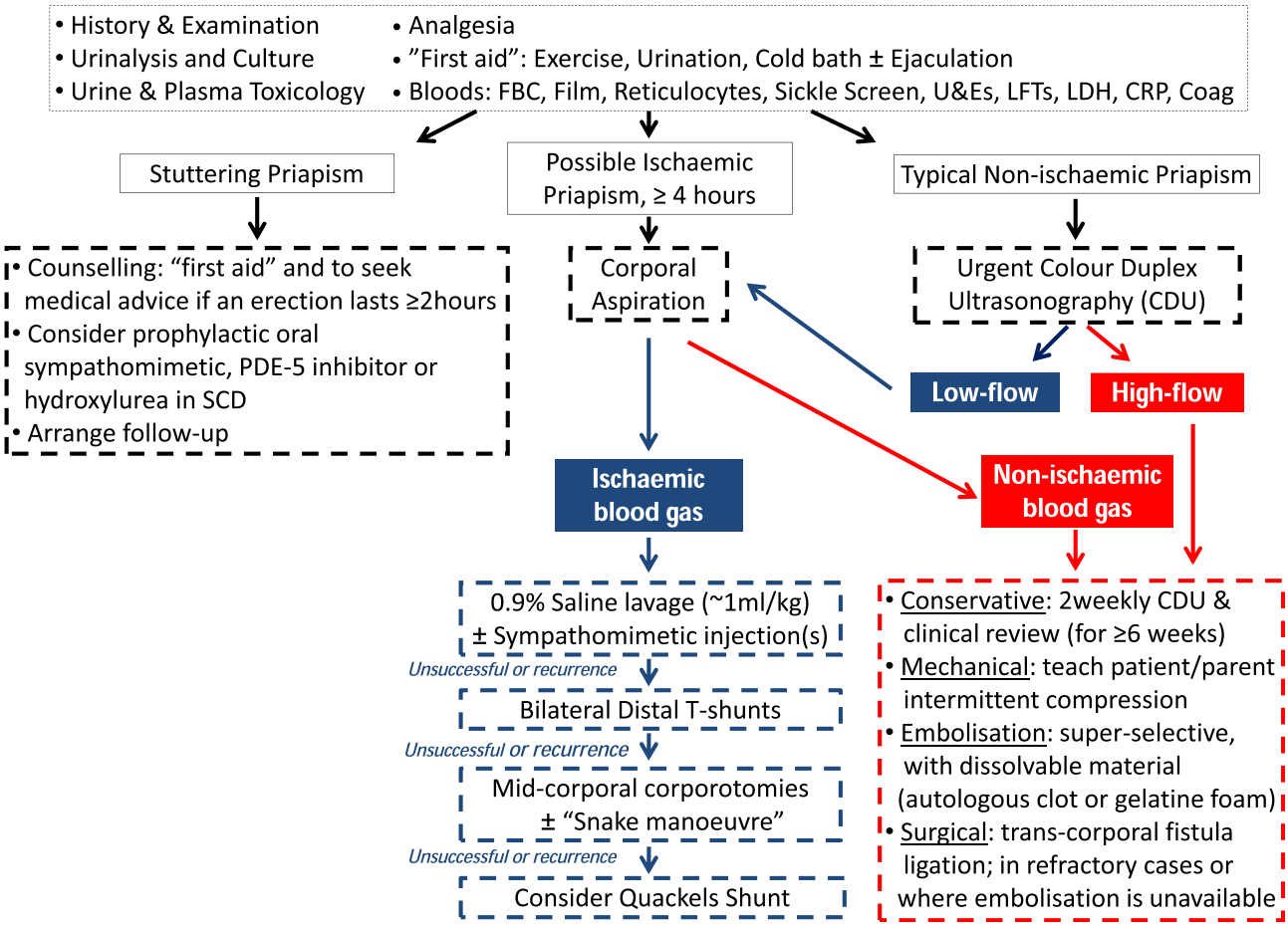

Figure 3 Management algorithm for priapism in children. Coag: coagulation studies; CRP: C-reactive protein; FBC: full blood count; LDH: lactate dehydrogenase; LFTs: liver function tests, U&Es: urea and electrolytes.

vasoconstriction decreasing penile blood flow [2]. However, cold may induce priapism in children with SCD [3]. We offer cold packs (where tolerated) to boys in whom SCD is unlikely.

For children responding to these measures careful counselling on what to do if a prolonged erection returns (including to seek urgent medical assessment $\geq$2 h) and potential sequelae are important. Prognosis is improved with early medical attention. Screening/follow-up must be arranged.

In persistent cases, clinical features and identification of an underlying cause will guide management (Table 2). A blood panel should be sent to exclude haemoglobinopathies and leukaemia; of which priapism may be a presenting feature [3].

The absence of pain is an unreliable indicator of non-ischaemic priapism [19]. Corbetta et al. [28] advocate routine corporal blood gas analysis. However, this nearly always necessitates general anaesthesia and is not therapeutic in non-ischaemic priapism. CDU has nearly 100% sensitivity and specificity in experienced hands [29]. In a classic case where a painless partial erection follows trauma, we advocate urgent CDU, if radiological expertise is readily available [9]. If unavailable or equivocal, urgent corporal aspiration should be performed.

Anaesthesia: local, conscious, or general?

General anaesthesia (GA) is superior to sedation for painful procedures in children [30]. Risks from GA are reduced above the age of 6—12 months [31]. However, children with SCD have increased GA risks (including acute chest syndrome), necessitating meticulous fluid balance and even preoperative transfusion [32]. When choosing the optimal anaesthesia modality, the risk of lasting psychological damage, aspiration complications, anaesthetic risk, and delay that anaesthesia may incur (and thus the probability of ED) must be considered.

Successful aspiration/injection is reported under conscious sedation with local anaesthesia (LA) in 4—18 year olds [33]. This may be an appropriate alternative, particularly in hospitals without rapidly available paediatric anaesthetic expertise. Dissociative sedation (low-dose ketamine, propofol, fentanyl, or morphine) may be provided by paediatric anaesthetists [30]. Ketamine is an established detumescence agent and may resolve priapism so should be used preferentially where appropriate expertise is available [3].

Table 2 Differentiating ischaemic and non-ischaemic priapism. Normal values for mixed venous blood (in room air) are pO_2 40 mmHg (5.3 kPa), pCO_2 50 mmHg (6.6 kPa), pH 7.35 [2]. Note corporal aspiration is not required in typical cases of non-ischaemic priapism where urgent CDU is available.

Pain	Ischaemic (low flow)	Non-ischaemic (high-flow)
	Yes	No
History	SCD Recreational drug use Stuttering priapism	Antecedent trauma
Examination	Rigid (non-compressible) Bi-corporal (CC)	Partial (compressible or pulsatile) May involve glans (tri-corporal) Ecchymosis (antecedent trauma)
Blood gas	Hypoxic: $pO_2 < 5.3$ kPa/40 mmHg Acidotic: pH < 7.25 Hypercarbic: $pCO_2 > 7.98$ kPa/60 mmHg Glucopenia (<3.5 mmol/L)	Oxygenated: $pO_2 > 12$ kPa/90 mmHg Neutral: pH 7.35—7.45 Hypocarbic: pCO_2<5.3 kPa/40 mmHg

CC: Corpora cavernosa; SCD: Sickle cell disease.

Corporal aspiration and lavage

Lateral (3/9 o'clock) mid-shaft corporal needle access is obtained, avoiding the urethra or dorsal neurovascular bundle (Fig. 4). A 23—21 G (blue/green) butterfly needle should be used in pre-pubescent boys and 19 G (white) needle in adolescents [3,33] although larger needles may be required to evacuate clots. Minimizing needle passages using a three-way tap and applying pressure for 5 min after removal reduces haematoma rates (13% using a 23 G needle) [33]. Rare complications include infection, urethral lesions and non-ischaemic priapism. Some authors advocate passing the needle through the glans which may reduce haematoma rates, although this could render a subsequent distal shunt procedure more awkward [34].

The presence of dark, deoxygenated blood ($pO_2 < 40$ mmHg) confirms ischaemic priapism (Fig. 5, Table 2) [33]. In ischaemic priapism the corpora should be immediately decompressed: 3—5-mL aliquots should be aspirated until bright red (oxygenated) blood is seen (not exceeding 10% of the circulating blood volume; 7.5 mL/kg in children aged ≥1 year) [3]. The corpora should then be flushed with warmed 0.9% saline.

Intracorporal injection

If aspiration and irrigation do not achieve detumescence, sympathomimetic ICI should be performed with

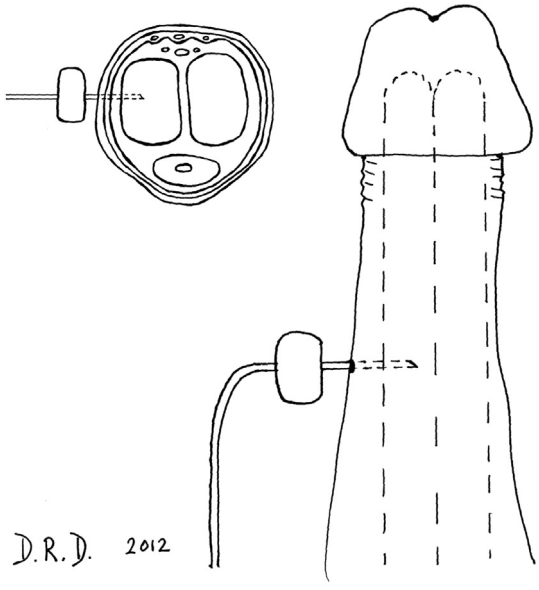

Figure 4 Illustration of corporal aspiration. A butterfly needle is inserted laterally at the 3 or 9 o'clock positions avoiding damage to the corpus spongiosum/urethra and the dorsal neurovascular bundles. Blood is aspirated in a heparinized syringe before blood gas analysis is undertaken to delineate if the priapism is ischaemic or non-ischaemic.

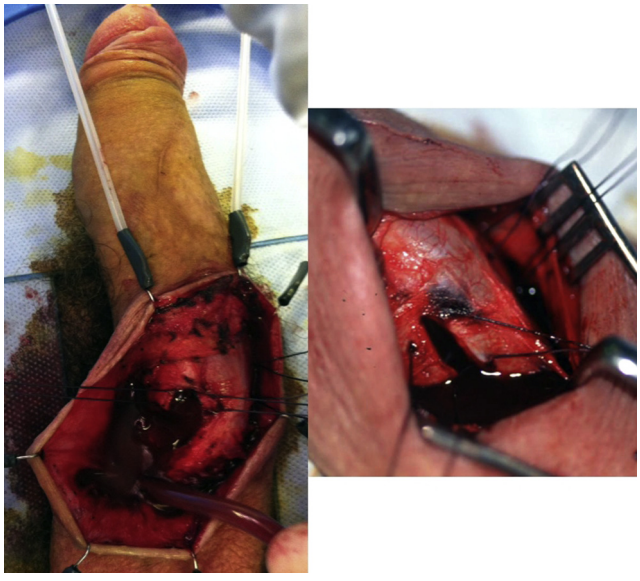

Figure 5 Deoxygenated blood. Two photographs of mid-corporal corporotomies demonstrating dark, deoxygenated blood seen in ischaemic priapism.

Table 3 Suggested sympathomimetic preparation for intracorporal injection (ICI). This is an unlicensed indication and route of administration. When available phenylephrine should be used in boys aged ≥11 years; epinephrine should be used in boys ≤10 years. There are no reliable data on ICI ≤2 years: we recommend using a reduced dose of epinephrine (adrenaline). Sympathomimetics should be prepared prior to corporal aspiration to avoid repeated needle passages. ICI should be performed in a level 2–3 environment with ≥30 min monitoring (electrocardiogram, blood pressure, heart rate and pulse oximetry); at which point the patient may be discharged if detumescence persists [33].

Drug	Available preparations	Concentration	Age and aliquot	Further doses
Phenylephrine	10 mg/mL (1%)	0.1 mL + 4.9 mL 0.9% saline (200 μg/mL)	≥11 yrs: 0.5 mL	≤10 at 5–10 min (≤1 mg)
Epinephrine (adrenaline)	1 in 10,000 (100 μg/mL)	1 mL + 99 mL 0.9% saline (1 in 1 000 000 or 1 μg/mL)	≥11 yrs: 15 mL, 3–11 yrs: 10 mL, <2 yrs[a]: 2.5–5 mL	≤4 at 10 min
	1 in 1000 (1 mg/mL)	1 mL + 1 L 0.9% saline (1 in 1 000 000 or 1 μg/mL)		
Etilefrine	10 mg/mL (1%)	None	0–18 yrs[a]: 0.5 mg/mL	≤2 at 10 min

[a] See separate guidance regarding neonatal priapism.

cardiovascular monitoring. The mechanism of action of sympathomimetics is strongly debated but is thought to be principally α-adrenergic-mediated cavernous smooth muscle contraction and cavernosal arterial vasoconstriction [2,27] Side-effects are rare but include headache, dizziness, hypertension, reflex bradycardia, tachycardia, arrhythmias, and (in overdose) a subarachnoid haemorrhage [36]. Injections must stop when detumescence is achieved.

The American Urological Association (AUA) guideline recommends ICI of phenylephrine, a selective α-1 adrenergic agonist which lacks β-mediated cardiac ionotropic and chronotropic effects [35]. This states that 100–500 μg should be injected in adults but "lower concentrations in smaller volumes" in children. There is no specific guidance on paediatric ICI dosages. We suggest using 100-μg aliquots of phenylephrine (0.5 mL of 200 μg/mL solution) at 5–10-min intervals in children aged ≥11 years (up to 10 times) (Table 3); there have been no reported complications.

Children as young as 3 years have received ICI epinephrine (adrenaline) in a dilute solution (5–20 mL of 1 in 1,000,000) [33,35]. This appears safe, having been reported in 45 children without side-effects. Etilefrine (available in Europe) has been used in 7 children as young as 2 years (5 mg, undiluted). Metaraminol (a pure α-agonist) should not be used because of its high cardiovascular complication rate [27]. Pragmatically, the sympathomimetic used must depend on what is readily available; suggested dosages, preparation and number of injections for different age groups are listed in Table 3.

Table 4 Characteristic findings of ischaemic and non-ischaemic priapism on penile colour Duplex ultrasonography. The following findings, in the absence of sexual stimulus, are diagnostic for ischaemic and non-ischaemic priapism.

	Ischaemic	Non-ischaemic
Sinusoids	Non-compressible, low echogenicity (mixed if incomplete thrombus)	Compressible, transonic (no thrombus)
Peak systolic velocity	Absent (may be low)	Normal or high
Diastolic velocity	Absent or low; typically with negative end-diastolic velocity	High end diastolic
Dorsal vein flow	Absent or low	Normal or high, with prominent veins
Other		Fistula: hypoechoic lesion with colour blush

Colour duplex ultrasonography

CDU is simple, reproducible, cheap, non-invasive, and involves no radiation or contrast. CDU should be performed in the frog-leg position, in a comfortable room with distraction therapy. This facilitates high frequency (≥7.5 MHz) linear array transducer access to the perineum and penis, visualizing the corpora in longitudinal and axial planes.

In ischaemic priapism cavernosal arterial flow typically demonstrates a "high resistance, low velocity" wave-form (Table 4, Figs. 6 and 7) [37]. Arterial flow is usually absent. The "high-resistance, high-flow" ischaemic sub-group has persistent cavernosal arterial flow (low: 1–2 m/s) but this is insufficient for small vessel perfusion and there is usually negative end-diastolic velocity [37].

In non-ischaemic priapism CDU demonstrates a low-resistance, high-flow arterial waveform. Reducing probe pressure demonstrates rapid engorgement. CDU detects nearly 100% of arteriosinusoidal fistulae [37]. A poorly circumscribed, hypoechoic cavernous lesion (greyscale) with a characteristic Doppler colour blush identifies the fistula, which later mimics a pseudoaneurysm [38].

CDU is also helpful in guiding ongoing management. Ischaemic priapism results in penile oedema, sometimes making it difficult to confirm detumescence by clinical examination. CDU can also assess the degree of fistula resolution or recanalization.

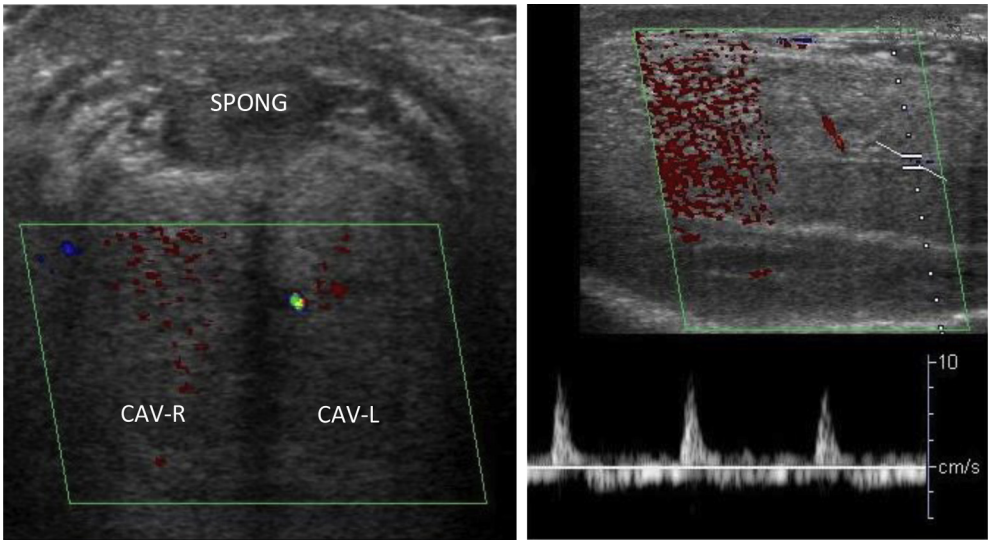

Figure 6 Penile colour duplex sonograph demonstrating the corpora cavernosa (CAV-L: left, CAV-R: right) and spongiosum (SPONG). Left cavernosal arterial flow is demonstrated with negative end-diastolic flow. The right cavernosal arterial flow was absent, consistent with ischaemic priapism. cm/s: centimetres per second (the reader is referred to the web version of this article for colour duplex interpretation).

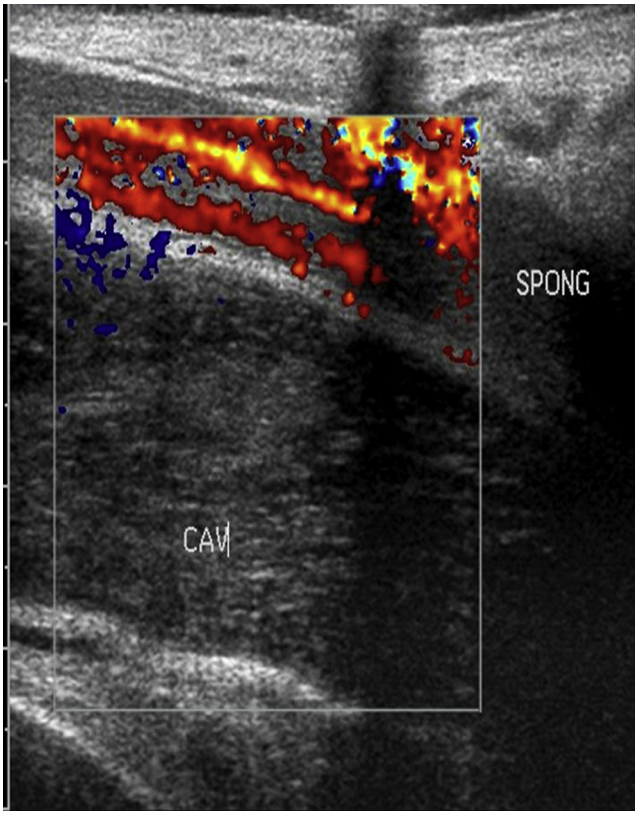

Figure 7 Penile colour duplex sonograph demonstrating the corpora cavernosa (CAV) and spongiosum (SPONG). Arterial flow (red) is seen in the corpus spongiosum but no flow is demonstrated in the corpus cavernosum, consistent with ischaemic priapism. (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

Digital subtraction angiography

Digital subtraction angiography (DSA) of the internal pudendal artery is invasive so should only be undertaken when concurrent embolization can be performed.

The ongoing management of ischaemic priapism

Corporal interstitial oedema is present at 12 h of ischaemic priapism and smooth muscle necrosis is seen by 48 h (in adults) [2]. Intervention beyond 48—72 h is unlikely to restore normal erectile function although it may provide relief of the erection and pain [2].

If repeated sympathomimetic ICI is unsuccessful a surgical fistula ("shunt") should be formed, bypassing the pathological veno-occlusion and allowing blood drainage [35]. Shunts can be distal (cavernoglanular: percutaneous — Winter/T-shunt, or open — Al-Ghorab), proximal (cavernospongiosal — Quackels) or cavernovenous (saphenous — Greyhack) [2].

Distal shunts are simpler to perform, have better functional outcomes and lower complication rates [35]. The

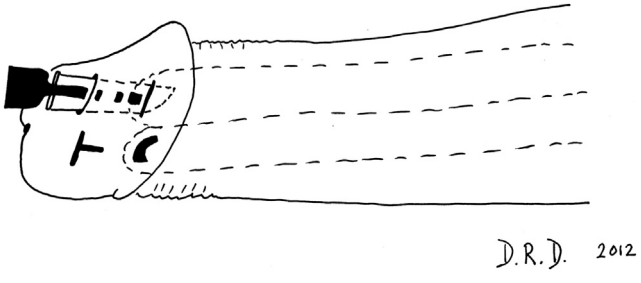

Figure 8 T-shunt formation. A number 10 blade scalpel is inserted percutaneously to puncture the glans (lateral to the urethra) and distal ends of the corpora cavernosa bilaterally. Ninety degrees of lateral rotation of the scalpel helps maintain shunt patency. Thrombus can be milked out of the corpora prior to superficial wound closure.

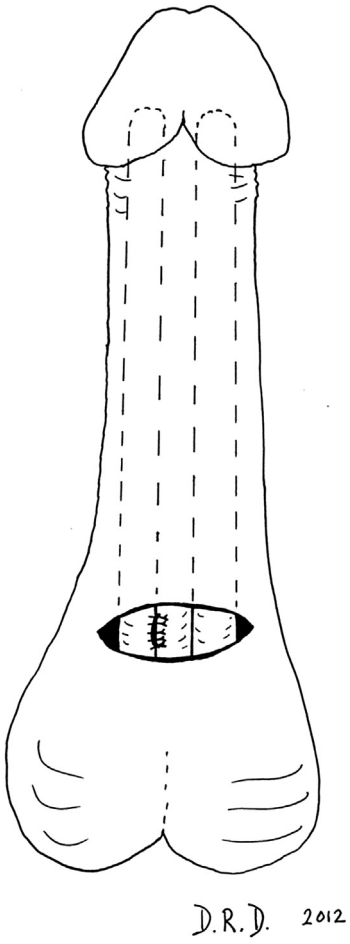

Figure 9 Proximal cavernospongiosum (Quackles) shunt. A transverse ventral penoscrotal skin incision is made allowing access to the corpora. Both cavernosa and the spongiosum are then incised vertically and then sutured together, forming a proximal shunt.

commonest initial procedure at our institution is percutaneous bilateral distal fistulae formation using T-shunts (a modification of Winter shunts) (Fig. 8). Favourable subsequent erectile function rates are reported (75%) in adults [35].

Classically proximal cavernospongiosal shunting procedures (Fig. 9) were undertaken when distal shunts proved unsuccessful. However, these have high ED rates (≥50%) and may result in a cavernourethral fistula, urethral strictures, and cavernositis [35]. Historically a cavernovenous shunt would be performed in refractory cases, although this risks thromboembolic complications. A 14-year-old regained potency following cavernovenous shunting after 72 h of painful priapism [39].

Alternatively, bilateral mid-corporal corporotomies (Fig. 10) can be performed in refractory cases. This allows corporal lavage and heparin flush. Gently expunging clot while assessing corporal stenosis using Hegar's dilators can be performed after corporotomy in a modification of the "corporal snake manoeuvre" [40]. Success rates of 80% have been reported in adults using this technique; however outcomes have not been reported in children [40]. Intermittent penile compression and/or anticoagulation to maintain fistula patency in adults is controversial and not reported in children.

Penile prosthesis insertion

Early penile prosthesis insertion in adults treats acute priapism and the ensuing ED and reduces complications and loss of penile length compared with delayed insertion, where fibrosis renders insertion technically difficult [41]. In adults after 48 h of ischaemia ED is deemed inevitable. However, it is harder to confidently predict ED in children. Physical and emotional immaturity may also preclude early prosthesis insertion. Prosthesis placement has been performed electively in teenagers (≥17 years) following priapism [42].

Ongoing systemic treatment in ischaemic priapism

Where a systemic cause of ischaemic priapism is identified, adult guidelines recommend concurrent systemic and corporal therapy [35]. However, this is controversial in children. The resolution of priapic pain (after discontinuation of analgesics) has been correlated with the resolution of ischaemia, leading some physicians to avoid corporal aspiration despite priapism lasting ≤13 days [22].

Lower subsequent ED rates are reported in adults and children with SCD-related ischaemic priapism. However, priapic episodes cause scarring of intracavernous venules, disrupting regulatory mechanisms, and predisposing to future episodes, emphasizing the importance of prompt treatment [20].

Sickle cell disease

The lifetime probability of ED secondary to priapism is over 30%, yet the management of ischaemic priapism in boys with SCD remains controversial. Initial management is similar to other sickle cell crises (hyperhydration, oxygen, and analgesia). Treatment with plasma alkalinisation may also be considered; however, systemic treatments must not delay corporal aspiration or surgical management [23].

Recent evidence suggests exchange transfusions have limited efficacy in priapism [23]. This is congruent with the recently described pathophysiology of SCD priapism. Furthermore, the 21% risk of cerebrovascular accident must be considered (the association of SCD, priapism, exchange transfusion, and neurological events: ASPEN syndrome) [43].

Leukaemia

There is a paucity of evidence on the treatment of childhood leukaemic priapism. Management without corporal aspiration but solely systemic anti-leukaemic therapies (chemotherapy and/or leucopheresis) and anticoagulation (low molecular weight heparin) has been advocated, with no ED reported in 4 cases with ≤8-year follow-up [22]. Penile radiotherapy was used historically [22]. However, in our experience surgical shunts and even prosthesis insertion have been required, illustrating the importance of concurrent cavernosal and systemic treatment.

Testosterone-induced priapism

Anti-androgens may be considered in the management of testosterone-induced priapism alongside penile treatments.

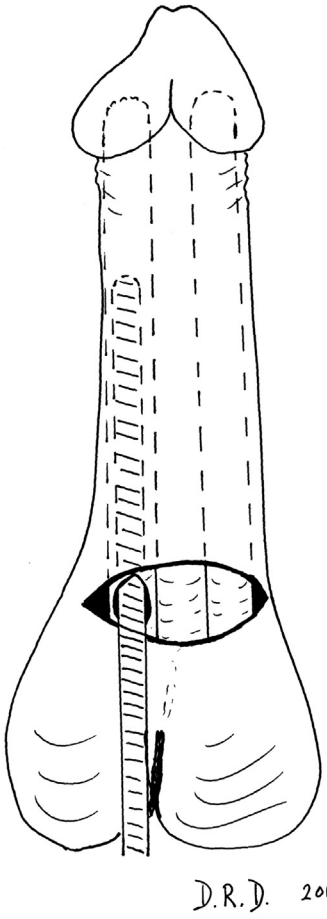

Figure 10 Mid-corporal corporotomy. A transverse skin incision is made on the ventral aspect of the penis, at the mid-corporal level. Bilateral vertical corporotomies allow the evacuation of clot and corporal washout. Further, a Hegar's dilator can be used, in a modified "corporal snake manouvre" to gently evacuate thrombus and encourage reperfusion.

However, anti-androgens are relatively contraindicated in boys who have not reached sexual or skeletal maturation [13]: the type of priapism, age, and pubertal status of the child must be considered. The half-life of the testosterone preparation should be considered in the duration of treatment.

The management of stuttering priapism

The management of each prolonged erection in stutterers should not differ from that of ischaemic priapism. Patients/parents must be counselled regarding simple "fist-aid" measures and to obtain medical advice if they experience an erection lasting $\geq$2 h [27].

The chronic management of stuttering priapism aims to reduce its frequency and prevent ischaemic priapism and its sequelae. How this is best achieved remains controversial in children and adults [20]. Compliance can be challenging in young boys who are often uncomfortable addressing their sexual health with health providers, particularly females [44]. The routine use of an erections diary (documenting duration and frequency of stuttering on a weekly basis) has been advocated in all boys with SCD to identify those at high risk [20].

α-Adrenergic sympathomimetics may augment corporal smooth muscle contraction: enhancing detumescence [45]. Pseudoephedrine or etilefrine 0.5 mg/kg up to 30 mg each night should be given at night with careful blood pressure and side-effect monitoring (palpitations and tachycardia) [27].

PDE-5 inhibitors have also been used with success, paradoxically, but should not be started during acute priapism [45]. It is postulated that surges of cGMP go unchecked in SCD (as basal PDE-5 levels are downregulated) [20]. Other proposed mechanisms include that chronic inhibition may enhance PDE-5 activity or improve penile oxygenation [45]. Daily sildenafil (short-acting) can be trialled before changing to thrice weekly tadalafil (long acting).

Hydroxyurea, which induces foetal haemoglobin synthesis, can also be used in SCD stuttering priapism: 10—35 mg/kg dose titration with 4-weekly blood counts (myelosuppression is a rare side-effect) [46]. There is also evidence that hydroxyurea may reduce other sickle crises and increase life expectancy in SCD [47].

Oral β-agonists (e.g. terbutaline) have success rates $\geq$62% in adults; their mechanism of action of is unclear; α-adrenergic effects are postulated. Baclofen, a gamma-aminobutryric acid (GABA) receptor inhibitor is reported to have treated priapism successfully in a few children [48], typically with spinal cord injury; however, its side-effect profile (drowsiness, nausea and ED) limits its use.

Gonadotropin-releasing hormone agonists, anti-androgens, oestrogens, ketoconazole (testosterone synthesis inhibitor), and 5-α-reductase inhibitors have been used in adults but are not recommended for use in children [35].

Phenylephrine ICI at home (by patients or parents) has also been advocated [2]. An implantable device used to administer intracaversonal phenylephrine has also been described in adults [45]. However, we suggest that prevention (prophylaxis) is better than cure (managing each episode individually). ICI may also carry a significant risk of physical/psychological trauma in children.

The management of non-ischaemic priapism

When non-ischaemic priapism has been confirmed, its management is not urgent. However, all children with perineal or penile trauma should undergo a full primary and secondary survey. Priapism may be a missed sign of spinal trauma [49]. Although no priapic child protection cases are reported in the literature, children with genital/pelvic trauma should be assessed by a paediatric specialist.

Fifty-five cases of childhood non-ischaemic priapism are reported; management options include conservative, mechanical, pharmacological, radiological embolization, or surgical ligation [9,28]. However, follow-up in children lacks longevity and reliably completed erectile function questionnaires [9].

Conservative

Initially expectant management is advised in both adults and children [35]. The natural history of non-ischaemic priapism is unclear (lasting 36 years in an adult). The social

unacceptability of priapism and hypothesized high oxygen tension-related fibrosis or secondary vascular changes may precipitate intervention [28]. We advocate fortnightly CDU and clinical examination up to at least 6 weeks prior to considering active management on an individual basis.

Mechanical

Perineal compression or ice packs may reduce blood supply through the fistula, heralding thrombus formation and resolution of priapism. Refinement using CDU with sedation has been reported in a few cases with successful outcomes [50]. These strategies are unlikely to cause harm, provided they are well tolerated.

Pharmacological

ICI of α-adrenergic agonists and methylene blue (a guanylate cyclase inhibitor) have been reported with limited success [9]. Androgen blockade reduces nocturnal erections and has good success rates in adults but is not advisable in most children [2].

Radiological embolization

Internal pudendal artery DSA (via femoral access) can accurately locate a fistula, allowing superselective embolization with autologous clot or synthetic material (e.g. gelatine foam). This allows temporary obstruction; allowing the fistula to heal before blood flow later returns, enabling future erectile function. Embolization is reported in 30 children, with an 80% first embolization success rate [9]. Failure was successfully treated with repeat embolization using non-absorbable material in 5 out of 6 cases (the remaining child was managed conservatively). Embolization therefore has an overall success rate of a 97%. No other complications are reported in children, although ED is not uncommon following embolization in adults [2].

Surgical

Open surgical transcorporal fistula ligation with intraoperative CDU in long-standing non-ischaemic priapism (where a pseudo-capsule around the fistula has developed) carries a high risk of subsequent ED and should only be considered in refractory cases or where embolization is unavailable [2].

Conclusion

Ischaemic priapism is a urological emergency. Left untreated it leads to necrosis, fibrosis and invariably future ED. Rapid resolution of ischaemic priapism prevents permanent cavernosal structural damage and is associated with improved prognosis for potency later in life [2]. Psychological sequelae may also be reduced with early intervention.

Where haematological or systemic disorders precipitate ischaemic priapism their treatment should not preclude or delay targeted corporal measures to reduce the child's priapism. We advocate timely assessment by experienced clinicians using the algorithm outlined in this article.

Children and their parents are often not comfortable discussing the child's sexual health. Coupled with a current lack of robust evidence for its management, treating paediatric stuttering priapism is challenging. Oral sympathomimetics or PDE-5 inhibitors should be trialled, coupled with hydroxylurea in SCD, with close monitoring.

Non-ischaemic and neonatal priapism may be treated less urgently if benign in aetiology, when initial expectant management is appropriate after confirmation with CDU. Compression may be trialled prior to embolization when the patient, parents, or clinician no longer find persistent priapism acceptable.

There is currently a paucity of understanding and evidence-based medicine in paediatric priapism, the management of which is, in part, therefore based around findings in adults. Differences compared to guidelines for adults include the volume of intracorporal blood aspirated, dosage of sympathomimetic ICI, anaesthesia modalities required for aspiration/injection, success rates of perineal compression in non-ischaemic priapism, and the management of neonatal priapism. Penile prosthesis insertion and anti-androgen therapy are inappropriate in most children.

Conflict of interest

None.

Funding

None.

Ethical approval

Ethical approval was not required for this review. All authors have read the policy of the journal on ethical consent and the standards of animal care.

Acknowledgements

We thank Mr Douglas R. Donaldson for providing all illustrations (Figures 4, 8—10). Thanks to Paul Manson and Craig Rore for their assistance with our electronic literature search.

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Commentary to 'Priapism in children: A comprehensive review and clinical guideline'

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The review article on priapism in the paediatric population is timely and comprehensive. It summarises current knowledge of the condition. Although meant for paediatricians, this is, in fact, relevant to those treating adults also. This review should be read by all paediatric urologists, and should be available in all paediatric emergency departments.

There are several interesting findings. Firstly, the quality of the evidence is poor, with only one known published randomised controlled trial, which was performed in the adult population. Secondly, much of the pathophysiology and treatments come from the adult world. Extrapolating this to younger males may be risky. Their recommendation of performing Doppler ultrasound on all cases is very sensible rather than first aspirating—as the converse tends to be the practice in adult medicine. Aspiration can cloud the issue and there have been reported cases of low-flow priapism being converted to a high-flow state by the passage of a needle. It is important for the Doppler study to survey the whole length of the copora cavernosa as there may be differences in flow proximally and distally. Patients who report a history of prolonged priapism several years prior can still have segmental woody feeling corpora when examined in the flaccid state. These patients also describe erectile dysfunction, which can be more marked in the fibrous-feeling segment. While this is typically distal, it can also be proximal. Hence, the needs to be thorough in the Doppler study on initial presentation.

The review also discusses the pros and cons of giving a general anaesthetic prior to any aspiration/injection. General anaesthetic is important for psychological/psychosexual reasons. I would also recommend a dorsal penile/caudal block once anaesthetised. I have seen adults who were treated without any form of anaesthesia develop symptoms of post-traumatic stress disorder, which add to the complexity of managing any subsequent erectile dysfunction.

Another benefit of performing any aspiration under general anaesthesia is that the anaesthetic team may be able to expedite blood gas analysis, for example in the nearby intensive care unit. An animal study from University College Hospital London [1] demonstrated that measuring glucose (often available as part of the blood gas analysis) was helpful in giving a prognosis of reversibility of the condition. A low glucose implies consumption of substrate and predicts smooth muscle damage. This was in a unit treating adults who may have a penile prosthesis placed acutely [2]. This is probably not appropriate in paediatric cases, although in severe cases in mature teenagers it may be considered.

All urology units should have rapid access to phenylephrine, not waiting for dispensing from the pharmacy at unsociable hours. Remember time is smooth muscle, so consider low-flow priapism like a "myocardial infarct of the penis". Patients at risk of priapism should be prompted not to wait for the arbitrary 4-h cut-off for the definition of priapism. They should be instructed to keep fasting, attend the emergency department urgently, and prompt staff to get them seen immediately by a urologist/paediatric urologist. The man's long-term sex life depends on prompt identification and treatment of priapism.

Hopefully, the quality of evidence will improve so more progress can be made in our understanding and management of this condition.

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DOI of original article: http://dx.doi.org/10.1016/j.jpurol.2013.07.024.