Diagnosis and management of priapism

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Priapism, a prolonged penile erection, is a rare urological emergency which can pose a significant management challenge. In this article the authors describe the pathophysiology and management of the condition.

Priapism represents a disorder of the normal physiological mechanisms that govern penile tumescence and detumescence, resulting in a prolonged penile erection that is unrelated to sexual stimulation and persists despite ejaculation. It is further subdivided into three categories: ischaemic or low-flow priapism (IP), non-ischaemic or high-flow priapism (NIP), and stuttering priapism, also known as recurrent ischaemic priapism (RIP).

In cases of IP there are specific high-risk groups, including patients with sickle cell disease or those undergoing antipsychotic treatment. The common risk factors for IP are listed in Box 1.

The aim of treatment is the immediate resolution of the painful erection and the preservation of cavernosal smooth muscle function so as to prevent long-term penile shortening and refractory erectile dysfunction due to the development of cavernosal fibrosis. However, the correct diagnosis must be made as the treatment options are very different for each subtype.

**DIAGNOSIS OF PRIAPISM**

Diagnosis is based on clinical history and examination, and supplemented by radiological imaging and corporal blood gas analysis. Further investigation using urine toxicology, haematological screening and abdominal cross-sectional imaging is performed to ascertain any underlying cause, although the majority of IP cases are idiopathic.

**CLINICAL HISTORY**

IP often presents well beyond the four-hour time frame commonly used as the time interval in guidelines whereby smooth muscle necrosis starts. This is partly due to a lack of patient awareness. Unfortunately, as the duration of IP increases, the greater the risk of smooth muscle necrosis. There may be a background of illicit drug use or prescription medications, which should be...
segments of the corpus cavernosum. In NIP, development of aberrant blood flow in Doppler studies can be difficult due to the performed, the interpretation of penile cavernosum. However, if aspiration of impaired perfusion of the distal corpus flow within the cavernosal arteries and studies will demonstrate reduced or absent subtypes. In cases of IP, penile Doppler cavernosum to differentiate IP from NIP the cavernosal arteries and the corpus is more rigid in NIP but the clinical picture overlying penile shaft skin rarely undergoes still able to void despite the ischaemic is obvious discomfort with a fully rigid ischaemic priapism in patients presenting with priapism there is obvious discomfort as opposed to severe penile pain, but it is important to accurately differentiate this condition from IP.

EXAMINATION
In patients presenting with priapism there is obvious discomfort with a fully rigid erection. Generally, patients with IP are still able to void despite the ischaemic blood within the corpus cavernosum. The overlying penile shaft skin rarely undergoes any colour or temperature change. The glans is more rigid in NIP but the clinical picture can change as treatment is instituted.

RADIOLOGICAL IMAGING
Colour penile Doppler ultrasonography is performed to assess the blood flow within the cavernosal arteries and the corpus cavernosum to differentiate IP from NIP subtypes. In cases of IP, penile Doppler studies will demonstrate reduced or absent flow within the cavernosal arteries and impaired perfusion of the distal corpus cavernosum. However, if aspiration of blood from the corpora has already been performed, the interpretation of penile Doppler studies can be difficult due to the development of aberrant blood flow in segments of the corpus cavernosum. In NIP, the Doppler studies will demonstrate a high velocity throughout the corpus cavernosum and often show a fistula. A more reliable investigation to distinguish between the two types of priapism is to aspirate blood from the corpus cavernosum for blood-gas analysis. Blood with low oxygen and acidosis indicates a diagnosis of IP; normal oxygen and normal pH indicate NIP.

MANAGEMENT OF ISCHAEMIC PRIAPISM
If the duration of priapism is between 4–24 hours at presentation, initial conservative management involves ejaculation, vigorous physical exercise and ice packs, with the aim of inducing smooth muscle contraction via sympathomimetic α-receptors and inducing detumescence. Failing this, more invasive approaches are required.

Aspiration and instillation of α-agonists
The next step involves aspiration of ischaemic blood from the corpora cavernosa using a large gauge needle (19G) placed either through the glans penis and into the corpora, or alternatively into the penile shaft at the 2 or 10 o’clock position, which avoids the neurovascular bundles. The needle can be used to inject α-adrenergic agonists (usually 200μg of phenylephrine repeated to a maximum of 1500μg) in an attempt to increase the smooth muscle tone and promote penile detumescence. Alternative α-adrenergic agonists include metaraminol and adrenaline. High-dose phenylephrine (total dose 2mg in aliquots of 200-500μg) has also been successfully used in small case series, although in refractory cases it is unlikely to be successful due to the development of irreversible smooth muscle dysfunction. Aspiration of ischaemic blood alone may resolve IP in up to one third of cases and should therefore always be attempted before injecting phenylephrine, as the smooth muscle contraction is impaired by the development of an ischaemic microenvironment. Phenylephrine injection should be performed with continuous monitoring of blood pressure, especially in patients with pre-existing hypertension, as it may potentially precipitate a cardiovascular event.

Although corporal blood aspiration and instillation of α-adrenergic agonists should be performed in all patients, irrespective of the time of presentation, priapism episodes lasting more than 24–36 hours are unlikely to respond to this intervention alone due to the presence of irreversible smooth muscle damage. The absolute time point at which irreversible damage to the smooth muscle occurs is unknown, although hypoxia, glucopenia and acidosis usually occur within six hours of the start of IP.

If IP is reversed within 24 hours, there is usually recovery of erectile function in approximately 50% of patients; a duration of more than 36 hours is invariably associated with a degree of corporal fibrosis and erectile dysfunction. The outcomes in the time period 24–36 hours is variable and governed by the degree of ischaemia and the amount of smooth muscle function recovered if penile detumescence is achieved.

Surgical shunts
Patients who do not respond to corporal blood aspiration and instillation of α-adrenergic agonists often require surgical shunts. The aim of surgically created shunts is to allow drainage of stagnant blood from the corpora cavernosa to either the corpus spongiosum or saphenous vein.

Winter and Ebbehoj shunts are minimally invasive, distal, percutaneous shunts. The Winter shunt uses a large-bore needle inserted into the glans penis and the tip of the corpus cavernosum. The Ebbehoj technique consists of a stab incision with a number 10 scalp into the corpora cavernosa through the glans penis. In cases where percutaneous shunt surgery fails, an Al-Ghorab shunt has been used. This is an open corporo-glanular shunt involving the excision of a segment of tunica albuginea from the tip of the corpus cavernosum. More recent shunt techniques involve the creation of a wider communication between the distal corpora and glans penis. Described as a T-shunt, the technique involves the insertion of a number 10 blade through
GENITAL PROBLEMS

the glans penis into the ipsilateral corpus cavernosum, followed by a 90-degree rotation laterally, away from the urethra, to create a fistula (Figure 1). The procedure can be repeated on the contralateral side if detumescence is unsuccessful (TT-shunt procedure). Where the TT-shunt fails, a tunnelling manoeuvre can be performed, with the aim of allowing the blood to drain from the proximal aspect of the corpora cavernosa. This procedure, also known as the corporal snake manoeuvre, uses a 22-French urethral sound through the previous T-shunt. 

Although initial reports showed that a combination of a distal shunt with the tunnelling manoeuvre is a safe technique, with almost all cases resolved, a recent series of 45 patients has shown that the success of the T-shunt and tunnelling manoeuvre is dependent on the duration of the priapism.

Penile prosthesis implantation – the traditional treatment in patients developing erectile dysfunction following a prolonged priapism – is now advocated as an alternative initial management for prolonged IP. Immediate penile prosthesis implantation in patients with refractory priapism and cavernosal smooth muscle necrosis reduces the painful priapism episode, guarantees the adequate long-term rigidity necessary for sexual intercourse, and prevents the otherwise inevitable penile shortening secondary to the development of corporal fibrosis (Figure 2).

As the potential overtreatment of patients with no evidence of necrosis in the cavernosal smooth muscle is one of the risks associated with this approach, the correct timing of surgery is paramount. Although irreversible smooth muscle necrosis has already occurred after 24–48 hours of a persistent erection, additional imaging and cavernosal smooth muscle biopsies can aid the decision as to whether an early penile prosthesis is suitable, or whether to adopt a conservative approach in the hope that there is cavernosal smooth muscle recovery in the long term (Figure 3). In a series of 23 patients, in which the radiological findings were correlated with biopsies from the corpus cavernosum, gadolinium-enhanced high-definition magnetic resonance imaging (MRI) of the penis had a sensitivity of 100% when used to detect the presence of necrosis of the cavernosal smooth muscle. This method may therefore represent an extremely useful way to support the clinical decision to proceed with acute penile prosthesis implantation rather than embarking on a conservative line of treatment.

Implantation of a malleable prosthesis is easier in the early period (ie the first two weeks) of refractory IP, as corporal fibrosis is limited and the cavernosal smooth muscle is softer. The risk of infection is higher when malleable implants are placed soon after acute priapism (up to 6% compared to 2% for later cases).

Delayed penile prosthesis
If patients are unwilling to undergo an acute insertion of a penile prosthesis, maintenance of penile length can be encouraged using a vacuum erection device for three to six months. In refractory cases, it is likely that dense fibrosis in the corpus cavernosum will ultimately occur. This represents a challenge for the surgeon as the fibrotic corpora renders insertion of the cylinders difficult, with a high risk of complications such as urethral injury, tunical/crural perforation and cylinder crossover.

Figure 2. Corporotomy showing smooth muscle necrosis during surgery for an acute penile prosthesis placement

Figure 3. Penile MRI performed in a case of ischaemic priapism showing no enhancement of the smooth muscle in the corpus cavernosum. Biopsy of the smooth muscle confirms necrosis
NON–ISCHAEMIC PRIAPISM
The presence of oxygenated blood in the corpora and lack of severe penile pain indicates NIP, indicating that the condition can be managed conservatively, provided that the diagnosis is accurate. After a period of conservative treatment, including regular clinical review, diagnostic angiography combined with embolisation of the fistula can be performed if the fistula has not closed spontaneously. Occasionally, patients can develop fibrosis within the distal corpora if the NIP is unresolved; early intervention is therefore advocated.17

CONCLUSION
Although priapism is a rare urological emergency, a prompt, accurate diagnosis and urgent intervention is required to preserve smooth muscle function and prevent the development of erectile dysfunction. The long-term outcome depends on the duration of the priapism. The initial management for ischaemic priapism involves corporal blood aspiration and instillation of pharmacological agents. If detumescence fails, then surgical intervention with the use of shunts or an early penile prosthesis is required.

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REFERENCES