Priapism: Current Updates in Clinical Management; Review article
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ABSTRACT
Priapism a status of continues erected penis more than four hours without erotic stimulus, which is referred to as a persistent erection. There are three recognised types of priapism at this time: ischemic or low-flow priapism, non-ischemic or high-flow priapism, and stuttering priapism. These are all based on the history and pathophysiology of the individual episodes.

It is characterised by a long-lasting, painful, and rigid erection produced by an abnormality in venous blood outflow from the corpora cavernosa, which is similar to penile compartment syndrome. Patients of ischemic priapism with sickle cell syndrome are more susceptible to stuttering priapism, which is characterised by self-limiting, recurring, and intermittent erections. When arterial blood drains excessively into the corporus cavernosus, a condition known as non-ischemic priapism results in an erection that is neither painful nor rigid. Because the emergency status and treatment choices for ischemia and non-ischemic priapism differ, it is necessary to make an accurate distinction between the two in order to begin appropriate therapeutic therapy. An important part of treating and managing priapism is ensuring that patients retain their ability to perform sexual functions even after the symptoms of priapism have disappeared. Medical and surgical advances in treating and preventing priapism are reviewed in this article, as well as scientific studies in this rapidly evolving subject.

Keywords: Erectile dysfunction; Penile erection; Priapism; Surgery; Urological.

INTRODUCTION
Priapism is considered a rare urological crisis that, if not recognised and treated immediately, could lead along run malfunctional erection. Priapism stands a malfunction in the physiological systems that regulate penile distension and swelling subsides, resulting in an abnormally extended erection of the penis which is not related to erotic stimulus (1).

There are three recognised types of priapism at this time: ischemic or low-flow priapism, non-ischemic or high-flow priapism, and stuttering priapism, which is sometimes referred to as recurrent ischemic priapism (4).

For purposes of consistency, the concepts ischemia and non-ischaemia are better over low drainage and high drainage, especially while understanding the penis scanning which follows the treatment procedures; this similarly confirms that physicians recognize the importance of interference (2).

The main approach of the therapy is to resolve the pain related to erecting penis immediately and in time stabilizing the cavernosal smooth muscle activity to evade the long term penis shortening and stubborn malfunction erection secondary to fibrosis of the cavernosa (3).

There hasn't been much significant advancement related to priapism, through the mainstream of original ideas pertaining to primary establishment of the penis prosthesis and a greater grasp of scanning techniques (4). There are three recognised types of priapism at this time: ischemic or low-flow priapism, non-ischemic or high-flow priapism, and stuttering priapism (sporadic, ischemic recurring). Every single case of priapism has its own unique set of aetiologic and pathophysiologic characteristics. As a direct consequence of this, numerous types of priapism each have their own unique options for therapy (5).

Definitions
Priapism requires immediate attention and may necessitate emergency management. (6)

1-priapism that is caused by a lack of blood-flow in the venous system
Priapism ischemic sort is accounting for 95 percent of priapism. A continuous erection, which is not related to sexual stimulation defined by a decrease or deficiency of intra-cavernous blood drainage and recognized of corporus cavernosus stiffness with little or no cavernous artery input. Clinical evidence of ischemic priapism is often painful and totally stiffed corpus cavernos with slight or with no association of the corpora spongiosa and the glans penis (6).

Acidosis and hypercarbia are seen in cavernous blood gas analysis. After 4 to 6 hours of ischemia, irreversible bodily damage ensues. Priapism ischemic type is considered a type of pouch syndrome characterised by a compression surrounded by the constrained space of the of corpus cavernosus, which rigorously impairs the circulation in the tissues of cavernous (7).

Stuttering (sporadic or recurring) priapism is something of an ischemic priapism. Stuttering priapism is characterised by recurring periods of detumescence followed by undesired, prolonged, and painful erections. This priapism frequently necessitates multi official visiting the urgency care room for the treatment. These outbursts are frequently self-limited and last less than three hours. (8)

If priapism continues and is not treated speedily, there is a danger of fibrotic damage of the corpora cavernosa. Because of hyper viscidness, augmented adherence of the blood to the endothelium of the vessels, and alteration the intravenous homeostasis, the incidence percentage of stutter priapism kind exists highly with the patients of sickle cells syndrome (9).
2. Priapism that is not ischemic (i.e., arterial, high-flow): An insistent erection not related to sexual reproduction that is not deemed urgency is known as non-ischemic priapism. An arteriolar-sinusoidal fistula causes uncontrolled arterial influx to the corpora cavernosa, bypassing the regulating, extremely resistant helicine arteries. The cavernous blood gases are neither hypoxic nor acidic, and the corpora cavernosa are typically not entirely stiff, sensitive, or in acute pain. The most prevalent cause is a history of penile or perineal injuries. Due to the fact that the cavernous tissues are well-oxygenated, non-ischemic priapism does not constitute an urgent medical situation (10).

3. Priapism of stuttering status:
Sickle cell disease (SCD) is supposed to stand the source of stuttering priapism, which is characterised by painful erections that come and go. Tribal India, Sub-Saharan Africa, and the Middle East have the highest prevalence of SCD, whereas Japan and Korea have the lowest prevalence.

Patients with SCD get their primary episode of priapism in more than 75% of cases of stutter priapism, and SCD seems to be the most common cause of priapism in children (11).

Stuttering priapism, on the other hand, can be idiopathic or drug-induced, eventually leading to ischemic priapism, and diagnostic and therapeutic treatment is primarily focused on ischemic priapism. The lower phosphodiesterase-5 (PDE5) levels in males with SCD suggest that the reduced endothelial nitric oxide (NO) activity leads to neuronal nitric oxide release, which can contribute to improper corpora cavernosa relaxation (12).

4. Malicious priapism
Malicious priapism status is a rare situation caused by metastases spread of the penile part primary from urinary bladder, prostatic, rectosigmoid, and tumours of the kidney (so-called “malignant priapism”). In 20–53 percent of instances, penile metastases cause priapism. The precise mechanism is unknown; however, infiltrating tumours, venous obstruction, and continuous neural circuit activation have all been postulated as plausible explanations (12,13).

Updates work up in priapism diagnoses:
When done in conjunction with thorough medical history and body built exam, corpus cavernosum blood gases investigations can help confirm a diagnosis and classify patients according to their specific subtypes. While many ischemic priapism patients are thought to be undiagnosed, in order to discover the underlying reason, further techniques such as urine toxicology, haematological screening, and abdominal imaging are utilised (13).

Another method of non-ischemic priapism management to be the regulation of low-flow priapism, which involves intromission. Cavernous arteries have been shown to be damaged or transected by the needles themselves, resulting in a new fistula and transforming an ischemic priapism into a non-ischaemic situation (14).

Causes of priapism are found in table 1.

**Table 1: Priapism causes**

<table>
<thead>
<tr>
<th>Priapism type</th>
<th>Causes</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ischaemic priapism</td>
<td>Idiopathic, haematological dyscrasias (e.g. sickle cell anaemia, thalassemia, and leukaemia), illegal drugs (such as cocaine and marijuana); medical drugs (such as psychotropic drugs, antidepression drugs, injection of prostaglandin E1 intra cavernosa, anti-coagulation drugs, and alpha blocker drugs too); malignant tumours of the pelvis; neurological conditions (such as injury of the spinal cord); toxic infections (such as scorpion stings); and metabolic disorders (e.g. gout arthritis, amyloid disorder).</td>
</tr>
<tr>
<td>Non-ischaemic priapism</td>
<td>Injuries of the penis/perineum and treatment of ischaemic priapism type.</td>
</tr>
</tbody>
</table>

**Clinical background:** Ischaemic priapism frequently manifests long beyond the four-hour timeframe typically cited in international guidelines as the duration during which corpus cavernosum smooth muscle necrosis occurs. There may have been sharp or piercing perineal or genitalia injuries prior to the commencement of non-ischaemic priapism type. Prior to the beginning of priapism, this trauma may have happened weeks before the actual event. It's crucial to distinguish this illness from ischemic priapism, which needs rapid medical attention despite the fact that most patients only have mild penile discomfort (14).

**Radiological imaging in priapism**
Color penile Doppler ultrasonography could be applied for assessing blood drainage inside the cavernosal arteries and the corpora cavernosa in order of identifying ischemic type from non-ischaemic type of priapism. Color penile Doppler imaging reveals weakened blood drainage of the corpora cavernosa distal end and decreased or nonexistent exudation inside the cavernosum blood vessels in the ischemic priapism instances. However, if blood has already been aspirated from the corpora, interpreting penile Doppler studies can be problematic because of the development of abnormal aberrant blood flow in sections of the corpus cavernosum (15). A high frequency in the corpus cavernosum and, in most cases, a fistula is shown by Doppler studies in patients of non-ischaemic priapism. Doppler waveforms might be difficult to interpret following therapy, as shown in recent investigations.

**Blood gases exam: (Table 2)**
To identify if a patient has ischemia or non-ischemic priapism, aspirating blood sample from the corpus cavernosum for doing blood gases investigation is more accurate. Injury that is caused by ischemic infarction may be confirmed by testing the blood pO2 and pH.
levels; cases of non-ischaemic can be proven if these values are within normal ranges (16).

Table 2: Results from blood gases test for ischemic and non-ischaemic priapism (17)

<table>
<thead>
<tr>
<th>Variable</th>
<th>Ischemic priapism (low flow)</th>
<th>Non-ischaemic priapism (high flow)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Etiology</td>
<td>SCD, different medications, malignancies injected intravenously, and other idiopathic causes</td>
<td>Antecedent trauma</td>
</tr>
<tr>
<td>Symptoms</td>
<td>Painful, stiff, and with a full erection.</td>
<td>Erection that is painless, not entirely stiff, and lacking in fullness</td>
</tr>
<tr>
<td>Corporal blood gas analysis</td>
<td>pH≤7.25, PO$_2$≤30 mmHg, and PCO$_2$≥60 mmHg</td>
<td>PO$_2$&gt;90 mmHg, PCO$_2$&lt;40 mmHg, pH 7.40</td>
</tr>
<tr>
<td>Compression signs</td>
<td>Not positive</td>
<td>Positive</td>
</tr>
<tr>
<td>Color Doppler</td>
<td>Cavernous blood flow is disrupted.</td>
<td>Arteriolar-sinusoidal fistula with turbulent cavernous blood flow</td>
</tr>
<tr>
<td>CT scan</td>
<td>Not commonly used</td>
<td>Arteriocoronal fistula, other pelvic injuries</td>
</tr>
<tr>
<td>MRI</td>
<td>Not commonly used</td>
<td>Arteriocoronal fistula</td>
</tr>
</tbody>
</table>

EVALUATION

The assessment consists of a reliable history, body examination, and other analytical studies in order to decide the scientific demonstration and to develop a treat strategy. It is vital to distinguish between ischemia and nonischemic priapism throughout the diagnostic process, as ischemia is a urological emergency. Even in the lack of laboratory and radiologic proof, early treatment can be initiated for ischemic priapism based on a range of clinical criteria (10).

A chest X-ray and a blood gas test are both necessary. Hyperacidity (pH= 7.25), anaemia (PO$_2$= 30 mmHg), excessive carbon dioxide (PCO$_2$ > 60 mmHg), and glucopenia on a blood gases test are all symptoms of ischemic priapism (18).

When the findings of the corporal blood gas examination are compatible with normal arterial blood gases values (pH= 7.4; PO$_2$>90 mmHg, PCO$_2$=40 mmHg), this is referred to as non-ischaemic priapism.

Ultrasonographic colour Doppler of the perineal pelvis and penis, in the hands of a competent practitioner, can aid in the identification of both forms of ischemic and nonischemic priapism. In most cases of ischemic priapism, the corpora cavernosa arterial flow is poor or nonexistent (19). An arteriolar-sinusoidal fistula or pseudoaneurysm, as well as a normal, high, uncontrolled, or unstable cavernous arterial vessels flow, strongly imply non-ischaemic priapism (18,19). The performance of an ultrasonographic colour Doppler may assist in detecting the impact of ischemic priapism treatment on a partially detumescent penis (19).

If chronic ischemia develops, or if alternative diagnostic facilities exist, such as resolved ischemia with edoema of the penis or conversion to high-flow priapism, extra intervention may be compulsory. The patient should be in the lithotomy or frog-leg position during the examination so that the perineum and the whole penile shaft may be scanned. This technical characteristic recognises the potential of a straddle injury or direct scrotal trauma causing an anomaly in the corpora cavernosa's perineal region. Penile arteriography is not utilised for the initial diagnosis of priapism. It is used to pinpoint the location of a suspected arteriocavernous fistula in males with nonischemic priapism. Penile arteriography is typically used in combination with embolization for fistula treatment (19).

There is currently interest in using magnetic resonance imaging (MRI) in priapism patients because MRI yields good findings of the corpora cavernosa. MRI, on the other hand, is not helpful in determining the cause of priapism, while it may be useful in imaging for rare cases of priapism.

Differential Diagnosis

Table 3 provides differential diagnosis for ischemia and non-ischaemic priapism.
### Clinical Management: (Figure 1)

The objective of clinical treatment for priapism is to alleviate the persistent erection and reserve the capability to achieve erection in the upcoming days. Consequently, emergent examinations are essential to evaluate the priapism, is ischemic or non-ischemic, in order to commence the correct treatment. Initial differentiation can be achieved based on the patient’s medical history and body exam.\(^{(20)}\)

Afterwards, confirmation must be achieved through capillary blood gas measurement and radiologic evaluation. The goal of the clinical care of ischemic priapism, an emergency that might lead to permanent erectile dysfunction (ED), is to eliminate the compartment state of the continuous cavernosal hypoxia. In contrast, non-ischemic priapism is not a surgical emergency since cavernosal hypoxia is not present. The intervention's main purpose is to treat the prolonged erection and restore ordinary erectile function as a consequence of sexual desires and interaction.

#### Variable | Ischemic priapism (low flow) | Non-ischemic priapism (high flow)
---|---|---
Angiography | Not commonly used | Arteriocorporal fistula, embolization

### Figure 1: Managing process for priapism

#### Ischemic priapism: (Figure 2)

Ischemic priapism is frequently associated with erectile dysfunction. Patients with sickle cell disorder are suffering up to five times more than healthy people to suffer from erectile dysfunction.

As a result, early detection and treatment are critical outcomes. During the first 24 hours, ischemic priapism is commonly treated with aspiration and an intravenous infusion of an alpha-adrenergic drug such as diluted phenylephrine.\(^{(21)}\)

Oral terbutaline, for example, has been shown to be less effective in increasing blood flow than phenylephrine.\(^{(22)}\) If conservative therapy fails within 24 hours or surgical intervention is not possible, detumescence is frequently required. A percutaneous (Ebbehoj or Winter shunt, T-shunt) or open (Al-Ghorab) approach is commonly utilised to create a
distal shunt between the corpora and glans. Even though surgeon familiarity has a role in decision-making, it is best to go from the least intrusive to the most invasive procedure\(^{(23)}\).

**Figure 2: Management of ischemic priapism**\(^{(24)}\)

Patients who have experienced ischemic priapism for more than two days may have considerable intracavernous tissue edema, which might hinder effective blood flow from the proximal to the distal penis. The proximal shunt \(^{(24)}\) and the cavernosum-venous shunt have historically been the gold standards for this surgery. Tunneling procedures, on the other hand, were developed by Burnett et al. \(^{(18)}\) and Hudnall et al. \(^{(24)}\) and have since supplanted the more complex and time-consuming processes that were previously utilised.

Consider that in all of the preceding treatments, the tunica albuginea must be sliced open in order to expose the collagen to coagulation factors in the penile blood, which initiates the clotting cascade. Priapism was successfully treated with perioperative anticoagulation. Patients who have had an ischemia episode lasting more than 48 hours previous to intervention should have a penile prosthesis implanted \(^{(25)}\).

Patients who receive therapy within two weeks after the ischemic event are less likely to experience postoperative problems including erosion or infection, as well as being happier and less likely to experience penile shortening as a result \(^{(25)}\). Rapid implantation of a flexible prosthetic penis enables easy overall capacity with an inflated prosthesis at a later date, perhaps saving money in the long run and reducing hospital resource use.

**Non-ischemic (arterial, high vascular drainage) priapism**

Because non-ischemia priapism seldom causes penile ischaemic impairment, delaying and if the treatment is not expected to have significant consequences. As a result, monitoring (clinical surveillance) is the primary intervention indicated \(^{(26)}\).

According to scientific trials clarified that two-thirds of instances spontaneously resolve themselves. Non-ischemic priapisms can also cause erectile dysfunction. Non-physiologically high intracavernosal \(\text{PaO}_2\) may be a contributing factor. \(^{(27)}\) Nonischemic priapism is best treated with angiography and super-selective embolization. With this treatment, the abnormal arteriovenous connection will be broken. Another choice is blockage of the cavernous artery. \(^{(28)}\)

Gluteal ischemia, penile gangrene, or purulent cavernositis can complicate these treatments. There is a 30% to 40% chance of further embolization, and the chances of erectile dysfunction are between 5% and 39% depending on whether the substance is absorbed or not \(^{(28)}\).

If non-absorbable constituents, such as application of gelatin foam or autologous clots, erectile dysfunction may be transitory. The preservation of erectile function will be achieved through vascular recanalization in this instance. Color duplex ultrasonography (CDU)-guided compression and super-selective embolization have been shown to improve success rates.

Erectile dysfunction can occur in as many as 50% of men who have undergone open surgery. As a result, open surgery isn't recommended as a first resort. When all other treatment options have failed, open surgery is the only alternative. \(^{(30)}\)

**Stuttering (intermittent or recurrent) priapism**

Because there is a chance of penile ischemia and the condition has a propensity to get worse, stuttering priapism should be treated with caution when it occurs.
There are both reactive and proactive treatment options available for this condition, including systemic hormone therapy also penile prosthesis surgery to prevent disorder recurrences in the future (31).

The antiandrogenic effects of most commonly used to be hormone usages (e.g., androgen receptor antagonists, gonadotropin-releasing hormone agonists, and 5-alpha reductase inhibitors) limit their efficacy. Adult height and sexual maturity are essential before a person may get hormone treatment, which can have serious side effects. A lack of predictability and reliance on case reports and case studies for the efficacy of various treatments currently restricts evidence-based recommendations for any one treatment (32).

Despite the fact that further research is needed, there has been an increase in the usage of long-term, low-dose PDE5 inhibitor therapy as part of a preventative, molecular-mechanism-based treatment approach (32).

Self-injection of intracavernosal sympathomimetics may be utilised to terminate a severe episode of acute ischemic priapism, therefore avoiding hospitalisation. This therapy is not suggested for stuttering priapism since it can only treat acute symptoms and does not prevent further attacks from occurring.

It is an alternate treatment for priapism in patients who cannot take systemic preventative medication or whose symptoms are resistant to it (33).

Ralph et al. (34), created a device of implant for delivering sympathomimetic stuffs to treat the stuttering priapism. Surgical methods, like as penile prosthesis implantation, may be explored for men with stuttering priapism who have not responded to pharmaceutical therapy.

Update on the management of ischaemic and non-ischaemic priapism

In a subset of patients, daily low-dose PDE-5 inhibitor therapy prevented recurrent priapism while preserving normal erectile function. Burnett et al. revealed a unique therapy to prevent recurrent ischemic priapism (stuttering) in a study that offered data to support the function of nitric oxide synthase/PDE-5 dysregulation as a significant factor in the genesis of priapism(12).

Due to the existence of oxygenated blood inside the corpora and the absence of immediate penile pain, non-ischaemic individuals can be treated conservatively if the initial diagnosis is accurate. Any fistula can be treated with diagnostic angiography and super-selective embolization if the fistula has not spontaneously healed following a conservative treatment plan that needs routine clinical evaluation. Patients with chronic non-ischaemic priapism may develop fibrosis inside the distal corpora; hence, early embolization is advised to prevent this from occurring. Distal flaccidity is a sign of distal corpus cavernosum fibrosis, which is best seen utilising a penile MRI scan. (35)

CONCLUSIONS

Treatment for priapism aims to detumesce persistent penile erections and maintain erectile function once priapism has been resolved. As a result, even though priapism is rare, quick examination and emergency management are typically necessary. Priapism’s pathogenesis, diagnosis, and treatment have all been improved thanks to a slew of earlier clinical research. Urological doctors should be prepared to treat this urgent situation in order to ensure successful outcomes. Among the several erectile dysfunctions, treating priapism remains a difficult task.

In order to prevent ischemic priapism, immediate attention should be given to all cases of acute priapism. Aspiration/irrigation with sympathomimetic injections, surgical shunts, and the placement of a penile prosthesis are all methods of treatment. Conservative treatment options exist for non-ischemic priapism. Preventative measures are the most important part of treatment; however acute episodes should be treated in accordance with guidelines for ischemic priapism. Priapism research created new treatments that could be used in conjunction with current medicines. Focusing on prevention and early intervention is critical to preventing this rare but clinically devastating disease.

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REFERENCES


