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Guidelines

European Association of Urology Guidelines on Priapism

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Abstract

Context: Priapism is defined as a penile erection that persists beyond or is unrelated to sexual interest or stimulation. It can be classified into ischaemic (low flow), arterial (high flow), or stuttering (recurrent or intermittent).
Objective: To provide guidelines on the diagnosis and treatment of priapism.
Evidence acquisition: Systematic literature search on the epidemiology, diagnosis, and treatment of priapism. Articles with highest evidence available were selected to form the basis of these recommendations.
Evidence synthesis: Ischaemic priapism is usually idiopathic and the most common form. Arterial priapism usually occurs after blunt perineal trauma. History is the mainstay of diagnosis and helps determine the pathogenesis. Laboratory testing is used to support clinical findings. Ischaemic priapism is an emergency condition. Intervention should start within 4–6 h, including decompression of the corpora cavernosa by aspiration and intracavernous injection of sympathomimetic drugs (e.g. phenylephrine). Surgical treatment is recommended for failed conservative management, although the best procedure is unclear. Immediate implantation of a prosthesis should be considered for long-lasting priapism. Arterial priapism is not an emergency. Selective embolization is the suggested treatment modality and has high success rates. Stuttering priapism is poorly understood and the main therapeutic goal is the prevention of future episodes. This may be achieved pharmacologically, but data on efficacy are limited.
Conclusions: These guidelines summarise current information on priapism. The extended version are available on the European Association of Urology Website (www.uroweb.org/guidelines/).
Patient summary: Priapism is a persistent, often painful, penile erection lasting more than 4 h unrelated to sexual stimulation. It is more common in patients with sickle cell disease. This article represents the shortened EAU priapism guidelines, based on a systematic literature review. Cases of priapism are classified into ischaemic (low flow), arterial (high flow), or stuttering (recurrent). Treatment for ischaemic priapism must be prompt in order to avoid the risk of permanent erectile dysfunction. This is not the case for arterial priapism.

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1. Introduction

Priapism is a pathologic condition representing a true disorder of penile erection that persists beyond or is unrelated to sexual interest or stimulation [1]. Overall, erections lasting up to 4 h are by consensus defined as *prolonged* (level of evidence [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 yr of age [4] increasing up to 42% in patients ≥18 yr of age [5–7].

2. Methodology

A systematic literature search of the Medline database was performed. The controlled vocabulary of the Medical Subject Headings database was searched using the term *priapism*. This search yielded 1199 articles (125 review articles, 404 original articles, and 670 case reports). The expert panel also identified critical problems and knowledge gaps, enabling priorities to be established for future clinical research. These European Association of Urology (EAU) guidelines on priapism were presented for the first time by the EAU Male Sexual Dysfunction Guidelines Panel.

3. Classification

3.1. 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 [8]. The patient typically complains of penile pain, and the examination reveals a rigid erection. Resolution of ischaemic priapism is characterised by the penis returning to a flaccid nonpainful state. However, in many cases, persistent penile oedema, ecchymosis, and partial erections can occur that may mimic unresolved priapism.

When left untreated, resolution may take days, and erectile dysfunction invariably results.

3.2. Arterial (high-flow or nonischaemic) priapism

Arterial priapism is a persistent erection caused by unregulated cavernous arterial inflow [8]. The patient typically reports an erection that is not fully rigid and 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.

3.3. Stuttering (recurrent or intermittent) priapism

Stuttering priapism, also termed intermittent or recurrent priapism, is a distinct condition characterised by repetitive, painful episodes of prolonged erections. Erections are self-limited with intervening periods of detumescence [9]. The duration of the erectile episodes in stuttering priapism is generally shorter than in the low-flow ischaemic type [1]. 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.

4. Epidemiology and pathophysiology

4.1. Ischaemic (low-flow or veno-occlusive) priapism

Ischaemic priapism is the most common form of priapism, accounting for >95% of all priapism episodes [8,10]. In ischaemic priapism, there are time-dependent modifications in the corporal metabolic environment, progressively leading to hypoxia, hypercapnia, and acidosis.

Ischaemic priapism beyond 4 h is considered a compartment syndrome, characterised by pressure within the closed space of the corpora cavernosa that severely compromises circulation in the cavernous tissues. A compartment syndrome requires emergency medical intervention to minimise potential irreversible consequences such as corporal fibrosis and permanent erectile dysfunction [11,12]. The duration of priapism represents the most significant predictor of the maintenance of premorbid erectile function; in this context, interventions beyond 48–72 h since onset may eventually help relieve erection and pain, but they have little benefit in preserving erectile functioning. Histologically, by 12 h, corporal specimens show interstitial oedema, progressing to destruction of sinusoidal endothelium, exposure of the basement membrane, and thrombocyte adherence at 24 h. At 48 h, thrombi can be found in the sinusoidal spaces, and smooth muscle necrosis with fibroblast-like cell transformation is evident [12].

In terms of pathophysiology (Table 1), ischaemic priapism has been identified as idiopathic in most cases [8,13]. Moreover, ischaemic priapism has been associated with sickle cell anaemia, haematologic dyscrasias, neoplastic syndromes, and the use of several different medications. Ischaemic priapism occurs relatively often (0.4–35%) after intracavernous injections of papaverine, phentolamine, and/or prostaglandin E1 [8,14–16] (Table 1). However, most of these cases were treated with papaverine-based combinations; the prevalence of priapism is <1% in the case of prostaglandin E1 [15]. Since their introduction on the market, a few cases of priapism have been described in men who have taken phosphodiesterase type 5 inhibitors (PDE5-Is) [8]. Most of these men had histories of increased risk for priapism including sickle cell disease, spinal cord injury, combined administration of PDE5-I and intracavernosal injection of vasoactive agents, a history of penile trauma, abuse of narcotics or psychotropic medication, or taking PDE5-I for recreational purposes without a medical reason [8].

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, with a lifetime probability of developing ischaemic priapism of 29–42% in men with sickle cell disease