**Priapism**

**Definition**
- **Abnormally prolonged erection** (>4 h) not resulting from sexual desire. It affects both corpora cavernosa (CC) and does not usually include the spongiosum tissue of the glans. The name comes from the Greek god *Priapus*.

**Types**
- **Veno-occlusive, ischemic, or low-flow priapism:**
  - The most common and dangerous type, this is a consequence of obstructed venous drainage of the penis, with little or no arterial blood flow.
  - The erection is complete and painful.
  - The accumulation of non-oxygenated blood produces ↓ PO₂ and pH, which leads to cell death and fibrosis with permanent ED within 24-36 h. Emergency treatment is required after the 4th h of priapism.
  - **Causes:** intracavernosal injection of vasoactive drugs (IIVD), medications (*antihypertensives*, *psychotropics*, *vasodilators*, *androgens*), drugs (*cocaine*), alcohol, sickle cell disease (more common in blacks), leukemia, hyperviscous states such as prenatal overfeeding or withdrawal of anticoagulants, traumatic hematoma with compression of venous drainage, vasculitis, dialysis, urinary retention, spinal cord injury, malignant priapism (bladder, prostate, kidney, and gastrointestinal tumors, in that order of frequency), idiopathic.

- **Arterial, nonischemic, or high-flow priapism:**
  - Traumatic fistula of the cavernous artery or one of its branches with lacunar spaces.
  - The erection is partial (60-75%) and not painful. May be very prolonged (days).
  - Because it is well-tolerated and reversible, treatment can be delayed.
  - **Causes:** blunt penile or perineal trauma, IIVD with laceration of the cavernosal artery, iatrogenic (from medical or surgical treatment of venous priapism or from surgeries such as internal urethrotomy or plication in cases of Peyronie's disease), idiopathic. Onset of priapism may occur hours or days after the initial trauma.

**Diagnosis**
- **Differential diagnosis between ischemic and nonischemic priapism:**
  - **Anamnesis:** to search for causes suggesting one or the other type of priapism.
  - **Symptoms:** complete, painful erection (venous); or partial, painless erection (arterial).
  - **Doppler ultrasound:** absence (venous) or presence (arterial) of arterial flow.
  - **Blood gas analysis of the CC:** in venous priapism ↓ PO₂, ↑ PCO₂, and ↓ pH; in arterial priapism, blood gas results are the same as for peripheral arterial blood.

- **Etiological diagnosis:**
  - **CBC:** to detect anemia, leukemia, platelet disorders, etc.
  - **Peripheral blood smear and Hb electrophoresis:** to detect sickle cell anemia and HbS.
  - **Detection of drugs in blood or urine:** when drug abuse is suspected.

**Treatment**
- **Venous, ischemic, or low-flow priapism:** if one option fails, the next should be tried.
  - **1st Suction-irrigation:** puncture one CC with a 19-21 G needle, followed by compression of the penis to extract dark blood until red blood appears and subsequent irrigation with 40 mL of cold NSS.
  - **2nd Intracavernous (IC) agents:** 1st choice is Phenylephrine with monitoring of BP and pulse. If SBP >200, oral administration of 10 mg Nifedipine (ADALAT®) or 25 mg Captopril (CAPOTEN®).
<table>
<thead>
<tr>
<th>Generic name</th>
<th>Brand name®</th>
<th>Dose</th>
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<tbody>
<tr>
<td>Phenylephrine</td>
<td>PHENYLEPHRINE 0.2 mg/mL solution (1 mL 1 mg/mL solution + 4 mL NSS)</td>
<td>1 mL every 5-10 minutes (max 5 mL) until detumescence occurs</td>
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<tr>
<td>Etilefrine</td>
<td>EFFORTIL 10 mg/mL solution</td>
<td>0.4 mL every 5 min (max 20 mg)</td>
</tr>
<tr>
<td>Methylene Blue</td>
<td>METHYLENE BLUE</td>
<td>50 mg every 5 min</td>
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- **3rd Shunts:**
  - **Percutaneous distal CC-glans shunts:** the Winter technique (with a biopsy needle) and the Ebbehøj technique (with a scalpel) are the procedures of choice as they induce less permanent ED. The simplest is the Winter technique: with a prostatic biopsy needle and under local anesthesia (penile block), multiple holes are made in the tip of each CC through the glans penis. In the postoperative period the patient should be monitored for recurrence of priapism due to shunt closure; if there is doubt as to whether the patient suffers a recurrence of ischemic priapism, postoperative penile edema, or development of high-flow priapism, a blood gas analysis of the CC or a Doppler ultrasound is performed. If a recurrence of ischemic priapism is identified, penile compression is used to permeabilize the shunt (pediatric BP cuff). If the condition persists, the Winter or Ebbehøj technique is repeated or open surgery is performed.
  - **Open distal CC-glans shunt (Al-Ghorab):** resection of the most distal part of the tunica albuginea of both CC. These close less frequently than percutaneous shunts.
  - **Proximal CC-spongiosum shunt (Quackels) or CC-saphenous vein (Grayhack):** the last recourse if distal shunts fail.

- **4th Penile prosthesis:** in cases of ED after prolonged priapism. Some authors recommend early placement (before severe fibrosis and penile shortening develop) in cases of proximal shunt failure, ischemia lasting over 36 h, or if an MRI shows necrosis of the smooth muscle of the CC.

- **Etiological treatment of venous priapism:** in addition to the aforementioned measures.
  - **Sickle cell anemia:** in these patients, episodes of dehydration, fever, or exposure to cold can trigger priapism. Treatment: abundant iv hydration + iv bicarbonate + hypertransfusion to reach Hb >10 mg/dL + supplementary O₂ + opioid analgesia.
  - **Leukemia, bladder, or prostate tumors:** pelvic chemo/radiotherapy.

- **Arterial, nonischemic, or high-flow priapism:**
  - **1st Conservative treatment:** 62% resolve spontaneously with observation only. Other conservative measures can be taken, such as the occlusion of the arterial flow with external compression or the topical application of ice to the perineum.
  - **2nd Arteriography and selective embolization:** less ED with autologous clot or reabsorbable gelatin than with permanent materials. If there is recurrence, a second embolization is required.
  - **3rd Surgery:** in cases of very prolonged priapism caused by the formation of a pseudocapsule around the fistula or if embolization has failed. The fistula is ligated after being located with the aid of intraoperative Doppler ultrasound.
Priapism

Differential diagnosis

- IIVD, drugs, sickle cell disease, hyperviscous states, tumors
- Trauma, iatrogenic, idiopathic

Anamnesis
- Complete, painful erection
- Partial, non-painful erection
- Absence of arterial flow
- Presence of arterial flow

Symptoms
- Doppler ultrasound

Blood gas analysis
- pO₂, pCO₂ and pH as in arterial blood

venous

Emergency treatment
- 1º Suction-irrigation
- 2º Phenylephrine
- 3º Distal shunt
- 4º Proximal shunt

arterial

Delayed treatment
- 1º Compression or ice.
- 2º Arteriography and selective embolization
- 3º Surgery
Recurrent prolonged erections

- **Definition**: painful, long-lasting erections with subsequent spontaneous detumescence that can occur daily and which typically wake the subject up at night.
- **Etiology**: this disorder is often idiopathic, but sometimes appears in blood diseases such as sickle cell anemia. Any patient who has suffered an episode of ischemic priapism is at risk for recurrent priapism.
- **Prophylactic treatment**: physical activity is recommended because it increases sympathetic tone. Also recommended instructing the patient to self-inject *Phenylephrine* if an episode occurs, and a daily oral preventive drug such as:
  - **Alpha-adrenergics**: *Etilerine* or *Phenylpropanolamine*. First choice.
  - **PDE5 inhibitors**: *Sildenafil* or *Tadalafil* oo. First choice.
  - **Hormonal manipulation**: antiandrogens, LHRH analogs, estrogens, Ketokonazole (*+ prednisone*). Effective, but unsuitable for chronic use due to ↓ libido and ED.
  - **5-α-reductase inhibitors**: *Finasteride/Dutasteride*. Effective, but with side effects.
  - **Others**: Baclofen, Diazepam, Gabapentin, Hydroxyurea (only in sickle cell disease). Alternatives to 1st choice treatments should these fail or be contraindicated.

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<thead>
<tr>
<th>Generic name</th>
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<th>Dose</th>
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<tbody>
<tr>
<td>Etilerine</td>
<td>EFFORTIL 25 mg prolonged release capsules</td>
<td>Initial: 25 mg/night (max 100 mg)</td>
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<td>Phenylpropanolamine</td>
<td>ORNADE 50 mg tablets</td>
<td>Initial: 50 mg/night (max 150 mg/night)</td>
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<td>Sildenafil</td>
<td>VIAGRA 25-50 mg tablets</td>
<td>25-50 mg/night</td>
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<td>Tadalafil</td>
<td>CIALIS 5-10 mg tablets</td>
<td>5-10 mg/night three times per week</td>
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<td>Bicalutamide</td>
<td>CASODEX 50 mg tablets</td>
<td>50 mg/day or every two days</td>
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<td>Leuprolidine</td>
<td>ELIGARD 7.5 mg solution</td>
<td>1 im/month</td>
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<td>Cyproterone acetate</td>
<td>ANDROCUR 50 mg tablets</td>
<td>50 mg/12 h (max 100 mg/8 h)</td>
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<td>PROSCAR 5 mg tablets</td>
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<td>Dutasteride</td>
<td>AVODART 0.5 mg capsules</td>
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<td>Diazepam</td>
<td>VALIUM 10 mg tablets</td>
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<td>Baclofen</td>
<td>LIORESAL 10 mg tablets</td>
<td>1 tablet/night (max 4 tab/night)</td>
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<tr>
<td>Gabapentin</td>
<td>NEURONTIN 400 mg capsules</td>
<td>400 mg/night (max 2400 mg/night)</td>
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<tr>
<td>Hydroxyurea</td>
<td>HYDREA 500 mg capsules</td>
<td>20-35 mg/kg/day</td>
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