- *β-2-agonists*

Oral terbutaline is a β-2-agonist with minor β-1 effects and some α-agonist activity; although its mechanism of action is not yet fully understood [1385-1387]. The main use of terbutaline is for prevention of recurrent episodes of prolonged erection. Oral treatment with terbutaline was tested in three placebo-controlled RCTs [1386-1388] showing a success rate of 30 to 60% in patients with ischemic priapism associated with intracavernous injection of erectogenic agents. Terbutaline should be given cautiously in patients with coronary artery disease, increased intravascular fluid volume, oedema or hypokalaemia [1387]. In a single multi-centre prospective study, another β-2-agonist, salbutamol, has been reported to induce detumescence in 34% of cases of prolonged erection (more than 3 hours) after intracavernous injection of erectogenic agents [1389]. However, more robust data are needed to recommend oral salbutamol for the treatment of ischaemic priapism.

Table 10.4: Medical treatment of ischaemic priapism

Drug	Dose/Instructions for use
Phenylephrine	• Intracavernous injection of 200 µg every 3-5 minutes. • Maximum dosage is 1 mg within 1 hour. • Lower doses are recommended in children and patients with severe cardiovascular diseases.
Etilephrine	• Intracavernosal injection at a concentration of 2.5 mg in 1-2 mL normal saline.
Methylene blue	• Intracavernous injection of 50-100 mg, left for 5 minutes. It is then aspirated and the penis compressed for an additional 5 minutes.
Adrenaline	• Intracavernous injection of 2 mL of 1/100,000 adrenaline solution up to five times over a 20-minute period.
Terbutaline	• Oral administration of 5 mg for priapism lasting more than 2.5 hours, after intracavernous injection of vasoactive agents.

10.1.3.1.5 Management of priapism related to sickle cell disease

The results of a systematic review on the overall management of priapism related to SCD found that few studies were conducted exclusively on patients with SCD and studies on mixed populations usually did not report separate data on SCD patients [1331]. Clear and systematic reporting of patient characteristics, interventions and outcomes was lacking, and the length of follow-up, if reported, varied significantly among the studies. Overall, the quality of studies was deemed poor to allow high-quality, evidence-based recommendations to be made.

Urgent intervention is essential and the general approach is similar to that described for other cases of ischaemic priapism and should be co-ordinated with a haematologist [1390-1392].

However, as with other haematological disorders, other therapeutic interventions may also need to be implemented [1390, 1392, 1393]. Specific measures for SCD-related priapism include intravenous hydration and narcotic analgesia while preparing the patient for aspiration and irrigation. Additionally, supplemental oxygen administration and alkalinisation with bicarbonate can be helpful [1347, 1391].

Haemoglobin S (HbS) percentage should be measured in all SCD patients with acute priapism. Exchange blood transfusion has also been proposed, with the aim of increasing tissue delivery of oxygen [1394]. The transfused blood should be sickle cell haemoglobin negative and Rh and Kell antigen matched [1395]; however, the evidence is inconclusive as to whether exchange transfusion itself helps to resolve priapism. A systematic review reported that the mean time to detumescence was eleven days with exchange transfusions compared to eight days with conventional treatment. Moreover, there were 9 cases of ASPEN syndrome (association of SCD, priapism, exchange transfusion and neurological events) as a consequence of blood transfusion [1396].

A series of 10 patients with SCD-related priapism showed that it was safe to perform exchange transfusion [1394]; however, several reports suggest that exchange transfusion may result in serious neurological sequelae [1396]. Therefore, routine use of exchange transfusion is not recommended as a primary treatment intervention in this group unless there is a risk of SCD-related symptoms. However, in patients who failed medical management, transfusion may be required to enable general anaesthesia to be safely administered prior to definitive surgery [1397].