

GSTFT Clinical Practice Guideline

Management of Priapism in Sickle Cell Disease

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Definition

A persistent penile erection that continues hours beyond, or is unrelated to, sexual stimulation. The subtypes include: **Ischaemic** (veno-occlusive) with little or no cavernous blood flow, the corpora cavernosa are rigid and tender; **Non-ischaemic** (arterial) due to unregulated cavernous arterial flow. Priapism may presently acute or stuttering if it lasts for more than a few minutes but less than four hours; stuttering episodes may recur and/or develop into more prolonged episodes. Priapism is well known in sickle cell disease and as many as 90 percent of males with SCD will have experienced one or more episodes of priapism by the age of 20 years. Priapism in SCD is usually of the Ischaemic subtype due to obstruction of the venous drainage of the penis. Other causes include thalassaemia, trauma, leukaemia and drug induced. Prolonged priapism is an emergency that requires urologic intervention.

Psychosocial and counselling considerations

In early childhood, males need to know that Priapism is one aspect of SCD and that they should tell their parents or other appropriate adult if it occurs. If untreated, Priapism can result in impotence in the future. It can be triggered by a number of factors such as full bladder and prolonged sexual activity. Future recurrence can be prevented by the use of medication.

- Prolonged Priapism is an emergency and requires urgent assessment and treatment.
- Document the time of onset of the episode.
- Precipitating factors, such as trauma, infections, or the use of drugs (e.g., alcohol, psychotropic agents, sildenafil, testosterone, cocaine).
- A careful history physical examination is essential, note the TCD status prior to surgery

The aim of therapy is to relieve pain, abort the erection and preserve future erectile function.

ONSET LESS THAN 2 HOURS Patients should be advised to drink extra fluids, use oral analgesics, and attempt to urinate as soon as Priapism begins. Walking and warm-baths may also help to avert early Priapism.

EPISODES LASTING MORE THAN TWO HOURS

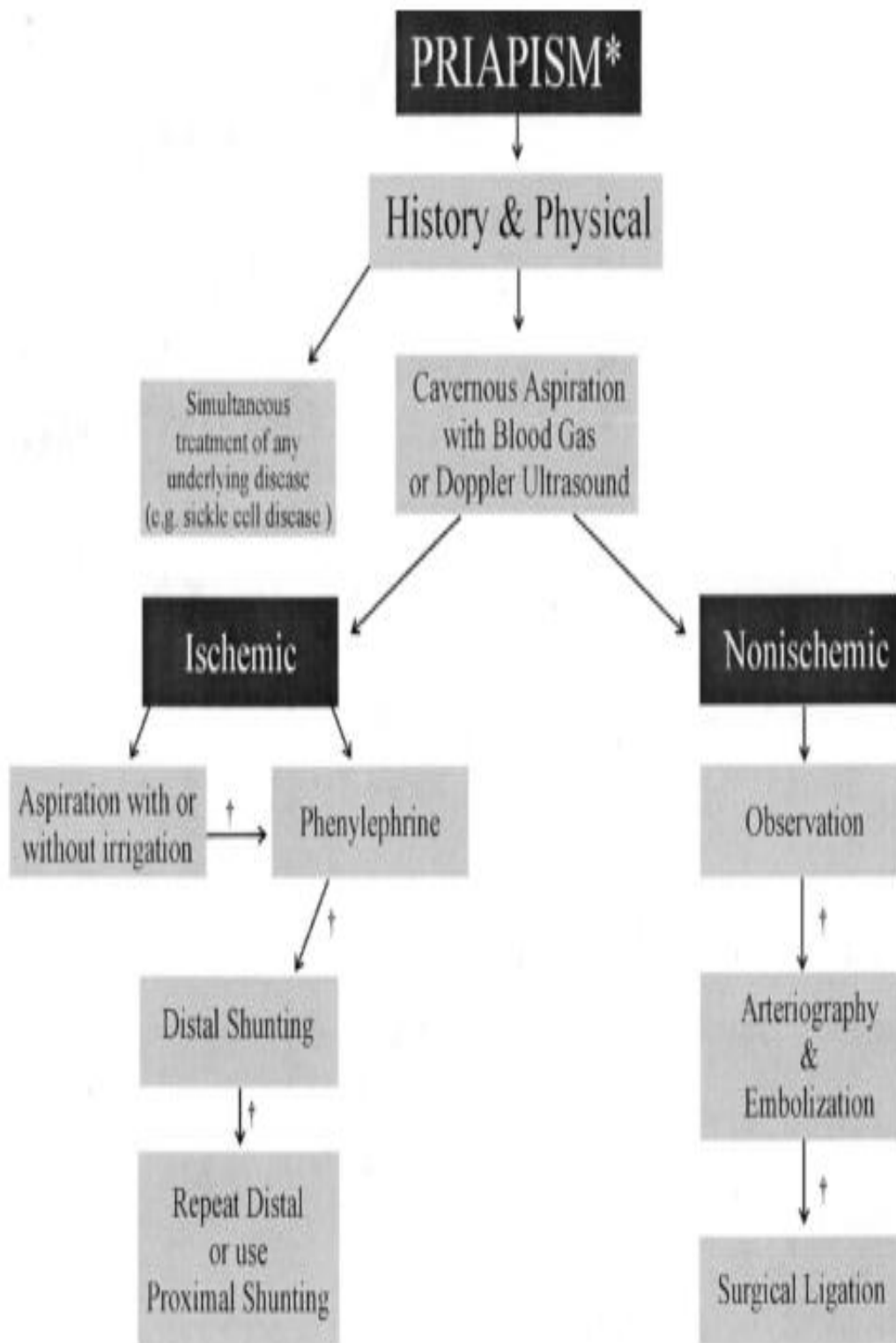
- Patient should be referred **directly** to the Paediatric Urology Team-02071887188 St Thomas Switchboard and for Paediatric Urologist on call as well as the Paediatric Registrar on bleep 0339 to liaise with Urologists and Dr Inusa's team (during working hours 02071887774 / 02071889432 / ask to page -CNS (Helen Appleby or Luhanga Musumadi) and / Or On-Call Paediatric Team (outside working hours) Consultant.
- While waiting to transfer to Evelina Hospital, the patient should be given intravenous fluids and adequate analgesia. The patient should be kept nil-by-mouth.
- Blood transfusion will not normally be necessary before any surgical procedure, but may be appropriate in patients with a history of chest crises, or post-anaesthetic problems.

- The initial treatment is usually penile aspiration and irrigation with an α -adreno-receptor agonist. If this does not relieve the Priapism, shunting procedures may be necessary. This plan is determined by the Urologist. Surgical Intervention should not be delayed by either medical treatment or the provision of blood products.
- If Priapism recurs and further surgery required, an exchange blood transfusion should be performed before the second anaesthetic in sickle cell disease patients, if this has not taken place already.
- Complications of Priapism and treatment include bleeding from the holes placed in the penis as part of the aspiration or shunting procedures, infections, skin necrosis, damage or strictures of the urethra, fistulae, and impotence.

Further Prevention

- Etilefrine 0.5mg/kg per day orally-(unlicensed prescription)
- Patients should be given a follow-up appointment with Paediatric Urology Team (Kalpana Patil or Arash Taghizadeh) as Paediatric Sickle Cell Clinic at Evelina

ERECTILE DYSFUNCTION GUIDELINE UPDATE



*Erection greater than 4 hours duration.

†Proceed upon treatment failure.

Management algorithm for priapism

References

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