The Management of Priapism in Children and Adolescents with Sickle Cell Disease

This guideline is intended for paediatricians caring for children with Sickle Cell Disease (SCD)… The urgent need to deal with priapism must be recognised, and delays avoided by ensuring urgent referral to the paediatric urology team at the Evelina. (Or appropriate local team in some children >13)

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Definition/Background

Priapism is a pathological condition of persistent and often painful penile erection that is beyond or unrelated to sexual interest or stimulation. In sickle cell disease, it is typically an ischaemic condition and may be classified as follows:

a) Prolonged; a single episode lasting more than four hours. This is a urological emergency.

b) Stuttering, if there are repetitive and painful episodes. The duration and frequency of these events are variable. Priapism can last anywhere from a few minutes to a few hours; stuttering episodes may recur and/or develop into more prolonged episodes.

Priapism is common in sickle cell disease and as many as 90% of males with SCD will have experienced one or more episodes by the age of 20 years. Priapism in SCD is due to vaso-occlusion, which causes obstruction of the venous drainage of the penis, as opposed to high flow priapism which can be common in other conditions. Prolonged priapism is an emergency that requires urologic intervention.

Psychosocial and Counselling Aspects of Priapism

From early childhood, male patients need to know that priapism is a complication of SCD and that they should tell their parents or other appropriate adult if it occurs. Parents and children should be given the information leaflet on priapism in the first year of life and again aged 11-12 years. If untreated, priapism can result in erectile dysfunction. It often occurs at night and it can be triggered by a number of factors such as having a full bladder or sexual activity. Recurrence can sometimes be prevented by the use of medication.

Evaluation and Treatment

PROLONGED PRIAPISM IS AN EMERGENCY AND REQUIRES URGENT ASSESSMENT AND TREATMENT.

- Document the time of onset of the episode.
- Identify any precipitating factors, such as trauma, infections, or the use of drugs e.g., alcohol, psychotropic agents, sildenafil, testosterone, cocaine.
- A careful physical examination should reveal a hard penis with a soft glans. If there is a full bladder, encourage micturition and consider catheterisation.
- If time allows, Doppler ultrasound may be used to confirm ischaemic priapism

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

The patient should be offered prompt analgesia. They should be kept NBM and started on maintenance IV fluids, in preparation for urgent surgical intervention necessary if the priapism has persisted for more than 4 hours.

Patient Pathway for Priapism Lasting more than 1-2 hours

- Contact the Paediatric Urology team at Evelina Hospital urgently. The Paediatric Urology SpR can be contacted via the Evelina Hospital switchboard Monday-Friday, 9am – 5pm (020 7188 7188, bleep 1103). Outside these hours contact the paediatric surgical registrar (020 7188 7188 bleep 2505). The urology consultant can be contacted via switchboard.
- The patient will be admitted under the Paediatric Urology team
- The paediatric haematology team at Evelina Hospital should also be informed. They can be contacted via the Evelina Hospital switchboard Monday-Friday, 9am – 5pm (020 7188 7188 Bleep 0339). Outside of these hours contact the on call haematology SpR (020 7188 7188 Bleep 0294). The paediatric haematology consultant at the Evelina should also be made aware of patients transferred out of normal working hours.
- Queen Elizabeth Hospital and University Hospital Lewisham patients <13 years old are referred to the Evelina as above. Patients > 13 years old are seen by the urologists at Queen Elizabeth Hospital

- While waiting to transfer to Evelina Children's Hospital, the patient should be given intravenous fluids, adequate analgesia, and be kept nil-by-mouth.
- Surgical Intervention should not be delayed by either medical treatment nor the provision of blood products, except in case of repeated, major surgical procedure**.**
- Blood transfusion will not normally be necessary before any urgent urological surgical procedure, but may be appropriate in patients with a history of acute chest syndrome, or post-anaesthetic problems.
- The initial treatment is usually penile aspiration and irrigation with a α–adrenoceptor agonist. If this does not relieve the priapism, shunting procedures may be necessary. This plan is determined by the Urologist.
- If priapism recurs and further surgery required, an exchange blood transfusion may be necessary before the second anaesthetic, 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.
- If impotence persists for 12 months, the patient may be referred to the andrology team at Guy's for consideration of implantation of a semi-rigid penile prosthesis.
- Patients should be given a follow-up appointment with one of the paediatric urology consultants at Evelina Hospital, following discharge.

Future Prevention

Etilefrine is first line treatment. It is unlicensed, but available via pharmacies at KCH and ECH. We advise the following with respect to use of this treatment:

· Monitor BP after one week, and then 4 weekly as etilefrine can cause hypertension, tachycardia

· Caution in cerebrovascular disease

· Aim to assess response over a period of 4 weeks initially

· Aim to withdraw gradually if the episodes of priapism fully resolve

If further episodes of stuttering or fulminant priapism occur despite Etilefrine, alternative treatment such as hormonal agents may be considered, in discussion with the paediatric urologists (see below). These are non-formulary drugs and require a Chairman's action request with supporting evidence in order to prescribe them.

Pseudoephedrine can be considered as second line therapy if Etilefrine is not available or not tolerated.

If further episodes occur despite Etilefrine, other options include:

Leuprorelein acetate is a gonadotropin-releasing hormone analogue that suppresses the hypothalamic-testicular axis and the production of testosterone, used with some degree of success.

Stilboestrol could abort episodes of priapism and smaller doses can be used to prevent recurrence.

Long-term Management

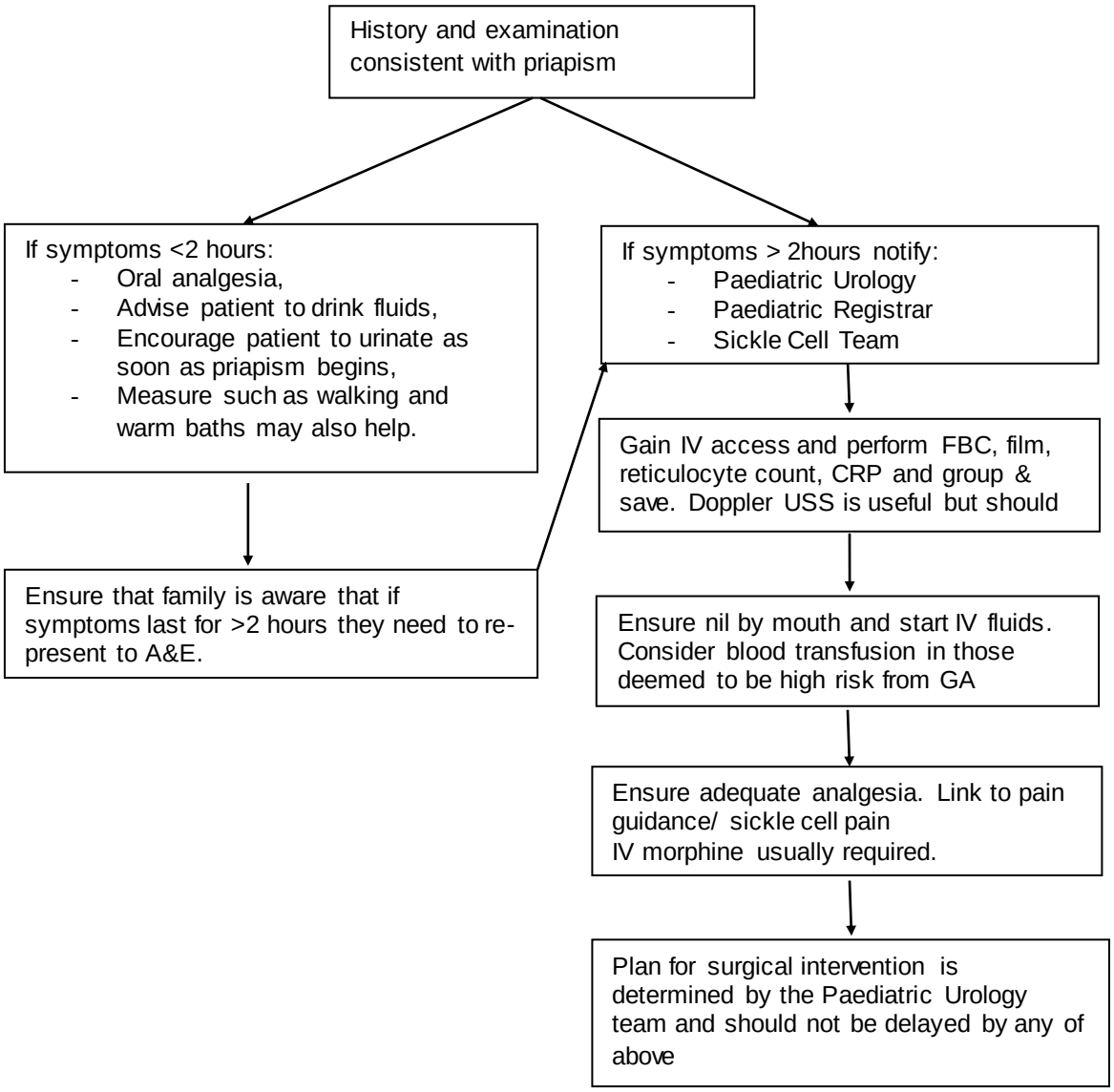
Patients should be referred to Paediatric urology
Consultant Paediatric Haematologist to discuss with Consultant Paediatric Urologist Arash Taghizadeh

Other information

Patient Information leaflet:

Priapism (painful erection) associated with sickle cell Disease

Acute Management Algorithm of Priapism



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Additional contacts can be found on the STSTN website (www.ststn.co.uk)

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