Priapism in Sickle Cell Disease

Reference: CG1466
Written by: Dr Emma Astwood
Peer reviewer Dr Jeanette Payne
Approved: October 2018
Approved by D&TC: 14th September 2018
Review Due: October 2021

Intended Audience
This document contains information and clinical guidelines for management of children with a diagnosis of sickle cell disease whose care is overseen by Sheffield Children’s NHS Foundation Trust. It is to be used by staff within the Trust for information whenever they are caring for these children.

Purpose
To provide clear guidance on the evaluation and treatment for Priapism for patients with Sickle Cell Disease who attend Sheffield Children’s NHS Foundation Trust.

Table of contents
1. Introduction
2. Psychosocial Implications
3. Evaluation and Treatment
4. Prevention of Subsequent Episodes
5. Summary of Management
6. References
1. Introduction

Priapism is a sustained, painful and unwanted erection. Priapism is a urological emergency and requires an immediate response.

Priapism is well known in sickle cell disease (SCD) and as many as 90% of males with SCD will have experienced one or more episodes of priapism by the age of 20 years.

Priapism in SCD is due to vaso-occlusion, which causes obstruction of the venous drainage of the penis.

Types of priapism

Major/fulminant
- Lasts for several hours
- Painful
- Can lead to impotence if untreated
- Most have a history of stuttering priapism
- Penile ischaemia occurs after 6 hours

Stuttering
- More frequent
- Less severe
- Lasts <1 hour
- Resolves on its own
- A cluster of stuttering attacks can lead to a major fulminant episode

2. Psychosocial Implications

In early childhood, boys 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.

It can be triggered by a number of factors such as a full bladder and sexual activity. Recurrence can be partially prevented by the use of medication.

Clearly this can be very upsetting and embarrassing for patients and they need to be treated with the utmost sensitivity, otherwise they will be unwilling to present appropriately early.
3. **Evaluation and Treatment**

Priapism is an emergency and requires urgent assessment and treatment. Document the time of onset of the episode.

Note precipitating factors such as trauma, infections, or the use of recreational or prescribed drugs (e.g., alcohol, psychotropic agents, sildenafil, testosterone, cocaine).

**The chances of response to medical and surgical treatment are highest when patients are managed promptly.**

Exchange transfusion (EBT) can be used in stuttering priapism that is increasing in frequency and severity. EBT usually does not resolve established fulminant priapism.

A careful physical examination should reveal a hard penis with a soft glans. The aim of therapy is to relieve pain, abort the erection and preserve future erectile function. Patients in the early stages of an episode can try plentiful fluids, a warm bath and analgesia. Attempting to urinate as soon as priapism begins can be helpful.

**Immediate management on presentation**

The patient should be referred directly to the Paediatric Urology Team **URGENTLY**.

Whilst making the referral, 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. **Discuss with a Consultant Haematologist.**

**Surgical Management**

Initial management is penile aspiration and irrigation with an α adrenoreceptor agonist. This is usually Phenylephrine (Phenylephrine should be diluted in sodium chloride 0.9% to produce a concentration of 100–500 micrograms/ml. Then, injections of 1ml aliquots should be performed intracavernously every 3–5 min for up to 1 hour or up to a dose escalation of 1000 micrograms of diluted phenylephrine [Montague et al. 2003]).

If this does not relieve the priapism shunting procedures may be necessary, to be determined by the urology team.

If priapism recurs and further surgery is required, an exchange blood transfusion may be performed 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.

In older patients if impotence persists for 12 months, the patient may wish to consider implantation of a semi rigid penile prosthesis. Patients should be given a follow-up appointment in the paediatric urology clinic following discharge.
4. Prevention of Future Episodes

Etilefrine 0.5mg/kg orally (30-100mg/day in adults) in 2 divided doses per day is the treatment of choice (available in 5mg tablets or 25mg modified release capsules).

Oral diethylstilbestrol in doses of 5mg daily for 3 to 4 days have been used to abort episodes of priapism in adults and smaller doses can be used to prevent recurrence. Gynecomastia is reported as a side effect with prolonged treatment. This should resolve 4 to 5 weeks after treatment with stilbestrol.

This is a rare situation in paediatric practice. These drugs are unlicensed for this indication. Their use would be a consultant decision on a named patient basis only and would require the drug to be ordered into pharmacy (not kept as stock).

5. Summary of Patient Pathway

- Patient presenting with priapism.

  - Contact paediatric urology team URGENTLY.
  - Commence IV fluids and IV analgesia
  - Ensure normal oxygenation
  - PATIENT MUST BE KEPT “NIL BY MOUTH”

  - FBC, U&E, G&S samples to labs for URGENT processing

  - Blood product administration should not delay initial surgery for priapism episode. (Exchange transfusion should be considered before repeat procedures)
6. References

Micromedex drug database

Martindale- The complete pharmacopeia (on line)


King’s College Hospital Joint Paediatric and Urology Clinical Guidelines for the management of Children with Priapism and Sickle Cell Disease April 2010