

Management of Children with Priapism and Sickle Cell Disease: Joint Paediatric and Urology Clinical Guidelines

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Consultation distribution (before ratification)

Sent to	Version	Date	Actions taken as a result
Mr Arash Taghazideh, Consultant Paediatric Urologist ECH	3	Dec 2013	Changes in contact details.
Mrs Kalpana Patil Consultant Paediatric Urologist ECH	3	Dec 2013	Agrees with new version
Dr Baba Inusa Consultant Paediatrican ECH	3	Dec 2013	No changes to current version

Reviews and updates (including CGG comments)

Date	New version no.	Summary of Changes	Author of change/s

Dissemination schedule (after ratification)

Target audience(s)	Method	Person responsible
KCH paediatric staff	Guidelines on Cliniweb	KCH Guidelines
Paediatric Teams in South Thames	South Thames Sickle and Thalassaemia Network – on website	Sue Height and Administrator

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Abstract Page

This guideline is relevant to all children in King's College Hospital with Sickle Cell Disease and priapism. The guideline applies to all patients with sickle cell disease who are currently under the care of the Paediatric Haematology team. It is mainly aimed at being a tool for the medical team managing these patients, but any member of the multidisciplinary team may find it useful.

Background

Priapism is a sustained, painful, and unwanted erection. It is classified into a) prolonged if it lasts more than three hours or b) stuttering if it lasts for more than a few minutes but less than three 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. Prolonged priapism is an emergency that requires urologic intervention.

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Definition/Background

Priapism is a sustained, painful, and unwanted erection. It is classified into

- a) prolonged, if it lasts more than three hours or
- b) stuttering, if it lasts for more than a few minutes but less than three 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. Prolonged priapism is an emergency that requires urologic intervention.

Psychosocial and Counselling Aspects of Priapism

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

Pre – Requisites for Practitioner

This guideline is intended for paediatricians caring for children with Sickle Cell Disease (SCD). No specific equipment is required. 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.

Guideline steps

Evaluation and Treatment

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 eg, alcohol, psychotropic agents, sildenafil, testosterone, cocaine.
- 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.

Episodes lasting 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. However, if the episode is not resolving despite these measures and lasting more than 2 hours, the patient must be kept NBM (see below)

Episodes lasting more than 2 hours

- Patient should be referred urgently to the Paediatric Urology Team at Evelina Children's Hospital using the direct number (020 7188 4610).
- While waiting to transfer to Evelina Children's 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 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 an α -adrenoreceptor 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 consultant's at Evelina Hospital, following discharge.

Future Prevention

Etilefrine is first-line treatment and usually effective.

Dose: <2 years: 1-2.5mg orally 3 times a day
2-6 years: 2.5-5 mg orally 3 times a day
>6 years: 5-10mg orally 3 times a day

If further episodes occur despite Etilefrine, other options include:

Leuprorelin acetate is a gonadotropin-releasing hormone analogue that suppresses the hypothalamic-testicular axis and the production of testosterone, used with some degree of success. Dose 3.75 mg every four weeks; The BNF states that the 3.75mg dose is sometimes halved in children weighing less than 20 kg.

Stilboestrol could abort episodes of priapism and smaller doses can be used to prevent recurrence. Dose 5 mg daily for 3 to 4 days, lower dose for prophylaxis.

Patient Pathway for Priapism Lasting more than 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). Also, within these hours; the on-call Paediatric Urology consultant can be contacted through the secretary (020 7188 4610). Outside these hours contact the paediatric surgical registrar (0207188 7188 bleep 2505).
- The patient will be admitted under the Paediatric Urology team, and will also be assessed by the paediatric registrar at Evelina Hospital (020 7188 7188 Bleep 0339), who will contact Dr Baba Inusa, consultant paediatrician, through St Thomas's switchboard (020 7188 7774), or Dr Sue Height or Professor David Rees through King's College Hospital switchboard if he is not available.
- The patient should be kept NBM and started on maintenance IV fluids and analgesia with the aim of urgent surgical intervention if the priapism has persisted for more than 3 hours. Surgical Intervention should not be delayed by either medical treatment or the provision of blood products, except in case of repeated, major surgical procedure.

Other information

Related guidelines (inc patient information)

Patient information leaflet - priapism

References

How I manage priapism due to sickle cell disease Olujohungbe A, Burnett A. British Journal of Haematology 2013 160; 754-765