Priapism in sickle cell disease

Introduction

Priapism **is a urological emergency.** It is defined as a persistent, prolonged, and painful erection and is one of the vaso-occlusive complications of sickle cell disease (SCD). The prevalence of severe priapism in SCD is unknown, hospital admission data suggest a rate of 25%, but a recent survey in young males suggested that 89% will experience priapism by 20 years of age.

Patient education and provision of written information should be part of the new patient and annual review processes. Patients should be made aware of the importance of contacting their local Haematology service or attending the emergency department urgently if their priapism does not settle within 2 hours.

Priapism can be acute/fulminant or stuttering:

Acute / fulminant priapism

- Severe pain
- Duration >4 hours
- Penis fully erect
- High risk of cavernosal fibrosis and impotence
- Urgent intervention indicated

Stuttering priapism

- Recurrent
- Pain of variable intensity
- Lasting 30 minutes to < 4hours
- Penis may not be fully erect
- Risk of subsequent fulminant attack

The optimal management of priapism is still a subject of debate however all management should be led by a Urologist with an interest in sickle cell disease.

Aims of Treatment

- Achieve rapid relief from pain and discomfort
- Preserve potency
- Prevent recurrence

Outcome is dependent on the pubertal status of the patient and length of time to detumescence. Poor long-term outcome in terms of impotence is associated with post pubertal males and a long duration of erection. The likelihood of responding to intervention is also related to duration of erection with most procedures being most effective within the first 6 hours and relatively ineffective after 24-48 hours.

Hence **priapism is a urological emergency** requiring rapid assessment and treatment to prevent irreversible ischaemic penile injury, corporal fibrosis and impotence.

Management

At home: early stages

Patient advice

- Attempt to urinate
- Try a warm bath/shower
- Hydration
- Analgesia
- Gentle exercise – such a pacing the room or walking up and down the stairs
- Attempt ejaculation

Acute/Fulminant Priapism

- **Ensure Haematology team are notified of admission immediately**
- **Ensure urgent urology review – this is a urological emergency** (if no urology service available, see below)
- Encourage patient to urinate (bladder catheterisation should only be performed if this is unsuccessful and the bladder is full)
- Intravenous hyper-hydration (1.5-2 x normal maintenance fluids) and analgesia (morphine is usually needed), ensure fluid balance and weight is closely monitored
- Sedation with a benzodiazepine may be useful in some cases, follow the local Trust guidance (take care with concomitant opiate use)
- Blood tests should include FBC, U+E and cross-match. Refer to SCD crisis protocol (link here).
- Treatment of choice is aspiration and irrigation, which should be performed within 4-6 hours of onset, by a urologist. **Facilitation of safe general anaesthesia may require full/partial exchange transfusion but should not delay definitive surgical treatment.** (See appropriate protocols on NSSG link here)

Acute Referral

- Local haematology services should develop links with their local Urology service for immediate management of acute/fulminant cases
- If there is no local service, links should be established with a nearby hospital, the OUH team or refer acutely to Urology at St Mary's Hospital, London (020 3311 3311)
- Reading patients: Refer to Urology at Royal Berkshire Hospital (Dr Shafi Wardak 0118 322 8629).

Discharge and follow up

- If aspiration is successful then the patient should be observed for a few hours and then, if well, discharged home by the Urologists with both Urology and Haematology follow up.
- If the patient has had previous episodes of priapism then medium to long term management should be discussed in Urology.

Management of Stuttering Priapism

Initial treatment at home should include:

- Increase fluid intake
- Oral analgesia
- Gentle exercise
- Attempts to urinate soon after onset

If the episode lasts more than 2 hours then the patient should be advised to attend hospital at once and acute priapism protocol should be followed.

Long term management and prevention

- Consider referral to specialist SCD Urology service at UCLH for patients with stuttering priapism or after admission with acute priapism (020 3447 9393)
- Reading patients: consider referral to Urology at Royal Berkshire Hospital (Dr Shafi Wardak 0118 322 8629)
- Hydroxycarbamide should be offered, if not already prescribed
- Haematology options may include Exchange transfusion programme, Hydroxycarbamide and Voxelotor.

References

Standards for the Clinical Care of Adults with Sickle Cell Disease in the UK (2018)

Link documents

S49 Priapism: patient information leaflet.

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Review

Name	Revision	Date	Version	Review date
Dr Wale Atoyebi	Pre-peer review	Jan 2013	1.0	Jan 2015
Dr Deborah Hay	Routine review	Aug 2105	1.2	Jan 2017
Dr Asif Khan Sandy Hayes, ANP	Removal of details on aspiration as this should only be carried out by a consultant Urologist. Patient education.	October 2016	2.0	October 2018
Dr Magbor Akanni Sandy Hayes	Pathway update	December 2019	3.0	December 2021
Dr Wale Atoyebi	Long term management and prevention – inclusion of Voxelotor. Logo update.	August 2021	3.1	August 2023
Manuela Sultanova	Acute referral and long-term management section: Referral to	January 2022	3.2	August 2023

	Royal Berkshire Hospital has been included			
Dr Wale Atoyebi Lesley McCarthy Faith Ehigie	Full review	May 2024	4.0	May 2026