

Paediatric Sickle Cell Service

Priapism and Sickle Cell Disease in Children

Information for parents, relatives and carers

Introduction

This leaflet has been provided to help answer some of the questions you may have about **priapism and sickle cell disease in children**. It is not meant to replace the consultation between you and your healthcare team, but aims to help you understand more about what you discussed together.

What is priapism?

Priapism is a prolonged and painful erection of the penis. It is normal for the penis to become erect for short periods of time in boys, of all ages, although this should not normally be painful or last more than 30 minutes.

What causes priapism?

It happens when sickled red blood cells block the flow of blood in the penis. This trapped blood causes an unusual hard erection. Because of lack of oxygen to the penile tissue, damage may then occur, which causes pain and, if prolonged and *untreated*, priapism may cause scarring of penile tissue, which can lead to loss of sexual function.

Priapism is mainly seen in teenagers and adults. It is less common in younger children but can still occur at almost any age. It is, however rare in children under the age of two years. Priapism occurs in all forms of sickle cell disease, but is most common in HbSS disease. Most episodes occur for no obvious reason however it can occur as part of a more general episode of acute pain.

Types of priapism

- **Stuttering priapism:** Episodes of reoccurring priapism, which lasts a few minutes to 2 hours, and can sometimes be painful.
- **Acute priapism:** A painful prolonged erection lasting for more than 2 hours. It can sometimes last from 6-10 hours or many days if left untreated.

What should I do if priapism occurs?

This is very embarrassing for children and teenagers but **it is vital that they tell someone..** The importance of telling a parent or their nurse, doctor or any responsible adult must be stressed from a young age. To help children/adolescents/ parents talk about priapism you can always use this leaflet to show someone. **Patients with priapism must attend hospital if the symptoms have not disappeared after 2 hours.**

Is there anything I can do to help?

- Treat priapism as a sickle cell crisis and drink extra fluids.
- Walking around or gentle exercise.
- Having a warm shower or bath
- Keeping warm, improves your circulation.
- Passing urine- this is sometimes easiest done in a warm bath
- Do not apply anything cold to the penis. This will make it worse as cold causes the blood vessels to constrict further.

Will I need medication?

If short episodes of priapism occur frequently, medication may help to stop further problems occurring. Your Doctor will discuss this with you, including side effects and benefits of treatment.

What happens if I do not get treatment?

Treatment and advice must be sought when priapism lasts for 2 hours as this will reduce the risk of later developing loss of normal sexual function. You must attend your nearest hospital.

Treatment will normally be analgesia for the pain and intravenous fluids to help with the sickling. In some cases surgery will be needed which involves a small operation under general anaesthetic to wash the sickled blood out of the penis. This surgery can only be done in specialist centres.

Whom can I contact for more information?

Clinical nurse specialist, Monday to Friday 9-5pm on 07795651153

Paediatric Haematology Registrar Monday to Friday 9-8pm and Saturday/Sunday 9-5pm via switchboard on 020 3313 6666

You can talk to your consultant about this problem during outpatient visits.

Further information and support

Sickle Cell Society, Tel: 020 8961 7795. Email: info@sicklecellsociety.org. Website: www.sicklecell society.org

How can I make a comment about my treatment?

We aim to provide the best possible service and staff will be happy to answer any questions you may have. However, if your experience of our services does not meet your expectations and you would like to speak to someone other than staff caring for you, please contact the patient advice and liaison service (PALS) on **020 3313 3322** or **020 3133 0088** (Charing Cross and Hammersmith hospitals), or **020 3312 7777** (St Mary's Hospital). You can also email PALS at pals@imperial.nhs.uk. The PALS team are able to listen to your concerns, suggestions or queries and are often able to help sort out problems on behalf of patients.

Alternatively, you may wish to express your concerns in writing to:

The chief executive
Imperial College Healthcare NHS Trust
Trust Headquarters
The Bays, South Wharf Road
London W2 1NY

Alternative formats

This information can be provided on request in large print, as a sound recording, in Braille, or in alternative languages. Please contact the communications team on **020 3312 5592**.