

Priapism in sickle cell disease

Information for patients, relatives and carers

Introduction

Priapism is the medical term for an erection that lasts a very long time. It is not always related to sexual intercourse or stimulation. It is often painful. If left untreated priapism can cause lasting damage to the penis. At its worst, it may stop you being able to have normal erection (impotence).

Priapism is common in boys and men who have sickle cell disease (SCD). This information leaflet will help you recognise it and explain what you can do about it.

During a normal erection blood flows into the penis and it becomes rigid. After ejaculation hormones are released which allow the blood to drain. If sickled red blood cells cause a blockage, the blood cannot drain, and the penis stays erect.

Normally, an erection:

- lasts for no more than 30 minutes
- is not painful

In priapism, an erection:

- can last for hours or even days
- is often painful
- the shaft of the penis is always rigid, but the head may sometimes be soft and tender if touched.

Sickle cell disease can cause two main types of priapism:

- **Recurrent acute or 'stuttering' priapism.** This is an erection that last for up to an hour and happens on several occasions. This may get better on its own or you may need the measures described below. It should not require further medical treatment.
- **Acute prolonged or 'fulminant' priapism.** This is painful erection which lasts for more than an hour. You need urgent medical assistance. Delaying treatment puts you at risk of permanent damage. This includes not being able to get an erection (impotence).

Who does priapism affect?

Priapism is most common in teenagers and adults – this leaflet is for you – though it can develop from an early age. Studies suggest up to half of men with SCD have an episode at some point during their life.

It occurs in all forms of SCD. It is more common in sickle cell anaemia (HbSS).

What to do if priapism happens

Though it can be embarrassing to talk about priapism, it is very important to tell the team caring for you. They can give the right advice and help you avoid long-term problems.

If you have an erection that is painful or lasts for longer than 30 minutes, do these things immediately:

- Drink lots of fluids
- Take pain killers such as paracetamol if you need them.
- Walk around or take gentle exercise (running on the spot can help).
- Have a warm shower or bath.
- Pass urine often to empty your bladder.
- Keep the area warm – DO NOT apply anything cold as this may make matters worse.
- Try to ejaculate.

If you have done these things and your penis remains erect after **TWO HOURS**, go to A&E **IMMEDIATELY**.

How we treat priapism in hospital

If come to the hospital with priapism, you will be cared for jointly by the haematology and urology teams. You will need to stay in hospital until the episode is completely over.

You may get the following treatment:

- Painkillers. You may need stronger painkillers like morphine or diamorphine if simple ones are not working. (It will follow your individual protocol.)
- Intravenous fluids. Pseudoephedrine tablet. This drug constricts the blood vessels, which helps to squeeze blood out of the penis. It also stops more blood from being pumped in.
- Draining some blood from the penis (aspiration). This is done with a small needle and syringe by a urology doctor. A local anaesthetic will be used to numb the penis. This usually helps to relieve the pain. Blood may be washed out with sterile salt solution at the same time. Sometimes more than one treatment may be needed.

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- An injection into the penis. This is often done at the same time as aspiration to help the erection go away. This works like pseudoephedrine but is more powerful.
 - Blood transfusion. A red blood cell exchange may help.
 - An operation. This may be considered if other measures fail. It will be carried out by a urology specialist. You may be transferred to a different hospital. There are several different types of operation. These involve implanting a small device called a shunt into the penis to reroute the flow of blood out of the penis. Some men who have surgery for priapism find it difficult to have an erection afterwards. It is difficult to estimate the exact risk of this because priapism itself can cause the same problem.

The team caring for you will talk to you about these treatments and procedures.

Treatment to prevent priapism

If you have had priapism more than once, you may be advised to have a treatment try to stop it happening again. The team caring for you will explain the options. They include taking a medication such as pseudoephedrine, daily or as soon as an episode begins. If this does not work, a regular exchange transfusion programme to reduce the number of sickle cells in the blood may help.

Contact details for advice/further information

Clinical Nurse Specialist for Haemoglobinopathies (Adults)

Phone: 020 33138553

020 8383 1000 Bleep 9513

How do I make a comment about my visit?

We aim to provide the best possible service and staff will be happy to answer any of the questions you may have. If you have any **suggestions** or **comments** about your visit, please either speak to a member of staff or contact the patient advice and liaison service (**PALS**) on **020 3313 0088** (Charing Cross, Hammersmith and Queen Charlotte's & Chelsea hospitals), or **020 3312 7777** (St Mary's and Western Eye hospitals). You can also email PALS at imperial.pals@nhs.net The PALS team will listen to your concerns, suggestions or queries and is often able to help solve problems on your behalf.

Alternatively, you may wish to complain by contacting our complaints department:

Complaints department, fourth floor, Salton House, St Mary's Hospital, Praed Street
London W2 1NY

Email: ICHC-tr.Complaints@nhs.net

Telephone: **020 3312 1337 / 1349**

Alternative formats

This leaflet can be provided on request in large print or easy read, as a sound recording, in Braille or in alternative languages. Please email the communications team:
imperial.communications@nhs.net

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