

Priapism in sickle cell disease

Information for patients, relatives and carers

Introduction

Priapism is the medical term for a prolonged erection. It is not always related to sexual intercourse or stimulation and is often painful. If left untreated priapism can cause lasting damage to the penis which at worst may result in loss of the ability to have a normal erection (impotence).

Priapism is common in boys and men who have sickle cell disease. This information leaflet will help you how to 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 there is a blockage caused by sickled red blood cells this does not happen and the penis stays erect. Normally an erection lasts for no more than 30 minutes and is not painful. In priapism an erection can last for hours or even days. The shaft of the penis is always rigid but the head may sometimes be soft and tender if touched.

Two main types of priapism can occur in sickle cell disease:

- recurrent acute or 'stuttering' priapism: erections that last for up to an hour and happen on several occasions. These may get better on their own or with the measures described below without needing medical treatment
- Acute prolonged or 'fulminant' priapism: a painful erection which lasts for more than an hour. This requires urgent medical assistance. Delayed treatment carries a risk of permanent damage, including impotence

Who does priapism affect?

Priapism can develop from an early age but is most common in teenagers and adults (for whom this leaflet is intended). It occurs in all forms of sickle cell disease though is more common in sickle cell anaemia (HbSS). Studies suggest that up to 50 per cent of males suffer an episode at some point during their life.

Who does priapism affect?

It can be embarrassing to talk about priapism but it is very important to report the problem to the team caring for you so appropriate advice can be given and long-term consequences avoided.

If you have an erection that is painful or lasts for longer than 30 minutes do the following immediately:

- drink lots of fluids
- take painkillers such as paracetamol, if needed

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- walk around or take gentle exercise (running on the spot can help)
 - have a warm shower or bath
 - pass urine frequently to empty the bladder
 - keep the area warm – **do not** apply anything cold as this may make matters worse
 - try to ejaculate

If, despite the above measures, the penis remains erect after **two hours** go A&E **immediately**.

How is priapism treated in hospital?

If you have to attend hospital for priapism you will be cared for jointly by the haematology and urology teams and will need to stay in hospital until the episode has completely resolved. The treatment you receive may include:

- painkillers - if simple pain killers don't work you may need stronger ones like morphine/diamorphine or as per your individual protocol
- intravenous fluids
- pseudoephedrine - a tablet which constricts the blood vessels. This helps to squeeze blood out of the penis while also preventing 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 after giving a local anaesthetic 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
- an injection into the penis – this is often done at the same time as aspiration to help the erection subside. This works like pseudoephedrine but has a more potent effect
- blood transfusion – a red blood cell exchange may be of benefit
- an operation - this may be considered if other measures fail and is carried out by a urology specialist. This may involve transfer to a different hospital. There are several different types of operation. These involve implanting a small device called a shunt into the penis to re-route 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.

These treatments and procedures will be discussed fully with you.

How is priapism treated in hospital?

If you have had more than one episode of priapism treatment may be advised to try to prevent this. The team caring for you will explain the options. They include taking a medication such as pseudoephedrine either on a daily basis 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): 020 8383 8372 or 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

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imperial.communications@nhs.net

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