

Priapism (painful erection) associated with sickle cell disease

This leaflet explains about priapism associated with sickle cell disease. If you have any further questions or concerns, please speak to a doctor or nurse caring for you.

What is a priapism?

Priapism is an uncomfortable, unwanted erection of the penis that persists despite a lack of sexual desire or stimulation. It is frequently painful and distressing. This is not associated with normal sexual function or desire.

The majority of males with sickle cell anaemia (HbSS) are likely to experience a priapism in their life time. Priapism is less common in males with sickle cell disease (HbSC) and sickle beta thalassaemia (HbSB).

There are two common types of priapism:

- stuttering
- acute (or fulminant).

What is a stuttering priapism?

The most common type of priapism is a stuttering priapism. It is called stuttering because it is usually transient but keeps reoccurring. Stuttering priapism attacks are self-limiting and get better by themselves, but may spontaneously reoccur again after a short period of time. They typically happen at night.

Some episodes of stuttering priapisms are not painful, but the longer the stuttering priapism lasts, the more painful the erections become. Stuttering priapisms may go on to develop in to a more severe attack of acute (fulminant) priapism.

What is an acute priapism?

This is a sustained painful erection lasting more than a few hours. This type of priapism will often not resolve spontaneously. Acute priapism is normally very painful and is considered a major complication of sickle cell disease. It needs emergency medical attention. Patients often experience episodes of stuttering priapism before an attack of acute priapism.

Why does priapism happen?

Sickled red blood cells block the supply and flow of blood in the penis. A priapism attack is a sickle cell crisis that is localised to the penis. The sickled red blood cells cause a disruption to the normal flow of blood within the penis, causing the blood to get trapped and keeping the penis in the erect state. The trapped blood in the penis becomes starved of oxygen and this causes pain and tissue damage the longer it goes on. Normal night-time and early morning erections are a common trigger for priapism in patients with sickle cell disease.

What are the risks from having priapism?

The tissues within the penis can become scarred and damaged because of frequent episodes of stuttering priapism, or, more commonly, following a prolonged episode of acute priapism (lasting four hours or more). These changes can become permanent the longer the priapism goes on. Scarring and damage to the tissue can then disrupt the normal flow of blood into the penis and this may result in poor quality erections or no erections when you want one (erectile dysfunction).

At what age am I likely to experience priapism?

Most patients report they first experience a priapism during early puberty (adolescence) and may continue still experience priapism into their 30s and 40s.

What time of the day or night am I likely to experience priapism?

Most priapism attacks occur during the night and in the early hours of the morning and are associated with the body's normal early morning erections. Sleep is often repeatedly interrupted causing tiredness, and this can significantly interfere with studies and work. If you are losing sleep because of priapism, discuss this with your haematologist or specialist nurse as they may be able to recommend treatments and helpful strategies.

Why did I not know about priapism?

Knowledge of this severe complication of sickle cell disease is often very poor. Many patients are not aware that priapism is a complication of sickle cell disease, or that there are risks associated with episodes of priapism. Patients are often reluctant to talk about priapism because of shyness and embarrassment.

What can be the consequences of priapism?

Some patients who have had stuttering and acute priapism report difficulties achieving normal erections when they want one. Erectile dysfunction is more likely to occur if priapism episodes are not treated promptly. Erectile dysfunction can cause severe psychological distress and affect relationships. Patients might hide the extent of the problem – particularly from nurses, doctors, partners and family members – because of embarrassment.

If you feel that you have developed erectile dysfunction, it is important that you are checked by an urologist who will be able to offer a number of treatment options. Your GP or haematologist can refer you to an urologist. Urologists often do regional clinics jointly with haematologists to manage both your sickle cell disease and its effect on your penile function. It is not advisable to try to treat erectile dysfunction yourself, as some treatments (including those that you can buy from the internet) may be harmful.

What can trigger a priapism?

There are a number of factors that can act as a trigger for priapism:

- Priapism is commonly triggered by the same factors that cause other types of sickle cell crisis and these include stress, infection, low oxygen levels, dehydration, alcohol, physical activity, and hot and cold environments.
- Early morning erections are an important trigger for sickle cell priapism. Early morning erections are normal and are associated with having a full bladder. These normal sustained erections normally resolve when you pass urine. However in sickle cell patients, early morning erections can develop into a priapism episode, particularly if the patients fail to resolve them normally by passing urine.
- Patients with a low oxygen level when well, or a history of breathing problems while sleeping, have an increased risk of priapism.
- Alcohol and smoking can act as triggers for all types of sickle cell crisis including priapism.
- Patients report that priapism can develop during sex. Having sex can be physically demanding and this may trigger a sickle cell crisis including a priapism.
- Certain drugs and treatments can increase your risk of priapism. If you have sickle cell disease, it is important to discuss new medications or treatments with your haematologist.
- Applying ice or cold water to the priapism will worsen the problem. Remember that being cold will trigger all types of sickle cell crisis, so applying ice or cold water to the penis is likely to increase the risk of localised sickling.

How do patients manage their priapism?

Many patients have developed different strategies for dealing with their own priapism.

Helpful strategies for dealing with priapism include drinking water, taking painkillers, heat packs and the application of warm water to the penis. Exercise such as doing squats, jogging, cycling or running up and down stairs can resolve a priapism attack. Emptying the bladder during the night to avoid early morning erections has been reported as a beneficial preventative measure in some cases.

Patients often report that the normal painkillers they use to treat their sickle cell crisis are not as effective in controlling the pain of priapism. Before trying a strategy for managing your own priapism it might be helpful to discuss it with your sickle cell nurse or haematologist.

Here are some helpful tips

- Treat a priapism as you would a normal sickle cell crisis including drinking extra fluids and taking pain relief.
- Empty your bladder at night and immediately on waking to help you avoid early morning erections that might trigger your priapism.
- Try exercise strategies to resolve your priapism.
- Seek support to manage stress, for example psychological support via the SCD team, SCD support group.
- Avoid sexual activity during priapism. Remember priapism will not be resolved by sex or masturbation
- Avoid taking lots of painkillers to manage your priapism. Discuss other treatment options with your SCD team.
- Avoid alcohol/cannabis/tobacco/Viagra®, as these can potentially worsen or trigger priapism.

You must attend you're the Emergency Department (A&E) urgently if the priapism does not show signs of resolving within two hours.

Seeking help

Many sickle cell patients report that they would not seek medical attention for priapism because of embarrassment, and practical difficulties such as getting dressed and travelling to the hospital in the early hours with a severe priapism. Many patients are unaware of the treatments available and are fearful of about what might happen when they come to hospital and how doctors and nurses might react.

Many patients therefore seek information about priapism from family members and friends with sickle cell disease. However, many of these people may be unable to recommend suitable treatments or strategies due lack of up-to-date medical knowledge. Many patients find it difficult to discuss the issue with female health care professionals, partners and friends, and are anxious about how their sexual function might be affected in the future.

We've put together some helpful information that you will need to know if you are coming to the hospital with a priapism:

- Ask for privacy in the clinic, ward or Emergency Department if you wish to discuss your priapism.
- Talk about your priapism and treatment options with your GP, sickle doctors, specialist nurse or psychologists.
- Ask to be referred to your region's joint haematology/urology sickle cell priapism clinic.
- Discuss helpful strategies and treatments with your haematologist or specialist nurse.
- Ask for written information about priapism and treatments.
- Call an ambulance if you cannot travel normally to the hospital because of the pain of priapism.
- Ask your haematologist or nurse specialist to explain this complication to your family or partner if you feel this would be helpful.
- If you are stressed because of priapism and the impact on your life, ask to be referred to the sickle cell psychologist as they are used to helping people live with this problem.
- Trying to find reliable information online about priapism may be difficult and could result in confusion. It's best to discuss this health issue with your haematologist, urologist or specialist nurse

Are there any treatments to prevent priapisms?

There are a number of common medicines that treat or prevent priapism. Your haematologist or urologist will talk to you about what is the best treatment to manage your type of priapism. It is important to understand that painkillers are not a direct treatment for your priapism, but will help you to manage the pain associated with this complication.

During your review with the haematologist, the amount of painkillers you use and why will routinely be reviewed and if you or your haematologist has concerns about the amounts you are using, other treatments options for both the priapism and your sickle cell will be discussed. If you are experiencing priapism episodes, mention this to your haematology team in clinic as they may recommended that you should be reviewed in our specialist priapism clinic.

Why and when should I attend the Emergency Department with a priapism?

- If your priapism fails to respond to your normal treatment or strategies at home after two hours, you should go to the Emergency Department. Priapism which goes on for more than four hours is more likely to result in some long-term damage and erectile dysfunction.
- The longer you remain at home with your priapism, the more likely it is that damage and severe pain in the tissue of the penis will occur. This increases the chances of tissue damage in the penis and loss of normal sexual function.
- Wear baggy jogging bottoms or loose underclothing because tight garments might aggravate the pain caused by priapism. Loose clothing will help you travel and will help you to disguise the nature of the problem when in public places.
- You must call an ambulance or arrange transport if the pain is severe and the priapism is not responding to your normal treatments and strategies.
- Once you arrive at the Emergency Department, inform the medical or nursing staff of the nature of the problem so that they can contact the SCD team and urologist. If you feel shy or worried ask for privacy.
- Priapisms that require you to attend hospital will be evaluated by your haematologist and a urologist and they will recommend treatments. These can include oral medications that can directly treat priapism and strong painkillers.
- In very severe attacks of acute priapism, the urologist may want you admitted for treatment injections directly into the penis, or aspiration (removal) of the sickle cell blood from the penis.

Although some of these treatments sound unpleasant, your urologist will make you as comfortable as possible. It is important to understand that your urologist is trying to prevent more severe and lasting damage, which can result in erectile dysfunction, and to treat this severe emergency.

While you are in hospital with a priapism you may also receive the common treatments that are used to manage a sickle cell crisis and this may include intravenous fluids, oxygen and antibiotics. In severe cases of priapism, exchange blood transfusions may be recommended by your haematologist. Surgery may also be required if the priapism fails to respond to the simple aspirations and injections by the urologist.

The earlier you go to the Emergency Department, the more likely it is that you will be treated successfully, which will reduce the risk of needing any surgery to solve the problem, or the risk of developing permanent erectile dysfunction.

Important note: In cases of priapism, time is function. Long delays will result in more damage and more severe erectile dysfunction.

Pharmacy Medicines Helpline

If you have any questions or concerns about your medicines, please speak to the staff caring for you or call our helpline. **t:** 020 7188 8748, Monday to Friday, 9am to 5pm

For more info: The telephone numbers below are available Monday-Friday, 9am-5pm. Outside these hours, please contact your GP or go to your local Emergency Department (A&E).

Guy's and St Thomas' Hospital

Consultant Haematologist: Jo Howard / Rachel Kesse-Adu Telephone: 02071882741
Out of hours – Haematology SpR or on call consultant via switchboard
(02071887188)

Clinical Nurse Specialists: Neil Westerdale/Luhanga Musumadi / Tolu Adeosin
Telephone – 020 7188 7188 (switchboard) then bleep 1843

Kings College Hospital

Consultant Haematologist: Moji Awogbade / Sara Stuart-Smith Telephone:
02032999000 Out of hours – Haematology SpR or on call consultant via switchboard
(020 32999 000)

Clinical Nurse Specialists: Giselle Padmore-Payne and Fester Ike Telephone – 020
3299 4968

St George's Hospital

Consultant Haematologist: Elizabeth Rhodes and Julia Sikorska Telephone: 020
87250885 Out of hours Haematology SpR via Switchboard 0208 6721255

Clinical Nurse Specialists: Carol Rose Telephone – Switchboard 0208 6721255 or
07500 835735

Additional contacts can be found on the STSTN website (www.ststn.co.uk)

Your comments and concerns

For advice, support or to raise a concern, contact our Patient Advice and Liaison Service (PALS). To make a complaint, contact the complaints department.

t: 020 7188 8801 (PALS) **e:** pals@gstt.nhs.uk
t: 020 7188 3514 (complaints) **e:** complaints2@gstt.nhs.uk

Language and accessible support services

If you need an interpreter or information about your care in a different language or format, please get in touch. **t:** 020 7188 8815 **e:** languagesupport@gstt.nhs.uk

NHS 111

Offers medical help and advice from fully trained advisers supported by experienced nurses and paramedics. Available over the phone 24 hours a day. **t:** 111

NHS website

Online information and guidance on all aspects of health and healthcare, to help you take control of your health and wellbeing. **w:** www.nhs.uk

Get involved and have your say: become a member of the Trust

Members of Guy's and St Thomas' NHS Foundation Trust contribute to the organisation on a voluntary basis. We count on them for feedback, local knowledge and support. Membership is

free and it is up to you how much you get involved. To find out more, please get in touch.
t: 0800 731 0319 e: members@gstt.nhs.uk w: www.guysandstthomas.nhs.uk/membership

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A list of sources is available on request

Our values: Put patients first | Take pride in what we do | Respect others | Strive to be the best | Act with integrity