

# Priapism (painful erections) associated with sickle cell disease

**This leaflet explains about priapism associated with sickle cell disease. If you have any further questions or concerns, please do not hesitate to contact the doctor or nurse caring for you.**

## What is a priapism?

Priapism is an unusual erection of the penis that is frequently painful and distressing. These are 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 boys and men 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 keeps reoccurring. Stuttering priapism attacks usually get better by themselves, but spontaneously reoccur again in a few minutes or within couple of hours.

Some episodes of stuttering priapisms are not painful; however the longer the stuttering priapism lasts the more painful the erections become. Stuttering priapism may go on to develop into a more severe attack of acute 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. Therefore 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. Consequently the penis becomes starved of oxygen and this causes pain and tissue damage within the penis. Normal early morning

erections that men and boys experience are an important 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). Scarring and damage to the tissue within the penis can then disrupt the normal flow of blood into the penis and this may result in a loss of normal sexual function (erectile dysfunction).

### **At what age am I likely to experience priapism?**

The majority of patients report they first experience a priapism during early puberty (adolescents) but some patients 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 in the early morning and are associated with normal early morning erections. Sleep is repeatedly interrupted causing tiredness and this can significantly interfere with studies and work. If you are losing a lot of sleep because of priapism please 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 report that they are not aware that priapism is a complication of sickle cell disease. Sickle cell patients are therefore not fully aware of the 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 a normal sexual erection (erectile dysfunction). 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 evaluated by a specialist (urologist) who will be able to offer a number of treatment options. Your GP or haematologist can refer you to an urologist. It is not advisable that you attempt to treat erectile dysfunction yourself as some treatments (including those that you can buy from the internet) are 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 this includes 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 attack, particularly if the patients fail to resolve them normally by passing urine.
- Patients with a low oxygen levels when well or a history of breathing problems whilst sleeping have an increased risk of priapism.
- Alcohol and smoking can act as trigger 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. Consequently 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 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. Do not use ice or cold water applications to the priapism as this is potentially harmful. 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 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.
- Find ways to avoid stress, for example counselling, support groups.

- Avoid sexual activity during priapism. Remember priapism will not be resolved by sex or masturbation
- Avoid taking lots of painkillers to manage your priapism; find out about other treatment options.
- Avoid alcohol/cannabis/tobacco/Viagra® as these can potentially worsen or trigger priapism.

**You must attend your usual 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 brothers, mothers and other males with sickle cell disease. However many of these people may be unable to recommend suitable treatments or strategies due lack of knowledge. Sickle cell patients have found it difficult to discuss the issue with female health care professionals, partners and friends. Many patients are anxious about how their sexual function might be affected in the future.

Therefore, 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 A&E if you wish to discuss your priapism.
- Talk about your priapism and treatment options with your doctor (haematologist) or specialist nurse or psychologist.
- 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 about priapism in resources such as the internet may be difficult and could result in confusion. Therefore, 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.

If you find you are using too many painkillers to manage your priapism it would be advisable that you discuss this with your haematologist so that he/she can consider other treatment

options. If you are experiencing priapism episodes please mention this to your haematology team in clinic as it may be recommended that you should be reviewed in our specialist priapism clinic.

## Why and when should I attend A&E with a priapism?

- If your priapism fails to respond to your normal treatment or strategies at home after two hours you should go to A&E.
- The longer you remain at home with your priapism the more likely damage and severe pain in the tissue of the penis can occur. This increases the likelihood of tissue damage in the penis and loss of normal sexual function (erectile dysfunction )
- Wear baggy jogging bottoms or loose underclothing as 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/arrange transport if the pain is severe and the priapism is not responding to your normal treatments and strategies.
- Once you arrive at A&E, inform the medical or nursing staff as to the nature of the problem so that they can contact the sickle cell 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 (specialist) and they will recommend treatments which can include oral medications that can directly treat priapism and strong painkillers,
- In very severe acute attacks of priapism the urologist may advise you are admitted for injections of treatment 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 try to make you as comfortable as possible. It is important to understand that your urologist is trying to prevent erectile dysfunction and to treat this severe emergency.
- Whilst 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 priapisms exchange blood transfusions may be recommended by your haematologist if the priapism fails to respond to treatments recommended by the urologist
- The earlier you go to A&E, the more likely it is that you will be treated successfully, which will reduce the risk of erectile dysfunction.

## Useful contacts for the sickle cell team

Consultant haematologists – Dr Jo Howard and Dr R. Kesse-Adu, **t:**020 7188 2741

Consultant urologists – Dr Majed Shabbir and Dr Z Evangelos, **t:** 020 7188 0243

Sickle specialist registrar, **bleep** 0248

Sickle senior house officer (SHO), **bleep** 2283

Advanced nurse practitioner – Mr Neil Westerdale

**t:** 0207 188 2710 / 07770 683 947 **bleep** 1843.

Clinical nurse specialist – Mr Luhanga Musumadi

**t:** 020 7188 2710 / 07770 678 851 **bleep** 2256

Adolescent clinical nurse specialist – Mrs Judith St Hilaire

**t:** 020 7188 2710 / 07920 711 266 **bleep** 2868

Consultant health psychologist – Dr Nicky Thomas

**t:** 020 7188 2725

Clinical health psychologist – Dr Heather Rawle

**t:** 020 7188 2718

Health psychologist – Mina Abedian

**t:** 020 7188 2718

Haematology Clinic, **t:** 020 7188 2743 / 2724

Haematology Day Unit, **t:** 020 7188 2745 / 2710

Florence Ward (urology), **t:** 020 7188 813

For more information leaflets on conditions, procedures, treatments and services offered at our hospitals, please visit [www.guysandstthomas.nhs.uk/leaflets](http://www.guysandstthomas.nhs.uk/leaflets)

## 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 9am to 5pm, Monday to Friday

## Patient Advice and Liaison Service (PALS)

To make comments or raise concerns about the Trust's services, please contact PALS. Ask a member of staff to direct you to the PALS office or:

**e:** 020 7188 8801 at St Thomas' **t:** 020 7188 8803 at Guy's **e:** [pals@gstt.nhs.uk](mailto:pals@gstt.nhs.uk)

## Language Support Services

If you need an interpreter or information about your care in a different language or format, please get in touch using the following contact details.

**t:** 020 7188 8815 **fax:** 020 7188 5953

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