

# Priapism (painful erections) in sickle cell disease

## Information for patients, parents or carers

This leaflet explains priapism experienced by men and boys with sickle cell disease: why it happens, how to manage it and when to contact the hospital.

Often, patients and or their parents are too embarrassed to discuss this subject with their specialist doctors or nurses. We have written this information booklet to increase information and understanding of priapism and to encourage all patients and their families to discuss priapism with their specialist team.

***You should seek urgent medical attention if your priapism does not resolve within 2 hours or if stuttering priapism keeps recurring for 2 hours.***

### What is a priapism?

Priapism is an unusual erection of the penis that can be very painful and distressing. It is caused by blocking of the blood vessels in the penis by sickled red blood cells. Blood then gets trapped in compartments of the penis causing it to stay erect. The penis then becomes starved of oxygen, this causes pain and tissue damage. It can also occur before a sickle cell crisis.

These erections are not associated with sexual function or desire and often occur during sleep or the early morning. Priapism is more common in some types of sickle cell disease (HbSS) than others (HbSC, HbS-beta thalassaemia).

There are two common types of priapism:

- stuttering
- acute (or fulminant)

### What is stuttering priapism?

The most common type of priapism is stuttering priapism. It is called stuttering because it keeps reoccurring. Stuttering priapism usually gets better by itself, but spontaneously reoccurs again in a few minutes or within a couple of hours.

Some episodes of stuttering priapism 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.

## Adult and Paediatric Haemoglobinopathy Services

## What is acute priapism?

This is a sustained painful erection lasting more than two hours. This type of priapism will often not resolve by itself. Acute priapism is normally very painful and is considered a major complication of sickle cell disease. It needs emergency medical attention.

## Why does priapism happen?

Priapism is a sickle cell crisis that is localised to the penis. Sickled red blood cells block the supply and flow of blood in the penis. The penis then becomes starved of oxygen, this causes pain and tissue damage, and further sickling. Normal early morning erections that men and boys experience are an important trigger for priapism in patients with sickle cell disease.

## At what age does priapism occur?

Children as young as three have been reported as having a priapism and one third of all teenagers have their first episode of priapism by the age of fifteen. Some men still experience priapism into their 30s and 40s.

## What time of the day or night does priapism occur?

Most priapism attacks occur in the early morning and are associated with normal early morning erections. Sleep is repeatedly interrupted causing tiredness which 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.

## What can trigger priapism?

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

- The same triggers that cause other types of sickle cell crisis including: stress, infection, low oxygen levels, dehydration, alcohol, recreational drugs, excessive physical activity, and hot and cold environments.
- Early morning erections. Early morning erections are normal and are associated with having a full bladder. The erections usually resolve when urine is passed. Occasionally early morning erections can develop into priapism.
- Low oxygen levels in the blood when well or a history of breathing problems whilst sleeping have an increased risk.
- Alcohol, recreational drugs and smoking.
- Priapism can develop during sex. Sex can be physically demanding and this may trigger a sickle cell crisis including priapism.
- Certain drugs and treatments can increase the risk. Therefore, 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 can trigger a sickle cell crisis, so applying ice or cold water to the penis is likely to increase the risk of localised sickling in the penis making priapism worse.

## What are the risks of 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).

## What is erectile dysfunction?

Erectile dysfunction is a term used to describe difficulty obtaining or maintaining an erection during sexual activity. 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. You should not attempt to treat erectile dysfunction yourself as some treatments (including those that you can buy from the internet) are harmful.

## Can my fertility be affected by having priapism?

Priapism does not affect your body's ability to make sperm but as described above it can affect the ability to have an erection, which is called erectile dysfunction.

## Are there any treatments to prevent priapism?

There are a number of common medicines that treat or prevent priapism. Your haematologist or urologist will talk to you about 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, please discuss this with your haematologist so that other treatment options can be considered.

## How do people manage their priapism?

If you are experiencing priapism episodes please tell your haematology consultant or specialist nurse. *They are unable to help you if they do not know you are having problems with priapism.*

Helpful strategies for dealing with priapism include:

- Treat priapism as you would a normal sickle cell crisis including drinking extra fluids and taking pain relief. However, some men report that the normal pain killers they use to treat their sickle cell crisis are not effective in controlling priapism. If this is the case for you, please discuss with your haematology team.
- Heat packs and the application of warm water to the penis, such as a warm bath.
- Exercise such as doing squats, jogging, cycling or running up and down stairs.
- Emptying the bladder at night and immediately on waking to avoid early morning erections.
- Find ways to avoid stress, for example counselling, support groups.

### Adult and Paediatric Haemoglobinopathy Services

- Wear baggy jogging bottoms or loose underclothing, as tight garments might aggravate the pain caused by priapism.
- Before trying a strategy for managing your priapism it might be helpful to discuss it with your specialist nurse or haematologist.

### What does not help?

- Do not use ice or cold water applications to the priapism as this is potentially harmful.
- 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.

### Seeking help

**You must attend your usual A&E or contact your triage or ward service urgently if your priapism does not resolve within 2 hours, or if stuttering priapism keeps reoccurring for 2 hours.**

Men and boys with sickle cell disease report that they do not seek medical attention for priapism because of embarrassment and practical difficulties such as getting dressed and travelling to the hospital at night with severe priapism. Most men and boys are unaware of the treatments available and are fearful of what might happen when they come to hospital and how doctors and nurses might react. This leads people to 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 that would be offered by the medical team. Also, trying to find reliable information about priapism on 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.

### Privacy

Another common concern is discussing priapism with female health care professionals, partners and friends. If you would prefer to discuss this with a male member of the medical team we will make sure that we honour your wishes. You can ask to see a male doctor and will not be asked why you have made that request.

Below is some helpful information if you need to come to the hospital urgently or if you would like to discuss priapism in the outpatient clinic.

### Urgent care

- Call an ambulance if you cannot travel normally to the hospital because of the pain of priapism. Inform the emergency service and/or on arrival to A&E/ triage/ ward service that you have sickle cell disease and you are experiencing priapism which is a medical emergency. Bring this leaflet with you if you have it available.
- Wear loose clothing such as track suit pants to travel to the hospital. Loose clothing will help you travel and will help you to disguise the nature of the problem when in public places.
- Ask for privacy in the A&E/triage/ward.

**Adult and Paediatric Haemoglobinopathy Services**

- You will be seen by a haematologist and 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. Most importantly, having this treatment done rapidly will prevent permanent damage and reduce the chances of developing erectile dysfunction (inability to obtain or maintain an erection) in the future.
- Whilst you are in hospital with priapism you may also receive the common treatments that are used to manage a sickle cell crisis such as intravenous fluids, oxygen and antibiotics. In severe cases of priapism, exchange blood transfusions may be recommended by your haematologist if the priapism fails to respond to treatments recommended by the urologist.

**Remember:** *The longer you remain at home with priapism, the more you are at risk of damage in the tissue of the penis. This increases the likelihood of permanent tissue damage in the penis and loss of normal sexual function.*

**Outpatient clinic**

- Ask for privacy in the clinic. You can request to be seen by a male doctor alone.
- Ask about priapism and treatment options.
- Ask about helpful strategies and treatments.
- Ask for written information about priapism and treatments.
- Ask your haematologist or nurse specialist to explain this complication to your family or partner if you feel this would be helpful.

**Who can I contact with any queries and concerns?**

If you have any questions or concerns, please talk to your Haematology consultant or specialist nurse.

**Oxford**

<b>Paediatric haemoglobinopathy service</b>	Telephone: 01865 226 654
Dr Amrana Qureshi	<b>Specialist Nurse</b> Lesley McCarthy <a href="mailto:Lesley.mcarthy@ouh.nhs.uk">Lesley.mcarthy@ouh.nhs.uk</a>
<b>Adult haemoglobinopathy service</b>	Telephone: 01865 227 907
Dr Wale Atoyebi Dr Kieran Burton Dr Noemi Roy	<b>Specialist Nurse</b> Faith Ehigie <a href="mailto:Faith.Ehigie@ouh.nhs.uk">Faith.Ehigie@ouh.nhs.uk</a>

Adult and Paediatric Haemoglobinopathy Services

**Milton Keynes**

<b>Adult haemoglobinopathy service</b>	Telephone: 01908 660033 Ext 85834 or Bleep 2134
Dr Magbor Akanni	<b>Specialist Nurse</b> Sophie Clarke <a href="mailto:Sophie.clarke@mkuh.nhs.uk">Sophie.clarke@mkuh.nhs.uk</a>

**Southampton**

<b>Paediatric haemoglobinopathy service</b>	Telephone: 02381204567
Dr Nyree Cole Dr Michael Roe	<b>Specialist Nurse</b> <a href="mailto:haemoglobinopathiescns@uhs.nhs.uk">haemoglobinopathiescns@uhs.nhs.uk</a>
<b>Adult haemoglobinopathy service</b>	Telephone: 02381204567
Dr Srin Narayanan	<b>Specialist Nurse</b> <a href="mailto:haemoglobinopathiescns@uhs.nhs.uk">haemoglobinopathiescns@uhs.nhs.uk</a>

<b>Local haematology service</b>	Telephone:
(Insert details)	

**Contributors:** Sandy Hayes, ANP; Dr Amrana Qureshi; Dr Wale Atoyebi; Dr Magbor Akanni; Dr Noemi Roy; Dr Deborah Hay.

**Acknowledgements and references:**

University College London Hospitals (2015) Patient information: Priapism in patients with sickle cell disease.

St George's University Hospitals (2015) Patient information: Priapism and sickle cell disease (SCD).

Guy's and St Thomas Hospitals (2015) Patient information: Priapism (painful erections) associated with sickle cell disease.

Thames Valley Haematology Network: Haemoglobinopathy service (2016) Priapism in sickle cell disease: clinical guideline, S14, V.2.0.

National Heart, Lung, Blood Institute (2014) Evidence based management of sickle cell disease. USA.

Olujohungbe, A., Burnett, A. (2013). How I manage priapism due to sickle cell disease. BJH, 160, pp.754-765.