

Priapism (painful erection) associated with sickle cell disease

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

What is a priapism?

Priapism is an uncomfortable, unwanted erection of the penis. It is not caused by sexual desire, arousal or stimulation. It can often be painful and distressing.

It is common for individuals with sickle cell anaemia (HbSS) to have priapism in their lifetime. Priapism is less common in sickle cell disease (HbSC) and sickle beta thalassaemia (HbSB).

There are two common types of priapism: stuttering and acute (or fulminant).

What is stuttering priapism?

The most common type of priapism is called stuttering priapism. It is called 'stuttering' because the erection does not last long but keeps recurring. Stuttering priapism erections get better by themselves but may spontaneously happen again after a short period of time. They usually happen at night.

Some episodes of stuttering priapism are not painful, but the longer it goes on for, the more painful the erections become. Stuttering priapism may develop into a more severe attack of acute (fulminant) priapism.

What is acute priapism?

This is a painful erection lasting more than a few hours. This type of priapism will often not get better without medical help. Acute priapism is normally very painful and is considered a major complication of sickle cell disease. **It needs emergency medical attention. Call 999 or go to the emergency department (A&E) if the erection lasts more than 2 hours.**

Why does priapism happen?

Sickled (crescent-moon shaped) red blood cells block the supply and flow of blood in the penis. This traps the blood and keeps the penis erect. The trapped blood in the penis becomes starved of oxygen and this causes pain and tissue damage the longer it goes on.

What are the risks from having priapism?

The tissues in the penis can become scarred and damaged because of frequent episodes of stuttering priapism, or, more commonly, after an 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 one is desired (erectile dysfunction).

At what age does priapism first happen?

Most people report their first experience of priapism during early puberty (adolescence), but it can happen at any age. So, it is important to encourage your child to talk to you about it.

What time of the day or night does priapism happen?

Most priapism attacks happen during the night and in the early hours of the morning and are associated with the body's normal early morning erections. Priapism can often repeatedly interrupt sleep causing tiredness, and this can significantly interfere with studies or school.

If your child is losing sleep because of priapism, talk about this with your haematologist or specialist nurse as they may be able to recommend treatments and helpful strategies.

What can trigger priapism?

- Priapism is commonly triggered by the same factors that cause other types of sickle cell crisis. These include stress, infection, low oxygen levels, dehydration, alcohol, physical activity, and hot and cold environments.
- Early morning erections are normal and are associated with having a full bladder, and usually resolve when urine is passed. However, in people with sickle cell disease, early morning erections can develop into a priapism episode, particularly if they don't resolve on passing urine.
- People with a history of low oxygen saturations when well, or breathing problems while sleeping, have an increased risk of developing priapism.
- Alcohol and smoking can act as triggers for all types of sickle cell crisis including priapism.
- Certain drugs and treatments can increase the risk of priapism. It is important to discuss new medications or treatments with your haematologist or specialist nurse.

How can I help my child with their priapism attack?

- Your child should drink extra fluid.
- Your child should do light exercises, such as running up and down stairs or squatting.
- Give your child pain relief.
- Don't apply ice or cold water to the penis as it may make it worse.
- If the penis remains erect for more than **2 hours** go to the emergency department **immediately**.

How do I manage my child's priapism?

- Your child should empty their bladder at night and as soon as they wake up to help avoid early morning erections that might trigger priapism.
- If your child might be stressed, get support from the Sickle Cell team, or talk to sickle cell support groups.
- Don't use painkillers repeatedly. Talk about other treatment options with your Sickle Cell team.

Why and when should I attend the emergency department (A&E)?

If your child's penis is still erect after **2 hours**, you should go to the emergency department right away. Priapism that goes on for more than four hours is more likely to result in some long-term damage and erectile dysfunction as well as severe pain.

Your child should wear loose clothing because tight garments might make any pain worse.

Once you arrive at the emergency department, tell the medical or nursing staff about what's happening so that they can contact the sickle cell team and urologist. If you or your child feels shy or worried, just ask for privacy.

What is the emergency treatment for priapism?

If you go to hospital, your child will be evaluated by a haematologist and a urologist who will recommend treatments. These can include oral medications that can directly treat priapism and strong painkillers.

In very severe case of acute priapism, the urologist may treat with injections directly into the penis, or remove the sickle cell blood from the penis. This will be done under general anaesthesia, so your child will be asleep and won't feel anything.

While in hospital your child may also receive common treatments that are used to manage a sickle cell crisis. This may include intravenous fluids, oxygen and antibiotics.

In severe cases of priapism, exchange blood transfusions may be recommended by your haematologist. Your child may need surgery if the priapism doesn't get better with other treatments.

It is important to understand that your urologist is trying to prevent more severe and lasting damage, which can result in erectile dysfunction.

The earlier you go to the emergency department, the more likely it is that your child will be treated successfully. This will reduce the risk of needing any surgery to solve the problem, or the risk of developing permanent erectile dysfunction.

Meet our team

Lead consultant for children with haemoglobin disorders and consultant in paediatric haematology

t: 020 7188 6203 (secretary)

Lead nurse and general enquiries

t: 020 7188 9432 or 07918 338 730

e: HaemoglobinopathyCNS@gstt.nhs.uk

Nurse specialist

t: 07771 345 847 or 074 4329 0973

e: HaemoglobinopathyCNS@gstt.nhs.uk

Research nurse

t: 020 7188 6637 or 077 4893 2743

Specialist nurse – community

t: 020 3049 5993

Therapy and psychology team

t: 020 7188 9125 or 078 1790 5844

Contact us

If you can any questions or concerns about Priapism, please contact the sickle cell team (Monday to Friday, 9am to 5pm). Out of hours, please call the hospital switchboard on 0207 188 7188 and ask them to bleep the haematology registrar.

For more information leaflets on conditions, procedures, treatments and services offered at our hospitals, please visit **w:** www.evelinalondon.nhs.uk/leaflets

Evelina London Medicines Helpline

If you have any questions or concerns about your child's medicines, please speak to the staff caring for them or contact our helpline.

t: 020 7188 3003, Monday to Friday, 10am to 5pm

e: letstalkmedicines@gstt.nhs.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

Leaflet number: 4993/VER1

Date published: June 2020

Review date: June 2023

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A list of sources is available on request