



Priapism

(painful erections associated with sickle cell disease)

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What is priapism?

Most boys and healthy adult males have erections every day. It is common for boys and men to wake up with an erection. This type of erection is painless and fades away on its own.

Priapism is a long-standing painful erection of the penis without physical, mental or sexual stimulation. It happens when blood becomes trapped in the penis and is unable to drain away. Most males with **sickle cell anaemia (Hb SS)** are likely to have priapism. It is not common in patients with other forms of sickle cell disease and thalassaemia.

If you ignore priapism it may lead to future problems. For example, you may not be able to have normal erections.

What are the symptoms of priapism?

- erections lasting longer than 30 minutes
- rigid erection without sexual stimulation
- penile pain or sensitivity

Types of priapism

There are three types of priapism:

- intermittent (stuttering)
- low-flow (ischaemic or acute)
- high-flow (non-ischaemic)

(these terms are explained on page 2)

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What is intermittent priapism?

This is the most common type of priapism. It is called intermittent because it keeps happening. It usually gets better by itself but then happens again in a few minutes or within a few hours. Some episodes are not painful, but the longer it lasts, the more painful the erection becomes. Intermittent priapism can develop into low-flow priapism.

What is low-flow priapism?

This occurs when the vein running from the penis is blocked. This may lead to tissue damage. This type of priapism is a continued and painful erection that may last for a few hours. It does not get better on its own and will need emergency medical attention.

What is high-flow priapism?

High-flow priapism is rare and usually less painful. It usually happens after an injury to the penis or the area between the scrotum and anus (perineum). The injury prevents blood in the penis from circulating normally.

Why does priapism happen in patients with sickle cell disease?

Priapism is a sickle cell crisis within the penis. Sickled red cells block the supply and flow of blood in the penis. The sickled red blood cells cause disruption to the normal flow of blood in the penis. The penis is deprived of oxygen, causing pain and tissue damage within the penis.

What problems are caused by priapism?

The tissue within the penis can become scarred and damaged due to frequent episodes of intermittent priapism or long episodes of low-flow priapism (lasting for hours).

Damage and scarring to the tissue within the penis can disrupt the normal flow of blood in the penis and can lead to loss of normal sexual function (erectile dysfunction).

At what age is priapism likely to happen?

Most patients first experience priapism during early puberty (when a child's body begins to develop changes as they become an adult). Some patients experience priapism into their 30s or 40s.

What are the triggers of priapism?

Factors that can trigger priapism include:

- Alcohol and smoking
- Certain drugs and treatment. It is important to discuss new medications and treatments with your doctor if you have sickle cell disease
- Sexual intercourse. Some patients develop priapism during sex
- Normal early morning erections (usually together with a full bladder)
- Priapism often happens first thing in the morning

How can I prevent it?

The following advice may help you to prevent priapism:

- Do not drink alcohol or use drugs (other than those your doctor has prescribed)
- Drink plenty of water
- Keep warm during cold weather

Priapism may happen even if you follow the above advice. If you have had priapism, your doctor may ask you to take a medicine such as etilefrin regularly to prevent further problems.

How do I manage priapism?

Each person will find different ways to manage their priapism. Some tips include:

- Go to the toilet to empty your bladder just before you go to bed and as soon as you wake up to make early morning erections less likely
- Drink lots of fluids
- Take simple pain killers such as paracetamol
- Have a warm bath or shower and wear loose-fitting underwear
- Go for a gentle walk
- Try exercises, such as squats or running on the spot

Do not:

- apply ice packs or cold water to your penis as this can make things worse
- have sex or masturbate as it will not make your erection go away
- drink alcohol
- smoke
- use cannabis, tobacco or Viagra, as these can trigger priapism or make it worse.

These strategies should help to take the problem away within an hour. It is important that you report the episode to your haematology team as soon as possible. If your problem does not go away within **two hours**, you must go to the Emergency Department (A&E) urgently.

Hospital treatment for priapism

If you are admitted to hospital you may be given fluids intravenously (into your vein), oxygen and pain relief. The urology team may see you and recommend treatment to help reduce your erection, such as:

- Medicine such as etilefrin or pseudoephedrine, which are usually given by mouth or by injection into the penis
- Drainage of blood from the penis. This is done after the area has been numbed with local anaesthetic
- Surgery to drain the blood through a thin cut. This is done under general anaesthetic (whilst you are asleep)
- In some severe cases, where these treatments have not worked, the haematology team may recommend a red cell exchange

Remember:

Call 999 or go to A&E if you have an erection that lasts more than two hours.

An erection that lasts this long requires hospital treatment to avoid permanent damage to your penis.

Useful contacts

Sickle Cell and Thalassaemia Service

Ground Floor, Osborne Building

Leicester Royal Infirmary

We are always pleased to hear from patients and carers. If you have any suggestions about how the service is run please contact us.

Telephone number: 0116 258 6081 Monday to Friday 09.00 to 17.00

Haematology Helpline (Adult)

Telephone: 0808 178 2212

UHL Medicines Information

(Monday to Friday 8.30 am to 5.30 pm)

Telephone: 0116 258 6491

Sickle Cell Society

www.sicklecellsociety.org

Telephone: 020 8961 7795

Email: info@sicklecellsociety.org

UK Thalassaemia Society

www.ukts.org

Telephone: 020 8882 0011

Email: office@ukts.org

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