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The Royal Liverpool
and Broadgreen
University Hospitals
NHS Trust

Patient information

Sickle Cell Disorders - Priapism in Sickle Cell Disease

Haematology Department

What is it and who is affected?

Priapism can occur in males of all ages, including newborn babies and is common in sickle cell anaemia. Most boys over the age of about ten years and healthy adults will have at least three to four erections a day. Many will happen at night and it is normal for older boys and men to wake up with an erection. Normal erections are not painful and fade away as the person gets on with their normal life.

How does an erection happen?

The artery to the penis opens wider and blood rushes into the penis. Veins carrying blood away from the penis shut down. Blood then becomes trapped inside the penis, making it hard and erect. An erection subsides when the veins open up and allow blood to flow out of the penis.

How do I recognise the problem?

If the erection is painful or if it does not fade away after about 30 minutes this is called priapism. Priapism is very common in boys and men with sickle cell anaemia and needs to be treated seriously. If the problem is ignored it may lead to future problems: at worst the man may not be able to have normal erections.

Types of Priapism

There are two types of Priapism:

➤ **Priapism associated with low blood flow**

This occurs when the vein running from the penis is blocked, and results in a high risk of damage to the tissues.

➤ **Priapism associated with high blood flow.**

This occurs after injury to the penis, when the cavernosal artery (vessel leading to the penis) ruptures. This is very rare in sickle cell disease.

The erection can occur as repeated short episodes (known as stuttering Priapism), which can last from a few minutes to several hours. Or it can last for four hours or more (known as fulminant Priapism).

Priapism can be caused or made worse by **alcohol** and by the use of illegal drugs, including:

- Marijuana, Cocaine, and Ecstasy.

Can I do anything to prevent it?

- Reduce the use of any of the above named drugs.
- Keep well hydrated.
- Sometimes Priapism may happen despite these precautions.

What can I do about it?

Do the following things immediately:

- Drink lots of fluids.
- Some patients find jogging or other exercise helps.
- Take simple pain killers such as Paracetamol.
- Try to ejaculate.

This may take the problem away within the next hour, if so you should arrange to report the problem to the Haematology medical team caring for you within the next few days.

If the problem does not go away you must go to the Emergency Department. Show them this leaflet and ask them to contact the Sickle cell team (details below) and on call Urology team.

The plan will include a series of steps which includes a referral to Urologist.

In most cases a sample of blood will be taken from the penis using a very small needle to check how much acid and oxygen is in the blood. This procedure is done by numbing the penis first using local anaesthetic and is very important to save future penile function.

Other steps may include:

- Medicines such as Etilefrine or Psuedoephidrine which are usually given by mouth or may be given by injection into the penis.
- Drainage of some blood from the penis/
- A different drug which will stop erections by lowering testosterone is used in some cases.
- In severe cases you may need an operation done by a specialist Urology team at The Royal Liverpool University Hospital (RLUH). There are several types of operation and these will be discussed with you.

If you are worried about anything you have read in this leaflet please ask your consultant haematologist or clinical nurse specialist for support and advise relating to any of your concerns.

Heamoglobinopathy / Sickle Cell Team:

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Dr D Simcox Consultant Haematologist

Dr V J Martlew Consultant Haematologist

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Feedback

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Following your discharge from hospital or attendance at your outpatient appointment you will receive a text asking if you would recommend our service to others. Please take the time to text back, you will not be charged for the text and can opt out at any point. Your co-operation is greatly appreciated.

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Review Date: October 2021

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