# Priapism and Sickle Cell Disease

# Information for people with sickle cell disease

Read this information to learn:

- what causes priapism
- what treatments can help
- who to contact for more information





# What is priapism?

Priapism is a painful erection that can last a long time. It may happen without any sexual arousal. Priapism has been linked to other serious long term health problems, such as kidney disease, leg ulcers and lung problems.

You may feel embarrassed and uncomfortable talking about priapism, but it is very important to talk to your doctor or health care provider about this painful condition. They will be able to tell you more about priapism, and give you treatments that may help reduce the pain, stop the episode sooner and prevent new episodes in future.

# What causes priapism?

During a priapism episode, blood does not flow properly inside the penis. The "sickled" red blood cells in the arteries inside the penis get blocked and stop the blood from flowing back out of the penis, which would normally reduce the erection. This causes a painful erection to last much longer than normal.

If the erection lasts longer than 6 hours, there can be serious long-term effects to your health.

Priapism is like any other type of painful sickle cell crisis. It can happen at any time without other symptoms of sickle cell disease. Most men with sickle cell disease will have priapism at some time in their life. Episodes usually begin in childhood.

Some triggers that may cause a priapism episode are:

- fever
- dehydration
- cold exposure
- a full bladder
- prolonged sexual activity
- cannabis use (smoking marijuana, pot)

#### I heard there are 2 kinds of priapism. What are they?

The 2 kinds of priapism are:

- 1. Stuttering Priapism
- 2. Prolonged (Fulminant) Priapism

The difference between them is how long the priapism episodes lasts and how they are treated.

#### Stuttering priapism

This is an episode of priapism that may last up to 4 hours and may come and go over several days or weeks. The penis may ache or feel full and heavy.

The erection does not last more than 4 hours at a time.

#### Ways to reduce the pain and help stop the priapism episode sooner

- Drink lots of fluids and urinate (pee)
- Take a warm bath or long shower
- Do gentle exercise
- Try to masturbate
- Try activities that can distract you like reading, watching TV, meditating, playing video games, listening to music
- Take your pain medication

#### Prolonged (Fulminant) priapism

This is an episode of priapism that **lasts longer than 4 hours**.

#### What to do if you have an erection that lasts longer than 4 hours

- If you have already tried everything you can to reduce the pain and make your erection stop, but nothing worked, go to your nearest hospital Emergency Room right away.
- Prolonged priapism may stop you from urinating (peeing). This is considered a medical emergency.

# What are the treatments for prolonged priapism?

The treatments that may be used to end a prolonged priapism episode are:

- 1. Intravenous fluids (IV) and strong pain medicines.
- 2. Seeing a urologist (a doctor with special training) who may try to remove the sickled blood directly from your penis with a needle and syringe.
- 3. Medication that may be injected directly into your penis by needle, or may be taken as a pill. This medication is very effective at stopping the priapism. Most patients handle this treatment well.
- 4. Very rarely, if no other treatments are working, a blood transfusion may be considered.

### How can I prevent another episode?

If you are getting priapism often, ask your doctor about medication to prevent future episodes.

The medication options are:

- Hydroxyurea, a medication used for patients with sickle cell disease
- The same medication injected into the penis or taken as a pill, as directed by your doctor or nurse practitioner

# What do I need to know about treating priapism?

If you have an erection lasting more than 4 hours, get medical help right away. Permanent damage to the penis happens when an erection lasts 24 to 48 hours. This may leave you impotent (a condition when you cannot get an erection). If you are treated within 4 to 6 hours, the erection can generally be stopped with medication and therapy.

The goal of priapism therapy is to:

- ease the pain
- make your erection go away as soon as possible
- preserve future erectile function

Priapism can be linked to other serious complications of sickle cell disease. See your health care team to make sure that there are no other problems that also need to be treated.

# Can women get priapism?

Right now there is little research or knowledge about women and priapism, but there have been reports of women having severe pain in the clitoris. This may be a type of priapism.

# Who can I talk to if I have questions?

You and your and family may find it hard to talk about priapism. You are not alone. Priapism happens to many men and boys with sickle cell disease. We will listen to you, keep your information private, and help you find the best way to manage this condition.

If you have questions about priapism, call your primary Nurse Practitioner or any member of the Red Blood Cell Disorders Program at 416 340 4882.



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