

# Priapism and Sickle Cell Disease



UHN

## Information for Patients

### What is Priapism?

Priapism is a prolonged, painful and unwanted erection of the penis. This erection lasts a long time and often happens without any sexual stimulation or desire. Priapism is a serious complication of Sickle Cell Disease. It has been linked to other serious long-term health problems, such as kidney disease, leg ulcers and lung problems.

Due to the personal nature of this condition, you may feel embarrassed and uncomfortable talking about it. But it is very important for you 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? When can it happen?

- An erection of the penis happens when there is blood flow into the penis, but decreased blood flow out of the penis.
- During a Priapism episode, there is a “bunching up” of the “sickled” red blood cells in the arteries inside your penis. This blockage of cells stops the blood from flowing back out of your penis, which would normally reduce your erection. This causes your erection to last much longer than normal and be quite painful.
- If this condition 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, but episodes usually begin in childhood.

Please visit the UHN Patient Education website for more health information:  
[www.uhnpatienteducation.ca](http://www.uhnpatienteducation.ca)

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Some triggers that may cause a Priapism episode are:

- Fever
- Dehydration
- Cold exposure
- A full bladder
- Prolonged sexual activity
- Cannabis use (smoking pot)

## **I heard there are two kinds of Priapism. What are they?**

There are two kinds of Priapism:

1. Stuttering Priapism
2. Prolonged (Fulminant) Priapism

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

### **1. 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, but the erection **does not last more than 4 hours at a time.**

To reduce the pain and to help stop the Priapism episode sooner:

- Drink lots of fluids and urinate
- Take a warm bath or long shower
- Do gentle exercise
- Try to self-ejaculate or masturbate
- Use distraction activities like reading, watching television, meditating, playing video games, listening to music
- Use your pain medication

### **2. Prolonged (Fulminant) Priapism:**

This is when you have a prolonged 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 it has not worked, **you need to go to your nearest hospital Emergency Room right away.**
- Prolonged Priapism may prevent passing of urine. This is considered a medical emergency.

### **What are the treatments that may be used for Prolonged Priapism?**

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

1. Intravenous fluids (IV) and stronger pain medicines
2. Being seen by an Urologist (a doctor with special training) who may try to remove the sickled blood directly from your penis with a needle and syringe
3. Drugs that may be injected directly into your penis, or may be given to you by mouth. This direct treatment is very effective at stopping the Priapism and most patients handle this treatment well.
4. Very rarely, if no other treatments are working, an exchange blood transfusion may be considered

### **How can I prevent another episode?**

If you are getting Priapism often, ask your doctor about medication to prevent further episodes.

The medication options are:

1. Hydroxyurea - a drug used for patients with Sickle Cell Disease
2. The same medication that is injected into the penis, in pill-form, to be taken by mouth as directed by your doctor or nurse practitioner

## **What is the Goal of therapy?**

- You and your family may be embarrassed or, may find it hard to talk about Priapism. Priapism happens to many men and boys with Sickle Cell Disease, and getting immediate medical assistance is the best thing that you can do to help yourself.
- Permanent damage to the penis happens after an erection lasts 24 to 48 hours. This may leave you impotent (a condition when you cannot get an erection). The goal of Priapism therapy is to ease the pain, make your erection go away as soon as possible, and preserve future erectile function. If you are treated within 4 to 6 hours, the erection can generally be stopped with medication and therapy.
- You should know that Priapism can be linked to other serious complications of Sickle Cell Disease. See your healthcare 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?**

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**.

We know that this may be a hard thing for you to talk about. We will listen to you, keep your information confidential and private, and help you find the best way to manage this condition.