

# Pain Crisis and Sickle Cell Disease



TGH

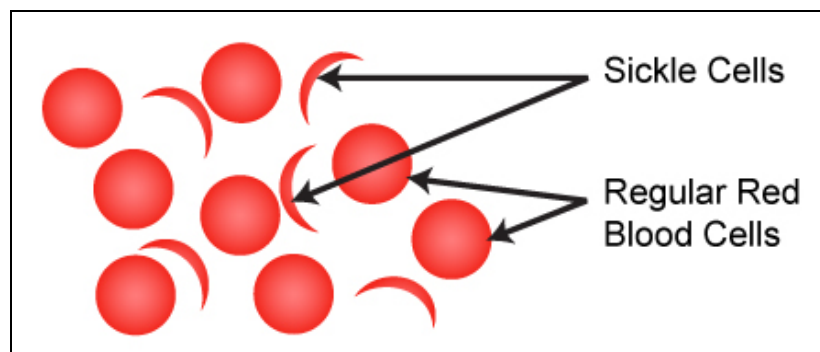
## UHN Red Blood Cell Disorders Program

### What Is a Pain Crisis?

A pain crisis may happen in patients who have Sickle Cell Disease (SCD). Pain happens when the tissues of the body do not get a normal amount of blood flowing to them. When this happens, a person in “crisis” may feel severe pain.

### What is Sickle Cell Disease (SCD)?

- Sickle cell disease is an inherited or genetic blood disorder that changes how the red blood cells (RBC) in our body look and work.
- Hemoglobin (Hb) is a protein (found in RBCs) that carries oxygen around the body. Oxygen is needed to keep all of the major organs of the body working well. The usual RBC is round, flexible and can pass through tiny blood vessels.
- In SCD, a different type of hemoglobin is made by the body. It is called HbS. In SCD, the RBCs turn into a “sickle”\* or curved shape and become ‘sticky’. During a pain crisis, more and more RBCs start to turn into a sickle shape. Since the RBCs are a strange shape and very sticky, they cannot pass through the blood vessels easily. This causes blockage of the blood supply to bones, tissues and organs and terrible pain.



\*a sickle is a kind of blade for cutting grass and hay

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## **How will I know if I am having a Pain Crisis?**

Signs and symptoms of a pain crisis may include:

- Pain in your arms, hips/pelvis, legs, chest and back
- Difficulty breathing with chest pain, shortness of breath, fever or both
- Stomach pain with or without nausea, vomiting, and diarrhea

## **What can cause a Sickle Cell Crisis?**

There are many conditions that may start a pain crisis. These include:

- If you are dehydrated (do not have enough water in your body)
- If you have an infection
- If you have been exposed to extreme cold or heat
- If you smoke, use drugs or drink too much alcohol
- If you are pregnant
- If you have psychological stress, are unable to sleep, or both
- If you have surgery and subsequent uncontrolled pain

## **What should I do if I think I am having a Sickle Cell Crisis?**

If you are at home:

1. Take your pain medications just as your doctor, nurse practitioner or pharmacist has told you to do
2. Drink lots of fluids
3. Get some rest
4. Keep warm
5. Keep taking your Hydroxyurea (Hydrea®) just as your doctor, nurse practitioner or pharmacist has told you to do

## **When should I go to the Emergency Room?**

- You should go to the nearest Emergency room if you have:
- Pain that will not go away or gets worse
- Pain that does not seem like your usual sickle pain
- Side effects from your pain medications
- Not been able to drink lots of fluids
- A fever (higher than 38° C)
- Chest pain or breathing problems
- Vomiting, diarrhea or both
- A severe headache, trouble with speech or walking, problems with your eye sight or are feeling confused
- A sense that there maybe “something else going on”
- Priapism (a painful erection that will not go away)

## **How will my Sickle Cell Crisis be treated?**

- In the Emergency room, a doctor or a nurse will ask you about your medical history and perform a physical examination of your body
  - You should provide the contact information for the TGH Red Blood Cell Disorder (RBCD) clinic and doctor to the Emergency room staff so that they may talk over your care with the RBCD clinic team
- You may be given:
  - Pain medicine to help control your pain
  - Antibiotics (to treat any infections) and other medicines (if needed)
  - Fluids through an intravenous (IV) line placed in your vein
  - Oxygen

**\*\* Let the doctor or nurse in the Emergency room know what you have been taking at home and how much you usually need to treat your pain. The doctors and nurses in the Emergency room may not be used to giving patients pain medicines in high doses.\*\***

## **Will I have to stay in the hospital?**

You may be admitted to hospital if:

- Your crisis is related to an infection
- Your crisis is affecting your lungs or neurological system
- Your pain does not go away after 4 to 6 hours
- You are not able to drink enough fluids at home
- You need some further tests to find out what started your crisis

## **When you go home:**

1. Drink large amounts of fluids such as a water or juice
2. Make an appointment to see your doctor in the Red Blood Cell Disorder clinic about one week after going home
3. Return to the hospital right away if you have:
  - A fever (higher than 38°C) that does not go away
  - Chest pain, shortness of breath, nausea and vomiting
  - Abdominal pain with nausea and vomiting
  - A new headache that does not go away

## **What can I do to prevent another Pain Crisis?**

Here are some things you can do on your own that may help to prevent a pain crisis:

- Talk to your doctor about taking Hydroxyurea if you are not taking this medication – Hydroxyurea has been proven to be helpful in preventing sickle cell crises
- Take your Hydroxyurea just as your doctor, nurse practitioner, or pharmacist has told you to do
- Prevent other illnesses - get your vaccinations for pneumococcus, H influenzae, meningococcus, influenza as well as your other routine vaccines
- Avoid extreme temperatures (very cold places)
- Avoid drinking alcohol, using non-prescribed drugs or medications, and smoking
- Drink lots of fluids

- See your doctor:
  - At least once a year for a check-up
  - OR
  - more often if you are having trouble managing your medications or illness
- Seek out the help of a social worker to help with any social issues you may have
- Engage the use of self-help groups and non-pharmaceutical pain management techniques. These include:
  - Distraction
  - TENS (Transcutaneous electrical nerve stimulation) – this involves using electrical current (through a device) to stimulate nerves for therapeutic needs
  - Yoga
  - Cognitive behavioural therapy
  - Breathing exercises

### **Who can I talk to if I need more information?**

Please feel free to call:

Red Blood Cell Disorder Program  
University Health Network – Toronto General Hospital  
NCSB 12th Floor – 1274  
Telephone 416-340-4882