

Sickle Cell Disease Pain Crisis

Patient and Family Education

This teaching sheet contains general information only. Your child's doctor or a member of your child's health care team will talk with you about specific care for your child.

What is a pain crisis?

A pain crisis happens when there is loss of blood supply to a part of the body. This usually happens in the bones or joints.

- Sickle cells can block blood vessels in the body tissues. This keeps the body tissues from getting the blood supply and oxygen it needs. If the blockage lasts too long, the tissues can be damaged.
- When tissue is damaged, the injury causes nearby nerves to send electrical signals to the brain. The signals are felt as pain.
- Pain can continue for some time until the tissues start to heal and the nerves calm down.
- In a few people, pain can continue for a longer time because the nerves do not calm down.
- How often pain happens also depends on such things as how often sickle cells get stuck" and how your child's body reacts to tissue injury or pain.

What are the symptoms of a pain crisis?

Everybody feels pain in a different way. This depends on many things such as:

- How well the body repairs itself
- How the body reacts to pain
- Past experiences with pain
- How well pain medicines work

Pain crises can occur:

- All of a sudden without warning
- With some warning signs
- With other sickle cell problems, especially in the lungs
- As a direct result of other sickle cell problems, such as a gall stone

Sometimes pain symptoms go away in a few hours. Other times they can last for a week or two. Some patients that have pain often have persistent pain (pain that stays all the time).

In babies and toddlers

A pain crisis is often called a hand-foot syndrome (dactylitis) in young children. This means that the symptoms are mostly in the hands or feet and include:

- Swelling, warmth or change in movement
- Pain or distress



In case of an urgent concern or emergency, call 911 or go to the nearest emergency department right away.

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Young children often have trouble telling where the pain is, what it feels like and how much they hurt.

In older children

Pain can occur in more places in the body and can occur in more than one place at the same time.

- The most common sites of pain are the arms, legs, joints, back and abdomen (stomach).
- Swelling, warmth and increased pain with movement or pressure (touching) can occur.
- Sometimes the pain can be deep in the body or bones without these other signs.

Older children learn how to tell where the pain is and how much they hurt. They often say that their sickle cell crisis pain feels "different" than other pain such as headaches, sore throats or muscle sprains.

What tests could my child have?

Pain can happen from any type of tissue injury or damage, such as after surgery or an accident. Pain in a person with sickle cell disease can be caused by many types of problems.

- There are no medical tests that check for pain or a pain crisis. There are also no tests that can tell when sickle cells are causing a blockage. This means that a pain crisis is not always easy to confirm and doctors need to take your word for it.
- If there are no other known reasons for the pain, and if the pain is like past pain crises, most doctors assume pain is from a sickle crisis.
- Sometimes other tests are needed like:
 - Blood tests to look for other sickle cell problems, such as anemia that is getting worse
 - Blood tests to look for signs of dehydration (drying out)
 - A chest X-ray or cultures if there is a fever with the pain crisis
 - Tests to help rule out other types of problems

How is a pain crisis treated?

The goal of treating a pain crisis is to make your child less uncomfortable and allow for as many normal activities as possible.

- This does not always mean being pain free.
- It does mean having a positive outlook, taking control and not giving up in spite of problems. These are important life skills that can help your child cope with pain.

Most pain crises are similar each time. Your child's doctor will talk with you about specific care for your child, including pain medicines based on your child's weight and usual level of pain. Work with your doctor to help choose treatments for mild or severe types of pain. Some general guidelines for treating a pain crisis are listed below.

Things you can do at home

Most pain can be managed a home with pain medicine, extra fluids and other comforting activities to help your child manage the pain. Make a plan that works best for you and your child's current level of pain. This includes such things as:

- Massage or apply heat to the painful area for 15 minutes every hour. To prevent skin burns, be sure place a cloth cover on the skin before you apply heat.
- Help your child find a quiet place where he can relax, rest and be comfortable. Many pain medicines cause children to feel sleepy.

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- Use distraction to help take your child's mind off his pain. Some ideas include: playing cards and video games, reading, watching TV, listening to music or talking with friends on the phone.
- Some children find quiet meditation or prayer helpful.
- Give your child plenty of extra fluids such as water, juice or Kool-Aid to keep him from drying out.

When you need to call the doctor

When you cannot handle your child's pain at home, call your child's doctor. Your child may need other types of pain medicines or stronger pain medicines. Also be sure to call if your child has a fever (temperature of 101°F or higher).

When your child needs to come to the hospital

You may need to bring your child to the hospital to receive stronger pain medicines. These medicines are usually given IV (intravenously or into a vein).

- They often work better when given with IV fluids.
- They may be given in the clinic, Emergency Department or in the hospital. This is based on how severe the pain is.
- Your child's doctor will let you know where to go for his treatment.

What do I need to know about pain medicines?

- Make sure you don't run out of pain medicine. Your child's doctor cannot call in a prescription for some
 of the stronger pain medicines. You will need to give a written prescription to the pharmacy for these
 stronger medicines.
- Be sure to keep medicines in a safe place. Some of them are dangerous for small children.
- Don't wait to see if the pain will go away by itself. Pain can get out of control, making it harder to treat.
- When your child needs medicine for pain, give it on a **set schedule**.
 - Most pain medicines only work in the body for 3 to 4 hours.
 - Follow your doctor's orders carefully for how much medicine to give your child and when to give it.
 This is very important when your child needs more than one medicine to control the pain.

Doctors usually use a step-up or "ladder" approach with pain medicines.

For milder pain or pain that is just starting:

- Give your child ibuprofen (Motrin, Advil or other less costly store brand) or acetaminophen (Tylenol or other less costly store brand). Be sure to check your child's temperature **before** giving him acetaminophen or ibuprofen.
- DO NOT:
 - Give more than 6 doses of acetaminophen or ibuprofen within a 24-hour period
 - Take more than the recommended dose each time.
 - Combine acetaminophen with other medicines that also contain acetaminophen, like Lortab. This is too much acetaminophen and can cause liver damage.

For more severe pain

Give your child a stronger pain medicine such as Lortab or similar medicine prescribed by your child's
doctor. There are even stronger pain medicines that can sometimes be used, especially for older children
or teens who have severe pain.

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SCD: Pain Crisis, continued

• Stronger pain medicines can cause some side-effects such as constipation, sleepiness, upset stomach and itching. If these are common problems for your child, talk with his doctor about things you can do to help prevent or reduce them.

What type of follow-up care is needed?

After your child goes home from the emergency room or hospital:

- Give the pain medicine and fluids as ordered by your child's doctor
- Make sure that you understand when to stop the pain medicines. If some stronger pain medicines are stopped too quickly he could have problems with withdrawal symptoms.
- Have your child resume his normal activities and school work as soon as possible.
- Follow-up with your child's doctor as advised. Your doctor can talk with you about how the medicines worked and make any changes needed. You will also be able to get new prescriptions for medicines to use in the future.

How can I help prevent a pain crisis from happening again?

Doctors do not know exactly why a pain crisis occurs. That makes it hard to find ways to prevent them. People seem to have their own triggers or patterns. Some things that seem to play a role include:

- Physical and mental stress
- Changes in the weather
- Being very tired
- Dehydration (drying out)
- For some older girls and women, pain crises happens around their menses (monthly period)

For some children, it is just part of their sickle cell disease. If this is true, there are a few things that may help reduce how often a pain crisis occurs such as:

- Blood transfusions
- A medicine called hydroxyurea
- A bone marrow transplant

What should I do if my child is sick?

- Follow your child's doctor's advice for what you should do.
- Call your child's health care provider anytime that your child has a fever of 101°F or higher and is sick. Your child will need to be seen **right away.**
- During the time when the clinic is closed (between 5PM and 8AM during the week, on weekends and holidays), call the hematologist on call. They will notify the Emergency Room and help with your child's care.
- **ALWAYS** tell the Emergency Room doctors and staff that your child has sickle cell disease. Also tell them which sickle cell clinic he visits.

For more information on sickle cell disease and services, visit Children's Healthcare of Atlanta website at www.choa.org/sicklecell.