

# Sickle cell disease (SCD): pain episodes

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## What is a pain episode?

Pain and other sickle cell problems happen when the red blood cells (RBCs) are sickle shaped. This blocks the flow of blood and oxygen to parts of the body, such as the bones, lungs, spleen, brain, eyes and kidneys. A pain episode (also called a vaso-occlusive episode) is the most common problem for children with sickle cell disease.

- 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 some children, pain can last longer because the nerves do not calm down.
- How often pain happens also depends on such things as how often sickle cells block blood flow in the body and how your child's body reacts to tissue injury or pain.

## What are the symptoms?

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

- How well the body repairs itself.
- How the body and mind react to pain.
- Past experiences with pain.
- How well pain medicines work.

Pain episodes can occur:

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

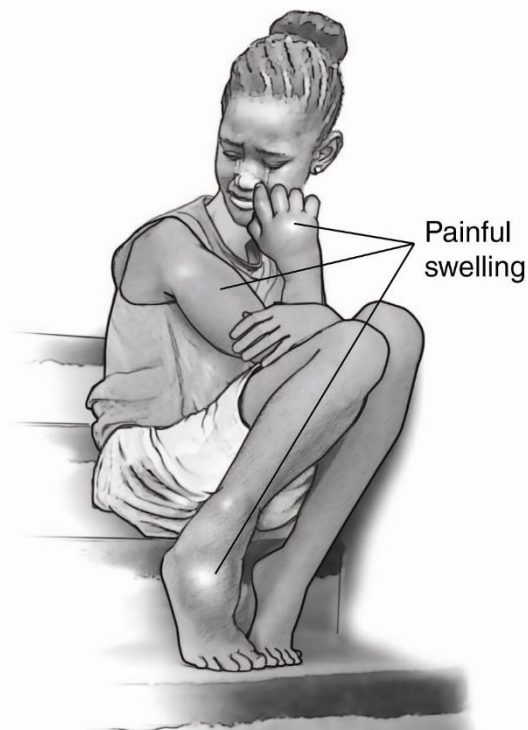
Sometimes, pain symptoms go away in a few hours. Other times, they can last for 1 or 2 weeks. Some children who have pain often have persistent pain (pain that stays all the time).

## In babies and toddlers

A pain episode in younger children can also be called hand-foot syndrome (dactylitis). This means the symptoms are mostly in the hands or feet and include:

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

Pain Episode



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# SCD: pain episodes, continued

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Young children often have trouble telling where the pain is, what it feels like, and how much they hurt. Some signs of hand-foot syndrome are when a child does not want to hold their bottle or have socks and shoes on their feet. They may also avoid walking or putting weight on their feet.

## In older children and teens

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

- The most common sites of pain are the arms, legs, joints, back and abdomen (belly).
- 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 and teens learn how to tell where the pain is and how much they hurt. They often say their sickle cell 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 episode. There are also no tests that can tell when sickle cells are causing a blockage. This means that pain is not easy to confirm, and providers need to listen to what your child says about their pain.
- If there are no other known reasons for the pain and if the pain is similar to past pain episodes, most providers assume pain is from sickle cell disease.
- 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
  - A chest X-ray or cultures if there is a fever with the pain episode
  - Tests to help rule out other types of problems

## What is the treatment?

The goal of treating a pain episode is to help make your child more comfortable and allow for as many normal activities as possible.

- This does not always mean being without pain.
- Having a positive outlook, taking control and not giving up despite problems are helpful life skills that can help your child cope with pain.

Most pain episodes are similar each time. Your child’s provider 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 child’s provider to help choose treatments for mild or severe pain. Some guidelines for treating a pain episode are listed on page 3.

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## Things you can do at home

Most pain can be managed at home with pain medicine, extra fluids and other comforting activities. Make a plan that works best for you and your child's level of pain.

This includes such things as:

- Massage or heat to the painful area for 15 minutes every hour. To prevent skin burns, be sure to place a cloth cover on the skin before you apply heat. **Do not use ice packs** because they could make the pain worse.
- Help your child find a quiet place where they can relax, rest and be comfortable. Many pain medicines cause children to feel sleepy.
- Use distraction to help take your child's mind off their pain. Some ideas include:
  - Playing cards and video games
  - Reading
  - Watching TV
  - Listening to music
  - Talking with friends on the phone
- Some children find quiet meditation or prayer helpful.
- Give your child plenty of extra fluids, such as water (best choice) or juice, to prevent getting dehydrated.

## For milder pain or pain that is just starting

- Give your child acetaminophen (Tylenol or less costly store brand) or ibuprofen (Motrin, Advil or less costly store brand).
- Be sure to check your child's temperature **before** giving either of these medicines.
- **DO NOT:**
  - Give more than 6 doses of acetaminophen or 4 doses of ibuprofen within a 24-hour period.
  - Give more than the recommended dose each time.
  - Combine acetaminophen with other medicines that also contain acetaminophen, like Lortab (hydrocodone + acetaminophen). This is too much acetaminophen and can cause liver damage.

## For more severe pain

- Give your child a stronger opioid 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 for older children or teens who have severe pain.
- Opioid pain medicines can cause side effects. If these are problems for your child, talk with their provider about things you can do to help prevent or reduce them. Some side effects include:
  - Constipation
  - Sleepiness

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- Upset stomach
- Itching

## When should I call the clinic about my child's pain?

Call your child's provider when you cannot handle your child's pain at home. Your child may need other types of pain treatment. Also, be sure to call the clinic **right away** anytime your child has a fever of 101°F (38.3°C) or higher.

## When should I take my child to the hospital for pain?

You may need to take your child to the hospital to get stronger pain medicines. These medicines are most often given through an I.V. (into a vein). These pain medicines:

- Often work better when given with I.V. fluids.
- May be given in the clinic, emergency department (ED) or hospital. This is based on how severe the pain is. Your child's provider will let you know where to go for their treatment.

Take your child to the hospital if they have other serious symptoms, such as:

- Fever:
- Trouble breathing
- Weakness
- Loss of consciousness

## What do I need to know about pain medicines?

- Make sure you do not run out of pain medicine. Your child's provider may not be able to call in a prescription for opioid pain medicines and you may need to give a written prescription to the pharmacy.
- Be sure to keep medicines in a safe place. It is dangerous for children to take too much.
- Do not wait to see if the pain goes away by itself. Pain can get very strong, 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 4 to 6 hours.
  - Follow the medicine directions carefully for how much 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.

**Providers most often use a step-up or "ladder" approach with pain medicines. Ask them what this means for your child.**

## What follow-up care does my child need?

After your child goes home from the ED or hospital:

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- Give pain medicines and fluids as directed.
- Make sure you know when to stop the pain medicines. If opioid pain medicines are stopped too quickly, your child could have problems with withdrawal symptoms.
- Have your child return to their normal activities and schoolwork as soon as possible.
- Follow-up with your child's provider as advised. They can talk with you about how well the medicines worked and make changes as needed. You will also be able to get new prescriptions for medicines to use in the future.
- Your child's provider can make a Pain Action Plan. This lists treatments that work best for your child at home and in the hospital

## Can we prevent pain episodes from happening again?

It is not known exactly why a pain episode 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 – dress your child warmly in cold weather
- Being very tired
- Dehydration
- Infections
- For some older girls and women, pain episodes happen around their monthly period.

Healthy lifestyle habits can help prevent some pain. Try to help your child:

- Drink enough water.
- Sleep 7 to 8 hours each night
- Reduce or treat their stress.
- Avoid cold water on their skin and use of ice packs.

For some children, pain is a part of their sickle cell disease. If this is true, there are a few things that may help reduce how often a pain episode occurs. This may include:

- Blood transfusions
- Medicines like hydroxyurea, L-glutamine or crizanlizumab (Adakveo)
- A bone marrow transplant

## What should I do if my child is sick?

- Follow the sickle cell provider's advice for what you should do.
- Call and **get medical care right away** anytime your child has a fever of 101°F (38.3°C) or higher.
- During the time when the clinic is closed (between 5 p.m. and 8 a.m. during the week and on weekends and holidays), call the hematologist on call before going to the hospital. They will call the emergency department (ED) and help with your child's care.

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- **ALWAYS** tell the ED doctors and staff that your child has sickle cell disease. Also tell them which sickle cell clinic they visit.

For more details on sickle cell disease and services, visit the Children's Healthcare of Atlanta website at [choa.org/sicklecell](http://choa.org/sicklecell).

**This teaching sheet contains general information only. Talk with your child's doctor or a member of your child's healthcare team about specific care of your child.**

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