

Sickle Cell Disease and Acute Chest Syndrome

Sickle cell disease is an inherited blood disorder. Children are born with the condition. It affects a part of the red blood cell called **hemoglobin**. Hemoglobin is the part of the red blood cell that carries oxygen to different parts of the body. A person with sickle cell disease makes a different kind of hemoglobin called **sickle** hemoglobin. Instead of being round and smooth, cells with sickle hemoglobin are shaped like a banana or a sickle. They become hard and sticky and have trouble moving through small blood vessels. Sometimes they clog up these blood vessels, preventing the blood from bringing oxygen to the tissues. This can cause pain or damage to the areas that are not getting oxygen.

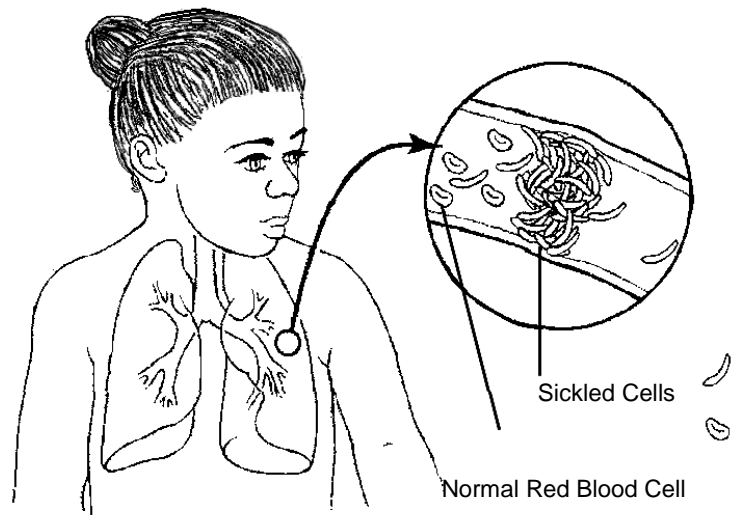
Acute Chest Syndrome (ACS)

Acute chest syndrome (ACS) is a group of symptoms that occur when sickled cells become clumped together in the lungs (Picture 1).

Children with ACS usually complain of chest pain. They may also have a fever, cough or trouble breathing. ACS can be triggered by a lung infection like pneumonia.

It may also happen before, during or after a pain episode. There is no way to tell the difference between ACS and pneumonia (new MOAN yuh).

Acute Chest Syndrome can be life-threatening if not treated promptly.



Picture 1 Sickled cells can clump together in the lungs, causing pain and trouble breathing.

Symptoms of ACS

- Chest pain
- Tightness in the chest
- Bad cough
- Fever (101 degrees Fahrenheit or higher)
- Breathing very fast
- Shortness of breath

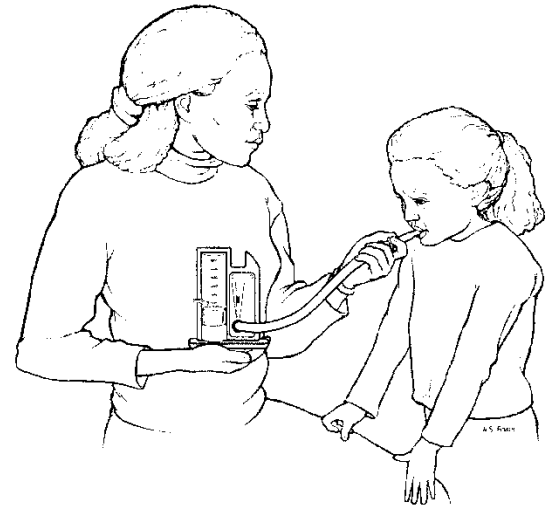
Treatment

If the doctors think that your child might have ACS, blood will be drawn to check the child's blood count and a chest X-ray will be done. Most children with ACS are admitted to the hospital so they can be watched very closely. IV fluids and antibiotics will be given to treat the infection. If the level of oxygen in your child's blood is low, oxygen will also be given. Sometimes a blood transfusion is needed. Your child may also be given aerosols (breathing treatments) and medicines to help with breathing.

Prevention of ACS

ACS often happens during a child's hospital stay for treatment of pain. It is also the most common problem after surgery and anesthesia.

A child who is hurting may not breathe as deeply as usual. Lying in bed all day, and the medicines given to treat pain, may also cause your child to breathe less deeply. Not breathing deeply can cause some of the small airways in the lungs to collapse. When this happens, it is more likely that your child will develop ACS.



Picture 2 Using an incentive spirometer.

Here are some things you and your child can do to help prevent ACS:

- Your child needs to be out of bed and walking at least 2 or 3 times per day while in the hospital. This is important even if he or she is hurting.
- An **incentive spirometer** is a tool your child uses to take big, deep breaths (Picture 2). The incentive spirometer should be used to take 10 breaths each hour during the day.

Taking big, deep breaths is very important. It helps prevent collapses in the small airways in the lungs. If your child is too young to understand how to use the spirometer, you can help him take deep breaths by having him blow bubbles or pretending to blow the candles out on a birthday cake.

Phone Numbers: The phone number for the Sickle Cell Clinic at Nationwide Children's Hospital is (614) 722-3250. If your child becomes ill during the day call the Sickle Cell Clinic for treatment advice. On evenings and weekends call (614) 722-2000 and ask for the hematologist on call. The Sickle Cell Nurse is available to answer non-urgent questions between 8 am to 4:30 pm, Monday through Friday, at (614) 722-8914.

When to call 911

The following situations are emergencies. Call 911 if:

- Child is unresponsive
- Child is unable to wake after a nap
- Child has trouble breathing
- Child has sudden weakness or loss of feeling in a body part
- Child is unable to move a body part