

Sickle cell disease (SCD): acute chest syndrome

What is acute chest syndrome (ACS)?

Acute chest syndrome (ACS) is a lung problem that happens in children who have sickle cell disease. It looks like pneumonia on a chest X-ray. It can be caused by 1 or more of these:

- Sickle red blood cells blocking blood vessels in the lungs, like a sickle cell crisis
- An infection in the lung
- Fat droplets in the lungs caused from a sickle cell bone pain or crisis
- Sedation (extreme sleepiness) from medicine being used to treat pain

Children with sickle cell disease who have ACS may develop it again. If they also have asthma, the risk for ACS again is even higher. Repeated ACS can cause lasting lung damage.

- ACS can get worse quickly and become life-threatening.
- It is important for you to know about symptoms of ACS so you can get medical help **right away**.

What are the symptoms?

Your child may have 1 or more of these:

- Trouble breathing (fast or heavy breathing)
- Fever [temperature of 101°F (38.3°C) or higher]
- Cough
- Chest pain
- A low oxygen level
- Back pain or belly pain

What should I do if my child has symptoms?

If your child has any of the symptoms listed above:

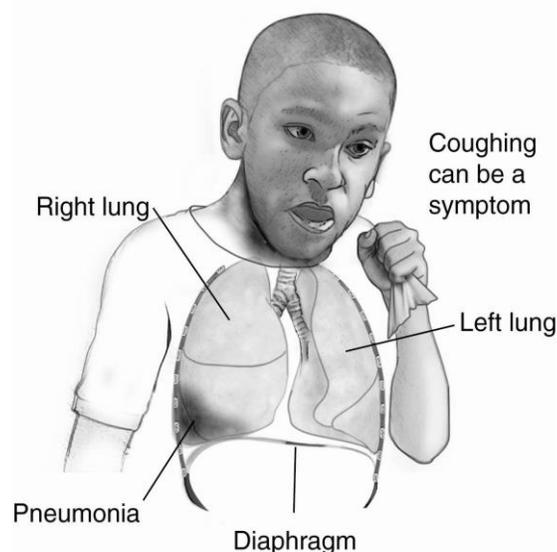
- **Get medical help right away.** Your child needs treatment as soon as possible.
- Call the sickle cell clinic, and then take your child to the emergency department (ED).

What tests could my child have?

Your child will need:

- A physical exam
- A chest X-ray
- Blood tests (labs), including blood counts
- Their blood oxygen level measured
- Blood cultures or other tests to check for bacteria (germs)

Acute Chest Syndrome



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

SCD: acute chest syndrome, continued

What is the treatment?

Your child will first need treatment in the hospital. This may include:

- Intravenous (I.V.) fluids.
- Incentive spirometry (breathing exercises) to help to keep the lungs open.
- Breathing treatments and chest physiotherapy (CPT) to help loosen mucus and thick fluids in the lungs.
- Checking blood oxygen levels by using a pulse ox (a soft probe that is attached to your child's finger or toe using a piece of tape).
- Treatment with oxygen if oxygen levels are low.
- Antibiotics to help kill germs.
- Pain medicine if needed.

Your child may also need:

- A blood transfusion to help the lungs heal and improve oxygen levels.
- BiPAP or CPAP (breathing machines to help get more air into the lungs if oxygen levels are too low or if your child has trouble breathing).
- An intensive care unit (ICU) stay where the care team can watch them more closely and provide more serious treatments if needed.

What follow-up care does my child need?

Your child needs a clinic visit after they go home from the hospital. It is very important that your child goes to their visit to make sure the lungs are healing and to help prevent further damage to the lungs.

During the visit, your child may need:

- A physical exam
- Blood tests (labs), including blood counts
- Their blood oxygen level measured
- A chest X-ray to see if the lungs are getting better

Many children also need to see a pulmonologist (lung doctor).

- They may need other medicines or treatments to help prevent ACS from happening again.
- A visit with the lung doctor is especially needed for those children with asthma or other lung problems.

When can my child return to school or day care?

Your child may be able to go back to school or day care soon after leaving the hospital if they feel better. It could take up to about 1 week for your child to feel back to normal.

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SCD: acute chest syndrome, continued

How can I help prevent ACS?

To help prevent ACS from happening again:

- Follow-up with the sickle cell clinic and lung doctor as advised. **This is very important.**
- You can help avoid infections when you:
 - Clean your hands often. Wash them well with soap and water for at least 20 seconds, or use an alcohol-based gel or foam. Teach your child and anyone who cares for them to clean their hands often, too.
 - Keep your child’s vaccines (shots) up-to-date, including the Influenza (flu) vaccine.
 - Give your child antibiotics if the doctor prescribes them. Do not skip doses. If you do, the infection could return.
- Give your child:
 - Any prescribed breathing treatments, such as inhalers or nebulizers.
 - Other medicines as prescribed to help treat or prevent problems.

What should I do if my child is sick?

- Follow your child’s sickle cell provider’s advice for what you should do.
- Call your child’s provider 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.
- **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.

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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