

## PATIENT INFORMATION: Acute Chest Syndrome

### What is acute chest syndrome?

Acute chest syndrome (ACS) is a problem that sometimes happens in children who have sickle cell disease. It is usually caused by one or more of the following:

- Sickle cells causing blockages of blood vessels in the lungs
- An infection like pneumonia
- Fat in the lungs, which comes from sickle cells causing damage in the bones

These problems can cause sudden damage to the lung tissue. When lung tissue is damaged, body fluid can build up in the area, much like the swelling that occurs from an injury.

This fluid build-up causes several specific symptoms:

- Makes it harder to breathe because the lungs get stiff from the extra fluid
- Blocks oxygen in the air from getting to the rest of the body through the lungs
- Is what the doctor looks for on a chest X-ray to check for acute chest syndrome

Nearly half of all sickle cell disease patients have ACS at least one time during their life. Once a child has ACS, he or she is more likely to get it again.

- Repeat episodes of ACS can cause permanent damage to the lungs
- Severe ACS can be life-threatening
- It is important for you to know about symptoms of ACS so you can get medical help right away

Children with asthma or other types of lung problems are at a higher risk of having ACS. If a child with both sickle cell disease and asthma starts to have ACS more often, stronger or different asthma medicines may be needed.

### What are the symptoms of ACS?

Your child may have one or more of the following:

- Fever (temperature of 101°F or higher)
- Cough
- Trouble breathing, including fast or heavy breathing
- Chest pain
- Back pain or stomach pain

### What should I do if my child has symptoms of ACS?

Get help right away. Your child needs to be evaluated by a medical professional. Call to make an appointment in the sickle cell clinic or go to the emergency room.

### How will I know if my child has ACS?

- Your child will be examined to see if he or she has ACS
- A chest X-ray will be performed
- Blood work will be performed
- Oxygen levels will be checked
- If your child has a fever, he or she will need blood cultures to check for infection in the blood

### How is ACS treated?

- IV fluids are given to help with dehydration
- Antibiotics are given to kill bacteria (germs)
- Breathing treatments help open up the lungs
- Blood transfusion may be given
- Oxygen or medicine may be given to help your child's breathing
- Pain medicine is provided, if needed
- More treatments may be needed if your child becomes more sick including admission to the Pediatric Intensive Care Unit

## How can I prevent my child from getting ACS again?

- Go to your follow-up appointments where your child will be examined to make sure his or her lungs are getting better
- Your child may need to see a pulmonologist (a doctor who treats lung problems); this is especially important for children with asthma or other lung problems

## What else should I know about ACS?

- Patients who are admitted for treatment of a pain crisis are at increased risk of developing ACS – this is because pain medicines can cause them to not take deep breaths and stay in bed
- It is important to do breathing treatments while your child is hospitalized for pain to prevent ACS
- A respiratory therapist will assess your child while he or she is in the hospital to determine what respiratory exercises are needed to prevent ACS

## Key points:

- If your child develops a fever greater than 101°F, he or she must be seen at the hospital and given IV antibiotics
- If your child develops a bad cough, chest pain or difficulty breathing, call the Rainbow Sickle Cell Anemia Center so your child can be evaluated
- When a child has ACS, the lungs can be injured, making it difficult to breath and stopping oxygen from being delivered to the rest of the body
- It is important to know the symptoms of ACS so you can get medical help right away
- It is important to attend all follow-up appointments including a lung doctor, if instructed

### Rainbow Sickle Cell Anemia Center

If your child has signs or symptoms of illness, please call 216-844-3345 and follow the prompts.

### Angie Fowler Adolescent & Young Adult Cancer Institute at University Hospitals

2101 Adelbert Road, Cleveland, Ohio 44106

For every question, every kid,  
There's only one Rainbow.



Have a question? Ask a Rainbow doctor. **216-UH4-KIDS** | **216-844-5437** | [Rainbow.org/AngiesInstitute](http://Rainbow.org/AngiesInstitute)

[Facebook.com/UHRainbowBabies](https://www.facebook.com/UHRainbowBabies) | [Twitter.com/UHRainbowBabies](https://twitter.com/UHRainbowBabies) | [Instagram: @UHRainbowBabies](https://www.instagram.com/UHRainbowBabies)