Sickle Cell Disease: Acute Chest Syndrome

What is Acute Chest Syndrome?

Acute chest syndrome (ACS) is a serious illness that can happen with sickle cell disease. ACS is usually caused by one or more of the following problems:

- Sickle red blood cells block the blood vessels in the lungs
- An infection in the lung
- A fat embolism (small piece of fat that goes into the bloodstream)
- Other unknown reasons

These problems can cause sudden damage to the lung tissue. If a person with sickle cell disease has ACS several times, it can cause permanent damage to the lungs. ACS can be life-threatening and is the most serious problem for people with sickle cell disease.

What are the symptoms of ACS?

Symptoms may include one or more of the following:

- Fever
- Cough
- Trouble breathing
- Shortness of breath
- Wheezing
- Chest pain

This handout is for informational purposes only. Talk with your doctor or health care team if you have any questions about your care.

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What tests are done to check for ACS?
You may have the following tests done to see if you have ACS:

- Your doctor will talk with you about your symptoms and do a physical exam.
- You will have a chest X-ray.
- A sputum culture will be collected. This test checks for bacteria in your lungs and airways.
- Blood tests may be done. This often includes blood cultures and arterial blood gas tests.

How is ACS treated?
The following may be done to help treat ACS:

- Intravenous (IV) fluids
- Antibiotics
- Oxygen may be needed if your oxygen levels are low
- Use of a breathing device (incentive spirometer) to help open the lungs
- Coughing and deep breathing
- Pain medicine
- Bronchodilator therapy (medicine to help open your lung passages)
- Blood transfusion therapy (simple or exchange)
- You may need to be admitted to the hospital for more care

What can I do to help prevent ACS?
The following are important ways to help prevent ACS:

- After you are discharged from the hospital, it is very important to take all of your medicines as directed by your doctor.
- Go to all of your follow-up appointments with your doctor.
- Good hand washing can help prevent infections.
- Do your breathing exercises including cough and deep breathing and use an incentive spirometer (breathing device).
- Talk to your doctor about the immunizations (pneumococcal and influenza) you should get to help prevent ACS.
Acute chest syndrome can be a very serious illness. Go to the nearest emergency room or call 911 right away if you have any of the following:

- Trouble breathing
- Chest pain
- Coughing
- Chills or fever of 100.4 degrees Fahrenheit (38 degrees Celsius) or higher