

Sickle Cell disease: Understanding the Facts

Sickle Cell disease affects red blood cells

Red blood cells carry oxygen throughout the body. Sickle cell disease changes the shape of the red cell to a hard, sickle shaped cell. This makes it harder for the blood to flow and deliver the oxygen. Also, the cells break up easily, causing anemia.

Sickle Cell is not restricted to one group

People in many ethnic groups can have sickle cell trait or disease. It is most common among African Americans, but also in people from the Caribbean, Middle East, Mediterranean, India, South and Central America.

Sickle Cell disease can cause serious health problems

It is very important for people with sickle cell disease to be followed by a comprehensive sickle cell program.

Sickle Cell disease is not contagious

A person cannot catch sickle cell disease through the air, water, skin, etc. The only way to get it is to have it passed on from your parents.

Other forms of Sickle Cell Disease

There are different types of sickle cell disease: Sickle cell disease (SS); Sickle-Hemoglobin C Disease (SC); Sickle Beta-Plus Thalassemia (SB⁺) and Sickle Beta-Zero Thalassemia (SB⁰).

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Supported in part by Project #2 H46 MC00240-04-00
Department of Health & Human Services/
Health Resources and Services Administration (HRSA)
/Maternal and Child Health Bureau



COMPREHENSIVE PEDIATRIC
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Sickle Cell & ACS



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What is Acute Chest Syndrome (ACS)?

Acute Chest Syndrome is a complication of Sickle Cell Disease where there is a new shadow seen on the lungs on a chest x-ray. It is a common cause of hospitalization.

What are the symptoms?

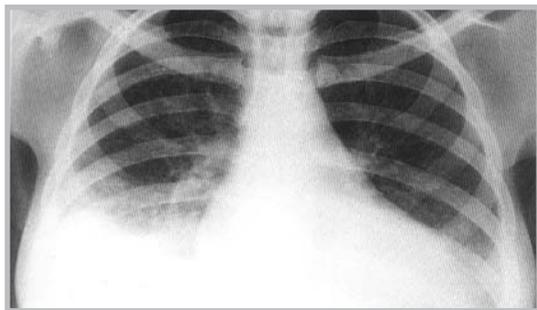
One or more of the following:

- Chest or back pain
- Cough
- Fever
- Difficulty breathing or fast breathing

If any of these occur, seek medical attention.

What tests will be done?

- Physical exam
- Oxygen level in the blood will be checked
- A blood count (CBC and reticulocyte count)
- Chest x-ray will show a new infiltrate (patch) on the lungs –this confirm ACS.



Why does this happen in sickle cell disease?

In sickle cell disease, the sickled red cells block the blood flow in parts of the lung and release damaging chemicals, both of which cause an area of the lung to not function properly. Additionally, there may be infection which can trigger or worsen ACS.

Why is this serious?

Lungs exchange air and obtain oxygen for the rest of the body. With ACS the person will not have enough good lung tissue to supply oxygen for the body.

What treatment is given?

Usually the treatment can be one or more of the following:

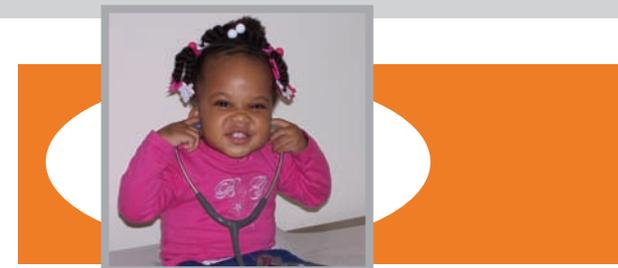
- Admission to the hospital
- Intravenous fluids in monitored amounts
- Oxygen through a mask
- Antibiotics given intravenously or by mouth to prevent or treat infections
- Pain medicine
- Breathing exercises to help expand the lungs
- Medication to open up the air passages
- Chest physiotherapy
- Transfusions



Oxygen through mask



Breathing exercise



Why are transfusions given?

- To give the patient normal red cells
- To improve oxygen delivery to the body
- To enable quicker recovery
- To prevent further complications

Can ACS happen again?

Yes, it can.

Why is ACS a serious complication?

- ACS is serious because it can reduce the amount of oxygen to the body.
- Repeated episodes of ACS can lead to chronic lung disease and affect the right side of the heart.

How can you prevent ACS?

- a) Treat pain adequately.
- b) Take prescribed antibiotics regularly
- c) Encourage breathing exercises like incentive spirometry
- d) Give adequate treatment for underlying asthma
- e) Seek early medical attention for symptoms
- f) Insure that Pneumococcal vaccine has been given
- g) Get a yearly flu vaccine
- h) Avoid smoking near the patient

Are there treatments to prevent ACS?

There are other treatments such as hydroxyurea, blood transfusions or bone marrow transplantation for patients with recurrent complications of sickle cell disease. These can be discussed with your hematologist.