

Facts about sickle cell anemia

Sickle cell anemia is an inherited blood disorder. If both your parents pass on the sickle cell gene, you are born with sickle cell anemia. If one parent passes on the sickle gene and the other parent passes on a normal gene, you are said to have **sickle cell trait**. This means you are a carrier for sickle **hemoglobin** (see “What Is Sickle Cell Anemia?”) and can pass on sickle cell trait to your children. You also can pass on sickle cell anemia to them if the other parent also has sickle cell trait or sickle cell anemia. Sickle cell trait usually does not cause any symptoms.

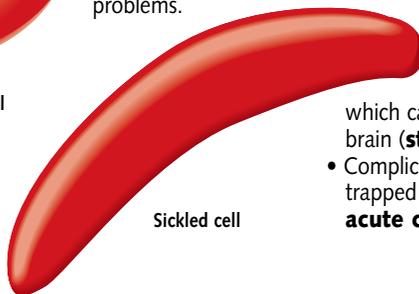
WHAT IS SICKLE CELL ANEMIA?

It is a blood disorder that causes **anemia** (shortage of red blood cells) and periodic pain. In people with sickle cell anemia, not all of their **hemoglobin** (a molecule in red blood cells that carries oxygen from the lungs to other parts of the body) works properly. Some of the hemoglobin forms long, rod-like structures that cause the red blood cells to be sickle-shaped and stiff. These cells can clog small blood vessels, preventing some organs or tissues from receiving enough oxygen. When this happens, it can cause

episodes of severe pain or damage to organs and tissues, and may lead to other serious medical problems.



Normal blood cell



Sickled cell

Sickle cell anemia is particularly common among people whose ancestors come from sub-Saharan Africa, South and Central America, Cuba, Saudi Arabia, India, Turkey, Greece, and Italy.

There is no cure for the disorder. Researchers, including those reporting in the May 12, 1999, issue of *JAMA*, continue to explore experimental techniques to enhance the chances that parents carrying sickle cell trait or sickle cell anemia can have children free from this debilitating disease.

SIGNS AND SYMPTOMS OF SICKLE CELL ANEMIA:

- Severe pain in bones, muscles, or the abdomen that can last for days or weeks (called **painful crisis**).
- Fatigue, paleness, and shortness of breath. All are symptoms of anemia, or a shortage of red blood cells.
- Pain that occurs unpredictably in any body organ or joint.
- Vision problems or blindness when the **retina** (back of the eye that receives visual images) fails to receive the nourishment it needs from circulating red blood cells.
- Yellowing of skin and eyes, caused by liver malfunction (**jaundice**).
- Delayed growth and puberty in children and a slight build in adults.
 - Higher vulnerability to infections.
 - Narrowed or blocked small blood vessels in the brain, which can cause damage to parts of the brain (**stroke**).
- Complications caused by infection or trapped sickle cells in the lung, called **acute chest syndrome**.

DIAGNOSIS:

Most states offer a simple blood test for sickle cell disease and/or sickle cell trait for all newborn infants that can be performed at the same time as other screening tests. Early diagnosis of sickle cell anemia is important so that children who have the disorder can receive proper treatment. The diagnosis in children and adults can be made with a simple blood test.

TREATMENT:

- Antibiotics help to prevent infections in infants, and painkillers (oral or intravenous), intravenous fluids, and breathing oxygen help to treat episodes of pain.
- Blood transfusions help to correct anemia by increasing the number of normal red blood cells in circulation.
- Other drugs and procedures are currently under investigation. **Hydroxyurea** may be beneficial in some adults but treatment in children is still under investigation.

FOR MORE INFORMATION:

- National Heart, Lung, and Blood Institute
NHLBI Information Center
301/592-8573
E-mail: nhlbiinfo@rover.nhlbi.nih.gov or www.nhlbi.nih.gov
- Sickle Cell Disease Association of America
800/421-8453 or sicklecelldisease.org

INFORM YOURSELF:

To find this and previous *JAMA* Patient Pages, check out the AMA's Web site at www.ama-assn.org/consumer.htm.

Additional Sources: National Heart, Lung, and Blood Institute, Emory University School of Medicine, March of Dimes

Mi Young Hwang, Writer

Richard M. Glass, MD, Editor

Jeff Molter, Director of Science News

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