



Educator's Guide to Sickle Cell Disease



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Sickle cell disease is an inherited blood disorder affecting about (1) one out of every 375 African Americans. Most children with sickle cell disease fall within the normal intelligence range and should be encouraged to reach their full potential. Advances in medical treatment have dramatically increased the lifespan of people with sickle cell disease. For this reason, it is even more important that children with sickle cell disease receive the full benefit of a primary, secondary, and college education. The goal of this booklet is to help educators understand sickle cell disease, its complications (health problems), and its treatment. This publication will also help educators assist children who have this condition to achieve their educational goals.

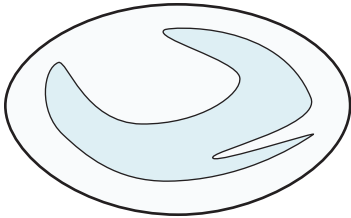
This document is not intended to take the place of the care and attention of your personal physician. Our aim is to promote active participation in your care and treatment by providing information and education. Questions about individual health concerns or specific treatment options should be discussed with your physician. For more general information on sickle cell disease, please visit our Web site at www.stjude.org/sicklecell.

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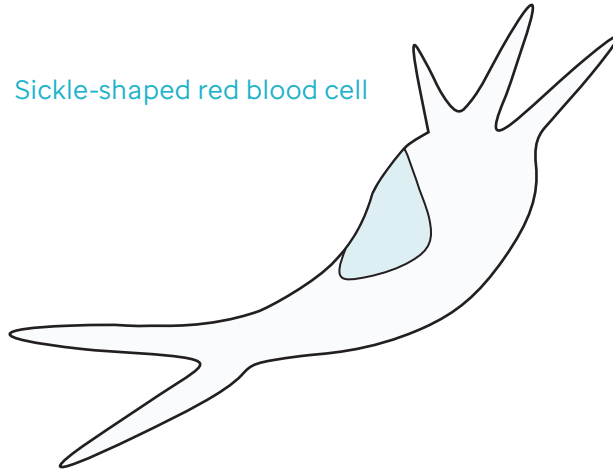
What is sickle cell disease?

Sickle cell disease is an inherited blood disorder affecting red blood cells. Normal red blood cells contain hemoglobin A. People with sickle cell disease have red blood cells containing mostly hemoglobin S, an abnormal type of hemoglobin. These red blood cells become sickle shaped (crescent shaped), and they have difficulty passing through small blood vessels. The most common types of sickle cell disease are hemoglobin SS, hemoglobin SC, and sickle-beta thalassemia.

Normal red blood cell

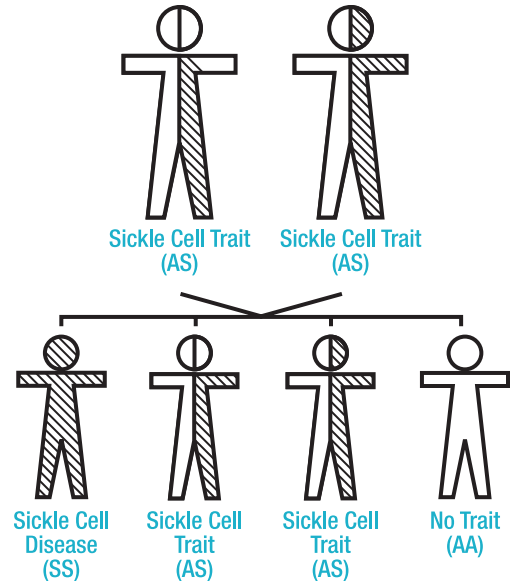


Sickle-shaped red blood cell



Who is affected by sickle cell disease?

- About one (1) out of every 375 African-American babies is born with sickle cell disease.
- Sickle cell disease affects about 70,000 people in the United States.
- People from Africa, Latin America, Asia, the Mediterranean, and other parts of the world can also have this disease.



How do people get sickle cell disease?

- Sickle cell disease is not spread like a cold and cannot be caught from another person.
- Sickle cell disease is an inherited (genetic) condition. It is inherited like eye color or hair color.
- Sickle cell trait is a carrier condition for sickle cell disease.
- People with sickle cell trait inherit one (1) gene for normal hemoglobin A and one (1) gene for abnormal hemoglobin S.
- People with the hemoglobin SS type of sickle cell disease inherit 2 hemoglobin S genes from their parents.
- If both parents have the sickle cell trait, they have a 25 percent (1-in-4) chance with each pregnancy of bearing a child with hemoglobin SS disease.

What are the most common complications of sickle cell disease?

- **Pain** – Pain episodes occur when sickle cells clog small blood vessels, depriving areas of the body of adequate blood and oxygen. Pain most often occurs in the arms, legs, chest, and abdomen. People with sickle cell disease are not affected equally by pain. Only 20 percent will have frequent pain episodes. Mild-to-moderate pain episodes can often be managed at home. Severe pain may require hospitalization and treatment with narcotic pain relievers. Drug dependency or addiction is rare when pain medications are used as prescribed.
- **Infection** – A common complication of sickle cell disease is abnormal function of the spleen. This results in an increased risk of infection in the blood, bones, lungs, and urinary tract.

What are some of the possible cognitive complications of sickle cell disease?

- **Increase risk of stroke** – When sickle cells clog small blood vessels in the brain, brain tissue can die from lack of oxygen. The symptoms of stroke depend on the part of the brain that is damaged.
- **Silent cerebral infarctions** – This occurs when brain tissue dies from lack of oxygen but does not cause symptoms. Normally, this is a risk factor for a stroke.
- **Abnormal school performance** The following areas can be affected:

- **Attention**
- **Executive functioning – planning, organization**
- **Language – receptive and expressive**
- **Visual-spatial and visual motor skills**
- **Working and short-term memory**
- **IQ**
- **Academic achievement**

What can teachers do to help students with sickle cell disease?

- Be aware that sickle cell complications may cause frequent absences from school.
- Provide parents with make-up work as needed. Discuss and explore possible needs for tutoring sessions.
- Communicate often with parents about academic performance, including excessive absences.
- Be aware of major changes in academic performance or behavior. These can be indicators of more serious sickle cell disease complications.
- Call parents right away if a student complains of significant pain, has a fever, or if a student develops weakness, numbness, or slurred speech.

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To help prevent pain episodes:

- Allow the child to take frequent trips to the water fountain and restroom.
- Encourage the child to take part in moderate exercise.
- Allow the student to rest when needed. A child with sickle cell disease may tire before others and might need to rest more often.
- Help keep the student's lifestyle as normal as possible.
- Help the child avoid temperature extremes (getting too hot or too cold).

Other important facts about sickle cell disease:

- A diagnosis of sickle cell disease can only be determined by a blood test.
- Comprehensive care includes early diagnosis, preventive measures, treatment of complications, and ongoing patient education.
- Many people with sickle cell disease live for long periods without having acute or severe symptoms.
- The average lifespan of people with sickle cell disease has increased from age 20 to the mid-40s and beyond.
- Individuals with sickle cell disease can pursue a variety of professions.
- Many adolescents with sickle cell disease experience delayed puberty (the average delay is about 2 years).
- Yellowing of the eyes is common in people with sickle cell disease. It should not be confused with hepatitis.
- The use of alcohol, “street” drugs, or tobacco can greatly increase the risk of developing serious complications.

Many students may have sickle cell trait.

Sickle cell trait is a carrier condition for sickle cell disease.

Facts about sickle cell trait:

- Sickle cell trait affects about one (1) out of every 12 African Americans.
- Sickle cell trait is inherited from one (1) parent.
- Sickle cell trait provides a “genetic window” into a family that may be at risk for having a child with sickle cell disease
- People with sickle cell trait have both normal hemoglobin A and abnormal hemoglobin S.
- Sickle cell trait can only be found with a blood test.
- It is believed that sickle cell trait first appeared long ago in areas of the world where people were sick with malaria. Usually, a person with sickle cell trait has increased protection against malaria.
- Sickle cell trait is not a disease. People with sickle cell trait do not develop sickle cell disease.
- People rarely have health problems caused by the sickle cell trait except under certain circumstances.



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