

# THE PEDIATRIC PATIENT LIVING WITH SICKLE CELL DISEASE

Sickle cell disease (SCD) is the most common genetic disorder in the United States, with about 100,000 American who have the disease and another 2 million who carry the trait. As people live with SCD for years, management of this disease and healthy living are important components of care, particularly for the pediatric population. Read below for more information about the diagnosis, treatment, and prognosis of SCD in pediatric patients.

This “Patient Education” tear sheet was produced in collaboration with the Children’s Sickle Cell Foundation, Inc. ([cscfkids.org](http://cscfkids.org)).

## What Is Sickle Cell Disease?

SCD is a group of inherited red blood cell disorders. Normal red blood cells are soft, round, and able to move through small blood vessels (veins and arteries) easily in the body to deliver oxygen. A person with SCD has red blood cells that, under certain conditions, become hard, sticky, and pointed. When they go through the blood tube, they clog the flow and break apart. This can cause pain, organ damage, and a low blood count. SCD occurs when a person inherits two abnormal hemoglobin genes from his or her parents. A person is born with abnormal “sickle hemoglobin,” and it is present for life.

## What Is Sickle Cell Trait?

Abnormal hemoglobin is inherited from parents who may be carriers of the sickle cell trait (SCT) or who have SCD. If an individual only inherits one sickle gene, it is SCT, while those who inherit two sickle cell genes have SCD. SCT is not a mild form of SCD. People with SCT usually do not have any of the symptoms of SCD and live a normal life.

## How Is Sickle Cell Disease Diagnosed?

Babies of all races should be screened for sickle cell hemoglobin at birth. A simple blood test, called the hemoglobin electrophoresis test, can be conducted to identify if a child is a carrier of the sickle cell trait or has the disease. Every state and the District of Columbia perform the sickle cell test at birth to help get babies born with SCD into care as soon as possible.

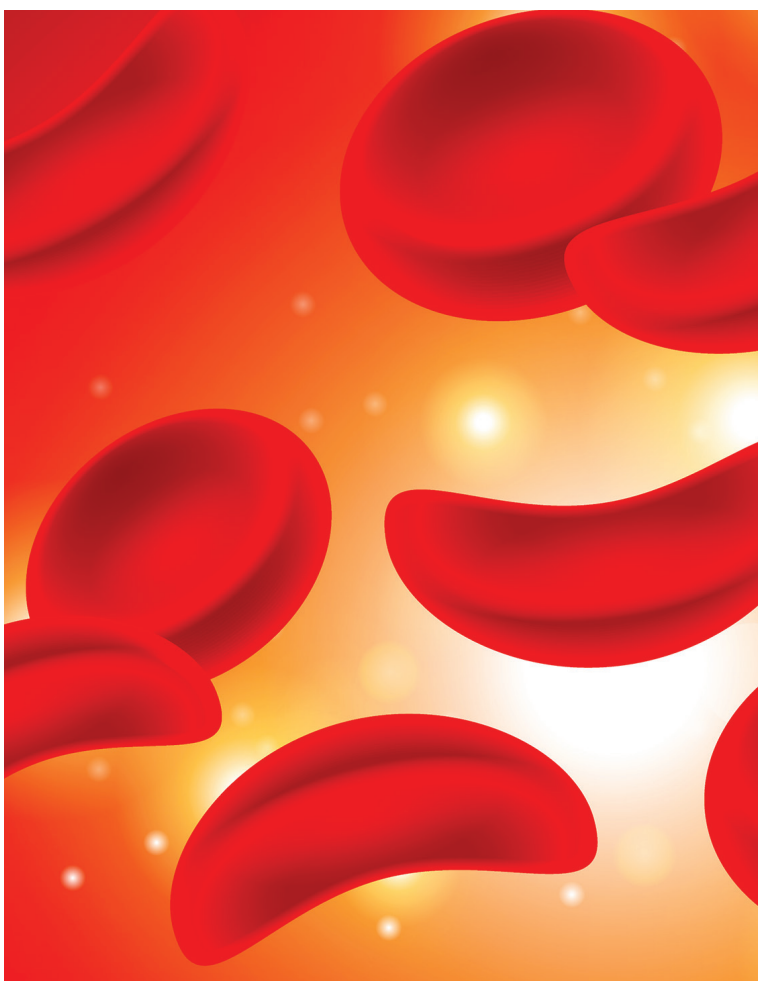
Other types of traits that may be identified on the test results include:

- hemoglobin S trait
- hemoglobin C trait
- hemoglobin E trait
- hemoglobin Barts, which indicates an alpha thalassemia trait
- beta thalassemia trait

## What Are the Complications Associated With Sickle Cell Disease?

Complications from the sickled cells blocking blood flow and breaking apart include:

- pain
- stroke
- increased risk of infection
- leg ulcers
- bone damage
- yellow eyes or jaundice
- early gallstones
- lung blockage
- kidney damage and loss of body water in urine
- eye damage
- low red blood cell counts (anemia)
- delayed growth



## Patient Education

**Patient Resources from the Children's Sickle Cell Foundation**

The Children's Sickle Cell Foundation, Inc., provides quality programs and services for children with SCD and their families to help them face the educational, social, and economic challenges caused by the disease. They provide educational support services for children with SCD, including educational advocacy, school-work retrieval while the child is absent from school, and loaner laptop computers to prevent children from falling behind when their illness interrupts their education. Both family and support programs are part of the Foundation's core values.

The following services are available to help families who have a child living with SCD:

- **Family Support Programs**  
[cscfkids.org/programs-and-services/family-support](http://cscfkids.org/programs-and-services/family-support)
- **Educational Support Programs**  
[cscfkids.org/programs-and-services/educational-support-program](http://cscfkids.org/programs-and-services/educational-support-program)
- **Transitioning to Adulthood**  
[cscfkids.org/living-with-sickle-cell/transitioning-to-adulthood](http://cscfkids.org/living-with-sickle-cell/transitioning-to-adulthood)

**How Is Sickle Cell Disease Treated?**

Every person with SCD should be under the care of a medical team that understands the disease. It is recommended that all newborn babies with SCD be treated with an antibiotic (penicillin) to prevent serious infections. All of the childhood immunizations should be given, as well as the pneumococcal vaccine. Parents must also know how to check for a fever because this signals the need for a quick medical check-up for a possible serious infection.

Patients and families will want to be mindful of the following conditions that may require an urgent medical evaluation:

- fever (101°F or higher)
- chest pain
- sudden, unusual headache
- shortness of breath
- increasing tiredness
- abdominal swelling
- any sudden weakness or loss of feeling
- pain that will not go away with home treatment
- priapism (painful erection that will not go away)
- sudden vision change

**Living with Sickle Cell Disease**

Living a healthy lifestyle is important for people with SCD. Minor illnesses may quickly turn into a medical emergency, so it is important for the person with SCD and his or her family members to keep a close eye on health status. People with SCD can live full lives and enjoy most of the activities that others do. The following tips are guidelines for living a safe and healthy lifestyle with SCD:

- **Find quality medical care:** Managing SCD takes a comprehensive team of doctors, nurses, and other health-care professionals working in collaboration. Every person with SCD should have a hematologist and a primary-care physician who knows the person's detailed medical history.
- **Make and keep doctor's appointments:** Consistent health check-ups with a primary-care physician can help prevent serious health problems. Every person with SCD should talk about any problems or pain he or she is experiencing.
- **Prevent infections:** Keep up-to-date on all vaccines, including the flu vaccine, pneumococcal vaccine, and any others recommended by a doctor. Washing hands with soap and clean water is a simple and essential way to prevent infections.
- **Take all medications, including penicillin, as prescribed.** Do not miss doses, and do not take more than recommended.
- **Stay hydrated and eat healthy foods:** It is critical that people with SCD stay hydrated. Eating balanced meals and making healthy choices are important. Avoiding food bacteria is also important; certain uncooked foods can carry harmful bacteria. Be sure to wash anything that comes into contact with contaminated food.
- **Avoid extreme temperatures:** Maintaining a balanced body temperature is very important, as patients with SCD have increased sensitivity to temperature extremes. Be sure to wear layers to adapt to changing temperatures.
- **Avoid reptiles:** The harmful Salmonella bacteria is present in some reptiles and is especially harmful to people with SCD. Children and adults should stay away from snakes, lizards, and turtles.

