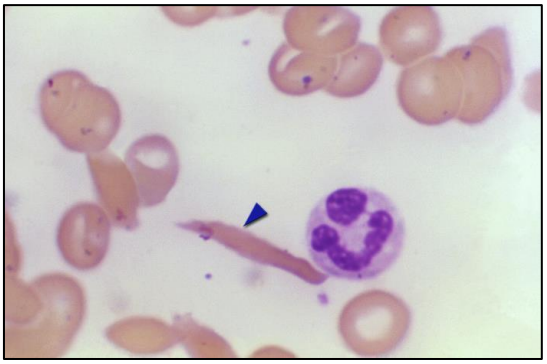
Priority Area – Sickle Cell Disease



Sickle-cell disease. CDC Dr. F. Gilbert

What is it? Sickle cell disease (SCD) is the most common inherited blood disorder. That means it's passed down through families. You're born with SCD. It is not something you catch or develop later in life.

The disease gets its name because SCD's red blood cells look like a sickle, which is a C-shaped farm tool. Red blood cells contain a molecule called hemoglobin, which carries oxygen through the body.

- In a healthy person, hemoglobin is smooth, round, and flexible allowing red blood cells to glide easily through your bloodstream.
- But in SCD, the hemoglobin's shape forms rods that clump together. That causes red blood cells to become rigid and curved.
- The odd-shaped cells block blood flow. It's dangerous, and can cause extreme pain, anemia, and other symptoms.

About 100,000 people in the United States have sickle cell disease. Most of them are African-Americans (WebMed).

- If only one parent passes the SCD gene on the child will have the sickle cell trait.
- With one normal and one defective form of the gene, people with the sickle cell trait generally don't have symptoms. But they are carriers.
- For a baby to be born with sickle cell anemia, both parents must carry a sickle cell gene (MayoClinic)

Signs and symptoms

SCD symptoms vary from person to person and change over time, including:

- **Anemia.** Sickle cells break apart and die, leaving a shortage of red blood cells (anemia) and oxygen leading to fatigue. Red blood cells usually live about 120 days before being replaced but sickle cells usually die in 10 to 20 days.
- **Episodes of pain.** SCD blocks blood flow to chest, abdomen, joints and bones causing pain. It varies in intensity and can last hours or weeks, a few times a year or many. Long-lasting pain can result from bone and joint damage, ulcers and other causes.
- **Painful swelling of hands and feet.** SCD may block blood flow to the hands and feet.
- **Frequent infections.** SCD can damage the spleen, which fights infections. Vaccinations and antibiotics help prevent infections, such as pneumonia.
- **Delayed growth.** A shortage of healthy red blood cells can slow growth and delay puberty.
- **Vision problems.** Tiny blood vessels to the eyes may become plugged with sickle cells. This can damage the retina, which processes visual images (MayoClinic).

Complications:

SCD can lead to many complications, including:

- **Stroke.** Sickle cells can block blood flow to the brain and cause seizures, weakness of arms and legs, speech difficulties and loss of consciousness. A stroke can be fatal.
- **Acute chest syndrome.** Sickle cells blocking blood vessels in the lungs can be caused by a lung infection and may require emergency antibiotics and medical treatment. It can be life-threatening and cause chest pain, fever and difficulty breathing.
- **Pulmonary hypertension.** SCD can develop high blood pressure in lungs (pulmonary hypertension) causing shortness of breath and fatigue, usually in adults not children.
- **Organ damage.** Sickle cells that block blood flow through blood vessels deprive the affected organ of blood and oxygen. In SCD, blood is also chronically low on oxygen which can damage nerves and organs, including kidneys, liver and spleen, which can be fatal.
- **Blindness.** Sickle cells can block blood supply to the eyes and lead to blindness.
- **Leg ulcers.** SCD can cause open sores, called ulcers, on your legs.
- **Gallstones.** Red blood cells breaking down produces bilirubin, which can lead to gallstones.
- **Priapism.** Men with SCD can have painful, long-lasting erections, called priapism due to sickle cells blocking blood vessels in the penis. This can damage the penis and lead to impotence (MayoClinic).

Resources

Support Services:

- I Care for Someone with Sickle Cell (CDC)
- Healthy Living with Sickle Cell Disease (CDC)
- Real Stories from People Living with Sickle Cell Disease (CDC)

Evidence-based resources related to Blood Disorders

- Evidence-Based Management of Sickle Cell Disease: Expert Panel Report, 2014
- Interventions for Patients and Caregivers to Improve Knowledge of Sickle Cell Disease and Recognition of its Related Complications
- Hydroxyurea (Hydroxycarbamide) for Sickle Cell Disease