

Sickle Cell Disease (SCD)

Sickle cell disease (SCD) is a life-long blood disorder that is passed down (inherited) from parents through genes. Sickle cell disease affects a part of the red blood cell called hemoglobin (hee-muh-**glow**-bn). It helps carry oxygen to different parts of the body. Normal red blood cells are round, smooth, and move through the body easily.

People with sickle cell disease have hard, sticky, banana-shaped red blood cells (Picture 1). These sickled cells have a hard time moving through small blood vessels. They can pile up and clog the vessels. When clogging happens, the blood cannot carry enough oxygen to certain parts of the body.

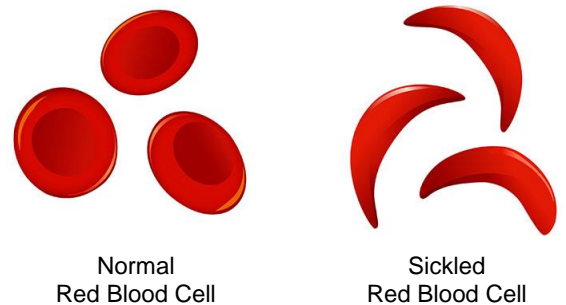


Image source: National Center on Birth Defects and Developmental Disabilities, Centers for Disease Control and Prevention

Picture 1 Sickled red blood cells are hard, sticky, and shaped like a banana.

Genetics of Sickle Cell Disease

Hemoglobin A is normal, non-sickled hemoglobin. **Hemoglobin S** is abnormal, sickled hemoglobin. Sickle cell disease is caused by hemoglobin S. All newborns are screened for both sickle cell disease and trait.

- If you inherit **1 normal, non-sickle gene (A)** and **1 sickle cell gene (S)**, you have sickle cell trait. You are a carrier for sickle cell disease. This does not mean you have sickle cell disease. If **both** parents pass down the hemoglobin S trait, their child will have hemoglobin SS. This is a type of sickle cell disease called sickle cell anemia.

AA (no sickled cells)	AS (sickle cell trait)	SS (sickle cell disease)
25% chance of having 2 normal genes	50% chance of having 1 normal gene (A), and 1 sickle gene (S)	25 % chance of having 2 sickle genes

- If 1 parent with sickle cell trait and 1 parent with abnormal hemoglobin have a child, that child could also have sickle cell disease. This abnormal hemoglobin can include beta thalassemia, hemoglobin C, or hemoglobin E.

Things That Affect Sickle Cell Disease

Sickle cell disease affects each person differently. There's no way to know how sick your child will be. Some things that affect it are:

- The type of sickle cell disease. Some types cause less problems than others.
- The quality of medical care your child gets.
 - Your child should go to a doctor or health care provider that specializes in sickle cell disease.
 - The medical practice should have a team of nurses, social workers, genetic counselors, and psychologists.
- Care at home. To keep your child healthy:
 - Have them drink a lot of water. For babies, formula and breast milk have enough water.
 - Make sure they take all medicines as prescribed.
- Outside care for your child. Explain to relatives, friends, and teachers what your child's health care needs are.
- Know when your child needs medical care for a fever. Seek help if your child is:
 - Less than 4 months with a fever at or over 100.4° Fahrenheit (F) or 38° Celsius (C).
 - Older than 4 months with a fever at or over 101°F or 38.3°C.

Problems Caused by SCD

- **Infections** are one of the most serious problems children with SCD can have.
 - Cause: Sickled cells can clog blood vessels which damage the spleen. The spleen's job is to help the body fight infections.
 - What you can do: Children 0 to 5 years old should take penicillin 2 times a day. Their immunization vaccines must be up-to-date. Remember a fever is a medical emergency and that your child needs to be seen right away for a fever at over 101°F or 38.3°C.
- **Pain** can come on suddenly, strongly, and without warning.
 - Cause: Clogged blood vessels block oxygen flow. Lack of oxygen can cause that part of the body to hurt.
 - What you can do: Have your child drink more fluids, use heat, massage the area, and give pain medicine like acetaminophen (Tylenol®) or ibuprofen (Motrin®, Advil®). Your child may have pain medicine prescribed by their provider. If pain can't be managed at home, it may need to be treated in the hospital with stronger medicine.

- **Anemia** can make your child get tired easily or sleep more soundly.
 - Cause: Sickled cells do not live as long as normal red blood cells. When there are less red blood cells, there is also less hemoglobin in the body. This is called anemia.
 - What you can do: Encourage frequent rest breaks and trying to follow a regular routine. Usually, the anemia does not need to be treated unless your child is not able to do their normal activities.

Surgery

If your child has any type of surgery, make sure doctors and health care providers know they have sickle cell disease.

- Call the sickle cell clinic when any type of surgery is scheduled.
- Your child may need extra blood tests before their surgery.
- Some children may need a blood transfusion if their hemoglobin level is low.

When to Call 911

Call 911 for emergency help if your child:

- Is breathing slowly or stops breathing.
- Is unresponsive and cannot talk to you.
- Cannot wake after a nap.
- Has sudden weakness, loss of feeling, or cannot move a body part.

If your child gets sick during the day, call the sickle cell nurses at (614) 722-8914 Monday through Friday, 8 a.m. to 4:30 p.m.

- If they are not available, please call the Sickle Cell Clinic at (614) 722-3250.
- On evenings, weekends, and holidays, call (614) 722-2000 and ask for the hematologist on call.