

Sickle Cell Disease

Questions and Answers

What is Sickle Cell Disease?

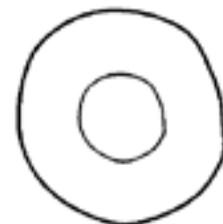
Sickle Cell Disease is an inherited condition that affects people of all ethnic backgrounds. This includes persons whose family came from Africa, the Caribbean, Latin America, Mediterranean or Middle Eastern areas, Southeast Asia or India.

Sickle Cell Disease affects the red blood cells. The red blood cells of people with Sickle Cell Disease contain an abnormal type of hemoglobin. There are three common types of Sickle Cell Disease in the United States:

1. Hemoglobin SS
2. Hemoglobin SC
3. Hemoglobin sickle beta-thalassemia

What is Hemoglobin?

The red material in the blood is called hemoglobin. The hemoglobin's job is to pick up oxygen from the lungs and carry it to different parts of the body. Normal red blood cells are shaped like a "doughnut". The normal red blood cells are soft and travel easily through tiny blood vessels and capillaries. They contain two genes for "A" type of hemoglobin.



The red blood cells of people who have Sickle Cell Disease contain an abnormal type of hemoglobin. The red blood cells are shaped like a "sickle or banana". The cells are rigid and do not travel easily through tiny blood vessels and capillaries. They contain two genes for "S" type

This handout is for informational purposes only. Talk with your doctor or health care team if you have any questions about your care.

of hemoglobin. The sickle-shaped red blood cells may clump together. When they clump, they clog small blood vessels. This causes periods of pain and cuts off oxygen to tissue and organs.



What is Sickle Cell Anemia?

Sickle Cell Anemia is the most common type of Sickle Cell Disease. Sickle Cell Anemia is a blood disorder in which there is an abnormality in red blood cells and hemoglobin. This is caused by the genes that are inherited from parents.

What is Sickle Cell Trait?

Sickle Cell Trait happens when a person gets one “S” hemoglobin gene from one parent and one “A” hemoglobin gene from the other parent.

People with Sickle Cell Trait have enough normal hemoglobin in their red blood cells to prevent them from sickling and to keep the red blood cells round in shape. There is usually no health problems associated with Sickle Cell Trait. People with Sickle Cell Trait can have problems in high altitudes or with extreme exertion. A person who carries genes for the Sickle Cell Trait does not develop Sickle Cell Anemia.

How do I know if I have the Sickle Cell Trait?

Your doctor can perform a special blood test to tell if you have Sickle Cell Anemia or carry the Sickle Cell Trait. You may decide to have this test done before you plan to have children.

In many states, the law requires babies to be tested right away for Sickle Cell Disease. This allows children born with Sickle Cell Disease to receive treatment to protect them from life threatening infections.

What is the chance a child will be born with Sickle Cell Anemia or Sickle Cell Trait?

- If both parents have the gene for Sickle Cell Trait, there is a 25% chance that their child will have Sickle Cell Anemia.
- If one parent has the Sickle Cell Trait and one parent has normal hemoglobin there is a 50% chance that their child will have the Sickle Cell Trait. In this case a child cannot inherit the genes for Sickle Cell Anemia, but the trait could be passed along.
- If one parent has Sickle Cell Disease and one parent has the sickle cell trait, there is a 50% chance that their child will have Sickle Cell Anemia.

What should I know about pregnancy and Sickle Cell Disease?

- Pregnant women with Sickle Cell Disease have a higher number of miscarriages (also called “spontaneous abortions”). A miscarriage may happen up to 25% of the time.
- A higher number of pregnancies (8% to 10% of the time) may result in the baby not surviving at birth (also called “stillborn”).
- The baby’s birth weight can be lower than average. About 1/3 of the babies born to mothers who have Sickle Cell Disease weigh less than 5.4 pounds.
- Infection is more common in women with Sickle Cell Disease during pregnancy - especially bladder infections.
- There is an increased chance of a condition called “pre-eclampsia” (also known as PIH or Toxemia) which can be dangerous for the mother and baby.
- It is more common to have a painful Sickle Cell Crisis during pregnancy.

Here are some of the things to expect when you see your doctor during pregnancy. The doctor may have you:

- Get a pneumonia vaccine
- Get a flu vaccine
- Eat well balanced meals
- Take a folate supplement (vitamin), 4 mg per day
- Have your blood iron levels checked every month
- Have a urine culture done each trimester (every 12 weeks)
- Begin non-stress testing in the third trimester (about 28 to 32 weeks)
- Get frequent ultrasounds to check the growth of the baby

Signs, Symptoms and Complications of Sickle Cell Disease

The following problems can occur for people who have Sickle Cell Disease:

- Leg ulcers
- Frequent infections
- Swelling in the hands and feet
- Jaundice – yellowish color on the whites of eyes
- Painful joints especially shoulders and hips
- Blood clots
- Anemia (feel tired or fatigue easily)
- Sickle Cell Crisis - severe pain in chest, abdomen, arms and legs
- Delayed growth

Complications are mostly caused from the sickle-shaped red blood cell blocking blood vessels. This can result in:

- Stroke
- Bone damage
- Kidney damage
- Breathing problems
- Painful erections in the penis
- Abdominal pain from blood blockage in the spleen or liver
- Eye damage

Treatment

Common treatments that are used to help with problems of Sickle Cell Disease are:

- Medicine for pain
- Use heat to lessen pain
- Medicine to treat infections (antibiotics)
- Apheresis treatment (also called pheresis or Red Blood Cell exchange)
- Blood transfusions
- Bed rest
- Surgery to repair damaged joints in the hips and shoulders
- Non-drug pain relief such as imagery (a way of picturing a thought or image that will distract you from your pain) and stress management
- In some cases patients may need:
 - ▶ To take a medicine called hydroxyurea to help control the disease
 - ▶ To have bone marrow transplant

What can be done to help prevent Sickle Cell Crisis?

- Drink 6 to 8 glasses of fluid each day.
- Do not smoke.
- Avoid getting chilled. Keep warm in winter and avoid getting chilled by air conditioning in the summer. Try to keep your body warm. Avoid swimming in cold water
- Keep immunizations up to date including the flu shot and pneumonia vaccine.
- Avoid high altitudes such as mountains.
- Avoid traveling on airplanes that are not pressurized.
- Avoid stressful situations. Learn good ways to manage stress.
- Maintain good general health by:
 - ▶ Eating a balanced diet
 - ▶ Getting enough rest
 - ▶ Doing moderate exercise

For More Information Contact:

- Ohio Sickle Cell & Health Association, Inc.
341 South Third Street, Suite 200
Columbus, Ohio 43215
 - ▶ Call: 614-228-0157
 - ▶ Visit: ohiosicklecell.org
 - ▶ Connect: https://www4.gvtsecure.com/reg_scdaa/

- Joint Center for Sickle Cell and Thalassemic Disorders
 - ▶ Harvard University at <http://sickle.bwh.harvard.edu>
 - ▶ Sickle Cell Information Center at www.scinfo.org