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# Sickle Cell Disease

## The Health Education Network

The Health Education Network is a Program of the Minority Organ Donation Education Program, Inc.

## Learning About Sickle Cell Disease

Sickle cell disease is the most common inherited blood disorder in the United States. Approximately 80,000 Americans have the disease.

In the United States, sickle cell disease is most prevalent among African-Americans. About one in 12 African-Americans and about one in 100 Hispanic Americans carry the sickle cell trait, which means they are carriers of the disease.

Sickle cell disease is caused by a mutation in the hemoglobin-Beta gene (red blood cells) chromosome.

Hemoglobin (red blood cells) transports oxygen from the lungs to other parts of the body. Red blood cells with normal hemoglobin (hemoglobin-A) are smooth and round and glide through blood vessels.

In people with sickle cell disease, abnormal hemoglobin molecules (blood cells) stick to one another and form long, rod-like structures. These structures cause red blood cells to become stiff, becoming a sickle shape. Their shape causes these red blood cells to pile up, causing blockages and damaging vital organs and tissue.

Sickle cells are destroyed rapidly in the bodies of people with the disease, causing anemia. This anemia is what gives the disease its commonly known name - sickle cell anemia.

The sickle cells also block the flow of blood through vessels, resulting in lung tissue damage that causes acute chest syndrome, pain and stroke. It also causes damage to the spleen, kidneys and liver.

A baby born with sickle cell disease inherits a gene for the disease from both parents. When both parents have the genetic defect, there's a 25 percent chance that each child will be born with sickle cell disease.

If a child inherits only one copy of the defective gene (from either parent), there is a 50 percent chance that the child will carry the sickle cell trait (not the full disease). People who only carry the sickle cell trait usually don't get the disease, but can pass the defective gene on to their children.

### Is there a test for sickle cell disease?

Doctors diagnosis sickle cell through a blood test that checks for the defective form of hemoglobin. To confirm the diagnosis, a sample of blood is examined under a microscope to check for large numbers of sickled red blood cells.

In more than 40 states, testing for the defective sickle cell gene is routinely performed on newborns. Sickle cell disease can also be detected in an unborn baby. Amniocentesis, a procedure in which a needle is used to take fluid from around the baby for testing, can show whether the fetus has sickle cell disease or carries the sickle cell gene.

### Treatment:

Treatments for sickle cell include antibiotics, pain management and blood transfusions.

### The Only Cure:

### Bone Marrow Transplantation:

Currently the only cure for sickle cell disease is bone marrow transplantation. In this procedure a sick patient is transplanted with bone marrow from healthy, genetically compatible sibling donors. However only about 18 percent of children with sickle cell disease have a healthy, matched sibling donor. Bone marrow transplantation is a risky procedure with many complications.

**Source: National Institute of Health.**