

MANAGING YOUR SICKLE CELL DISEASE

What Is Sickle Cell Disease?

Sickle cell disease is a hereditary blood disease (passed from parents to children). It affects red blood cells. Normal red blood cells are round and contain hemoglobin. Hemoglobin helps carry oxygen to parts of the body. People with sickle cell disease have red blood cells with abnormal hemoglobin-S (Hb-S). Red blood cells become crescent or sickle shaped and cannot pass through small blood vessels. Vessels get blocked, tissues and organs may become damaged because they cannot get enough blood, and acute illness (crisis) results. Problems can affect every organ.

People may have sickle cell trait. Sickle cell trait means having one sickle cell gene and one normal gene, so both abnormal and normal hemoglobin are made. People with the trait are healthy but can have children with sickle cell disease if their sexual partner also has sickle cell trait.

More than 50,000 people in the United States have the disease, which is lifelong.

What Causes Sickle Cell Disease?

The cause is a mutation in DNA for beta-globin, a protein that makes up hemoglobin. The disease is most common among African Americans but people of Mediterranean, Caribbean, South and Central American, Arabian, and East Indian descent can have it.

What Are the Symptoms of Sickle Cell Disease?

Common symptoms include chronic anemia (low blood cell count), fast heartbeat, yellow skin (jaundice), tiredness, and severe bone pain during a sickle cell crisis. Symptoms get worse during a crisis (sickle cell crisis is sudden pain occurring throughout the body). Pregnancy, surgery, infection, and high altitudes make symptoms worse. Complications include kidney and eye

disease, heart disease, leg ulcers, and strokes. Infections such as osteomyelitis (bone infection) and pneumonia (acute chest syndrome with cough and shortness of breath) can occur. In aplastic crisis, bone marrow stops making red blood cells.

How Is Sickle Cell Disease Diagnosed?

The health care provider makes a diagnosis from a family history of the disease and blood tests. Blood tests look for sickle-shaped red blood cells and abnormal hemoglobin called hemoglobin S which is the defective form of hemoglobin underlying sickle cell anemia. A simple screening test in early pregnancy or after birth can check for the disease in a baby.

How Is Sickle Cell Disease Treated?

Treatment involves controlling pain. Oral medicine can help mild crises. Severe pain may need intramuscular or intravenous narcotics. Hydroxyurea is a medication that can reduce the number of crises. Fluids are given to prevent dehydration. Sometimes, blood transfusion is needed. Abnormal blood cells are replaced with normal cells.

Children usually receive penicillin to prevent bacterial infection.

Bone marrow transplantation from a brother or sister may cure the disease, but this treatment is experimental.

DOs and DON'Ts in Managing Sickle Cell Disease

- ✓ **DO** drink fluids to avoid dehydration.
- ✓ **DO** eat a healthy diet with plenty of green, leafy vegetables (high in folate). Take a daily folate supplement.
- ✓ **DO** mild to moderate exercise. Rest if you feel tired and drink enough fluids.
- ✓ **DO** consider genetic counseling to check your risk for having an affected child.

- ✓ **DO** get prenatal care from your obstetrician and primary care health care provider.
- ✓ **DO** call your health care provider if you have fever, shortness of breath, severe abdominal pain, or neurological symptoms (such as trouble speaking).
- ✓ **DO** get appropriate vaccinations as recommended by your physician.
- ⊗ **DON'T** fly on airplanes without pressurized cabins.
- ⊗ **DON'T** overuse pain drugs.
- ⊗ **DON'T** drink alcohol in excess.

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FOR MORE INFORMATION

Contact the following sources:

- Sickle Cell Disease Association of America, Inc.: Tel: (800) 421-8453; Website:
<http://www.sicklecelldisease.org>
- National Heart, Lung, and Blood Institute Information Center: Tel: (301) 592-8573;
Website: <http://www.nhlbi.nih.gov>

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