Health Basics

Sickle Cell Anemia

What is sickle cell anemia?
Sickle cell anemia is an inherited form of anemia—a condition in which the body doesn't produce enough healthy red blood cells or hemoglobin, a protein in red blood cells that carries oxygen through the body. Healthy red blood cells are round and flexible, but those of someone with sickle cell anemia are rigid and sticky and shaped like crescent moons. This irregular shape causes the cells to get stuck in small blood vessels, and it slows or blocks the flow of blood and oxygen to the body, resulting in numerous problems. For most people, there's no cure for sickle cell anemia. But the condition can be managed with pain-relieving treatments that also work to help prevent further problems associated with the condition.

According to the Centers for Disease Control and Prevention, sickle cell affects approximately 100,000 Americans. In the United States, most people with sickle cell are Black. It occurs among about 1 out of every 365 African-American births and about 1 in 13 Black babies is born with sickle cell trait.

What are the symptoms of sickle cell anemia?
Signs of sickle cell anemia usually appear after an infant is 4 months old. Some of the symptoms may include anemia, periodic episodes of pain (called crises), swollen hands and feet (called hand-foot syndrome), frequent infections, delayed growth and vision problems.

What causes sickle cell anemia?
Sickle cell anemia is caused by a gene mutation that tells your body to make abnormal hemoglobin. (Hemoglobin is the oxygen-carrying, iron-rich protein pigment in the blood.) Abnormal hemoglobin causes red blood cells to become rigid, sticky and misshapen. The sickle cell gene is passed from generation to generation, but for a child to develop sickle cell, both biological parents must pass on the defective gene. If only one parent passes the sickle cell gene to the child, that child will be a carrier of the disease without developing the condition. The gene is more common in families from Africa, India, the Mediterranean, Saudi Arabia, and South and Central Americas. In the United States, it most often affects blacks and Hispanics.

What are the complications of sickle cell anemia?
Sickle cell anemia can create the following medical problems:

> Stroke, caused when blood is blocked to an area of the brain; strokes are characterized by seizures, weakness or numbness of your arms and legs, sudden speech difficulties and loss of consciousness
> Acute chest syndrome, a life-threatening condition that causes chest pain, fever and difficulty

breathing
Pulmonary hypertension, high blood pressure in the lungs that causes difficulty breathing and possibly fatal shortness of breath
Organ damage, caused by the sickle cells blocking blood flow through blood vessels, which stops blood and oxygen from reaching an organ
Blindness, caused by damage to the retina—the part of the eye that processes visual images
Leg ulcers, open sores on the legs
Gallstones, usually found in the gallbladder when the hemoglobin from destroyed red blood cells forms a chemical called bilirubin. This chemical can then combine with calcium and fat to form pebble-like gallstones.
Priapism, painful, long-lasting erections, which can occur when sickle cells block the blood vessels in the penis. This can damage the penis and eventually lead to impotence.

How is sickle cell anemia diagnosed?

Genetic screening: This is done when a baby is born. Your family doctor or pediatrician gets these test results. Once the condition is diagnosed, parents usually get a referral to a doctor who specializes in blood disorders (hematologist) or a pediatric hematologist.
Blood testing: This screening checks for the presence of hemoglobin S—the abnormal form of hemoglobin that causes sickle-cell anemia. In the United States, this blood test is part of routine newborn screening done at the hospital. But older children and adults can be tested, too. If the blood test is negative, there is no sickle-cell gene present. But if the test is positive, doctors order further tests to determine whether one or two sickle-cell genes are present. People who have one gene are diagnosed as having sickle-cell trait only. People with two genes are diagnosed as having sickle-cell disease.
Amniotic fluid sampling: This test allows doctors to diagnose sickle cell anemia in an unborn baby. The test involves doctors taking a sample of the liquid (called amniotic fluid) surrounding the baby in their mother's womb to look for the sickle cell gene. Doctors recommend this test if a prospective parent has sickle cell anemia or sickle-cell trait.

How is sickle cell anemia treated?

Bone marrow transplant: This is the only potential cure for sickle cell anemia, but finding a donor is difficult and the procedure has serious risks including death.
Drug therapy: Medications, such as antibiotics, pain-relievers and hydroxyurea (Droxia, Hydrea), can reduce pain and lessens frequency of sickle cell crises.
Blood transfusions: These procedures help increase the number of normal red blood cells in circulation and decrease the risk of stroke in children with sickle cell anemia. But regular blood transfusions can cause excess iron to build up in the body. People with sickle cell anemia who have regular transfusions may need treatment to reduce iron levels to avoid damage to their heart, liver and other organs.
Lifestyle and behavior modifications: Maintaining a healthy diet, drinking plenty of water, using supplements, participating in regular sessions of mild to moderate exercise and reducing stress all help people with sickle cell anemia avoid complications.

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