Sickle cell anemia

Overview

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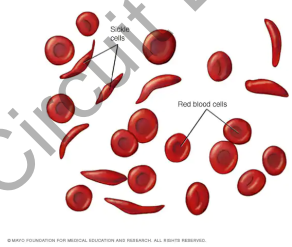
Sickle cell anemia is one of a group of disorders known as sickle cell disease. Sickle cell anemia is an inherited red blood cell disorder in which there aren't enough healthy red blood cells to carry oxygen throughout your body.



Sickle cell anemia

Normally, the flexible, round red blood cells move easily through blood vessels. In sickle cell anemia, the red blood cells are shaped like sickles or crescent moons. These rigid, sticky cells can get stuck in small blood vessels, which can slow or block blood flow and oxygen to parts of the body.

There's no cure for most people with sickle cell anemia. But treatments can relieve pain and help prevent complications associated with the disease.

Symptoms

Signs and symptoms of sickle cell anemia usually appear around 5 months of age. They vary from person to person and change over time. Signs and symptoms can include:

- **Anemia.** Sickle cells break apart easily and die, leaving you with too few red blood cells. Red blood cells usually live for about 120 days before they need to be replaced. But sickle cells usually die in 10 to 20 days, leaving a shortage of red blood cells (anemia).

  Without enough red blood cells, your body can't get enough oxygen, causing fatigue.

- **Episodes of pain.** Periodic episodes of pain, called pain crises, are a major symptom of sickle cell anemia. Pain develops when sickle-shaped red blood cells block blood flow through tiny blood vessels to your chest, abdomen and joints. Pain can also occur in your bones.

  The pain varies in intensity and can last for a few hours to a few weeks. Some people have only a few pain crises a year. Others have a dozen or more pain crises a year. A severe pain crisis requires a hospital stay.

  Some adolescents and adults with sickle cell anemia also have chronic pain, which can result from bone and joint damage, ulcers, and other causes.

- **Swelling of hands and feet.** The swelling is caused by sickle-shaped red blood cells blocking blood flow to the hands and feet.

- **Frequent infections.** Sickle cells can damage your spleen, leaving you more vulnerable to infections. Doctors commonly give infants and children with sickle cell anemia vaccinations and antibiotics to prevent potentially life-threatening infections, such as pneumonia.

- **Delayed growth or puberty.** Red blood cells provide your body with the oxygen and nutrients needed for growth. A shortage of healthy red blood cells can slow growth in infants and children and delay puberty in teenagers.

- **Vision problems.** Tiny blood vessels that supply your eyes can become plugged with sickle cells. This can damage the retina — the portion of the eye that processes visual images — and lead to vision problems.

When to see a doctor

Sickle cell anemia is usually diagnosed in infancy through newborn screening programs. If you or your child develops any of the following problems, see your doctor right away or seek emergency medical care:

- **Fever.** People with sickle cell anemia have an increased risk of serious infection, and fever can be the first sign of an infection.
- **Unexplained episodes of severe pain,** such as pain in the abdomen, chest, bones or joints.
- **Swelling in the hands or feet.**
- **Abdominal swelling,** especially if the area is tender to the touch.
- **Pale skin or nail beds.**
- **Yellow tint** to the skin or whites of the eyes.
- **Signs or symptoms of stroke.** If you notice one-sided paralysis or weakness in the face, arms or legs; confusion; trouble walking or talking; sudden vision changes or unexplained numbness; or a severe headache, call 911 or your local emergency number right away.

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Causes

Sickle cell anemia is caused by a mutation in the gene that tells your body to make the iron-rich compound that makes blood red and enables red blood cells to carry oxygen from your lungs throughout your body (hemoglobin). In sickle cell anemia, the abnormal hemoglobin causes red blood cells to become rigid, sticky and misshapen.

Both mother and father must pass the defective form of the gene for a child to be affected.

If only one parent passes the sickle cell gene to the child, that child will have the sickle cell trait. With one normal hemoglobin gene and one defective form of the gene, people with the sickle cell trait make both normal hemoglobin and sickle cell hemoglobin.

Their blood might contain some sickle cells, but they generally don't have symptoms. They're carriers of the disease, however, which means they can pass the gene to their children.

Risk factors

For a baby to be born with sickle cell anemia, both parents must carry a sickle cell gene. In the United States, sickle cell anemia most commonly affects black people.

Complications

Sickle cell anemia can lead to a host of complications, including:

- **Stroke.** Sickle cells can block blood flow to an area of your brain. Signs of stroke include seizures, weakness or numbness of your arms and legs, sudden speech difficulties, and loss of consciousness. If your child has any of these signs and symptoms, seek medical treatment immediately. A stroke can be fatal.
- **Acute chest syndrome.** A lung infection or sickle cells blocking blood vessels in your lungs can cause this life-threatening complication, resulting in chest pain, fever and difficulty breathing. It might require emergency medical treatment.
- **Pulmonary hypertension.** People with sickle cell anemia can develop high blood pressure in their lungs. This complication usually affects adults. Shortness of breath and fatigue are common symptoms of this condition, which can be fatal.
- **Organ damage.** Sickle cells that block blood flow to organs deprive the affected organs of blood and oxygen. In sickle cell anemia, blood is also chronically low in oxygen. This lack of oxygen-rich blood can damage nerves and organs, including your kidneys, liver and spleen, and can be fatal.
- **Blindness.** Sickle cells can block tiny blood vessels that supply your eyes. Over time, this can damage your eye and lead to blindness.
- **Leg ulcers.** Sickle cell anemia can cause open sores on your legs.
- **Gallstones.** The breakdown of red blood cells produces a substance called bilirubin. A high level of bilirubin in your body can lead to gallstones.
- **Priapism.** In this condition, men with sickle cell anemia can have painful, long-lasting erections. Sickle cells can block the blood vessels in the penis, which can lead to impotence over time.
- **Pregnancy complications.** Sickle cell anemia can increase the risk of high blood pressure and blood clots during pregnancy. It can also increase the risk of miscarriage, premature birth and having low birth weight babies.

Prevention

If you carry the sickle cell trait, seeing a genetic counselor before trying to conceive can help you understand your risk of having a child with sickle cell anemia. They can also explain possible treatments, preventive measures and reproductive options.

By Mayo Clinic Staff

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Diagnosis & treatment

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