Sickle cell anemia

Sickle cell anemia is one of a group of inherited disorders known as sickle cell disease. It affects the shape of red blood cells, which carry oxygen to all parts of the body. Red blood cells are usually round and flexible, so they move easily through blood vessels.

In sickle cell anemia, some red blood cells are shaped like sickles or crescent moons. These sickle cells also become rigid and sticky, which can slow or block blood flow.

The current approach to treatment is to relieve pain and help prevent complications of the disease. However, newer treatments may cure people of the disease.

Symptoms

Symptoms of sickle cell anemia usually appear around 6 months of age. They vary from person to person and may change over time. Symptoms can include:

- **Anemia.** Sickle cells break apart easily and die. Typical 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. This is known as anemia. Without enough red blood cells, the body can't get enough oxygen. This causes fatigue.
- **Episodes of pain.** Periodic episodes of extreme 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 the chest, abdomen and joints.

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

Some people with sickle cell anemia also have chronic pain from bone and joint damage, ulcers, and other causes.

- **Swelling of hands and feet.** Sickle-shaped red blood cells block blood circulation in the hands and feet, which can cause them to swell.
- **Frequent infections.** The spleen is important for protecting against infections. Sickle cells can damage the spleen, raising the risk of developing infections. Babies and children with sickle cell anemia commonly receive vaccinations and antibiotics to prevent potentially life-threatening infections, such as pneumonia.
- **Delayed growth or puberty.** Red blood cells provide the body with the oxygen and nutrients needed for growth. A shortage of healthy red blood cells can slow growth in babies and children and delay puberty in teenagers.
- **Vision problems.** Tiny blood vessels that supply blood to the eyes can become plugged with sickle cells. This can damage the portion of the eye that processes visual images, called the retina, and lead to vision problems.

Causes

Sickle cell anemia is caused by a change in the gene that tells the body to make hemoglobin. Hemoglobin is the iron-rich compound in red blood cells that allows these cells to carry oxygen from the lungs to the rest of the body. The hemoglobin associated with sickle cell anemia causes red blood cells to become rigid, sticky and misshapen.

Diagnosis

1-Complet blood test

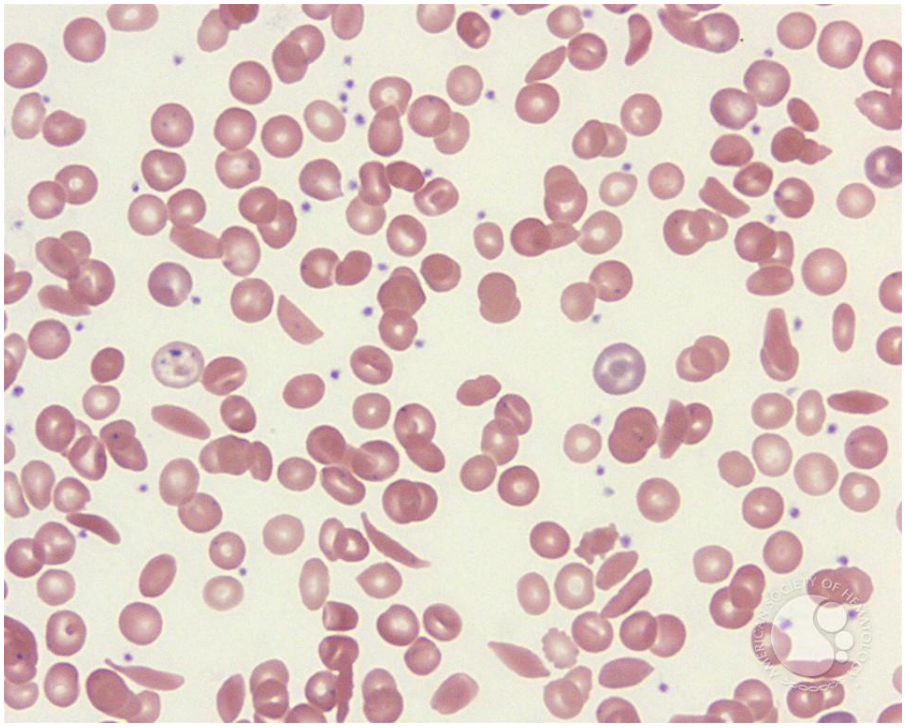
A blood test can check for the form of hemoglobin that underlies sickle cell anemia.

In adults, a blood sample is taken from a vein in the arm. In young children and babies, the blood sample is usually collected from a finger or heel. The sample then goes to a laboratory to be screened for the sickle cell form of hemoglobin.

2-Complet blood picture

3-Genetic tests

Genetic testing can help determine which type of sickle cell disease you have or confirm a diagnosis if results from blood tests are not clear. Genetic testing can also tell whether you have one or two copies of the sickle hemoglobin gene.



Peripheral smear from a patient with sickle cell disease illustrates the spectrum of RBC findings in this disorder including sickle cells, polychromatophilic RBCs, target cells, and Howell-Jolly bodies.