Sickle Cell Anemia

Sickle cell anemia is a severe form of sickle cell disease (SCD), a group of inherited red blood cell disorders. Sickle cell anemia is a blood disorder that causes your red blood cells (RBCs) to have abnormal hemoglobin and an abnormal shape. Normal RBCs are disc-shaped. In sickle cell anemia, however, the RBCs attain a crescent (an old farm tool), or sickle shape. The abnormal sickled RBCs also become hard and sticky. These sickle cells can't carry oxygen as well as normal, round blood cells.

Sickle cell anemia runs in families (inherited). It often affects people of African descent, including African-Americans as well as certain other ethnic groups. You get it from a changed (mutated) gene passed down from each parent. The abnormal gene causes problems with your hemoglobin, a protein in RBCs that carries oxygen throughout the body. The abnormal hemoglobin stiffens into rod-like structures inside the RBCs which alters their shape. There's also something called sickle cell trait. This happens when you get 1 mutated gene from 1 of your parents. Most newborns are now tested for sickle cell disease at birth.

Sickle cell trait usually doesn't cause symptoms and isn't serious. Sickle cell anemia and sickle cell trait are not contagious. This means it can't be passed from person to person by coughing or touching.

Until recently, SCD could be cured only with a bone marrow transplant. However, new gene therapies for SCD have been approved. Talk to your provider (or your child's provider) about these treatments.

People with sickle cell anemia may have several complications during their life. One is an acute pain crisis (also known as a sickle cell crisis or vaso-occlusive crisis). This happens when many sickle cells stick together and pile up in the blood vessels. This may block blood flow and oxygen delivery to the tissues. During a sickle cell crisis, you may have severe pain in the chest, lower back, abdomen, arms, and legs. A crisis may not have a known cause or may be triggered by dehydration, infection, stress, high altitudes, and temperature changes. The crisis can last for hours, or even days. It can happen several times a year.

Home care

Tips for taking care of yourself at home include:

- Watch for sores (ulcers) on your legs. These are caused by poor blood flow. They're a sign that the sickle cell anemia isn't under control.
- If you snore or have trouble breathing when lying down or during sleep, tell your healthcare provider.
- Get treatment for any other health condition, such as diabetes. This is important to prevent complications of sickle cell anemia.
- Get early prenatal care if you're pregnant or plan to get pregnant.
- If you plan to travel by air, go in pressurized aircraft only. Check with your healthcare provider about any needed safety steps if you must travel in a nonpressurized aircraft.
- Talk to your healthcare provider about what kind of pain medicine you can use.
- Drink plenty of water. This is even more important during warm weather.
- Get treated for any infection as soon as it happens. This includes cold, flu, and skin infections.
- Wear warm clothes in cold weather or in air-conditioned rooms.
- If you know which factors trigger acute pain crises, work with your healthcare provider to plan how to prevent and manage them.

Lifestyle changes

Suggestions for changes include:

- Limit alcohol intake to no more than 1 drink per day.
- Stop smoking. Go to a stop-smoking program to improve your chances of quitting.
- Exercise regularly but not to the point that you become extremely tired. Drink plenty of fluids when you exercise.
- Don't do very strenuous activities, such as rough contact sports. Ask your healthcare provider what type and level of physical activity if best for you.
- Get good quality sleep every day of at least 7 to 9 hours.
- Learn ways to manage your stress, emotions and cope with problems.
- Avoid sudden extreme changes in temperatures - both cold and heat.
- Choose a healthy diet that includes a variety of fruits, vegetables, and whole grains.

Follow-up care

Make a follow-up appointment as advised. Regular follow-up visits are very important. Know the best way to contact your healthcare provider, including outside clinic hours, weekends, and holidays.

When to call your healthcare provider

Call your healthcare provider right away if you have any of these:

- Swollen hands or feet
- Sudden paleness in the skin or nail beds
- Yellow color of the skin or eyes (jaundice)
- Fever, or any other sign of infection
- Belly (abdomen) swelling
- Unable to drink fluids to keep hydrated
- Sudden tiredness with no interest in what's going on
- Erection that won't go away or is painful
- Trouble hearing or seeing
- Weakness on 1 side of the body
- Sudden change in speech
- Headache
- Trouble breathing
- Joint, abdomen , chest, lower back, or muscle pain
- New skin sores
- Limping