

What is sickle cell disease? — Sickle cell disease is a condition that affects the red blood cells. There are a few different types of sickle cell disease. A common type is called "sickle cell anemia."

The red blood cells carry oxygen to organs in the body. Normal red blood cells are round. When people have sickle cell disease, some of their red blood cells can have an abnormal shape. They look like a crescent (or like a tool called "sickle"). The abnormally shaped cells cannot flow easily and get stuck in the blood vessels, and so they don't bring enough oxygen to the body's organs. This can cause pain or organ damage.

Sickle cell disease is a life-long condition that people are born with. It is caused by an abnormal gene which is inherited from parent(s)

- If a person gets the sickle cell gene from both their mother and father, they will have a form of sickle cell disease called "sickle cell anemia."
- If a person gets the sickle cell gene from 1 parent, and a different abnormal gene from the other parent, he or she will have one of the other forms of sickle cell disease.
- If a person gets the abnormal gene from only 1 parent, and a normal gene from the other parent, this is called "sickle cell trait." People with sickle cell trait generally do not have any of the symptoms of sickle cell disease. But they can pass the gene along to their children.

What are the symptoms of sickle cell disease? — Symptoms usually start after a baby is a few months old. Most commonly, people with sickle cell disease have episodes of pain. Doctors often use the term "sickle cell crisis" or "sickle cell pain episode." The pain can be in the bones, chest, or other parts of the body. It can be mild or severe, and last hours to days. People can have other symptoms with their pain, too. The symptoms depend on how old they are:

- Babies can have pain in the hands and feet with swelling and redness.
- Older children and adults can have pain in the chest with trouble breathing, fever, or coughing.
- Older children and adults can have pain in the bones.

Most episodes of pain just happen because of the abnormal red blood cells, and can't be predicted. But sometimes, the pain can be related to other things, such as:

- Infection – People with sickle cell disease are more likely than others to get certain infections.
- Dehydration – Dehydration is when the body loses too much water.
- Weather or air conditions

- Travel to high-altitude places – These are places where there is less oxygen in the air, such as mountains. Flying in an airplane might also cause symptoms for people with sickle cell disease.

Sickle cell disease can also cause other symptoms and problems, including:

- Severe anemia – Anemia is the medical term for having too few red blood cells. It makes people tired or weak. Most people with sickle cell disease have anemia. But in some cases, it can be severe and need treatment.
- Very severe infections, which can be life threatening.
- Lung problems – Symptoms can include trouble breathing or chest pain.
- Strokes – A stroke is when a part of the brain is damaged because of a problem with blood flow.
- Open sores on the skin (usually on the legs)
- A problem with the bone in the shoulder or hip – This happens when the bone doesn't get enough blood, leading to pain in the bone or joint. In some cases this can cause permanent damage. Doctors call this problem "osteonecrosis" or "avascular necrosis."
- In men, an erection that lasts too long and is very painful

Is there a test for sickle cell disease? — Yes. A blood test can show if someone has sickle cell disease.

Some parents are tested for the sickle cell gene before getting pregnant. This is to find out their risk of having a baby with sickle cell disease. Talk to your doctor about whether you should be tested.

How is sickle cell disease treated? — Sickle cell disease involves life-long treatment. Some treatments help prevent the different problems sickle cell disease can cause. Others are aimed at relieving pain or treating other problems, like infection. Treatments include:

- Antibiotics – They help prevent infections in young children.
- Vaccines – Getting all vaccines helps prevent infections. This is especially important in children and also true for adults.
- Hydroxyurea – This medicine helps the body make more normal red blood cells.
- Blood transfusions – A blood transfusion is when a person gets blood that was given by another person. People who have a high chance of having a stroke might get blood transfusions to help prevent strokes. People might also get blood transfusions to treat low hemoglobin.
- Folic acid – A vitamin called folic acid (also called folate) is needed for people with sickle cell disease to make red blood cells.

- Pain medicines – Doctors can use different medicines to treat pain. Your doctor can help you make a plan for treating pain at home. If the pain is severe, people might need treatment in the hospital or regular pain medicines every day.

- Medicines to treat infection (antibiotics) – It is important for people with sickle cell disease to be treated for infections right away. It is very important to call your doctor if you think you might have an infection.

A procedure called a "bone marrow transplant" or "stem cell transplant" can cure sickle cell disease. This procedure replaces the cells that make your blood with healthy cells. But this treatment is not done very often for sickle cell disease. That's because it has many side effects and can be done only if people meet certain conditions.

When should I call the doctor? — Your doctor will tell you what to look for and when to call for help. In general, you should call right away if the person with sickle cell disease has any of the following problems. Do not wait to call, even if it's the middle of the night, because it's important to get treatment right away. Call for help if the person:

- Gets a fever or has other signs of infection (for example, feeling very tired, loss of appetite, stiff neck, headache, trouble breathing, or cough).
- Has pain that doesn't get better after it is treated at home
- Has an erection that lasts longer than 2 to 4 hours
- Has trouble breathing
- Has symptoms of a stroke. Symptoms might include:
 - Severe headache
 - Seizure
 - Not being able to wake up
 - Trouble speaking
 - Drooping of the face on one side
 - Weakness in an arm or leg

What else can people with sickle cell disease do to stay healthy? — People with sickle cell disease should:

- See their doctor for regular check-ups, and follow his or her instructions about treatments and tests.
- Get the vaccines that the doctor or nurse recommends. Vaccines can prevent certain serious or deadly infections.

- Take good care of themselves, including staying well hydrated, exercising, eating a healthy diet, and getting enough sleep.

What if I have sickle cell disease and want to get pregnant? — If you have sickle cell disease and want to get pregnant, talk with your doctor. He or she might recommend that you see a "genetic counselor." This person can tell you your baby's chance of having sickle cell disease or sickle cell trait.

If you have sickle cell disease and are pregnant, your doctor will follow you and your baby closely throughout your pregnancy. That's because sickle cell disease can cause problems for both you and your baby.