

# WHAT YOU SHOULD KNOW ABOUT SICKLE CELL DISEASE

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## What Is Sickle Cell Disease?

Sickle cell disease is an inherited blood disorder that causes normally round red blood cells to become crescent or “sickle” shaped, hard, and sticky. When this happens, the abnormal cells can clump together in small blood vessels and block blood flow. This can cause pain and other serious problems.

## What Are Common Symptoms?

Pain episodes are the most common symptom. The pain can start suddenly and can be mild or severe. When the pain is severe, it is called a sickle cell crisis. A crisis might be triggered by weather, dehydration, infection, or stress. Pain is the most common reason that people with sickle cell disease go to the emergency room.

Other symptoms and complications of sickle cell disease include:

- Anemia (not enough red blood cells), which causes a very tired feeling
- Acute chest syndrome (chest pain, cough, fever, and difficulty breathing)
- Leg ulcers
- Stroke
- Infections
- Jaundice (yellow skin and eyes)
- Swelling

## Who Is at Risk?

Sickle cell disease happens when a child inherits the sickle cell gene from both parents. Sickle cell trait happens when a gene is inherited from only one parent. People with sickle cell trait usually do not have pain crises and do not have most of the complications of sickle cell disease.

Sickle cell disease is more common among people of African, Mediterranean, Middle Eastern, or Indian descent. In the United States, most people with the disease are African American.

## How Is It Diagnosed?

- Since 2006, all babies in the United States have been screened for the disease at birth.
- If you were born in the United States before 2006 or outside the United States, your doctor can do a simple blood test to see if you have sickle cell disease or sickle cell trait.
- If you are thinking about having children, you can have a test to see how likely it is that your child will have the disease. Doctors can also test to diagnose the disease before the baby is born.



## How Is It Treated?

When babies are diagnosed with sickle cell disease, early treatment is important. Children are treated with antibiotics for the first 5 years of life. This reduces the likelihood of infections and complications. Regular vaccines are also important for children with sickle cell disease to stay healthy.

The most common treatment for sickle cell disease is a medicine called hydroxyurea. Adults with 3 or more pain episodes a year, pain that interferes with daily activity, or a history of acute chest syndrome should be taking hydroxyurea. It decreases pain episodes and acute chest syndrome and increases survival rates in patients with sickle cell disease. It has been used safely in adults for many years and was recently approved for use in children. Newer medicines are also available.

Severe pain episodes can often be managed at home. It is important to have a chronic pain management plan to follow during a crisis. Nonprescription pain medicine, increased fluids, rest, warm baths, heating pads, and massage can help. If your pain is not relieved, talk to your doctor about other options, such as prescription pain medication.

Blood transfusions are used to treat stroke, severe acute chest syndrome, liver failure, and multisystem organ failure.

Newer treatments can cure sickle cell disease but are not yet widely available. These include stem cell transplant and gene therapy.

## Questions for My Doctor

- What new treatments are available to me?
- What are the risks and side effects of the treatment?
- Would you help me develop a pain management plan?
- How often should I have follow-up visits?
- If I am pregnant or plan to become pregnant, what should I be aware of?
- Will I need to see any other doctors?

## For More Information



### MedlinePlus

<https://medlineplus.gov/sicklecelldisease.html>

### Sickle Cell Disease Association of America

[www.sicklecelldisease.org/sickle-cell-health-and-disease/types](http://www.sicklecelldisease.org/sickle-cell-health-and-disease/types)