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## Sickle Cell Crisis

### Sickle Cell Crisis Overview

[Sickle cell disease](#) is the most common of the hereditary blood disorders. It occurs almost exclusively among black Americans and black Africans.

Sickle cell disease in black Americans occurs in 3 of every 1,000 (or about 1 in 375) live births. Estimates indicate that the severe form of sickle cell disease affects more than 50,000 black Americans.

The first account of what was then called sickle cell [anemia](#) in the medical literature was in 1910. James B. Herrick, a Chicago physician, described the symptoms of a 20-year-old black male student from the West Indies. The man had reported "shortness of breath, [palpitations](#), and episodes of icterus [yellow eyes]. He had an anemia." Dr. Herrick described the patient's blood smear as showing "thin, sickle-shaped and crescent-shaped red cells."

Red blood cells deliver oxygen to working or active tissues. In the lungs, [hemoglobin](#) (the molecule in the red blood cell) takes on oxygen and, at the same time, releases carbon dioxide. This process is called oxygenation. At the tissue level, this activity is reversed. The same hemoglobin molecule releases oxygen and takes on carbon dioxide. This process is called deoxygenation.

In sickle cell disease, certain red blood cells become crescent-shaped (the sickle cell Dr. Herrick described). These abnormal red blood cells, carrying an abnormal hemoglobin known as hemoglobin S, are fragile. A person who has sickle cell disease can become more likely to get infections because the damaged cells eventually clog the spleen. A severe attack, known as sickle cell crisis, can cause pain because blood vessels can become blocked or the defective red blood cells can damage organs in the body.

### Sickle Cell Crisis Causes

Sickle cell disease results from mutation, or change, of certain types of hemoglobin chains in red blood cells (the beta hemoglobin chains).

The changes in the building of normal hemoglobin result in the abnormal hemoglobin of sickle cell disease. These mutated molecules do not have the smooth motion needed for oxygenation and deoxygenation. When the oxygen concentration in the blood is reduced, the red blood cell

assumes the characteristic sickle shape. This causes the red blood cell to be stiff and rigid, and stops the smooth passage of the red blood cells through the narrow blood vessels.

It does not take much imagination to see sharp-end "sickled" red cells stacking up in narrow blood vessels known as capillaries. When this happens, red blood cells are not able to carry oxygen to tissues, and tissue cell injury or death occurs. Someone with sickle cell disease would be experiencing pain with this process—the sickle cell crisis.

### **Sickle Cell Crisis Symptoms**

The sites most often affected by the blocking or stacking action of sickled cells are found in the lungs, liver, bone, muscles, brain, spleen, penis, eyes, and kidneys.

The immune system of a person with sickled cells dramatically weakens. People with sickle cells are highly susceptible to infections from certain forms of bacteria. Some of the most common infections are from [flu viruses](#), [pneumonia](#), and [salmonella](#) (a type of bacteria).

Severe pain is the most common of sickle cell disease emergencies (acute sickle cell crises). A person may not know what brought on the pain, but one or more of the following situations may have contributed to the start of the painful sickle crisis:

#### [Dehydration](#)

Infection

Fever

Hypoxia (decrease in oxygen to body tissue)

Bleeding

Cold exposure

Drug and alcohol use

#### [Pregnancy](#) and [stress](#)

Four patterns of an acute sickle cell crisis are now recognizable. They are based on the part of the body where the crisis occurs.

**Bone crisis:** An acute or sudden pain in a bone can occur, usually in an arm or leg. The area may be tender. Common bones involved include the large bones in the arm or leg: the humerus, tibia, and femur. The same bone may be affected repeatedly in future episodes of bone crisis.

**Acute chest syndrome:** Sudden acute chest pain with coughing up of blood can occur. Low-grade fevers can be present. The person is usually short of breath. If a cough is present, it often is nonproductive. Acute chest syndrome is common in a young person with sickle cell disease. Chronic (long-term) sickle cell lung disease develops with time because the acute and subacute lung crisis leads to scarred lungs and other problems.

**Abdominal crisis:** The pain associated with the abdominal crisis of sickle cell disease is constant and sudden. It becomes unrelenting. The pain may or may not be localized to any one area of the abdomen. Nausea, vomiting, and [diarrhea](#) may or may not occur.

**Joint crisis:** Acute and painful joint crisis may develop without a significant traumatic history. Its focus is either in a single joint or in multiple joints. Often the connecting bony parts of the joint are painful. Range of motion is often restricted because of the pain.

Many other organ systems are often injured or impaired.

**Central nervous system:** Two-thirds of all strokes in people with sickle cell disease occur in children, at an average age of 8 years. About 10% of people with sickle cell disease have strokes or other brain bleeding when younger than 8-10 years. As the population ages, the incidence of these events also increases. Repeat strokes occur in two-thirds of all survivors within 3 years of the first [stroke](#). Blood clots affect the large vessels in the brain. Bleeding may occur in the small vessels damaged by sickle cell disease.

**Eyes:** The effect of sickle cell disease on the eyes comes from the increased viscosity, or "sludging," of blood and the narrowness of the eye's blood vessels. Retinopathy (disease of the retina in the eye) is common and causes problems with vision. [Retinal detachment](#) is frequent. [Hyphemas](#), bleeding in the eye, occur at the same rate as the general population, but complications are more common because of the increased sickling effect that the waterlike fluid in the eye promotes.

**Kidneys:** Some amount of kidney damage occurs in nearly every person with sickle cell disease.

**Genitals:** Priapism (a constant erection of the penis) is common. It affects about 40% of all men with sickle cell disease. Severe episodes are a frequent cause of impotency.

**Infections:** People with sickle cell disease have weakened immune systems and are at increased risk for developing infection, especially in the lungs, kidneys, bones, and central nervous system.

**Blood problems:** People with sickle cell disease can develop [anemia](#)—a reduction in the number of red blood cells. Symptoms of anemia are shortness of breath (oxygen is not getting to tissues), lightheadedness, and fatigue.

## **When to Seek Medical Care**

If certain conditions develop in a person with sickle cell disease, the person must contact a physician. If the physician is not quickly available or cannot see the person right away, the person with sickle cell disease may choose to go to a hospital's emergency department. Contact the physician in the following cases:

Many people with sickle-cell disease have pain with enough frequency that they need to take pain medications at home. If the pain is unrelieved by the medication, or the pain is significantly different from previous episodes, contact the health care provider.

If experiencing nausea, vomiting, and diarrhea; losing a lot of fluid; and having inability to drink and keep it down, the person with sickle cell disease is in danger of becoming dehydrated. This is a serious concern with sickle cell disease. The physician or the hospital may provide IV fluids to replace the lost fluids.

It is important to control infection. If it appears that a person with sickle cell disease is getting an infection, even if using antibiotics to prevent infection, contact the physician immediately.

A sickle cell crisis can often be managed efficiently and quickly in a hospital's emergency department with fluids and pain medicines. A person with sickle cell disease should not delay going to the hospital. Delay can only make the condition worse and might require hospitalization for treatment.

Go to a hospital's emergency department if these conditions develop:

Uncontrollable pain even with the use of narcotics

Continued loss of fluid leading to dehydration (if vomiting)

Uncontrollable fever

Chest pain or shortness of breath

Severe [abdominal pain](#)

## Exams and Tests

The health care provider will take the complete medical history of a person with sickle cell disease. This history should include whether any infections are present. The health care provider will ask about other problems that are common starters of sickle cell crisis. These problems would be a lack of oxygen in the tissue, bleeding, dehydration, alcohol and drug use, pregnancy, and other concerns.

During a physical exam, the physician will check the nervous system, lungs, bones, eyes, and abdomen, in particular.

The physician will perform blood and urine tests. If indicated, the physician may have a CT scan of the head taken and perform a spinal tap to check for problems in the spinal fluid and brain.

If the physician suspects sickle cell disease in an adult, or more commonly a child not previously diagnosed with this disease, attention will first be paid to getting a family history of sickle cell disease. The physician then performs a blood test to diagnose the disease.

## Sickle Cell Crisis Treatment

### Self-Care at Home

Even tiny changes in the red blood cells can begin a cascade of symptoms leading to a sickle cell crisis. Therefore, home care, even when a person is careful about drinking plenty of fluids and avoiding infection, is difficult. The best home care is understanding the illness and knowing when and where to seek immediate medical care.

## **Medications**

Sickle cell pain crisis

Pain medications, often narcotics, will be given.

IV fluids are an important part of therapy.

Infection: If the physician diagnoses or suspects a bacterial infection, antibiotics are prescribed.

Anemia: If there is a significant decrease in the red blood cell count, a red blood cell transfusion may be needed.

## **Other Therapy**

Chronic therapy: New developments in regularly scheduled transfusion therapy have shown promise in decreasing the following:

Symptoms of acute chest syndrome

Incidence of stroke

Severity of pain crises

Bone marrow transplantation holds promise for a very small percentage of people with sickle cell disease. Discuss this with the physician.

## **Next Steps**

### **Follow-up**

Considering the many body systems involved and the likelihood that sickle cell crisis will occur time and time again, strong consideration should be given to follow-up with a hematologist (a physician with a specialty in treating blood disorders).

Most uncomplicated cases of sickle cell crisis can be treated in community emergency departments. People with this condition can be safely sent home when their pain is under control and their dehydration is eliminated. A short observational period in the emergency department helps to prevent acute relapse and admission for pain and rehydration.

Outpatient treatment centers on the control of infection, reduction of pain, and prevention of dehydration. The use of narcotics is often necessary and should not be limited for fear of turning someone with sickle cell disease into a drug addict.

## Prevention

Sickle cell disease is an inherited disorder that is common among African Americans. If you have sickle cell disease, you received the gene from each of your parents. If you received the gene from just one parent, you are a carrier. Genetic counseling and testing can give you information about the likelihood that you might pass on this gene to your children.

### Prevention of infection

Infants and young children are at greatly increased risk of becoming infected with severe types of bacterial infections. Because of this increased risk, infants with sickle cell disease are placed on daily penicillin therapy until at least aged 5 years.

It is also extremely important that infants receive all their immunizations on schedule to prevent these life-threatening infections.

Subjecting yourself to conditions in which the oxygen concentration in the air is low may worsen sickling. This may include traveling in unpressurized airplanes or going to high altitudes.

## Outlook

In spite of the tremendous improvements in diagnosis, genetic counseling, neonatal screening, and medical care, the ever-increasing body of information may have falsely raised expectations for a specific cure. Progress on genetic manipulation, which has the potential to cure the disease, is slow.

Developments in long-term transfusion therapy have given promise in decreasing the severity of pain crises and the incidence of stroke; however, exposure to blood-borne diseases due to multiple transfusions is a risk.

Support and cure of infection are still the mainstays of treatment.

### For More Information

#### Web Links

[Sickle Cell Disease Association of America, Inc.](#)

[The Sickle Cell Information Center](#)

[American Sickle Cell Anemia Association](#)

MedlinePlus, [Sickle Cell Anemia](#)

### Authors and Editors

Author: John R Krimm, DO, FAAEM, Associate Professor, Department of Emergency Medicine, Albert Einstein University.

Editors: Brian F Chinnock, MD, Assistant Professor, Department of Emergency Medicine, Texas Tech University Health Sciences Center at El Paso; Francisco Talavera, PharmD, PhD, Senior Pharmacy Editor, eMedicine; Thomas Rebbecchi, MD, FAAEM, Program Director, Assistant Professor, Department of Emergency Medicine, University of Medicine and Dentistry of New Jersey.

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