

# Sickle Cell Disease

(Sickle Cell Anemia, Hemoglobin SS Disease, Sickle Cell Trait, Sickle Cell Thalassemia)

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## *Description of the Disability*

Sickle Cell Disease is an inherited disease affecting the red blood cells. The disease is caused by a gene for an abnormal form of Hemoglobin – “Hemoglobin S” (HbS) rather than Hemoglobin A, B, or O. Hemoglobin proteins are produced inside red blood cells, where they carry oxygen from the lungs to the cells of the body and then deoxygenate (drop off the oxygen) and return to the lungs. However, when Hemoglobin S deoxygenates, it causes changes in the blood cell that make the cell bend into a curve, or sickle shape. These sickle cells can’t move through the blood vessels as easily and tend to become damaged, burst, or create a block in the blood vessels. This drastically lowers the number of blood cells in the body and reduces blood flow to parts of the body.

The most serious form of the disease, Sickle Cell Anemia, occurs in individuals who have inherited the sickle cell gene from both parents (homozygous for HbS, or HbSS). Individuals who only carry one copy of the sickle cell gene and a copy of a “normal” hemoglobin gene can have one of several subtypes of Sickle Cell Disease. Individuals with Hemoglobin A and S (HbSA) will have **Sickle Cell Trait**, a milder version of Sickle Cell Anemia. People who are heterozygous for HbS with other versions of Hemoglobin (HbSB, HbSC, HbSD, HbSO, etc.) may or may not have milder versions of HbSS symptoms. A few of these heterozygous individuals may have symptoms equivalent to HbSS.

Other possible subtypes include **Sickle Cell-Beta Zero Thalassemia** and **Sickle Cell-Beta Plus Thalassemia**. In these subtypes there are small amounts of Hemoglobin A (HbA) present in the blood, but not enough to really matter, so the conditions are clinically identical to HbSS.

The main symptom of Sickle Cell Disease is sudden pain “crises” in joints or organs. Which joints or organs are affected can vary from person to person. The chest, back, and torso are most common, leading to difficulty breathing during the crisis. The pain can also occur in the hands and feet, or almost any other part of the body. Some individuals have only one or two crises a year, others have them more than once a month. Some crises last only a few hours, others last for weeks and require hospitalization for pain control.

There are a number of other symptoms associated with Sickle Cell Disease. People who have the disease are nearly always anemic (fatigue, shortness of breath) and have mild jaundice (yellowish skin and eyes), both of which are consequences of low blood cell counts. Most also have an enlarged spleen or a spleen that no longer functions. This makes them more susceptible to infections since the spleen acts to fight off infections. Sickle Cell Disease can cause vision problems by blocking blood flow to the retinal tissue, damaging the eye. In serious cases, blindness may occur. The liver and heart may also enlarge over time causing more problems. Other possible complications include stroke, leg ulcers, bone damage, early gallstones, lung

blockage, and kidney damage. Acute Chest Syndrome is a respiratory infection, similar to pneumonia, that can be associated with Sickle Cell Disease.

Anything that reduces the amount of oxygen in the blood can trigger an attack of pain and worsening of anemia. Possible triggers include vigorous exercise, flying at high altitude, mountain climbing, hot weather, etc. During these crises, abdominal pain may be severe and vomiting may occur.

### ***Incidence Statistics***

- In the general population, 8 in 100,000 people have Sickle Cell anemia.
- In the African American population in the US, 1 in 600 have the disease.
- In the Hispanic population, 1 in 1,000 have the disease.
- 2 million Americans have the sickle cell trait.

### ***Common Treatments, Medications, and Side Effects***

Individuals with this disease usually need regular medical checkups every 3 to 6 months. At times they may need more frequent checkups. During a crisis they need immediate medical attention. Signs of a crisis include: high fever, unstoppable or continuous cough, difficulty breathing, pain that can't be treated at home, weakness or dizziness.

Because pain episodes are the most common problem, many individuals keep a small supply of an oral narcotic analgesic (such as codeine) on hand and for use with pain attacks that do not respond to aspirin, acetaminophen or ibuprofen.

Less severe pain episodes can usually be treated at home with gentle heat, increased fluids, and pain medications. If the individual has to be hospitalized for a severe crisis, they are typically given IV fluids, pain drugs, and oxygen. Blood transfusions are another common treatment, since the transfusion will temporarily replace the HbS cells with normal hemoglobin cells. However, as soon as the fresh blood cells age and are removed by the liver, the body will replace them with HbS cells again. In severe cases, doctors may perform a bone marrow transplant to permanently improve the situation, but the donor for the transplant must be a healthy sibling or close relative and that is not always possible.

Bacterial infection is another serious complications of Sickle Cell Disease. Individuals are encouraged to routinely take preventive measures such as flu shots. In addition they should avoid working in hospitals or in settings where they are routinely exposed to infectious materials.

A few preventive strategies help reduce the frequency of crises. To improve fluid levels in the blood, individuals are encouraged to drink eight to ten glasses of water a day. A good diet, regular exercise, healthy sleep, and avoidance of smoking and alcohol are important strategies.

Many individuals need daily vitamin and folic acid supplements to help form new red blood cells.

The most common drug therapy is for pain management. These include several Narcotic Analgesics (see drug entry), such as Morphine, Codeine, Buprenorphine, Nalbuphine, Methadone, and Oxycodone (see drug entry for Narcotic Analgesics for side effects). Home treatment of pain often includes Non-Steroidal Analgesic Drugs (NSAIDs, see drug entry) such as ibuprofen or Acetaminophen. See the entry on Chronic Pain for more pain control information.

One relatively new drug, an anticancer drug named Hydroxyurea, seems to improve the root cause of Sickle Cell Disease, HbS. The drug reduces the frequency of pain crises and Acute Chest Syndrome in individuals with the disease. No one knows exactly how it works, but it seems to stimulate the production of Fetal Hemoglobin, a type of hemoglobin that most of us stop making after birth. Fetal Hemoglobin somehow reduces the severity of Sickle Cell Disease, possibly by providing an alternative form of Hemoglobin to replace HbS.

Gene therapy for Sickle Cell Disease is under development.

### ***Possible Functional Issues***

- Need to avoid strenuous physical activity
- Need to avoid temperature extremes
- Reduced stamina
- Difficulty walking long distances or running
- Need to avoid exposure to infection disease
- Reduced vision (occasionally)
- Daytime drowsiness (possible side effect of pain medication)
- Increased thirst
- Reduced mobility or dexterity because of joint pain

### ***Initial Interview Considerations***

#### Initial Questions

- What effect has the disease had on their employment?
- How often do they have crises? How long do the crises last?

- What is a crisis like for them? Where does the pain strike? What other symptoms do they have? What actions do they take to reduce the symptoms?
- What if anything makes a crisis worse? What makes it better?
- How often do they see their doctor for checkups?
- How much trouble do they have getting sick with a cold or similar illness? Have these kinds of infections been problems for them?
- How is their vision? Has it been affected?
- How often do they have trouble with joint pain?
- What if any medications are they taking? Are there any side effects that might cause work problems for them?
- What is a typical weekend like for them? (gets at leisure activities, social activities, hobbies)
- What kind of exercise do they get? Do they participate in any sports? How long can they maintain exercise like that? (minutes, hours?)

#### Initial Observations

- Does the person favor one leg or seem to protect one arm (or any other body part)? This may reflect joint pain that needs to be accommodated.

#### Interview Accommodations (if any)

- Find out if they need any visual accommodations – large print or audio versions of forms and literature, etc.

### ***Possible Accommodations and Assistive Technology***

- A flexible work routine, especially regarding an alternation between physically demanding tasks and low activity tasks.
- Assistive devices to help with lifting or other physically demanding tasks. (Note that “physically demanding” is relative to each person.)
- Frequent rest breaks.
- Flexible deadlines to accommodate pain crises.
- Mobility devices such as scooters if the job involves walking long distances.
- Cell phone or cordless phone to reduce walking on job site.
- Vision accommodations (see entry on Low Vision).
- Access to drinking water.

### ***Career Planning Issues***

- The frequency of pain crises may affect how well they can meet routine work hours.
- Physical demands of the job should be considered, but physically demanding jobs are not necessarily a problem. It will depend on the individual. Also, strenuous activity interspersed with low-impact activities may be workable.
- Consider whether any other aspects of a job might affect the levels of oxygen the person gets. The person will know his or her own triggers best.
- Cognitive skills, judgment, problem-solving skills, memory and learning skills should not be affected.
- Written skills and language skills should not be affected.
- Social skills should not be affected.
- Dexterity, coordination and strength (for short bursts) should not be affected.
- Jobs with high exposure to infections diseases, such as hospital work, should be approached cautiously because of reduced immune system.
- Self-employment or home-based employment may be an option.

### ***Emerging Issues***

- Gene therapy
- Pain control and treatment issues

### ***Additional Information Resources***

- Sickle Cell Society Home Page: [www.sicklecellsociety.org/](http://www.sicklecellsociety.org/)
- Sickle Cell Disease Association of America: [www.sicklecelldisease.org](http://www.sicklecelldisease.org)
- Harvard Center for Sickle Cell Disorders: [sickle.bwh.harvard.edu/](http://sickle.bwh.harvard.edu/)
- The Sickle Cell Information Center Emory University: [www.emory.edu/PEDS/SICKLE](http://www.emory.edu/PEDS/SICKLE)