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SICKLE CELL DISEASE



SICKLE CELL DISEASE

Caring for a loved one with sickle cell disease is no easy task. Not only does it require caring for someone you love at home, but it also means facing your own concerns about the disease. It may be frightening to discover that there is no cure, and you may worry about other family members becoming ill with sickle cell disease.

Physical and emotional care can be time-consuming and exhausting. Many caregivers of people with sickle cell disease have concerns about their loved ones' illness and future health prospects. Practical concerns, such as worries about financial issues and time management, are also common. The goal of this chapter is to address some of these concerns.

Learning more about sickle cell disease may help you feel more at ease in your caregiver role. This can be important for new caregivers of small children with the disease. The feeling of helplessness from caring for a sick child or relative of any age can be reduced by becoming knowledgeable about the disease.

It can be difficult to learn about a disease when you do not know where to start. This section provides information about sickle cell disease, including what caregivers can do to help their loved ones.

What Is Sickle Cell Disease?

Sickle cell disease refers to a genetic illness caused by an abnormality of hemoglobin. Hemoglobin is a molecule in red blood cells. A person can have the gene for sickle cell but not be sick; some people, however, develop sickle cell anemia.

In most people, red blood cells are round and smooth. This round and smooth shape allows the cells to move easily through the blood vessels. The sickle cell hemoglobin can cause the red blood cells to change shape, from the normal round shape into a crescent (or sickle) shape. The sickle-shaped red blood cells are hard and sticky and do not easily pass through blood vessels. Therefore, blood vessels can sometimes become clogged, preventing blood from flowing through them. A clogged blood vessel is called an "occlusion." Occlusions in blood vessels can lead to severe pain, called a "sickle cell crisis."



Normal Red Blood Cells



Sickle Cells

Sickle cell crises

Sickle cell crises can be frightening for both you and the patient. They can last hours to days, becoming so painful that people have to go to the emergency room for immediate treatment with pain medication and intravenous fluids. Your loved one may cry or scream during a painful crisis. At the beginning of the crisis, you may also notice other changes in his/her behavior like decreased appetite or loss of interest in regular activities.

The frequency of sickle cell crises can range from every few years to many times per year. The good news is that not all people with sickle cell disease have crises. About 30% rarely or never experience a crisis. About 50% have only a few crises throughout their lives. About 20% have frequent and severe sickle cell crises.

It may be useful for you to know that there are certain factors that may produce a crisis.

Factors That May Cause a Crisis

- n Having an infection, such as a cold or the flu
- n Cold weather
- n Being overly tired or fatigued
- n Exercising too hard
- n Being dehydrated (not drinking enough water and other fluids)
- n Not having enough oxygen in the blood

(Adapted from American Academy of Family Physicians, 2000)



Who Gets Sickle Cell Disease?

In the United States, sickle cell disease is most common among African Americans and Hispanic Americans. The disease affects approximately one in 500 African Americans and one out of 1,000 to 1,400 Hispanic Americans. About 72,000 people in the U.S. currently have sickle cell disease, which affects people all over the world.

Family members and friends may be afraid they will "catch" sickle cell disease and get sick themselves. The disease is not contagious. It cannot be spread from person to person through the air, by contact or via blood transfusions. Rather, sickle cell disease is inherited. This means that the disease is passed through families, from parents to their children. Each time a baby is born, he or she has two genes that determine the make-up of the hemoglobin -- one that comes from the mother and one that comes from the father. In families with a history of sickle cell disease, there are three possibilities:

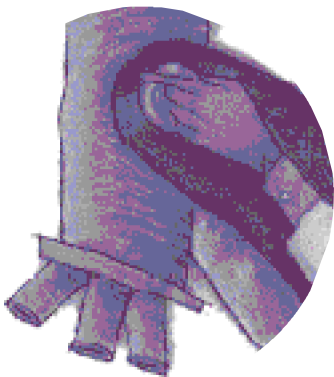
1 Family members may have two genes for healthy hemoglobin. These family members will have no sign of sickle cell disease.

2 Family members may have one gene for healthy hemoglobin and one gene for sickle hemoglobin. This is called having the "sickle cell trait." People with sickle cell trait are not sick and often do not know they have the sickle cell gene, unless they are tested.

3 Family members may have two genes for sickle hemoglobin. These family members will have sickle cell disease.

Diagnosing Sickle Cell Disease

Doctors use a simple blood test to diagnose sickle cell disease. This test can be done at birth along with the standard screening tests.



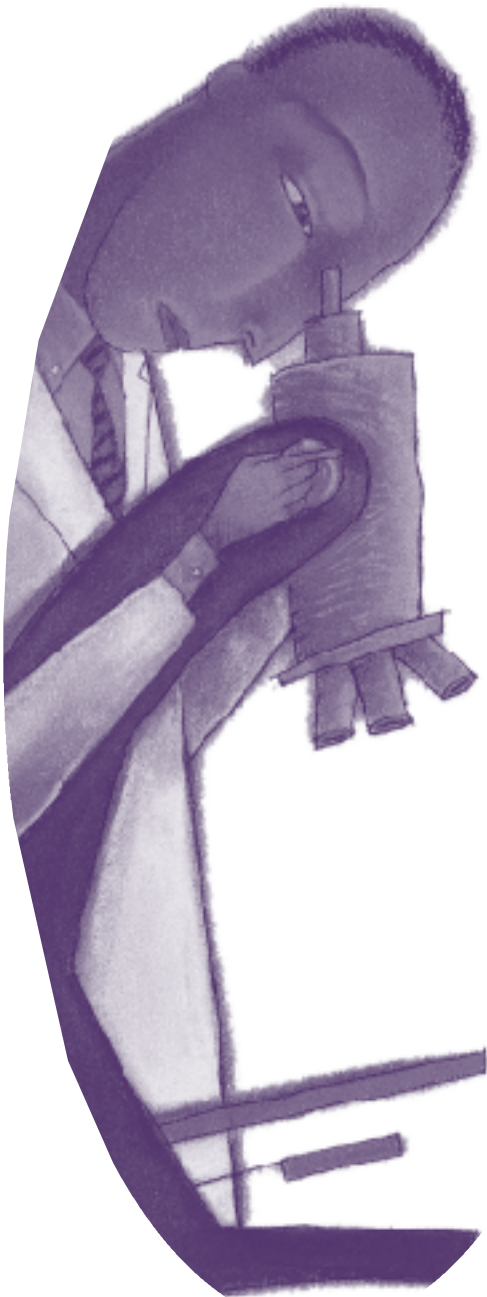
Common Symptoms of Sickle Cell Disease

Children may start to have symptoms of sickle cell disease as young as 6 months of age. Babies who are suffering from sickle cell symptoms may be irritable or cranky, even when their parents do everything possible to provide comfort. Also, children with sickle cell disease may develop more slowly and have difficulties in school because of the pain from the disease. Parents who learn about the disease are more likely to be able to help their children.

The most common symptom of sickle cell disease is pain. During a crisis, pain may seem to come from the bones, usually in the arms, hands, legs, feet, or back. Also, there may be pain in the stomach or chest.

Other health problems for people with sickle cell disease include:

- n Infection
- n Anemia (or a low blood count)
- n Fatigue
- n "Acute Chest Syndrome" (severe chest pain with breathing difficulties)
- n Eye problems such as bad eyesight and blindness
- n Skin ulcers or skin sores that do not heal well, especially on the lower legs
- n Jaundice, or yellowing of the whites of the eyes or skin
- n Damage to the organs, such as the liver, kidneys, or lungs
- n Brain damage or stroke



What You Can Do to Help

- n Learn as much as possible by reading and asking experts about sickle cell disease.
- n Find medical practitioners who understand the disease.
- n Ensure regular visits to a family doctor, pediatrician (children's doctor), a hematologist (blood specialist), and/or a specialized sickle cell clinic.
- n Ask questions of your loved one's doctors and discuss your concerns.
- n Help the patient avoid factors that bring on crises.
- n Find ways to avoid and manage stress in the household.
- n Make sure that the patient drinks plenty of fluids.
- n Administer pain medications as prescribed.
- n Offer to massage sore or painful areas for your loved one, if the physician approves.
- n Encourage your loved one to use relaxation techniques, and other pain management strategies.
- n Watch for signs of a sickle cell crisis.
- n Help the patient rate and record their pain and fatigue in a daily journal.

Please see the "Symptom Management at Home" section for other information on pain, fatigue, and difficulty breathing. This includes details on keeping pain and fatigue journals. For more information on relaxation techniques, see the "You Have Needs, Too" section of this directory.



Treatment for Sickle Cell Disease

While there is no cure for sickle cell disease, there are precautions for preventing painful sickle cell crises and treatments to provide relief during pain crises. Some are used during a crisis, some are used to reduce the number of crises, and some are used to treat other complications of the disease.

General medical treatments

These general medical treatments are recommended by physicians:

- n Antibiotics – medication given to prevent infections
- n Erythropoietin – medication given to improve anemia (low blood count)
- n Blood transfusions – increases blood count to improve anemia
- n Hydroxyurea – medication given to reduce the chance that the red blood cells will change into the sickle shape
- n Folic acid – a vitamin which helps to treat anemia

- n Hydration (increasing the level of fluids in the patient's body) – counters the frequent problem of dehydration (decreased level of body fluids)
- n Oxygen – provided via an oxygen mask, nasal catheter (tube connected to nose), or oxygen tent, increases the level of oxygen in the blood
- n Treatment for pain – drug and non-drug approaches

(Adapted from Harrison's Principles of Internal Medicine, 14th Edition. Edited by Fauci, A.S., Braunwald, E., Isselbacher, K.J., et al. New York: McGraw Hill, 1998)

Preventing crises

It is important for you to know that some sickle cell crises can be prevented. You can help by encouraging the patient to follow these steps:

- s Limit intake of alcohol and avoid smoking.
- s Keep warm and avoid cold or rainy weather.
- s Get plenty of sleep and rest, and let the patient's health practitioner know about any difficulties with sleep.
- s Exercise regularly, but not too hard.
- s Drink plenty of fluids (at least 8 glasses of water a day in warm weather).
- s Learn to manage emotional and physical stress effectively.
- s Treat infection immediately.
- s Monitor pain medication use with a trained medical practitioner.

(Adapted from American Academy of Family Physicians, 2000)

When to call the doctor

Be aware of the warning signs for a sickle cell crisis. Contact the patient's medical practitioner immediately if any of these occur.

Crisis Warning Signs

- s Fever of 101 degrees Fahrenheit (38 degrees Celsius) or higher
- s Pain not relieved by medication
- s Shortness of breath or fast breathing
- s Severe headaches or dizziness
- s Severe stomach pain or swelling
- s Jaundice (yellow color in whites of eyes or skin) or very pale skin
- s Painful erection in males
- s Sudden change in vision
- s Seizures
- s Weakness



Children and Adults Who Have Difficulty Communicating

Children or adults who have difficulty communicating can't always tell you about their symptoms. This can make your job as caregiver more difficult and leave you feeling helpless and out of control. Learning about what to look for in patients suffering from a crisis can help you take better care of them and get them the help that they need.

Watch For These Signs of a Sickle Cell Crisis:

- n Changes in breathing pattern
- n Frequent coughing
- n Unusual crankiness or crying
- n Screams or sudden movements when touched
- n Loss of appetite
- n Vomiting or diarrhea
- n Changes in urination
- n Unusual tiredness or weakness
- n Swollen hands or feet
- n Pale blue or gray lips or skin

For more information on how and when to effectively communicate with your loved one's health care team, please see the "Navigating 'The System'" section.

Pain Management

When a sickle cell crisis is severe, patients often go to the emergency room. Sickle cell patients sometimes have bad experiences in emergency rooms and hospitals when health care professionals question their pain and their need for medications. Some may be labeled as "addicted" to their pain medications. You should know that wanting medications to help the severe pain of a sickle cell crisis does not make a patient a drug addict, or mean that they are trying to "get high." These bad experiences may lead sickle cell patients to distrust health care professionals. It is important that patients be able to find health practitioners whom they trust. You can help by locating a health care team that is knowledgeable about sickle cell disease and sickle cell pain. Getting to know the members of the team and making sure they know your loved one is also important.

Still, you and other family members may worry about addiction to pain medications, particularly opioids (also called narcotics). It can help to understand the difference between addiction and physical dependence:

- s As tolerance increases the drug becomes less effective and higher doses are required.
- s Physical dependence means that a person will have symptoms of withdrawal when the drug is suddenly stopped.

- s Addiction occurs when drugs are used compulsively for reasons other than pain relief, and when their use is continued despite harm.

(Adapted from Guideline for the Management of Acute and Chronic Pain in Sickle-Cell Disease, American Pain Society, 1999)

For more information on opioid dependence versus addiction and other facts and myths about pain medications, read the section on "Symptom Management at Home."

For many patients, opioid medications can be extremely helpful, particularly during a painful crisis. Patients who have many crises can sometimes benefit from taking opioid medications daily, along with additional pain medication during crises. The daily opioids can help reduce the number of crises and make the pain less severe. Other medications may also be prescribed by a doctor for relief from sickle cell pain.

Non-Opioid Medications Prescribed for Pain

- n Hydroxyurea (Hydrea and Droxia are examples)
- n Anti-inflammatory medications (such as aspirin, ibuprofen, or Vioxx)
- n Steroids (such as prednisone)
- n Tricyclic antidepressants (such as amitriptyline)
- n Anticonvulsants (such as gabapentin)

Non-drug treatments

- n Transcutaneous electrical nerve stimulation (TENS) – a low intensity electrical current can sometimes be effective for pain relief.
- n Physical therapy – gentle exercises and heat and cold treatments can help with sickle cell disease pain.
- n Relaxation techniques – learning how to relax can help patients cope better with the illness and pain.
- n Massage – especially at the first signs of a crisis, massage can decrease muscle stiffness and potentially reduce pain.
- n Acupuncture and acupressure – can be useful in helping to relieve stress and manage pain.
- n Distraction – engaging activities (such as hobbies, video games and movies) that change the patient's focus can help relieve stress and pain.
- n Psychotherapy – speaking with a mental health professional about the stress and frustration of sickle cell disease and/or learning cognitive behavioral techniques for coping with the disease can be beneficial for both you and the patient.



How you can help obtain treatment for pain

- s Find medical practitioners who understand your loved one's sickle cell-related pain.
- s Ensure regular visits to sickle cell pain specialists.
- s Ask questions of doctors and discuss your concerns.
- s Administer pain medication as prescribed.
- s Offer to massage sore or painful areas, if the patient's physician approves.
- s Encourage the use of relaxation techniques to relieve stress and decrease pain.
- s Find ways to avoid and manage stress in the household.
- s Distract your loved one with activities he/she enjoys.
- s Watch for signs of pain or other discomfort.
- s Help the patient rate and record pain in a daily pain journal.
- s Contact the patient's medical practitioner if pain is not relieved by medication.

Please see the "Symptom Management at Home" section for other information on pain. This includes facts and myths about pain medication as well as details on a daily pain journal. For more information on relaxation techniques, read the section on "You Have Needs, Too."

HELPFUL SICKLE CELL DISEASE RESOURCES

Organizations

n American Sickle Cell Anemia Association
(ASCAA)

10300 Carnegie Avenue

Cleveland Clinic / East Office Building

(EEb18)

Cleveland, OH 44106

(216) 229-8600

<http://www.ascaa.org/ascaa.htm>

- Provides a nationwide directory of support groups that includes groups for both patients and parents. Other services performed by ASCAA include testing, education, counseling, support services, newborn testing, lead screening, genetic counseling, and an online bulletin board where you can discuss issues related to sickle cell with peers.

n National Institutes of Health National Heart,
Lung, and Blood Institute

2 Rockledge Centre

6701 Rockledge Drive

MSC 7950, Room 100420

Bethesda, MD 20892-7950

(301) 435-0055

<http://www.nhlbi.nih.gov/health/public/blood/index.htm#scd>

- This is the primary National Institutes of Health organization for research on sickle cell anemia.

n Sickle Cell Advocates for Research and Empowerment,
Inc. (S.C.A.R.E.)

P.O. Box 630127

Bronx, NY 10463

(718) 884-9670

<http://www.defiers.com>

- S.C.A.R.E. is an advocacy group working to empower sickle cell patients and their families. Their Website also provides an online bulletin board and e-mail listserv where you can communicate with peers.

n Sickle Cell Disease Association of America

4221 Wilshire Boulevard

Los Angeles, CA 90010

(800) 421-8453

<http://sicklecelldisease.org>

- This Website provides answers to frequently asked questions about sickle cell disease and offers an online chatroom where patients and caregivers can communicate with peers.



Websites

n MEDLINEplus Health Information

<http://www.nlm.nih.gov/medlineplus/sicklecellanemia.html>

- MEDLINE is a service of the National Library of Medicine that provides excellent links to Websites for a variety of topics related to sickle cell disease.

n Sickle Cell Information Center

Georgia Comprehensive Sickle Cell Center at Grady Health System

<http://www.emory.edu/PEDS/SICKLE/>

- This is one of the most comprehensive sickle cell sites and has information provided and reviewed by an advisory board composed of physicians and other health care professionals. The types of information provided include medical, newborn screening, research, and educational materials for health care professionals.

n Nemours Foundation

http://www.kidshealth.org/parent/medical/heart/sickle_cell_anemia.html

- This site, which is geared toward the parents of sickle cell patients, provides medical information on the disease.

n The Department of Pain Medicine & Palliative Care
Beth Israel Medical Center

<http://www.stoppain.org>

- This site provides a multimedia presentation with good information on sickle cell pain.

Español/Spanish Information

n Anemia Falciforme (March of Dimes Birth Defects Foundation)

<http://www.nacersano.org>

n Datos Sobre la Anemia Falciforme (Facts About Sickle Cell Anemia)

http://www.nhlbi.nih.gov/health/public/blood/sickle/fasca_sp.htm

