

Fact Sheet

Sickle Cell Pain

About

Sickle cell disease is a chronic, potentially life-threatening disease that is presently incurable. Pain is the hallmark clinical manifestation of sickle cell disease. A variety of factors makes pain from sickle cell disease unique among pain syndromes.

Sickle cell disease is a group of inherited red blood cell disorders. Normal red blood cells are round like doughnuts, and they move through small blood tubes in the body to deliver oxygen. Sickle red blood cells become hard, sticky and shaped like sickles used to cut wheat. When these hard and pointed red cells go through the small blood tube, they clog the flow and break apart. This can cause pain, damage, a low blood count, or anemia.¹

“According to CDC, In the United States sickle cell disease affects an estimated 70,000 to 100,000 Americans.”⁴”

“Pain episodes are among the most troublesome and frequent complications of sickle cell syndromes. The patient develops severe pain in the extremities, back, and abdomen. A sickle pain episode is both uncomfortable and frightening to the patient, however, in itself almost never results in mortality. Pain episodes may be

spontaneous or occur in association with physical or psychological stress.” —The Sickle Cell Information Center²

The terrible pain of a sickle cell crisis often requires a trip to the emergency room. Some sickle cell patients have had bad experiences in emergency rooms and hospitals when health care professionals questioned their pain and why they wanted pain medications. Patients and health care professionals should know that wanting medications to help the severe pain of sickle cell crisis does not make a patient a drug addict and does not mean that they are trying to “get high” from the medications. It is important that patients are able to find a doctor who they trust to believe them and take good care of them.³

Facts

In the United States sickle cell disease affects an estimated 70,000 to 100,000 Americans:⁴

- The disease occurs in about 1 out of every 500 African Americans births
- The disease occurs in about 1 out of every 36,000 Hispanic Americans births
- Sickle cell trait occurs in about 1 in 12 African Americans

The average life expectancy for a person with sickle cell disease in America has improved; it has risen from age 14 in 1973 to around 50 today.⁵

Sickle cell disease is a major public health concern. From 1989 through 1993, there were an average of 75,000 hospitalizations due to sickle cell disease in the United States, costing approximately \$475 million.⁴

Complications from the sickle cells blocking blood flow and early breaking apart include:⁶

- Pain episodes
- Strokes
- Increased infections
- Leg ulcers
- Bone damage
- Yellow eyes or jaundice
- Early gallstones
- Lung blockage
- Kidney damage and loss of body water in urine
- Painful erections in men (priapism)
- Blood blockage in the spleen or liver (sequestration)
- Eye damage
- Low red blood cell counts (anemia)
- Delayed growth

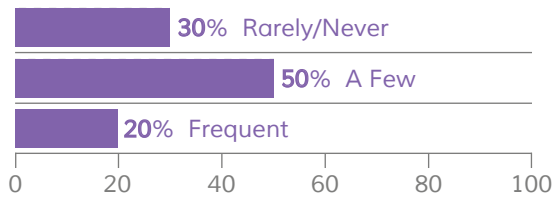
Severe pain as a result of sickle cell disease can be experienced as early as six months of age, and can occur unpredictably at varying intervals throughout a person's life.⁷

Sickle cell pain can be acute, chronic, or mixed, according to temporal characteristics, and consists of pain in the extremities, back, abdomen, or chest.⁷

Acute painful sickle cell episodes vary in intensity and frequency among individuals, ranging from isolated, mild, and infrequent to severe and frequently recurring.⁷

Patients with sickle cell anemia who experience frequent painful crises exhibit problems with self-concept and low self-esteem, anxiety, depression, dissatisfaction with body image, poor school performance, social isolation, decreased participation in normal activities of daily living, and poor peer and family relationships.⁸

Sickle Cell Crisis



Not all people with sickle cell disease have crises. About 30% rarely or never have the pain caused by a crisis. About 50% have only a few crises throughout their lives. And about 20% have frequent, severe sickle cell crises.³

Concerns about drug abuse, reluctance to prescribe pain medication, and disbelief of patients' report of pain severity are primary barriers to optimal sickle cell pain management reported by clinicians.⁹

When physicians were surveyed anonymously at 7 National Institutes of Health-funded sickle cell centers and asked to rank their beliefs about barriers to optimal management of sickle cell pain, "fear that the patient is drug abuser," and "disbelief in

patient's report of pain severity" ranked in the top 5 of 15 possible respondent choices. In a similar study of nursing attitudes about sickle cell pain management, 63% of the 77 respondents believed that "drug addiction frequently develops in the treatment of sickle cell pain episodes," and the majority of respondents (86%) believed that "drug addiction should not be a primary nursing concern when caring for a patient with sickle cell pain episodes."¹⁰

Smoking has been strongly associated with pain events among adults with sickle cell disease.¹¹

Additional Resources

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Resources verified July 2011.

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