Sickle Cell Disease

What is sickle cell disease?

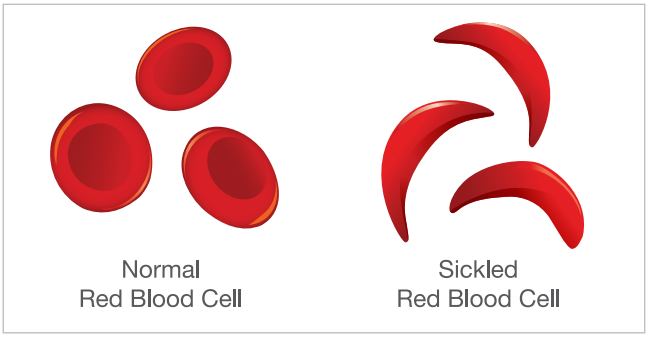
- Sickle cell disease is a condition through which children have inherited, from their parents, one or more of the abnormal forms of hemoglobin. Hemoglobin is a protein in red blood cells (RBCs) that carries oxygen throughout the body. Abnormal hemoglobin makes RBCs take on a pointy shape that, in turn, makes them clog blood vessels. Instead of being round and smooth, they form a *c* shape like a crescent moon (see figure to the right). These sickle cells can get stuck in blood vessels and block blood flow. These RBC blockages can cause pain or swelling and interfere with the body's ability to fight infection.
- Abnormally shaped RBCs do not live as long as regularly shaped cells, so children with sickle cell disease have a low RBC count and must make new RBCs more quickly.
- Children are born with this condition and have it for life. Some children are more severely affected by it; some have a milder form.
- There are different kinds of sickle cell disease, depending on the type of abnormal hemoglobin causing the disease: sickle cell anemia, sickle cell–hemoglobin C disease, and sickle cell–thalassemia disease are the most common.
- Stem cell or bone marrow transplantation can sometimes be used to replace the cells in the child that make the abnormal hemoglobin. Stem cell or bone marrow transplantation is limited by the need to find a genetically matched donor (usually a sibling) and the risks involved in using the treatment.

How common is it?

- About 90,000 to 100,000 people in the United States have sickle cell disease. It is most common in people of African or Mediterranean descent.
- People with sickle cell *trait* inherited an abnormal hemoglobin from only one parent. They do not have sickle cell disease and are generally healthy. Children with sickle cell trait usually have no special requirements. About 2 million Americans have sickle cell trait. They can pass the abnormal hemoglobin gene to their children.

What are some common characteristics of children who have sickle cell disease or of sickle cell disease as children present with it?

- Children with sickle cell disease may have a yellow tint to their eyes as a by-product of the breakdown of RBCs. They may be small or slender for their ages.



Normal red blood cell and sickled red blood cell

- Some children with more-severe disease require regular blood transfusions.
- Children with sickle cell disease may have increased absences because of complications and may need to be hospitalized for treatment.
- Some complications include
  - Pain
    - Pain can happen in any part of the body but often occurs in the hands, feet, or joints. Severe pain can start suddenly, without warning, when the sickle cells fail to deliver oxygen to body tissues.
    - Chest pain can be especially serious. Acute chest syndrome
      - Results from blockage of blood flow to the lungs and from infection
      - Is like pneumonia and includes symptoms such as cough, difficulty breathing, chest pain, and fever
      - Can worsen quickly and be a serious problem
    - Dactylitis, or hand-foot syndrome, is swelling and pain in the hands and feet. It usually affects infants and toddlers and can be associated with fever.
  - Infection
    - Children with sickle cell disease can have a hard time fighting infection. The abnormal cells can interfere with the body's ability to clear out and destroy bacteria.
    - Fevers must be evaluated urgently by the child's health care professional. The child should undergo laboratory blood studies and antibiotic administration to prevent complications.
    - Pneumonia can be very serious in children with sickle cell disease.

Sickle Cell Disease (*continued*)

- Splenic sequestration
  - Splenic sequestration is an emergency.
  - The spleen is an organ in the upper-left section of the abdomen next to the stomach. The spleen acts to strain the blood and remove damaged cells and infection. Sickle cells can clog the spleen and keep it from working properly. Sometimes, sickle cells get especially clogged in the spleen and cause the blood to back up. The spleen can get very big if this backup happens and can sometimes break open, which is a life-threatening emergency.
  - Patients who have splenic sequestration may have sudden weakness, fast breathing, pale lips, and abdominal (belly) pain on the left side of the body.
  - Over time, the spleen becomes injured and does not function.
- Aplastic crisis
  - Normal RBCs live about 90 to 120 days, but sickle cells last only 10 to 20 days.
  - If something such as a viral infection prevents the body from keeping up with making new RBCs, RBC counts can drop and the child can get a dangerously low RBC count very quickly. If this drop happens, the child can appear very pale and tired.
- School performance
  - Children with sickle cell disease may have difficulty in school.
  - Some children with sickle cell disease have silent cerebral infarctions, such as "mini-strokes," that do not show obvious neurologic signs but can both affect learning and increase the risk for a larger stroke. These children might need extra academic supports.
- Strokes
  - If sickle cells block the blood flow to the brain, a stroke can occur.
  - Signs of stroke include headache, weakness of a body part, seizure, and speech problems.
  - Strokes require emergency evaluation and treatment.
- Skin ulcers
  - Skin ulcers can develop.
  - Skin ulcers need to be treated promptly, if they develop.
- Priapism
  - Boys with sickle cell disease may experience painful penile erections that last for a long time because of poor blood flow. This type of erection is called *priapism*.
  - Priapism requires emergency treatment.

- Other blood diseases may share some characteristics of sickle cell disease. These are called hemoglobinopathies, in which RBC proteins are abnormal.

Who might be on the treatment team?

- Children with sickle cell disease may receive their primary medical care within their medical homes, from their pediatricians/primary care providers, or within a specialty clinic. Check with parents/guardians about who the first point of contact should be.
- Hematologists are the specialists who care for children with blood diseases.

What adaptations may be needed?

Medications

- Children with sickle cell disease may take penicillin from 2 months until at least 5 years of age to help prevent infection. Erythromycin may be substituted in children who are allergic to penicillin. Pain crisis is treated with pain medications such as acetaminophen (eg, Tylenol), codeine, and ibuprofen (eg, Motrin, Advil). Extra amounts of folic acid may be required because of the extra RBCs that are needed. If the pain is severe, stronger narcotic (pain-relief) medications given by mouth, such as oxycodone, may be needed. When the pain is severe and uncontrolled by oral medication, the child may need to be seen in the emergency department or admitted to the hospital and administered intravenous pain-relief medications.
- Children with severe forms of sickle cell disease may be taking hydroxyurea. Hydroxyurea is a medication given by mouth daily to decrease complications of sickle cell disease such as pain, hospitalizations, and the need for blood transfusions. It also increases the energy of these children by raising their hemoglobin levels. It may also prevent damage to the brain and other organs.
- All staff who will be administering medication should have medication administration training (see Chapter 6).
- Children who have certain complications that require long-term blood transfusions may be taking medications to get rid of the excess iron that accumulates from frequent transfusions.

Sickle Cell Disease (*continued*)

Immunizations

- It is especially important for children with sickle cell disease to undergo all the routine immunizations and receive some special vaccines. *People with sickle cell disease should receive all recommended childhood vaccines.* They should also receive additional vaccines to prevent other infections to which they are especially susceptible. In addition to the pneumococcal conjugate vaccine, known as PCV13, children with sickle cell disease should receive the pneumococcal polysaccharide vaccine known as PPSV23 after 2 years (ie, 24 months) of age and again 5 years later. A child with sickle cell disease should receive the meningococcal conjugate vaccine at 2, 4, 6, and 12 to 15 months of age, a booster dose 3 years after finishing the initial series, and another booster dose every 5 years after that.
- Everyone should be fully immunized with influenza vaccine at the start of each influenza season. Being fully immunized with influenza vaccine is especially important for children with sickle cell disease and for the people with whom they have contact, to protect themselves and the children in their care.

Dietary Considerations

Children with sickle cell disease should have at least 8 cups of water or a healthy fluid per day.

Physical Environment and Other Considerations

- Hydration helps prevent sickling, so children who have sickle cell disease should be allowed to have a water bottle. Encourage good hydration at all times, especially during physical activity.
- Children with sickle cell disease may need more bathroom breaks, as they are prone to frequent urination.
- Most children with sickle cell disease have normal activity, but allow them to rest if they tire easily from anemia.
- Avoid extreme temperatures, both hot and cold, and temperature fluctuations. Never apply an ice pack to an injury of a child with sickle cell disease.

Transportation Considerations

Special consideration should be given to transport to and from the child care center or school because vehicles can be very warm or cold, either of which can increase sickling of RBCs.

What should be considered an emergency?

- Call parents/guardians immediately for
  - Fever
  - Pain that does not improve with medication and rest
  - Cough or mild chest pain
  - Abdominal pain or swelling
  - Paleness or increased tiredness
- Call emergency medical services (911) if the child has
  - Difficulty breathing
  - Seizure or loss of consciousness
  - Severe headache or dizziness
  - Change in vision
  - Numbness or inability to move a body part
  - Slurred speech
  - Severe pain
  - Abdomen enlargement on the left side, just below the ribs, indicating that RBCs may be dangerously backed up in the spleen (sequestration)
  - Prolonged, painful erection (priapism)

What types of training or policies are advised?

- Recognizing an impending crisis and signs and symptoms of complications
- Responding to emergencies
- Medication administration

What are some resources?

- Centers for Disease Control and Prevention: "Facts About Sickle Cell Disease" (Web page), www.cdc.gov/ncbddd/sicklecell/facts.html
- National Heart, Lung, and Blood Institute: www.nhlbi.nih.gov, 301/592-8573
- Regional sickle cell disease centers (Contact local resources or children's hospitals for more information.)
- Sickle Cell Information Center: www.scinfo.org, 404/727-7857

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