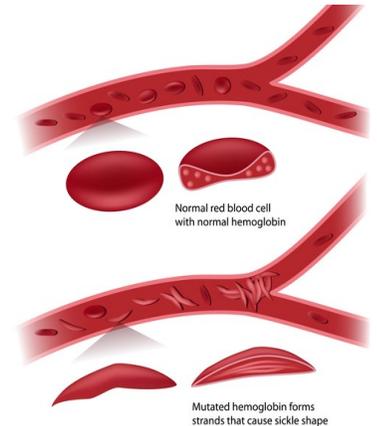


What is it?

Sickle cell disease (SCD) is defined as a group of inherited, lifelong red blood cell disorders. Healthy red blood cells that contain normal hemoglobin are shaped like a round disk or doughnut; allowing them flexibility as they move through both large and small blood vessels. Hemoglobin is a protein that carries oxygen from the lungs throughout the body. Individuals with SCD have red blood cells that contain abnormal hemoglobin (hemoglobin S). When hemoglobin is abnormal, these red blood cells become “sickled” or shaped like a crescent moon. They are inflexible and sticky, causing them to adhere to vessel walls and clog blood flow. Oxygen then cannot reach the intended target. Without proper tissue oxygenation, the child can experience sudden and severe pain. The unpredictable nature of pain is one of the biggest challenges of SCD. While normal red blood cells live about 90-120 days, sickled cells tend to hemolyze and only survive 10-20 days. The body struggles to produce new red blood cells and the individual will have an unusually low red blood cell count; a condition called anemia. There are various forms of SCD, ranging from mild to severe. The most common, and most severe form of SCD, is sickle cell anemia (HbSS). Other common types of SCD include HbSC and HbS beta thalassemia. HbSD, HbSE, and HbSO are considered rare.



What are the signs and symptoms?

Symptoms usually begin once the child is about 5-6 months old. Early symptoms can include painful swelling of the hands and feet, fatigue, fussiness, anemia, and jaundice. Other symptoms could include:

- Pain
- Weakness
- Lethargy
- Abdominal pain
- Fever
- Joint pain
- Frequent urination
- Perspiration
- Headache
- Shortness of breath
- Delayed growth
- Vision problems

Other symptoms should be considered warning signs that require immediate action. These include fever, noticeable change in complexion, rapid or difficult breathing, rapid heartbeat, sudden onset blurry vision, sudden onset pain, swelling of hands/feet/joints, or muscular weakness.

What conditions can precipitate a painful crisis?

- Stress
- Dehydration
- Increased physical activity
- Air pollution
- Illness
- Caffeine
- Lack of sleep
- High altitudes
- Infection
- Alcohol
- Sudden temperature changes
- Cold or damp conditions

Exposure to cold can constrict blood vessels and leave them prone to blockage and pain. Exposure to heat increases the likelihood of dehydration and overexertion; affecting blood flow and sickling of cells.

Complications

- Pain (sharp, intense, stabbing, or throbbing)
- Acute Chest Syndrome
- Stroke
- Pulmonary hypertension
- Organ damage
- Ulcers
- Delayed growth and puberty
- Mental health complications



Individuals with SCD have an increased susceptibility to infection. Common illnesses, like the flu, can be dangerous. They may follow a special vaccination schedule because they are considered ‘high risk.’ Individuals may also take prophylactic penicillin every day until at least age 5 to prevent infection.

What is the treatment?

There is no cure for SCD and it can worsen over time. Treatment medications can include hydroxyurea, analgesics, and antibiotics. Other treatments include increasing fluid intake, massage, relaxation, distraction, and use of heating pads.

Please note a cold pack should never be applied to an injury or pain site in a child with SCD.

Suggested school accommodations

- Water bottle with student at all times
- Encourage fluid intake based on plan
- Liberal bathroom privileges (fast/flash pass)
- Extra set of books
- Copies of notes
- Accommodations/considerations during extreme temperatures (consider temperature >80 or <45F)
 - Considerations for classroom temperatures or preferential seating locations (consider windows, vents, fans, heaters)
 - Note transportation needs including bus stops location and routes
 - Allow wait time indoors before/after school
 - Avoid exercise in extreme conditions
 - Permit layered clothing, jackets, hats
- Use of elevator
- Extra time to/from class (consider location/distance between)
- Modified physical education and recess (per provider orders)
- Allow student to self-limit activity (as appropriate)
- Allow rest breaks or designate a rest area
- Monitor for changes or declines in academic performance (learning difficulties may be associated with stroke)
- 504/IEP considerations
- Consider chronic health illness program
- Coping skills, opportunities to increase self-esteem
- Staff training for SCD and student specific plans
- Emergency planning, including warning signs that require action

SHNIC school nurses information:

Specific health issues for individual health care plans

- Sickle cell disease diagnosis and genotype
- Student's health complications related to SCD (such as history of blood transfusions, stroke, etc.)
- Student specific treatment plan including caregiver contact information, student symptoms, triggers
- Health consents (including SCD specific treatment team)
- Student specific pain scale with parameters (mild, moderate, severe) and check-in plan to assess pain
- Current medications including scheduled and PRN, also noting side effects (both home and school)
- When to contact caregivers, 911 services, and preferred hospital for treatment needs
- Baseline vital signs including heart rate, blood pressure, respiratory rate, capillary refill time, and pulse oximetry
- Emergency action plan
- Education of appropriate school staff for emergency action (including signs and symptoms of stroke, respiratory distress, modified first aid measures, etc.)
- Bus transportation needs and temperature considerations (type of bus, temperature control, route, and locations of pickup)
- Medical alert bracelet or AAP emergency card recommendations

Resources & Manuals

CDC– Tips for Supporting Students with Sickle Cell Disease

https://www.cdc.gov/ncbddd/sicklecell/documents/tipsheet_supporting_students_with_scd.pdf

NASN– Individualized HCP for a student with Sickle Cell Anemia

http://portal.nasn.org/text/6905_W22_6_001-003.pdf

Maryland Sickle Cell Disease Association

<http://www.marylandsicklecelldisease.org/>

Children's Mercy: Educator's Guide to Sickle Cell and School

https://www.childrensmercy.org/library/uploadedFiles/childrensmercyorg/Clinics_and_Services/Clinics_and_Departments/Hematology_and_Oncology/2015%20SickleCellGuide-Educators-HemOnc.pdf