SICKLE CELL DISEASE

What is it?

Sickle cell disease (SCD) is defined as a group of inherited, lifelong red blood cell disorders. Healthy red blood cells containing 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).

What are the symptoms or characteristics?

Initial 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 of SCD could include pain, headache, weakness, shortness of breath, lethargy, frequent urination, delayed growth, abdominal pain, and vision problems.

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

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.

Kennedy Krieger Institute

The **Specialized Health Needs Interagency Collaboration (SHNIC)** program is a collaborative partnership between the Kennedy Krieger Institute and the Maryland State Department of Education.

Suggested school accommodations

Students with sickle cell disease can have various symptoms since it is a chronic condition and can vary from other students with the same disease. Supporting students with these conditions in the school require educators and parents/guardian to work as a team. Some accommodations to consider for a 504/IEP could include:

- PT/OT/SLP evaluations
- Modified or flexible school day
- Plan for absences and make-up work
- Water bottle with student at all times
- Encourage fluid intake based on plan
- Liberal bathroom privileges (fast/flash pass)
- Accommodations/considerations during extreme temperatures (consider temperature >80 or <45F) including classroom temperatures and bus stops location and routes
- Allow wait time indoors before/after school
- Permit layered clothing, jackets, hats
- Preferential seating locations (consider windows, vents, fans, heaters)
- Extra time to/from class (consider location/distance between)
- Extra set of books
- Copies of notes
- Use of elevator
- Modified physical education and recess, avoid exercise in extreme conditions
- 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)
- Coping skills, opportunities to increase self-esteem
- Staff education/training as appropriate
- Emergency Evacuation Plan (EEP)

Specific health issues for Individualized Healthcare Plan

- Sickle cell disease diagnosis and genotype, student specific treatment plan
- 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
- 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
- Bus transportation needs and temperature considerations (type of bus, temperature control, route, and locations of pickup)
- Medical alert bracelet or AAP emergency card recommendations
- Communicate with school staff, parents/guardian, and provider any changes or concerns about the disease
- Emergency Care Plan (ECP) related to medical needs in the school setting and staff training as appropriate for each

Resources & Manuals

Kennedy Krieger Institute: Sickle Cell Neurodevelopmental Clinic
https://www.kennedykrieger.org/patient-care/centers-and-programs/sickle-cell-neurodevelopmental-clinic

Centers for Disease Control and Prevention: Tips for Supporting Students with Sickle Cell Disease
https://www.cdc.gov/ncbddd/sicklecell/documents/tipsheet_supporting_students_with_scd.pdf

Sickle Cell Disease News
https://sicklecellanemianews.com/

Maryland School Health Services Guideline: Management of Sickle Cell Disease in Schools
http://marylandpublicschools.org/about/Documents/DSFSS/SSSP/SHS/SHSGuidelines/SickleCellDiseaseGuidelines.pdf