

# SICKLE CELL DISEASE

## Background

Sickle cell disease (SCD) is a group of inherited red blood cell disorders characterized by sickle-shaped RBCs due to abnormal hemoglobin formation. Hemoglobin is a protein in red blood cells that carries oxygen from the lungs throughout the body. 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. Individuals with SCD produce abnormal hemoglobin known as hemoglobin S (HbS). The abnormal hemoglobin in SCD cause the red blood cells to become “sickled” or shaped like a crescent moon. These cells are inflexible and sticky causing them to adhere to blood vessel walls, clog blood flow, and prevent oxygen from reaching the intended target. Without proper tissue oxygenation, the child can experience sudden and severe pain, anemia, or other serious complications. There are various forms of SCD, ranging from mild to severe. The most common and most severe form of SCD is sickle cell anemia (Hb SS).

Initial symptoms usually begin once the child is about five to six 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, including:

- Fever
- Noticeable change in complexion
- Rapid or difficulty breathing
- Rapid heartbeat
- Sudden onset blurry vision
- Sudden onset pain
- Swelling of hands, feet, joints
- Muscular weakness

Common sickle cell crisis triggers include infection, stress, dehydration, cold and/or damp conditions, air pollution, extreme physical activity, sudden changes in temperature, and air altitude. Other factors include use of caffeine, alcohol or smoking.



Individuals with SCD have an increased susceptibility to infection. Fever is a sign of infection and should be taken seriously. Individuals may take prophylactic penicillin to prevent infections or follow a special vaccination schedule because they are considered high risk. Other complications of SCD include pain, acute chest syndrome, stroke, pulmonary hypertension, organ damage, ulcers, delayed growth and puberty, and mental health complications.

## Top Takeaways for School Considerations

The student’s emergency plan should communicate how to recognize, treat, and/or seek medical attention for signs and symptoms of SCD crises. Be responsive to complaints of pain.

It is important for the student to stay hydrated to prevent pain episodes and other health problems. Unlimited access to water throughout the school day or carrying a water bottle is encouraged.

Changes in temperature conditions can trigger pain crisis. Preferential seating may help alleviate a drafty location that is close to a vent, fan, or window.

SCD is associated with cognitive effects both from direct effects of SCD on brain function (e.g., seizure, stroke) or indirect effects of chronic illness and absenteeism.

Specific learning disabilities, language issues, and executive dysfunction may also be seen.

## Kennedy Krieger Institute’s Specialized Health Needs Interagency Collaboration

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



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## Considerations for the Individualized Healthcare Plan (IHP)

- Nursing diagnosis of risk for ineffective tissue perfusion, risk for altered cardiovascular tissue perfusion, risk for activity intolerance, risk for injury, chronic pain, acute pain, fatigue, risk for peripheral neurovascular dysfunction, impaired sensory perception, and potential for impaired social interaction
- Current diagnosed health condition including date of diagnosis, progress of disease process and other chronic health conditions
- Current medication and treatment orders (consider schedule, equipment needs and side effects)
- Student-specific triggers, avoidance, or intervention strategies
- Temperature regulation considerations in school setting and transportation
- Use of specialized equipment, adaptive equipment, and orthotics
- Activity, positioning, transferring (consider precautions and/or restrictions)
- Consider emergency care plan(s) (ECP) and emergency evacuation plan(s) (EEP) as related to medical needs in the school setting, and staff education/training, as appropriate

## Discussion Starters for Educational Team

1. Has the school staff been trained to implement the student-specific emergency plan?
2. Would the student benefit from evaluations or assessments in any of the following areas: physical therapy, occupational therapy, speech and language therapy, assistive technology, adapted physical education, functional behavior, psychology, hearing and vision?
3. Would the student benefit from additional academic support and/or modified education (e.g., copies of notes, extra time, reduced workload, simplified instructions, alternative formats for presentation of material, 504/IEP)?
4. Can strategies be implemented to assist the student with executive function (e.g., plan, prompts, organizers, agendas)?
5. Would schedule flexibility support the student?
6. Does the student require activity precautions to prevent injury?
7. Is the physical school environment safely accessible for the student's mobility needs (e.g., location of classes, access to elevator)?
8. Does the classroom environment support the student's needs and/or equipment (e.g., desk/seating options, flash pass for bathroom or nurse)?

## Resources

Kennedy Krieger Institute: Sickle Cell Neurodevelopmental Clinic  
[kennedykrieger.org](http://kennedykrieger.org)

Maryland Sickle Cell Disease Association  
[marylandsicklecelldisease.org/](http://marylandsicklecelldisease.org/)

St. Jude Children's Research Hospital: Educational Resources for Sickle Cell & Other Blood Disorders  
[stjude.org/treatment/disease/sickle-cell-disease/educational-resources.html](http://stjude.org/treatment/disease/sickle-cell-disease/educational-resources.html)



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