Sickle Cell Disease

Background

Sickle cell disease (SCD) is a group of inherited red blood cell (RBC) disorders characterized by sickle-shaped RBCs due to abnormal hemoglobin formation. Hemoglobin is a protein in RBCs that carries oxygen from the lungs throughout the body. Healthy RBCs 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 RBCs 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.

A sickle cell crisis is an acute, often painful complication of SCD caused by the obstruction of blood flow. A vaso-occulsive pain crisis is the most common. Other types include an aplastic crisis, splenic sequestration crisis, acute chest syndrome, and hemolytic crisis. 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

Triggers for a SCD crisis 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.

Top Takeaways for School

The student's emergency plan should communicate how to recognize, treat, and/or seek medical attention for signs and symptoms of SCD crisis. 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 minimize exposure to drafty areas near vents, fans, or windows.

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.

Considerations for the Individualized Healthcare Plan (IHP)

- Nursing diagnoses: 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
- Student-specific triggers, avoidance, or intervention strategies
- Temperature regulation considerations in school setting and transportation
- Activity, positioning, transferring (consider precautions and/or restrictions)
- Use of specialized equipment, adaptive equipment and orthotics
- Consider emergency action plans (EAPs) and emergency evacuation plans (EEPs) related to special health care needs, including staff education/training

Discussion Starters for the Educational Team

1. 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?
2. 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)?
3. Can strategies be implemented to assist the student with executive function (e.g., plan, prompts, organizers, agendas)?
4. Is the physical school environment safely accessible for the student's mobility needs (e.g., location of classes, access to elevator)?
5. Will staff receive education/training to implement the student-specific emergency plan?

Resources

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

Your Child's Brain Podcast
kennedykrieger.org/stories/your-childs-brain-podcast

Maryland Sickle Cell Disease Association
marylandsicklecelldisease.org/

St. Jude Children's Research Hospital: Educator's Guide to Sickle Cell Disease
treatment.stjude.org/content/dam/sj-treatment/hematology-resources/educators-guide-to-scd.pdf

For more information, please scan the QR code or visit: KennedyKrieger.org/SHNIC

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