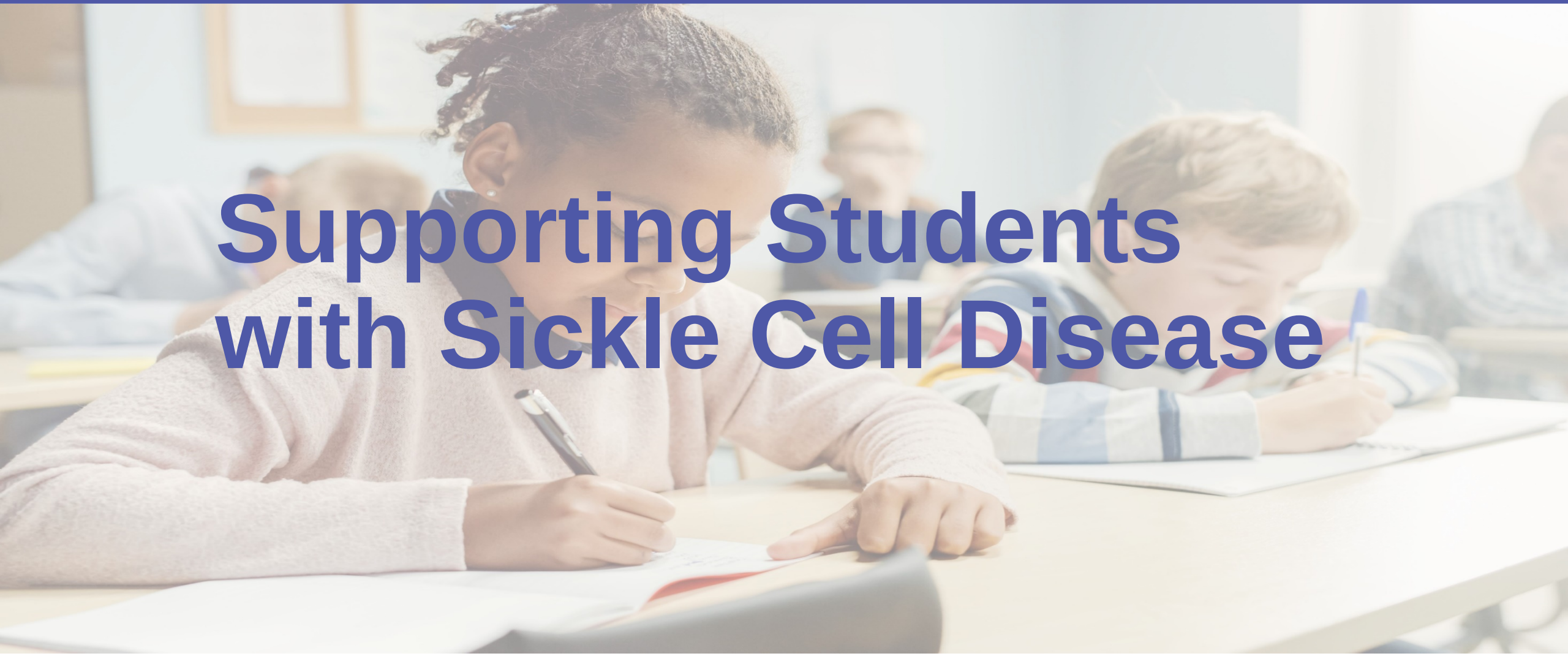

Supporting Students with Sickle Cell Disease

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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Objectives

1. Describe basic etiology and common symptoms of sickle cell disease (SCD).
2. Discuss management of the student's health care needs including potential complications and interventions to keep the student safe in school.
3. Identify the educational impact of the disease and potential strategies and accommodations to promote academic success.

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Sickle cell disease 1 of 3

- Group of red blood cell (RBCs) disorders affecting hemoglobin, the protein that carries oxygen through the body
- Abnormal hemoglobin causes the cells to become hard, sticky, sickled or banana-shaped
- Sickled RBCs adhere to vessel walls and clog or block blood flow, causing less oxygen to reach these areas
- Lack of oxygen causes pain and damage throughout the body
- Various types

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Sickle cell disease 2 of 3

- Most common genetic disorder identified in African Americans.
- Also found in people who are descendants of South and Central America, the Mediterranean, and the Middle East.
- Inherited blood disorder
- No universal cure

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Sickle cell disease 3 of 3

- Main clinical feature is the "sickle cell crisis" or vaso-occlusive crisis (VOC) causing an acute painful event
- Hydroxyurea is an FDA approved medication to treat children
- Complications requiring rapid initiation of emergency treatment:
  - Acute chest syndrome
  - Spleen crisis
  - Fever
  - Stroke

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Symptoms

Severity and presentation of symptoms vary widely from person to person and change over time.

- Pain (bone, joint, hip, abdominal)
- Headache
- Fatigue, weakness
- Vision problems
- Jaundice
- Frequent infection
- Anemia
- Swelling of hands and feet
- Shortness of breath
- Frequent urination
- Perspiration
- Fussiness
- Delayed growth
- Coldness in extremities

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Triggers

The exact cause of a SC crisis is not always understood. It is believed that anything that causes your blood vessels to constrict can trigger a crisis.

- Infection
- Stress
- Dehydration
- Cold and/or damp conditions
- Air pollution
- Extreme physical activity
- Sudden changes in temperature
- Air altitude
- Caffeine
- Alcohol, smoking

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Pain

- Vaso-occlusive crisis (VOC)
- Acute episodes of severe pain
- Can occur without warning
- Frequently affects extremities, chest, back
- Young children can have severe pain and swelling of both hands and feet (dactylitis)
- Musculoskeletal pain is the most common complaint in older children and adults

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Acute chest syndrome

- Caused by infections and/or a blockage of blood flow to the chest and lungs
- May develop during the treatment of VOC
- Creates a pneumonia-like illness, including symptoms such as fever, cough, chest pain, labored or rapid breathing, shortness of breath, tachypnea
- Can lead to respiratory distress without intervention
- Using an incentive spirometer is the best way to prevent
- Children prone to repeated attacks

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Spleen crisis

- Damage to the spleen and poor function occurs early on in childhood for those with SCD
- RBC's become trapped in the spleen causing it to fill with blood and enlarge
- Loss of blood in circulation can lead to hypovolemic shock
- Symptoms include:
    - Increasing pain left side
    - Abdominal pain
    - Increasing pallor
    - Lethargy
    - Increased heart rate

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Stroke

- Ischemic stroke caused by blockage in blood flow is more common in children
- Hemorrhagic stroke is more common in adults with SCD
- Symptoms include:
    - Severe headaches
    - Facial drooping
    - Slurred speech
    - Dizziness
    - Sudden onset of weakness or numbness "painless limp"

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“Silent” stroke

- "Silent stroke" or silent injury to white matter of the brain more common in children
- No outward physical motor symptoms
- Symptoms such as difficulty paying attention, struggle to stay awake, problems with memory
- Debilitating effects on executive function
- Symptoms may be first reported by classroom teacher

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Other health complications

- Bone disease caused by lack of blood flow to joints (hips and shoulders most common)
- Eye disease caused by lack of oxygen to retina (annual screening recommended starting at age 10)
- Gallstones, jaundice related to increased breakdown of RBC's
- Priapism-painful obstruction of blood vessels in the penis
- Fatigue related to anemia, sleep apnea
- Delayed puberty by about 2 years

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Symptoms requiring immediate action

Acute illness characteristics of common childhood illnesses may rapidly progress and become life-threatening

- Fever
- Vision changes
- Rapid heartrate
- Difficulty breathing
- Facial asymmetry
- Severe headache
- Chest pain
- Difficulty speaking, slurred speech
- Muscle weakness, inability to use extremities
- Nausea/vomiting
- Change in alertness/confusion
- Stomach pain or swelling

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Role of the school nurse

- Nursing appraisal and assessment including baseline vital signs, pain scale
- Medical releases to speak with providers
- Treatment plan from hematologist for emergency plan
- Parameters for contacting parent/guardian, 911
- Preferred hospital for treatment needs
- Education of appropriate school staff for emergency plan
- 504/IEP team member
- Social and emotional needs of the student

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Management goals

- Prevent fatigue
- Prevent infection
- Prevent injury
- Avoid dehydration
- Utilize pain management strategies
- Plan for extreme temperatures
- Encourage medication and vaccine compliance

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Management in school

- Follow student's emergency plan
- Rapid pain assessment (use of consistent and appropriate pain scale)
- Initiation of pain medication (type, route, and dose individualized to the student)
- Monitor of vital signs, oxygen saturation
- Comfort measures (heat, distraction)
- Rule out other causes of pain that may need additional treatment
- Do not initially administer any fever-reducing medication until nurse or parent communicates with healthcare provider

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Impact on education

- Pain is biggest antecedent of school-related challenges
- “Silent strokes” may result in learning and/or behavior problems
- Other negative brain affects related to anemia, decreased oxygen to brain, fatigue
- Increased absences due to specialist appointments and monthly transfusions
- Learning accommodations in the classroom may be necessary
- Development of 504/IEP
- Staff should be trained on student’s emergency plan

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Accommodations to consider

- Flash pass for nurse or health suite
- Encourage fluids, access to water bottle
- Unrestricted access to bathroom
- Plan and place for rest breaks, frequent breaks
- Extra set of books, rolling backpack, convenient locker location, distance between classes
- Preferential seating
- Extended time to travel between classes

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Accommodations to consider

- Temperature regulation considerations
  - Classroom seating (note location of windows, vents, fans, heaters)
  - Curb-to-curb transportation
  - Indoor activities instead of outside (safe temperature range is between 40 and 80 degrees)
  - Allow to dress in layers, hats, gloves, blanket
  - Possible blanket for fire drills in cold weather
- Partial days, flexible scheduling, hybrid learning

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Summary

- Any report of pain, fatigue, shortness of breath and headache should be immediately addressed for a student diagnosed with SCD.
- Staff should be educated on SCD and implementation of the student's emergency plan.
- Pain is subjective and has a profound impact on a student's ability to function in the school setting. It should be monitored and treated as needed.

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Resources

Maryland State School Health Services: Management of SCD in School
marylandpublicschools.org/about/Documents/DSFSS/SSSP/SHS/SHSGuidelines/SickleCellDiseaseGuidelines.pdf

CDC Tips for Supporting Students with Sickle Cell Disease
cdc.gov/ncbddd/sicklecell/documents/tipsheet_supporting_students_with_scd.pdf

Children's Mercy: Educators Guide to Sickle Cell and School
childrensmercy.org/contentassets/586bc7627fa148c6871864ef187d09e2/2015-sicklecellguide-educators-hemonc.pdf

Viriginia Sickle Cell Awareness Program School Handbook
vdh.virginia.gov/content/uploads/sites/65/2020/10/Sickle-Cell-School-Handbooklet-2020WEB.pdf

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