School Accommodations for Sickle Cell Disease

INDIANA HEMOPHILIA & THROMBOSIS CENTER, INC.

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What is Sickle Cell Disease?
Responding to Complications at School
Other Potential Health Problems
Prevention: What Schools Can Do
What is Sickle Cell Disease?

- Group of hereditary blood disorders that affect hemoglobin
- Normal red blood cells contain Hemoglobin A
- Sickled red blood cells contain Hemoglobin S
Hemoglobin S

- Polymerization of Hemoglobin S results in red blood cells that are rigid, sticky, and fragile.
- RBCs with Hgb S have shorter life span and trouble passing through blood vessels.
Why is it Called Sickle Cell Anemia?

- Because of their fragility, sickle cells burst (hemolytic anemia)
  - This lowers the red blood cell count
- Sickle cells live 10-20 days in the bloodstream
  - Normal red blood cells live 120 days
- All blood counts may drop if the bone marrow is suppressed by
  - Infection
  - Folic acid deficiency
Disease Burden

- 1 in 12 African-Americans have Hgb S trait
- 90,000-100,000 Americans have sickle cell disease
  - ~30 children are born in Indiana each year with a hemoglobinopathy
- Mutations arose in countries where malaria is endemic
How is SCD Inherited?
Types of Sickle Cell Disease

- **Hgb SS (Sickle Cell Anemia)**
  - ~60% of U.S. cases
  - Sub-Saharan Africa, Mediterranean, Middle East, India

- **Hgb SC**
  - ~25%
  - West and north Africa

- **Sickle β+/0 Thalassemia**
  - ~10-15%
  - Sub-Saharan Africa, Middle East, India

- **Other compound heterozygous states (Hgb SD, Hgb SE, Hgb SO)**
  - ~5%

- Frequency and severity of complications vary by type of hemoglobinopathy
What About Sickle Cell Trait?

- People with Sickle Cell Trait inherited Hemoglobin S from one parent and Hemoglobin A from the other.
- Trait is a condition of the red blood cell—NOT a disease.
- Health problems may occur under extreme conditions:
  - Dehydration, low oxygen, high altitudes.
RESPONDING TO COMPLICATIONS OF SICKLE CELL DISEASE
Pain

- Caused by sickled cells “clogging” the blood vessels
- Hallmark manifestation of SCD
- Pain can vary from mild to severe
- Ways to treat pain:
  - Pain medication provided by parent
  - Heating pad
  - Hydration and rest
The frequency of pain episodes will vary from person to person.

- 30% of those with SCD will never or rarely have pain.
- 50% will have occasional episodes of pain.
- 20% have frequent, severe pain.
  - 6% of patients account for 30% of all pain episodes.
Pain: What to Do

- Reports of pain should be taken seriously
  - There are usually no physical manifestations of the pain
  - Pain usually occurs in back, abdomen, arms, or legs
- Not always necessary for child with pain to go home
- May only need rest, pain medication, heating pad, etc. until he or she is feeling better and able to rejoin class
- Distractions can also help, especially with younger children
  - Looking at books, listening to music, toys
- Parents should always be contacted when pain is first reported
Acute Chest Syndrome

**What Can Happen**

- Serious complication that can lower levels of oxygen in the blood
- One of the leading causes of death in people with sickle cell disease
- Symptoms include: Fever, cough, chest pain, dyspnea, hypoxia, tachypnea

**What to Do**

- Follow asthma action plan if available
- Call parent immediately
- If symptoms are severe, call 911

Once a child has had one episode of ACS, they are more likely to have it again.
Asthma and Sickle Cell Disease

- Asthma prevalence in SCD population is similar to prevalence in African-American population
- Asthma is associated with an increase in SCD-related morbidity and premature mortality
- Studies show increased incidence in painful episodes and ACS for children with asthma
- Children with asthma:
  - More likely to have ACS
  - More likely to have multiple episodes of ACS
  - More likely to have ACS at a younger age
  - More likely to be hospitalized for longer with ACS
Asthma and Sickle Cell Disease

- Asthma triggers are often sickle cell pain triggers
  - Cold weather
  - Exertion
  - Strong emotions

Optimizing asthma control can help to control symptoms of sickle cell disease
Headache

What Can Happen

- Sickled cells tend to “clump up” along the walls of the large arteries going to the brain
  - Damages vessel walls
  - Exposes tissue that collects more sickled cells and narrows the vessels even further
- 5-10% of children with Hgb SS will have an overt stroke

What to Do

- F.A.S.T.
  - Face: Any facial weakness or drooping?
  - Arm: Can the student lift both arms above their head?
  - Speech: Can the student speak clearly and understand what you say?
  - Time: To call 911 if any of these are present
- Call parent immediately
**Priapism**

- **What Can Happen**
  - Sustained, painful, unwanted erection
  - Failure of venous outflow due to sickling of blood cells
  - ~30% of males with SCD under age 20 have had at least one episode of priapism
  - Can cause impotence

- **What to Do**
  - Give pain medication provided by parent
  - Heating pad
  - Push fluids
  - Call parent if erection does not go down within 30 minutes
What Can Happen

- SCD impairs splenic function, resulting in eventual functional asplenia
- Without the spleen’s filtering function, local infections can readily become systemic

What to Do

- Any fever of 101° or higher is a medical emergency
- Do not give fever reducers
- Call parent immediately
OTHER POTENTIAL HEALTH PROBLEMS
Gallstones and Jaundice

- Gallstones occur in about a third of children with SCD
- Students with SCD may be self-conscious about jaundiced skin or eyes
Other Potential Health Problems

Delayed Growth and Puberty
- Puberty is often delayed in children with SCD
- Children and teens with SCD may be small and thin for their age
- Usually reach full height by age 20

Retinopathy

Sleepiness
- Anemia/Sleep apnea
Other Potential Health Problems

Avascular Necrosis

- Caused by reduced blood flow to the joints
- Usually affects hip or shoulder joints
- Can cause severe pain
- May affect student’s ability to participate in gym activities or walk to and from classes
PREVENTION: WHAT SCHOOLS CAN DO
Plan for Extreme Temperatures

- Alternative indoor recess when 40° F or below
- Access to coat or jacket during fire drills
  - Students may need to return to the school sooner than other students, or have access to climate-controlled vehicle
- Access to sweater or extra layers when in the classroom
- Offer at least one cup of water per hour when outside in the heat
- Bus schedules or routes may need to be changed to ensure that children with SCD do not have to walk long distances or wait a long time for the bus in cold
Avoid Dehydration

- Unlimited access to water or other fluids
  - Some children may need to be encouraged to drink

- Unlimited restroom breaks
  - Needed due to increased fluids as well as damage to the kidneys from the sickled cells
Prevent Fatigue and Pain

- Allow frequent breaks during gym activities and sports events, as requested by the student
- Listen to and act quickly upon reports of pain or headaches
- Never apply ice to cuts or bruises
- Use caution with water activities/swimming
  - Consult with patient’s hematologist for guidelines
- **Provide two sets of textbooks**
- **Sickle Cell Trait**
  - Strenuous activities
Special Accommodations

- Studies have shown that children with SCD miss an average of 20-40 school days a year
  - Participants in one study missed an average of 12% of the school year, with 35% of students missing at least one month of school
  - Absences may be due to pain crises treated at home, serious health complications requiring hospitalization, or frequent medical appointments
Special Accommodations

- **Students may require 504/IEP**
  - May include accommodations such as providing two sets of textbooks (one at home, one at school), plans for getting homework assignments to the child, testing/ISTEP accommodations
  - Educational services in the home/hospital

- **Due to the unpredictable nature of SCD and the varied disease course, accommodations should be tailored for each child, but every child should have an individualized health care plan**
SCD and Learning Difficulties

- Children with SCD may struggle in school due to damage to brain tissue
  - Overt strokes
  - Silent strokes (up to 30% of children with SCD)
  - Hypoxia from anemia, poor pulmonary functioning, sleep apnea

- Report unexplained sleepiness or trouble concentrating to parents

- May need a referral for neurodevelopmental testing
Living with Sickle Cell Disease

- 83% Medicaid
- 83% African American
- 80% WIC
- 61% under 20,000 income

Sickle SAFE Program, March 2014
Living with Sickle Cell Disease

- School personnel must be aware of the stigma surrounding sickle cell disease
  - Students may be ashamed of diagnosis
  - Students may feel self-conscious about physical manifestations of SCD, such as jaundice or delayed growth and puberty
  - Students may worry about being different than their peers, or being a burden to their families

- People who experience frequent painful crises are more likely to have:
  - Low self-esteem
  - Anxiety and depression
  - Poor school performance
  - Isolation
  - Decreased participation in activities of daily living
  - Poor peer and family relationships
The Child with Sickle Cell Disease...

- Should be taken seriously when they present with any reports of pain or fatigue
- Should be encouraged to participate in school activities as much as possible
- Should be discouraged from thinking of themselves as “sick” or less capable than other children
Handout Requests

- Please email me at ebloom@ihtc.org to request electronic copies of today’s handouts
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