Comprehensive Sickle Cell Disease and Thalassemia Program

Facts about Sickle Cell Disease for Teachers, Coaches and School Nurses

SICKLE CELL DISEASE IS A GENETIC DISORDER that affects the hemoglobin in the red blood cells. Hemoglobin is a protein that carries oxygen to different parts of the body. The three most common types of Sickle Cell Disease are hemoglobin SS disease (also called sickle cell anemia), hemoglobin SC disease and sickle beta thalassemia.

Teachers, coaches and school nurses need to help a student who has sickle cell disease function as effectively as possible. Take a proactive approach by knowing what to do if a child with Sickle Cell Disease becomes ill at school.

EVERYDAY NEEDS OF THE STUDENT WITH SICKLE CELL DISEASE

- *Provide lots of fluids.* Make sure that your student always has fluids readily available in class. We recommend keeping a water bottle at his/her desk and during physical education. Fluids assist the red blood cells in moving more easily throughout the blood vessels, which ultimately decreases the number of pain crises your student may experience.

- *Provide liberal bathroom privileges.* Due to high fluid intake and because their kidneys do not function as well as those of healthy children, allow your student to use the bathroom when needed.

- *Avoid physical exhaustion.* Students with Sickle Cell Disease may tire more easily than other children. Allow your student to self-limit activity and rest when needed. However, we encourage inclusion in most activities without restrictions. If in doubt about the student's participation in certain activities, consult with the parents and the Comprehensive Pediatric Sickle Cell Program.

- *Avoid extreme temperatures.* Temperature extremes can trigger a pain crisis. Students should not be exposed to temperature extremes for long periods ($<40^{o}$ or $>80^{o}$). In hot weather, your student may need more frequent breaks for rest and water or juices to avoid dehydration. In cold weather, layered clothing, including boots and hats, should be worn and available during outside emergency drills. Temperatures in the classroom must also be moderately regulated. Classrooms that are too cold or too hot may cause increased red blood sickling and precipitate a pain crisis.

- Monitor for social distress or bullying. Students with Sickle Cell may experience poor growth, yellowing of the eyes and skin, and other body changes. They may experience concerns with self-image or self-esteem. It is important for adults to be mindful of teasing by other students. The school counselor or school psychologist may be helpful. Any concerns should be discussed with parents.

COMMON MEDICAL COMPLICATIONS OF SICKLE CELL DISEASE

Sickle Cell disease causes the red blood cells to become abnormal. Some red blood cells lose their oxygen, become sticky and stiff, and become shaped like a sickle or 'c'. The sickled cells do not move through the blood stream easily and can cause problems in multiple body systems.

- *Pain Crises*. A pain crisis may occur either gradually or suddenly. The crisis occurs when normal blood flow is blocked by the sickled cells and can happen anywhere in the body. The following strategies can be used to help address the pain.
    - A child with Sickle Cell Disease may be treated with acetaminophen or ibuprofen and opioids.
    - Give medications as soon as pain begins.
    - Give at least 8 ounces of water or juice to drink each hour.
    - Apply warm compresses and offer distractions, such as quiet activities or music.
    - If the pain persists for more than an hour or two without relief, call your student's parents. A visit to the hospital may be necessary.

- *Acute Chest Syndrome.* This is a medical emergency. It is believed to be caused by sickled red blood cells clogging blood vessels in the lungs. Sometimes, acute chest syndrome is caused by pneumonia or can lead to pneumonia. If your student experiences any of the symptoms listed below, contact his/her parents immediately.
    - Fever > 101 degrees
    - Chest pain
    - Congestion
    - Cough
    - Trouble breathing or shortness of breath
    - Rapid breathing

- *Anemia*. Normal red blood cells live for 90-120 days. Sickled cells live only 10-20 days. Having fewer healthy red blood cells causes anemia. Some symptoms of anemia are listed here.
    - Tiredness or decreased energy
    - Weakness
    - Paleness/pallor
    - Headache
    - Dizziness

- *Fever.* Fever, or temperature greater than 101 degrees, is often the first sign of infection. Sickle Cell Disease often causes damage to the spleen so children with Sickle Cell Disease are not able to fight infection normally. The student may need to receive IV antibiotics. Call the student's parents immediately. **Do not give acetaminophen or ibuprofen so as not to mask the fever.**

- *Stroke.* Sickled cells can block the blood vessels in the brain and keep the brain from getting enough oxygen. About 12% of children with Sickle Cell Disease have a stroke by age 18, with the average age between 4 and 6 years old. If you notice any cognitive changes, no matter how subtle (i.e. less attentive than usual or changes in school performance) discuss these concerns with the parents as soon as possible.
  If you notice any of the following, call 911 and his/her parents right away!
  - Weakness
  - Confusion
  - Difficulty swallowing
  - Unsteady gait
  - Inability to move one side of face or arms/legs
  - Seizure or loss of consciousness
  - Slurred speech
  - Severe headache

- *Jaundice.* Jaundice, or yellow coloring of the eyes or skin, is caused by bilirubin, a by-product of red blood cell destruction. Since sickled red blood cells are destroyed more rapidly than normal cells, the liver cannot filter them. The destroyed cells build up in the blood stream (bilirubin) and cause the yellow color.

- *Priapism.* This is a painful obstruction of blood vessels in the penis. The pain crisis should be managed in the way other pain is managed and parents should be notified if there is a consistent concern.

ACCOMMODATIONS FOR DISABILITY OR SPECIAL EDUCATION

The *Rehabilitation Act of 1973* is a civil rights law that includes protections against disability discrimination in the school setting. *Section 504* of the Act defines disability as a physical or mental *impairment that substantially limits one or more major life activity*. Major life activities include learning, reading, thinking, writing and concentrating. The student must have an impairment (or history of an impairment) that is not temporary. The impairment must interfere with the identified life activity.

When a student has Sickle Cell Disease, he or she would be eligible for a 504 Plan at school when the disease has a negative impact on a major life activity (learning, thinking). Due

to the nature of Sickle Cell, nearly all children with the disease are eligible for 504 Plan accommodations at school.

The Individuals with Disabilities Education Act (IDEA) is the federal education law that provides for specialized instruction when a child has a disability that interferes with academic success. Many children with Sickle Cell Disease qualify for special education under the category of *Other Health Impaired* when their learning is negatively impacting by the disease. Sickle Cell Disease does increase the risk of cognitive/learning difficulties.

CONTACTS AT NATIONWIDE CHILDREN'S HOSPITAL

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SICKLE CELL DISEASE INFORMATION FOR SCHOOL PERSONNEL

https://www.nhlbi.nih.gov/health-topics/sickle-cell-disease
http://scinfo.org
https://www.sicklecellsociety.org/resource/paediatricstandardsresource/
https://kidshealth.org/en/parents/sickle-cell-anemia.html
https://rarediseases.info.nih.gov/diseases/10333/sickle-beta-thalassemia

Brown, R (Ed.). (2006). *Comprehensive handbook of childhood cancer and sickle cell disease: A biopsychosocial approach*. New York, NY: Oxford University Press.

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