UNDERSTANDING THE CHILD WITH

Sickle Cell Disease

A Handbook for School Personnel

Distributed by
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Introduction

Sickle cell disease is an inherited condition that can cause chronic anemia, unexpected episodes of illness, and frequent medical appointments, which may impact a student's school performance. Similar to other children with chronic illnesses, children with sickle cell disease are at increased risk for school absenteeism. The teacher is closely involved with the daily behavior, activities, and functions of a child in the classroom. The following sections will give you a closer look at the physical and psychosocial complications of sickle cell disease and how they might be addressed in the school setting.

About Sickle Cell Disease

Sickle cell disease (SCD) is the most common genetic disorder identified in African Americans. People from South and Central America, the Mediterranean, the Middle East, and India can also be affected. When people have SCD, their red blood cells change from the usual soft, round shape to a hard and twisted sickle-like shape. Sickled red blood cells stick together, blocking the flow of blood and oxygen, causing pain and other serious complications. The most common types of sickle cell disease are hemoglobin SS disease, hemoglobin SC disease, hemoglobin S beta^{+} thalassemia, and hemoglobin S beta^{0} thalassemia. Hemoglobin SS and S beta^{0} thalassemia are severe forms, while SC and S beta^{+} thalassemia are typically milder.

The only cure for SCD is bone marrow transplantation for those with suitable donors. For individuals with severe forms of SCD, there is a daily medication called hydroxyurea that is very effective in ameliorating most symptoms. As a result of medical advances, people with SCD are living longer and children with SCD lead healthier lives, making academic achievement and career planning even more vital.

Table 1: Physical Complications Caused by SCD

ORGAN/TISSUE INVOLVED	PROBLEMS
BRAIN	• Headache • Stroke • Possible learning disabilities or delays
BONES	• Arm or leg pain • Hip Pain • Back pain
LUNGS	• Pneumonia • Asthma
KIDNEY	• Frequent urination
SPLEEN	• Abdominal pain
EYES	• Yellow Eyes
PENIS	• Priapism (painful unwanted erection)

Table 2: Warning Signs and Action Plan for Students with SCD

Signs	Symptoms	Plan
Fever	101 degrees or higher	Call parent/guardian
Pallor/Jaundice (pale or yellow skin)	Noticeable change in complexion, lips, eyes (some children have yellowish eyes all the time)	Call parent/guardian
Breathing	Rapid or difficulty breathing	**Emergent** call 911 Non emergent call school nurse/parent/guardian
Headache	Sudden Acute blurry vision	**Emergent** call 911 Non emergent call parent/guardian
Heartbeat/Pulse	Rapid heartbeat/pounding	**Emergent** call 911 Non emergent call school nurse/parent/guardian
Pain	Chest, arms, legs, back abdomen.	Do Not Apply Ice Call school nurse/parent/guardian May administer pain meds if arranged by parent
Swelling	Hands, feet, joints	Call parent/guardian
Muscular weakness	Sudden, either side of body Facial asymmetry Difficulty speaking	Call 911

Common treatments for children with sickle cell disease

Hydroxyurea – a medication that improves the symptoms of sickle cell disease. It must be taken regularly to reduce the complications of sickle cell disease. It decreases pain crises, hospitalizations, and improves the lifespan of patients with sickle cell disease. Children on hydroxyurea have more energy, do not get sick often, and can handle school work similar to other students, but still need to be cognizant of extreme hot or cold weather and may still have limitations in PE.

Blood transfusion – can be given for acute complications including aplastic crisis, acute chest syndrome, and stroke. Some patients require chronic transfusion on a monthly basis and would miss school on those days. A major side effect of chronic transfusion is iron overload. There are medications that need to be taken regularly to help lower the iron in the body.

Pain medications – pain medications such as acetaminophen (Tylenol), ibuprofen (Advil, Motrin) and sometimes narcotics (e.g. morphine, codeine, hydrocodone, hydromorphone, etc.) are needed to help relieve pain. Parents may leave pain medications at school in case their child has pain during school and needs medication.

Bone marrow transplant – this is the only cure for sickle cell disease currently. Only a few patients with sickle cell disease undergo bone marrow transplantation, as most do not have a sibling who is matched to be a donor. If a child undergoes a bone marrow transplant, he/she will be out of school for many months and may have different restrictions upon their return for some period of time to due potential complications from the bone marrow transplant.

Physiological Complications

Chronic Anemia 

Children with SCD have chronic anemia, or low red blood cell count, causing organs to have low oxygen. Anemia may contribute to the following:

- Yellow eyes and skin
- Reduced stamina and endurance
- Small stature
- Delayed puberty
- Learning delays

Children who are treated with hydroxyurea typically have better blood counts and are less likely to have these complications.

Care in School setting:

- Be aware of the need to rest. For many children, admitting to fatigue and taking a break from sports and PE activities can be embarrassing and draw unwanted attention. While participation should be encouraged, make it easy and as inconspicuous as possible for the child with sickle cell disease to take regular breaks and have ready access to fluids.
- Encourage the child to participate up to his level of tolerance.
- Allow recovery time after vigorous physical activity, with rest breaks as needed. If active participation is not realistic, involve the child in the activity in other ways, e.g. scorekeeper.
- Small stature, delayed puberty, and jaundice could contribute to the student being a target for teasing or bullying. School personnel should watch for this and implement appropriate action and/or counseling if necessary.

Sensitivity to Heat and Cold:

Exposure to extreme temperatures can precipitate pain crises.

 ***Care in School setting:***

- The child should not sit in drafts or directly in front of fans or air conditioners.
- Remind the child to wear a jacket outside during cold weather or to take off a layer of clothing if it is hot.
- Avoid exposing child to extreme temperatures. May excuse from strenuous physical exercise if outside temperature >90 F or < 40 F.

Pain crisis:

Pain episodes can be associated with over exertion, dehydration, variation in temperature (especially during change of seasons), infection, or they can occur seemingly out of nowhere. Pain can be in the arms, legs, back, or chest, usually without outward signs (no swelling, no redness).

 ***Care in School setting:***

- Increase fluid intake with water, juice, broths, popsicles
- If mild pain, rest and return to class when pain subsides
- Administer Tylenol, Motrin, or prescribed medications if okay with parent and proper school forms submitted with administration instructions.
- Moist heat
- NEVER apply cold packs/ice, as this can exacerbate pain crisis
- Notify parents of pain episodes and treatments

Increased urinary frequency

The kidneys in children with SCD do not concentrate urine effectively, causing more dilute urine to be produced.

Care in School setting:

- Allow extra water and bathroom breaks.

  (Hydration can help prevent pain/organ damage)
- Allow child to have access to unlimited fluids
- Allow the child to keep a water bottle at his/her desk.
- A special bathroom pass can be provided.

Fevers

Children with SCD are at increased risk of serious infections. Fever is a sign of infection and needs to be taken seriously.

Care in School setting:

- Do not give medication for fevers before checking temperature with a thermometer.
- Fever in a person with SCD of 101 degrees or higher is considered a medical emergency and urgent medical attention is required.
- Please contact child's family immediately and have them contact the hematology provider immediately.

Stroke

Stroke is a sudden and severe complication of sickle cell anemia. A stroke happens when blood supply to a part of the brain is cut off. It affects about 10 percent of patients, usually between the ages of 2 and 8. Stroke can cause brain damage, paralysis, coma and even death. Frequently, the child/young adult who has a stroke may return to normal activity in several days. However, there may be physical weakness and/or disability requiring rehabilitation. Children who have suffered a stroke often have learning problems as well as physical disabilities. These students may benefit from an IEP or 504 plans.

Care in School setting:

Note the Signs and Symptoms of Stroke:

If these symptoms occur call 911 and notify the family immediately.

- Difficulty with memory
- Difficulty with speaking or comprehension
- Defective or absent language
- Difficulty with balance
- Muscular weakness, typically on half the body
- Sudden loss or blurred vision
- Fainting/Dizziness
- Sudden weakness or tingling in extremity

If a stroke occurs, afterwards the student will need testing and appropriate 504 and/or IEPs.

Section II: Other School Related Issues

Modern treatment has significantly improved the lives of children with sickle cell disease. Children who are treated with Hydroxyurea well may be indistinguishable from other children at school for the most part, but others may continue to have problems.

Emotional and Social Adaptation

Many children are well adapted emotionally and socially to handle the stresses produced by their illness, while others are not. By creating opportunities for independence and accomplishment, and emphasizing progress, the teacher can foster the child's coping abilities and increase self-esteem.

 ***Care in School setting:***

Helpful interventions:

- Choose child for classroom jobs
- Assign leadership roles to the child in classroom activities
- Encourage participation

Physical Activity

 ***Care in School setting:***

Children should take an active part in physical activity, but let them set their own pace and allow for ***frequent water breaks, approximately every 10-15 minutes***. Students should not be *required* to participate in physical education activities that involve strenuous exercises and long distance running, or the Presidential Physical Fitness Testing, but *may and should* participate to the extent he/she is able.

Absenteeism:

Children may miss school due to routine clinic visits or due to an acute issue.

Care in School setting:

IEP or 504 plans could be helpful to ensure success of students. Make classwork and homework assignments available to the parents to prevent the student from falling behind. If the child is in the hospital, communicate with the hospital's teachers, if available, and give them the classwork. If necessary, help the parent arrange tutoring for your student.

An In-service or educational session can be very beneficial in helping teachers, staff, and classmates learn about sickle cell disease. An educational consultant, member of the medical team, or parent advocate could help provide these programs to the school.

School Performance

Brain function speed may be slower in children with SCD and they may require extra help or tailored instruction for effective learning.

Care in School setting:

- Be on the lookout for signs of decreased school performance
- Meet with parents about student's abilities and performance
- Be ready to refer the student for educational or neurocognitive testing
- Call for IEP when learning issues are present

Vocational Counseling:

Students with this disease should set their educational and career goals in accordance with their ambitions and innate abilities. Guidance is especially important. Military careers with challenging physical demands and jobs requiring heavy manual labor are not good options for persons with sickle cell disease. With proper vocational and professional counseling, exciting and rewarding careers can be realized. Role models are important and can demonstrate to the young adult that there are people with chronic illnesses who have succeeded.

Stay Informed:

School is where a child spends a majority of their waking time. The best thing a teacher, counselor, or nurse can do is to be informed about sickle cell disease and then create a plan to help a child stay involved, free of complications, and engaged in learning. Children with this disease are just like other children in many respects, but they do face particular challenges because of their life-long disease. You can play an important role in offering them the chance to lead relatively normal and productive lives.

Care in School setting:

- **Awareness**. Learn more about sickle cell disease. Comprehensive web sites with good information on sickle cell can be found at www.scinfo.org or the CDC website, http://www.cdc.gov/ncbddd/sicklecell/index.html
- **Partnership**. Facilitate a family conference to assess both the family and the student's adjustment to illness and school expectations.
- **Psychosocial support**. Involve the school social worker or counselor in your plan to understand the psychosocial aspects of living with a chronic disease.

Section III: Accommodations for Children with Sickle Cell Disease

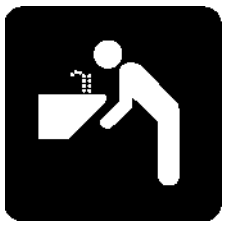
A child with sickle cell disease may qualify for special education through the Individuals with Disabilities Education Act (IDEA), under the category of Other Health Impairment, or under Section 504 of the Rehabilitation Act. If a child does not qualify for accommodations under these programs the school nurse (RN) can write accommodations under an Individual Health Plan (IHP).

The following represent accommodations that should be considered for a child with SCD. Other accommodations may be considered based on the student's individual needs.

1 Accommodation: Two sets of books, one for home and one for school.

Justification: Students with sickle cell disease often have unplanned absences due to their chronic illness. Having an extra set of books at home allows the students time to study and stay caught up with their assignments.

2 Accommodation: Permission to carry a water bottle to drink throughout the day



Justification: Drinking water throughout the school day prevents dehydration, which can trigger a pain episode. A student should drink one liter of water a day.

3 Accommodation: Bathroom and clinic passes when needed.

Justification: Children with sickle cell disease drink more fluids and pass urine more frequently than other children. When a child drinks less than usual, he/she can get dehydrated. Dehydration is a major cause of pain episodes. A laminated pass to use when needed allows the student to leave the classroom without drawing attention from the other students.

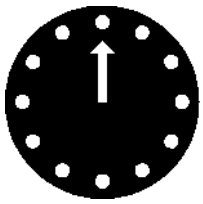
4. Accommodation: The student should not be required to participate in physical education activities that involve strenuous exercises and long-distance running. Participation in the Presidential Physical Fitness Testing should not be required, but allowed to the student's comfort level. The student should take water breaks every 10-15 minutes during physical exercise.

Justification: Strenuous exercises and long-distance running can often trigger the onset of lactic acidosis leading to sickle cell pain.

5. Accommodation: Student may be exempt from outdoor activities when temperature is less than 40 degrees or greater than 90 degrees.

Justification: Extreme temperatures can change the blood flow in the body and precipitate pain episodes. Allowing the student an alternative activity during recess or P.E. class is recommended in very hot or very cold weather.

6. Accommodation: If necessary, student will be allowed to leave class five minutes early to get to his next class.



Justification: Extra time will allow the student to get water, go to the restroom and go to his locker for books.

7. Accommodation: The student will be assigned a moderate workload with limited assignments requiring quality vs. quantity when absent from school for clinic visits, hospitalizations or complications due to sickle cell disease.

Justification: Stress can precipitate a pain episode. Often returning to school after an extended absence, the student may feel overwhelmed and worried about missed work and assignments; shortening assignments and allowing modifications will reduce stress and make the task of completing missed assignments feasible.

8. Accommodation: Student will be granted extended time to complete class work, tests, quizzes.

Justification: The student may process information more slowly due to sickle cell disease, hospitalization, medical treatment schedule, pain medications or stroke.

9. Accommodation: The student will be assigned an intermittent homebound teacher to help with assignments when he misses school.

Justification: Students with sickle cell disease often have frequent absences. Over time this may cause them to become skill deficient in one or more of the core subjects. An intermittent homebound teacher can work with the student on a routine basis and prevent this from

10. Accommodation: The student will be assigned a case manager or buddy to keep him informed of class activities and school functions.

Justification: Students with sickle cell disease and other chronic illnesses often have frequent absences that cause them to miss important school events, such as picture day or field trips. The case manager or peer buddy can e-mail or call students to keep them informed.

11. Accommodation: Medication during the school day.

Justification: Some children may have chronic pain due to SCD. Pain medication may allow the student to complete the school day. The school can sometimes assist in administering daily medications as well when issues at home do not permit regular dosing of daily medications such as hydroxyurea or iron chelators.

There are Pediatric Comprehensive Sickle Cell Centers located throughout the Commonwealth with teams of specialists who work cooperatively with each child's school. If you have further questions, please contact the Sickle Cell Center in your locality.

Table 3: Pediatric Comprehensive Sickle Cell Centers

Center	Address	Contact Information
Children's Hospital of the King's Daughters	Children's Cancer and Blood Disorders Clinic 601 Children's Lane Norfolk, Virginia 23507-1971	Phone: (757) 668-8260 Fax: (757) 668-7811 Edu. Consultant: Shannon Northrup – birth to 14yrs Phone: (757) 668-7515 Christine Booth–15 yrs to transition Phone: (757) 668-9624
University of Virginia Hospital	Department of Pediatrics Division of Hematology/Oncology P.O. Box 800386 HSC, University of Virginia Charlottesville, Virginia 22908	Phone: (434) 924-8499 Fax: (434) 924.5452 Edu. Consultant: Megan Shifflet Phone: (434) 243-5431
Virginia Commonwealth University Health System	Department of Pediatrics Division of Hematology/Oncology P.O. Box 980121 Richmond, Virginia 23298-0121	Phone: (804) 828-9300 Fax: (804) 828-8615 Transition Edu. Consultant: Alma Morgan Phone: (804) 828-4679 Edu. Consultant: Renee' Blackshear Phone: (804) 828-0426
Pediatric Specialists of Virginia, Inova Fairfax Hospital and Children's National Medical Center	Center for Cancer and Blood Disorders of Northern Virginia 6565 Arlington Blvd, Suite 200 Falls Church, VA 22042	Phone: (703) 531-3627 Fax: (703) 531-1590 Edu. Consultant: Ed Schnittger Phone: (703) 531-1508
Carilion Roanoke Community Hospital	Department of Pediatrics Division of Hematology/Oncology 102 Highland Avenue, Suite 435 Roanoke, Virginia 24029-2946	Phone: (540) 985-8055 Fax: (540) 985-5306 Edu.Consultant: Joyce Blount (540) 224-4660 Jennifer Franco (540) 224-6988

Additional Resources

Websites

Center for Disease Control and Prevention (CDC) Sickle Cell Disease
http://www.cdc.gov/ncbddd/sicklecell/index.html

Sickle Cell Information Center https://scinfo.org/

National Institute of Health http://www.nhlbi.nih.gov/health/health-topics/topics/sca

Virginia Department of Health
https://www.vdh.virginia.gov/ofhs/childandfamily/childhealth/cshcn/sickleCell/

Books

Hope and Destiny: The Patient and Parent's Guide to Sickle Cell Disease and Sickle Cell Trait (2011) by Allan F. Platt, Jr., James Eckman, Lewis L. Hsu.

Hope & Destiny Jr.: The Adolescent's Guide to Sickle Cell (2013) by Lewis L. Hsu, Carmen C. M. Rodrigues, Silvia R. Brandalise.