Understanding the Child with SICKLE CELL

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School Handbook

VDH VIRGINIA DEPARTMENT OF HEALTH

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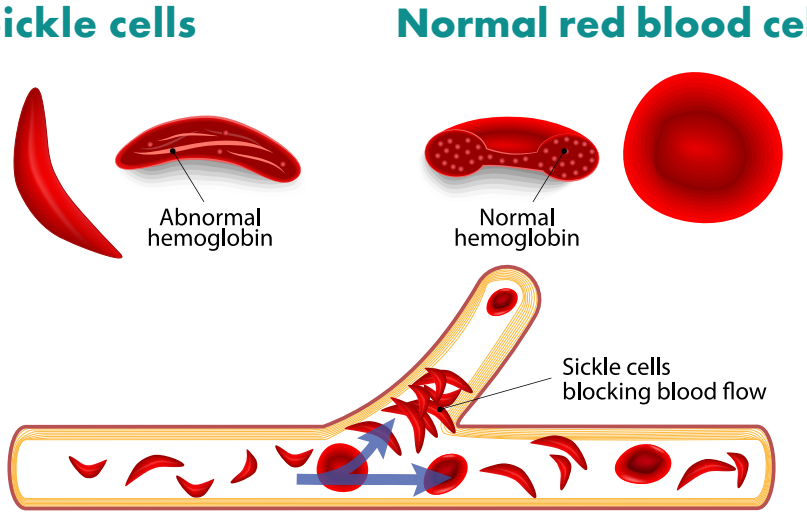
Introduction

This handbook will cover a range of topics regarding the care and treatment for students with sickle cell disease; not all students with sickle cell disease will experience these problems. Sickle cell disease is a group of inherited conditions 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 it 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 SCD 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.

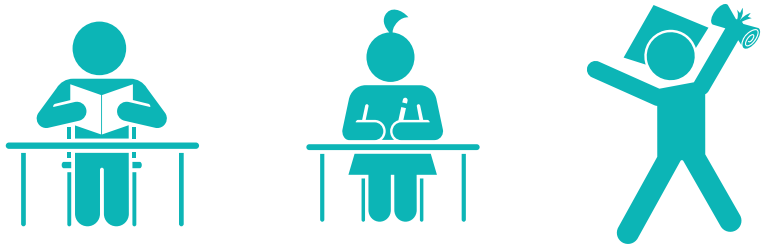


Table 1: Physical Complications Caused by SCD

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	● Jaundice (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	**Emergency** call 911 Non-emergent call school nurse/parent/ guardian
Headache	Sudden Acute blurry vision	**Emergency** call 911 Non-emergent call parent/guardian
Heartbeat/ Pulse	Rapid heartbeat/ pounding	**Emergency** 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 during PE.

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.

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.

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 due to 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 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 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).

Physiological Complications

Care in School Setting:

- Increase fluid intake with water, juice, broths, or popsicles
- If mild pain, rest and return to class when pain subsides
- Administer Tylenol, Motrin, or prescribed medications with parent's permission 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, which means more hydration is as well as more frequent urination are needed.

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 child's sickle cell doctor.

Stroke

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.

Modern treatment has significantly improved the lives of children with sickle cell disease. Children who are treated well with Hydroxyurea 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 illness.

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 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.

Other School Related Issues

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, www.cdc.gov/ncbddd/sicklecell/
- **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.

Other School Related Issues

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.

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

1 **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 to study and stay caught up with their assignments.

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

2 **Justification:** Drinking water throughout the school day prevents dehydration, which can trigger a pain episode. An elementary school student should drink one liter of water a day, while a middle or high school student should drink 2-3 liters a day.

Accommodation: Bathroom and clinic passes when needed

3 **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.

Accommodation: Tailor PE participation to the student

4 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.

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

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

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

6 **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. When 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 more manageable.

8 **Accommodation: Student will be granted extended time to complete class work, tests, and 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 they miss school.**

Justification: Students with sickle cell disease often have frequent absences.

Accommodations for Children with Sickle Cell Disease

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 falling behind.

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 email 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 when home situations make it difficult for regular dosing of daily medications such as hydroxyurea.

Table 3: Pediatric Comprehensive Sickle Cell Centers

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.

Center	Address	Contact Information
Children's Hospital of the King's Daughters	601 Children's Lane Norfolk, VA 23507-1971	Phone: 757-668-8260 Fax: 757-668-7811
University of Virginia Hospital	Pediatric Hematology/ Oncology 1204 W. Main St., 5th Floor Charlottesville, VA 22908	Phone: 434-924-8499 Fax: 434-924-5452
Children's Hospital of Richmond at Virginia Commonwealth University	1000 E. Broad St. Richmond, VA 23219	Phone: 804-828-9605 Fax: 804-628-5848
Pediatric Specialists of Virginia	8081 Innovation Park Dr. Fairfax, VA 22031	Phone: 571-472-1717 Fax: 571-472-1718
Carilion Roanoke Memorial Hospital	1906 Belleview Ave Roanoke, VA 24014	Phone: 540-981-7376 Fax: 540-985-5306

Additional Resources

Websites

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

Sickle Cell Information Center www.scinfo.org/

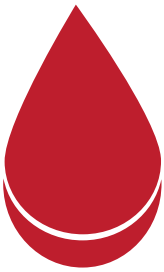
National Institute of Health
nhlbi.nih.gov/health-topics/sickle-cell-disease

Virginia Department of Health
vdhlivewell.com/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.



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