UNDERSTANDING THE CHILD WITH

Sickle Cell Disease

A Handbook for School Personnel

Distributed by
The Virginia Sickle Cell Awareness Program
Virginia Department of Health
109 Governor Street
Richmond, Virginia 23219
(804) 864-7769

VDH VIRGINIA DEPARTMENT OF HEALTH
Protecting You and Your Environment

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Introduction

Similar to other children with chronic illness, children with sickle cell disease are at increased risk for school absenteeism related to their illness. Unexpected episodes of pain, increased risk for stroke and chronic anemia are just a sample of the factors complicating school performance. If you have a child with sickle cell in your classroom, you no doubt have questions regarding this disorder and how it affects your pupil. Because the teacher is a direct observer of 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, it is also found in people from South and Central America, the Mediterranean and the Middle East. When people have sickle cell disease, 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 three most common types of sickle cell disease are hemoglobin SS disease (also called Sickle Cell Anemia), hemoglobin Sickle-C disease and Sickle Beta-Thalassemia. While some types of sickle cell disease are milder and cause fewer physical complications, it is important to remember that every child remains at risk for sickle-related complications.

Currently there is no universal cure for SCD; however, those affected are living longer, healthier lives; making academic achievement and career planning even more vital.

Table 1: Physical Complications Caused by Sickling

ORGAN/TISSUE INVOLVED	PROBLEMS CAUSED
KIDNEY	• Inability to control urination • Hematuria (blood in the urine) • Unconcentrated urine • Frequent urination • Kidney disease
SPLEEN	• Splenic sequestration (pooling of blood in the spleen) • Scared spleen becomes non-functional by age two contributing to increased risk for serious infections • Abdominal pain
LUNGS	• Pneumonia • Acute Chest Syndrome (sickling in the chest)
BONES	• Infection • Aseptic necrosis (breakdown of the bone)
BRAIN	• Stroke • Headache
SKIN	• Slow healing leg ulcers
PENIS	• Priapism (painful unwanted erection)
EYES	• Sickle cell retinopathy (changes in the blood vessels in the eye)
LIVER	• Enlarged liver • Gallstones • Jaundice (yellowing of eyes and skin)
• Not all these complications occur in every child with SCD. You need to know, however, that they **can** happen. Ask parents about their child's history regarding complications.	

Table 2: Physical Complications: Warning Signs

SIGNS	SYMPTOMS
FEVER	• 101 degrees or higher
PALLOR	• Noticeable change in complexion, lips, fingernails
BREATHING	• Dyspnea (difficulty breathing) • Tachypnea (fast rate of breathing) • Stertorous breathing (labored breathing)
HEADACHE	• Sudden or constant • Dizziness
HEARTBEAT	• Tachycardia (rapid heart beat) • Pounding
PAIN	• Head • Chest • Joints • Abdomen (abdominal distention) • Penis (prolonged erection)
SWELLING	• Hands • Feet • Joints (with redness)
MUSCULAR WEAKNESS	• Either side of the body
• Contact the child's family if you notice any of these signs and symptoms. Any change from what you feel is normal for the child should be reported to the family.	

Section 1: Physical Complications of Sickle Cell Disease

Chronic Anemia

Chronic anemia is a fact of life in children with sickle cell disease; most adjust well and are without symptoms. However, chronic anemia may contribute to the following complications:

- Jaundice (yellowing of eyes and skin)
- Reduced stamina and endurance
- Delay in skeletal maturation
- Delayed onset of puberty

Care in the School Setting

Be aware of the need to rest. For many children, admitting to fatigue and taking a break from sports and gym 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.

- Encourage the child to participate up to his level of tolerance
- Allow recovery time after vigorous physical activity
- If active participation is not realistic, give the child a duty related to the activity, such as scorekeeper

Other School-Related Issues

On average, children with sickle cell disease maintain a lower average height and weight than unaffected children. Puberty is delayed about two years compared to peers without SCD. Small statue and jaundice are factors that could contribute to the child being a target for teasing. Watch for compensating behaviors such as:

- Aggression to compensate for size

- Association with younger children
- Isolation and avoidance behaviors

Aplastic Event

An aplastic event is a sudden severe drop in the hemoglobin. In other words, a person becomes more anemic than usual. This is typically due to an infection with parvovirus, also the cause of Fifth Disease in childhood. The infection causes the bone marrow to stop making red blood cells. The symptoms of a very low blood count are:

- Headache
- Irritability
- Poor appetite
- Jaundice that is greater than normal for the child
- Unusual tiredness
- Rapid heart beat
- Change in complexion resulting in pale color.

  If the palms of the hands do not have any pink color even when the hands are warm, the child is pale.

Care in the School Setting

Contact the parent. If a child has symptoms of a low blood count, consult a doctor immediately. Blood transfusions may be necessary.

Pain

Pain is the most common complication of sickle cell disease. It occurs when sickled cells block the flow of blood and oxygen.

Common triggers include:

- Fever
- Infection
- Exposure to extreme cold
- Physical exhaustion
- Unusual stress or anxiety

Sometimes there is no identifiable precipitating event. Pain may occur at any time and in any part of the body. Each event differs in terms of severity and duration. The pain may be so mild that no analgesic is needed, or may be so severe that hospitalization is required.

Swelling may be seen in the area of the pain. In children under 2 years of age the swelling usually occurs in the hands and/or feet. Swelling and pain may also be caused by an infection in the bone.

Signs and Symptoms

- Chest pain
- Difficulty breathing
- Fever 101 or higher
- Priapism (prolonged, painful erection)
- Nausea, vomiting
- Severe abdominal pain
- Bone pain and swelling

Care in the School Setting

Pay attention to temperature control. Simply becoming overheated or too cold could trigger a pain episode. 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. Treatment to relieve mild to moderate pain may include any or all of the following:

- Fluids – water, fruit juice, popsicles, clear broth, jell-o
- If pain is mild, the child should be allowed to rest and return to the classroom when pain subsides
- Analgesics as directed by the physician
- Relaxation exercises to help relieve pain (if the child is familiar with these techniques)
- Moist heat applications
- Never apply cold packs as these could increase vessel constriction
- If a child has a fever, phone parents immediately

Kidney

Dehydration

Children with sickle cell disease drink more fluids and pass urine more often than other children. When a child becomes sick and drinks less than usual or looses more fluid by vomiting, diarrhea or fever, he can get dehydrated. Dehydration is a major cause of pain episodes.

Kidney and Bladder Infection

Bladder infections are fairly common. If they are not treated promptly, they can move from the bladder to the kidneys and cause kidney damage. Parents should instruct the child to alert the teacher or school nurse if they experience any of the following:

Signs and Symptoms

- Irritating or painful urination

- Hematuria (blood in the urine)
- More frequent urination than usual for this child
- Enuresis (inability to control urination)

Care in the School Setting

Allow extra water and bathroom breaks. In busy classrooms, it is not uncommon to limit water and bathroom breaks. For children with sickle cell disease, however, these rules need to be relaxed a little. The simple act of staying well hydrated can help prevent pain episodes and avoid organ damage. Children with sickle cell disease also need to take bathroom breaks more frequently, not only because of increased water intake but because their kidneys produce more urine, even when they're dehydrated.

- The child should be allowed to use the bathroom frequently
- A special bathroom pass should be provided if possible
- The child should have access to unlimited fluids throughout the day
- The child should be allowed to have a water bottle or juice box in their backpack.

Table 3: Fluid Requirements For Children With SCD

CHILD'S WEIGHT	Number of 8 oz. Cups Per Day
25 lbs.	5 cups
50 lbs.	8 cups
75 lbs.	10 or more cups
100 lbs.	10 or more cups

Spleen

Splenic sequestration is a possible complication of sickle cell disease that occurs most commonly in children under 5 years of age. It occurs when there is a sickling in the splenic vein resulting in the sudden entrapment of a large amount of blood in the spleen. The spleen suddenly becomes very large and the child can have signs and symptoms of a very low blood count (he/she becomes very anemic) resulting in shock in extreme instances. This is usually treated with hydration, analgesia, and red cell transfusion. In some children, a splenectomy (removal of the spleen) is necessary.

Signs and Symptoms

- Sudden weakness
- Pallor (especially of lips, gums and nails)
- Abdominal pain
- Increase in abdominal girth
- Enlargement of the spleen
- Fever

Care in the School Setting

Should you observe any of these signs and symptoms, notify the child's family immediately. Shortness of breath and rapid heartbeat should be evaluated in a hospital emergency room as soon as possible.

Infection and Fever

Phone the parent immediately if the child has a temperature greater than 101.

The child with sickle cell anemia will get colds, sore throats and ear infections just like other children. However, some infections can represent a life-threatening complication in these children. Prophylactic penicillin has decreased, but not eradicated, this problem. The potentially serious infections that are more likely to occur in the child with sickle cell anemia are:

- Septicemia (infection of the blood)
- Meningitis (infection around the brain)
- Pneumonia (infection in the lungs)
- Osteomyelitis (infection of the bone)

Because their spleen does not function correctly, children with sickle cell develop infections more easily and more frequently than other children. The child with a serious infection may not appear to be acutely ill in the early stages. Therefore, it is important to learn the early warning signs and to know when to contact the family and when to advise referral to the primary care provider, treatment center or emergency room.

Signs and Symptoms

- Fever 101 degrees or greater is the most common symptom of infection
- Lethargy (unexplained tiredness)

Care in the School Setting

- Do not give medication for fever before checking the temperature with a thermometer
- Fever in a person with sickle cell disease of 101 degrees or higher is a medical emergency
- The child's family and physician should be contacted immediately
- The child should be seen in a hospital (emergency room) as soon as possible

Lungs

Respiratory complications are common in children with sickle cell disease. These can be secondary to either an acute lung infection (pneumonia) or sickling in the lung. Difficulty breathing and chest pain are typical symptoms of respiratory complications. The combined symptoms are called Acute Chest Syndrome, and can be life threatening.

Signs and Symptoms

- Chest pain
- Cough
- Dyspnea (difficulty breathing)
- Tachypnea (rapid breathing)
- Fever

Care in the School Setting

- Recognize that chest pain is an emergency. Hospitalization of the child with acute chest syndrome is imperative
- Encourage deep breathing exercises. Ask parents if they have an incentive spirometer (machine that measures the amount of inhaled and exhaled air) that can be used at school
- Encourage compliance with medications for asthma if the child also has this diagnosis
- Report any of the above signs and symptoms to the child's family

Bones and Joints

Osteomyelitis (an infection of the bone) causes fever, pain, swelling and/or redness over a bone. A child will often have a fever of 102 degrees. If this complication

occurs and treatment is delayed, serious and permanent damage to the bone or joint can occur.

Signs and Symptoms

- Pain
- Swelling
- Tender red joint
- Fever
- Localized "warmth"

Care in the School Setting

If a child exhibits these symptoms contact parents immediately, infections in the bone can be life threatening.

Aseptic Necrosis

Aseptic Necrosis occurs when sickled cells block vessels supplying bone. Physical therapy may be necessary. In severe cases surgery will be required. Treatment depends on the extent of the problem. Sometimes a person needs to use crutches for a few months to take the weight off the joint. The hip may eventually need to be replaced.

Signs and Symptoms

- Chronic hip, extremity or lower back pain that does not resolve
- Pain with no fever
- Occurs most commonly in the hip bone
- Persistent pain in groin or buttocks
- Gait (manner of walking) disturbance

Care in the School Setting

- Notify the child/young adult's family if these symptoms are present
- Provide physician-prescribed pain medication
- Increase fluids
- Set appropriate limitations for physical education activities
- Accommodations should be put in place to allow use of elevators if they are present and extra time allowances to get to classes

Gallstones

About a third of children with sickle cell disease have gallstones by the age of seven. This is due to excess bilirubin caused by red blood cell destruction. Bilirubin collects in the gallbladder and form stones. When stones pass they may cause pain in the right side of the abdomen. Surgery to remove the gallbladder is often necessary. This is the most common surgery in children with sickle cell disease.

Signs and Symptoms

- Right upper abdominal pain
- Shoulder pain
- Nausea and vomiting

Care in the School Setting

- Notify the child's parents as hospitalization and surgery may be necessary
- Make preparations for homebound instruction

Stroke

Stroke is a sudden and severe complication of sickle cell anemia. It affects from six to 12 percent of patients, usually between the ages of 5 and 10. If a child is not put on transfusion therapy, the incidence of a repeat event is 60 percent. A stroke may occur with a painful episode or an infection, but in most cases there are no related illnesses. 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.

Signs and Symptoms

- Difficulty with memory
- Difficulty speaking or understanding what other people are saying
- Defective or absent language
- Difficulty with balance
- Muscular weakness on one half of the body
- Sudden loss of or blurred vision
- Persistent headaches or sudden strong headache
- Fainting
- Dizziness
- Sudden weakness or tingling of an arm, leg or the whole body

Minor strokes and other brain problems can happen without any signs or symptoms. These events often cause learning problems and are believed to affect a child's cognitive learning skills.

Care in the School Setting

Sickle cell disease is one of the few conditions that can cause brain injury in children. An uncharacteristic slip in academic performance may be a subtle sign.

Notify the child's family if any of the listed symptoms appear without another explanation. If a child has had a stroke, an IEP (Individual Education Plan) should be developed.

Priapism

Priapism is a painful swelling of the penis due to trapped red blood cells. This usually occurs without relation to sexual excitement and can occur in young children as well as in adults. Prolonged episodes can take several days to resolve and may require hospitalization.

Signs and Symptoms

- Sustained unwanted erection

Care in the School Setting

- Parents should counsel their child regarding the importance of reporting signs and symptoms in an effort to dispel embarrassment
- Notify the family immediately when symptoms are present as hospitalization may be required

Eyes: Sickle Cell Retinopathy

Children with sickle cell disease may develop eye problems that can occasionally lead to blindness. This condition manifests slowly and rarely occurs before the age 10.

Care in the School Setting

- Notify the child's family immediately of any vision problems or trauma to the eye.

Section II: Other School Related Issues

Socialization and Peer Support

While many children are well adapted emotionally to handle the stresses produced by their illness, others are not. Some children enter school from an environment in which they have been overprotected as a result of their illness. Their behaviors may be characterized by over dependency, a sense of helplessness, excessive and unpredictable levels of frustration and a reluctance to try new tasks. By creating opportunities for independence and accomplishment, and emphasizing progress, the teacher can foster the child's coping abilities and increase self-esteem.

Interventions

- Choose the child for classroom jobs
- Assign leadership roles to the child in classroom activities and projects
- Encourage participation in extracurricular activity in which the child should excel

Nutrition and Diet

There is no special diet for children with sickle cell disease, however recent research shows that children with sickle cell need about 20 percent more calories than other children to fuel their production of red blood cells. Not getting enough calories may lead to delays in growth and maturation.

Constipation is an unfortunate side effect of some of the pain medications these children take. Fiber is critical, whole grains and fruits will help prevent or treat constipation.

Children with sickle cell disease need extra folic acid in order to produce red blood cells more quickly. These can be found in foods such as grains, fruits, and leafy green vegetables.

Physical Activity

Children should take an active part in any physical activity that interests them. Let them set their own pace. The student should not be required to participate in physical education activities that involve strenuous exercises and long distance running. The student should not be required to participate in the Presidential Physical Fitness Testing.

Interventions

- Emphasize the child's assets and strengths; give positive reinforcements with praise and attention
- Assess hobbies and pastimes to encourage socialization and to avoid boredom
- Assist the child to retain roles and tasks as much as possible within existing limitations

Absenteeism

Absenteeism may be the largest obstacle to school success for some children with sickle cell disease. Children may be absent a lot due to clinic visits, pain crises or other health problems. 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 and give them the classwork. If necessary, help the parent arrange tutoring for your student. Absenteeism may also contribute to social isolation and disturbances in peer relationships.

In-service Education Sessions for Classmates

Classmates may begin to wonder, question and worry about their missing classmate. A class in-service can be very beneficial in helping teachers, staff and classmates learn about sickle cell disease. Educational in-services help to eliminate rumors and offer ideas on how to be a supportive friend.

An educational consultant or member of the medical team that is serving the child, a parent, or the child himself can provide these programs.

Learning Problems

Learning problems can affect how well children do at school and in their social lives. Children with sickle cell disease are more prone to silent brain infarctions that can result in cognitive deficits. If a teacher notices a change in a child's school performance parents should be informed.

Depression

Depression is no stranger to those coping with chronic disease and unpredictable episodes of pain. The young adult with sickle cell disease may experience particular feelings of fear and anxiety or be depressed by the prospect of being ill for the rest of his life or having a potentially fatal disease. Depression may affect an adolescent's ability to manage his disease and personal life.

Interventions

- Provide avenues for the student to express feelings
- Assist the student to understand his or her own way of coping
- Give positive reinforcement for efforts to cope
- Maintain hope and share it with the student

Vocational Counseling

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

Stay Informed

The teacher is a direct observer of daily behavior, activities and functions of a child in the classroom. The best thing you 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.

Interventions

- **Awareness**. Learn more about sickle cell disease. A comprehensive Web site, filled with valuable information on sickle cell can be found at www.scinfo.org
- **Partnership**. Facilitate a family conference to assess both the family and the student's adjustment to illness and school pressure.
- **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

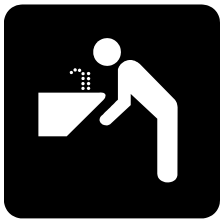
A child with sickle cell disease may qualify for special education through the Individuals with Disabilities Education Act (IDEA), under the category of Other Health Impairment, or under Section 504 of the Rehabilitation Act. If a child does not qualify for accommodations under these programs the school 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 bottle of water to drink throughout the school day.

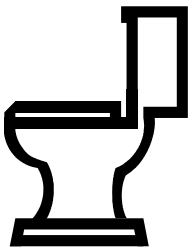


Justification: Drinking water throughout the school day prevents dehydration, which can trigger a pain episode. A student should drink one liter of water a day.

(See page 9.)

3. Accommodation: Bathroom and clinic passes when needed.

Justification: Children with sickle cell disease drink more fluids and pass urine more often than other children. When a child becomes sick and drinks less than usual or looses more fluid by vomiting, diarrhea or fever, he 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 will not be required to participate in physical education activities that involve strenuous exercises and long-distance running. The student should not be required to participate in the Presidential Physical Fitness Testing.

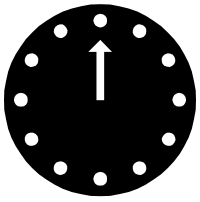
Justification: Strenuous exercises and long-distance running can often trigger the onset of lactic acidosis leading to sickle cell pain.

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

6.

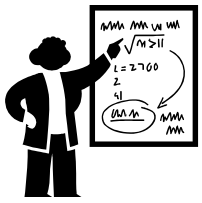
Justification: Extreme temperatures can change the blood flow in the body and precipitate a pain episode. Dressing in layered clothing or allowing the student an alternative activity during recess or P.E. class is recommended.

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: Student will be assigned a moderate workload with limited assignments requiring quality vs. quantity when absent from school for clinic visits, hospitalizations or complications resulting from the diagnosis of 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 classwork tests and quizzes.

Justification: The student may process information more slowly due to 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 happening.

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 multiple complications of SCD. Non-narcotic pain medication may allow the student to complete the school day.

Sample: Educational Modifications Form

Pediatric Comprehensive Sickle Cell Center

Educational Modifications Form

I request that the school provide all of the modifications/ accommodations listed. I also give permission for the school to contact the clinic educational Consultant listed.

Parent/Legal Guardian signature: _____
Date: ____/____/____

Child's Name	Last	Middle	First

School	DOB: ——/——/——	Grade

Home Address	City	Zip

Contact Information	Home Phone	Work Phone	E-mail

Primary Diagnosis:

Secondary Diagnosis

Modifications
To be completed by the Educational Specialist

The Student Needs/requires	Yes	No
Extra set of books		
Frequent bathroom and water breaks		
Length of assignments modified during periods of illness Extended time to complete classroom tests and quizzes		
Bathroom and clinic passes when needed		
Physical Activities to tolerance Exemption from outdoor activities during periods of Extreme temperatures (<40F and >90 F) Exemption from presidents physical fitness tests		
Seating away from drafts and air-conditioning ducts		
Student's medical condition may result in absences in excess of 10 days/semester. Attendance policy waiver may be needed		
Home Bound Services for ____ Short Term ____Long Term ____Intermittent		

Referral to child study team for:
____School health care plan ____504 Plan _____IEP

Medication during the school day: Medication must be brought to school by a parent/guardian in a container appropriately labeled by a pharmacist or MD.

Other Accommodations:

Signatures

Educational Specialist	Phone	Date

School Rep.	Phone	Date

Table 4: Parent/Teacher Sharing

ISSUE	PARENT	TEACHER
Information Sharing	• Visit school and talk with teacher • Take updated literature for teacher reference • Share a copy of IEP • Explain special classroom needs such as bathroom and water fountain privileges • Discuss disclosure of illness with child and teacher	• Listen and review information • Ask specific questions about child's behavior patterns • Discuss with the parent the possible need to foster classmates' understanding
Academic Performance	• Inform the teacher if your child has had a stroke or complication that might affect their ability to do school work • If your child is not performing as you might expect, contact the teacher to try and determine the cause • Do no assume all problem areas are due to SCD	• Check with parent to see if child has had any neurological complications or is on any medication that could cause drowsiness • If child is not performing consistent with ability, you should conduct an analysis of contributing factors
Absenteeism	• Make sure child has IEP with exclusion to mandatory attendance regulations and extra set of books for home • Inform the school when child is sick and if possible estimated date of return • Ask for lesson plan and homework • Ask about homebound teachers	• Share as much information as possible about current and future classroom projects • Consider reduced assignments that would ensure child has mastered the concept of the work but is not overburdened by length of assignment
Gym and Physical Activities	• Child should be excused from President's Physical Fitness Tests • Request that activities be modified so that child can participate to the best of their ability • Discuss alternative activities	• Observe child for over-compensation as some children do not like to draw attention to themselves • Observe child for avoidance behaviors and encourage participation • Redirect activities if necessary
Self-Esteem and Career Goals	• Praise, encourage and congratulate your child everyday • Be positive and supportive in your statements about school and planning for the future • Encourage child to set career goals • Identify role models and mentors	• Help child identify special interests and talents • Help child identify realistic aspirations and career goals • Assist family in identifying community resources and scholarship opportunities

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 5: Pediatric Comprehensive Sickle Cell Centers

Center	Address	Contact Information
Children's Hospital of the King's Daughters	Department of Pediatrics Division of Hematology/Oncology 601 Children's Lane Norfolk, Virginia 23507-1971	Phone: (757) 668-9783 Fax: (757) 668-7811 Ed. Consultant: Gus Guardino Phone: (757) 668-9499
University of Virginia Hospital	Department of Pediatrics Division of Hematology/Oncology P.O. Box 800386 HSC, University of Virginia Charlottesville, Virginia 22908	Phone: (434) 492-8499 Fax: (434) 924.5452 Ed. Consultant: Ginger Ellingwood Phone: (434) 924-2084
Virginia Commonwealth University Health System	Department of Pediatrics Division of Hematology/Oncology P.O. Box 980121 Richmond, Virginia 23298-0121	Phone: (804) 828-4879 Fax: (804) 828-0504 Ed. Consultant: Alma Morgan Phone: (804) 828-0426
INOVA Fairfax Hospital Women and Children's Center Comprehensive Sickle Cell Center	3299 Woodburn Road Suite 220 Annandale, Virginia 22003	Phone: (703) 876-2715 Fax: (703) 876-2716 Ed. Consultant: Yvonne Chapman Phone; (703) 876-2714
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 Ed. Consultant:

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The Sickle Cell Slime-O-Rama Game, CD Rom: Starbright Foundation, 2004

An interactive "quiz show" where players are challenged to answer questions about sickle cell disease and learn fundamental principles about disease management and adherence. The goal of the game is to provide children and teens with basic skills about communicating pain to adults, coping with a pain episode and preventing more severe pain episodes. Designed for ages 6 to 14. Web site: http://www.starbright.org/projects/hes/index.html

This guide reflects the state of knowledge, current at the time of publication, on effective and appropriate care. Given the inevitable changes in the state of scientific information and technology, periodic review, update and revision will continue to be done.

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Glossary of Terms

Acute - beginning quickly, sharp or severe

Acute Chest Syndrome - pneumonia and sickle cell crisis in chest

Afebrile - no fever

Analgesic - a drug that relieves pain.

Anemia - low blood count

Aphasia - defective or absent language

Aplastic Crisis - bone marrow temporarily stops making red blood cells

Aseptic Necrosis - gradual breakdown or destruction of bones usually involving the hips, knees or elbow joints

Bilirubin - yellow substance that comes from the breakdown of red cells

Bone marrow - the place inside your bones where blood cells are made

Carriers - persons who carry a gene for a disease but do not have the disease

Chronic transfusions - a blood transfusion that is given about once each month to hopefully prevent further complications from sickle cell disease

Crisis - an emergency related to sickle cell disease

Dehydration - loss of body fluids from sweating, diarrhea and urination

Diarrhea - frequent and watery bowel movement

Dyspnea - difficulty breathing

Dysuria - irritating or painful urination

Enuresis - the inability to control urination

Genes - message carriers that are passed on to children through the mother's egg and the father's sperm. Genes carry the messages for things such as eye color, hair color, blood type and the kind of hemoglobin a person will have.

Hematologist - a doctor who specializes in blood disorders

Hematuria - blood in the urine

Hemiparesis - muscular weakness of one half of the body

Hemoglobin - the part of blood that carries and delivers oxygen to all parts of the body

Hemoglobin SC Disease - a variant of sickle cell disease

Hemoglobinopathy - abnormal hemoglobin

Hemolytic - breakdown of red blood cells and release of hemoglobin

Infection - invasion of the body by germs causing disease

Inherited - passed on from the father and mother to their child

Jaundice - yellow pigment (color) to eyes and skin

Meningitis - brain infection

Morbidity - disease

Mortality - death

Osteomyelitis - bone infection

Pallor - paleness of skin

Pneumonia - lung infection

Priapism - prolonged, unexpected painful erections without sexual desire.

Prophylactic - preventive

Retinopathy - an eye disorder without redness or swelling, caused by changes in the eye blood vessels

Sepsis - serious blood infection

Sickle beta-thalassemia - a variant of sickle cell disease.

Sickle Cell Anemia (SS) - most common sickling disease in the United States

Sickle Cell Crisis - an emergency related to sickle cell disease where blood flow stops due to plugging of blood vessels with sickled cells

Sickle Cell Trait - a normal hemoglobin gene is inherited from one parent and a single hemoglobin gene from the other

Sickle Cells - red blood cells that look sickle-shaped ( )) under a microscope

Sickling Disease - sickle cell anemia (SS), sickle-C (SC), and sickle beta-thalassemia (S-B-thalassemia)

Spleen - an organ on the left side of the belly. It acts as a filter for blood

Splenic Sequestration - spleen overfills with blood

Stertorous breathing - labored breathing

Stroke - clogging of blood vessels to the brain

Tachycardia - rapid heart beat

Tachypnea - fast rate of breathing

Thalassemia major - an inherited disease with abnormal hemoglobin that requires lifelong transfusions but does not put a child at risk for infection or crisis

Thalassemia minor (trait) - an abnormal type of hemoglobin that is sometimes inherited along with sickle hemoglobin to produce sickle-beta thalassemia. Having the trait by itself is not harmful.

Transfusion - giving blood to replace blood lost through injury, surgery or disease

Ulcers - sores on the skin

Urination - passing water or peeing

Vessels - the tubes that blood flows through such as veins, arteries, and capillaries