NEPSCC
New England Pediatric Sickle Cell Consortium

Sickle Cell Disease: A Resource for the School Nurse and Educator

INTRODUCTION

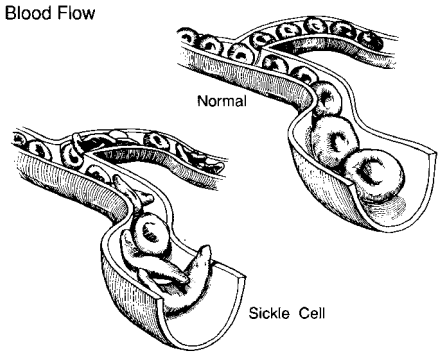
This resource is designed to provide teachers, school nurses, and other school personnel, with information about sickle cell disease. Having this information may make you more comfortable with the child with sickle cell disease in your classroom or school. This information may also give you some insights into the school-related needs of these children, allowing you to provide the best opportunities and services to fit each child's needs.

Sickle cell disease (SCD) is an inherited blood disorder of which there are several types, the most common being Sickle Cell Anemia (Hb SS Disease). Approximately 1 in 400 African-American babies are born with Sickle Cell Anemia. Another type is Sickle C Disease. Approximately 1 in 1000 African-American babies have Sickle C Disease. Sickle Beta Thalassemia is another type of SCD.

SCD is found predominantly in the African-American population but is also seen in people of other ethnic groups. These ethnic groups include individuals from parts of the Middle East, Central India, and countries bordering the Mediterranean Sea, especially Italy and Greece.

In light of these statistics, you are likely to have a student with some form of Sickle Cell Disease in your school or classroom.

WHAT IS SICKLE CELL DISEASE (SCD)?



SCD is an inherited blood disorder caused by an abnormality in the red blood cell. The red blood cell is responsible for carrying oxygen to all parts of our body. When a person has SCD, the red blood cells are abnormal. They may become misshapen into a "sickle" shapped which causes the cells to clog in the blood vessels. This clogging can lead to pain and organ and tissue damage.

Another abnormality of the sickle cell is the life span of the cell. A normal red blood cell lasts approximately 120 days. A sickle cell lasts approximately 6 to 14 days. This results in the child having chronic anemia.

COMPLICATIONS OF SICKLE CELL DISEASE

SCD can affect or damage many organs and tissues in the body. Some of the major complications are listed below.

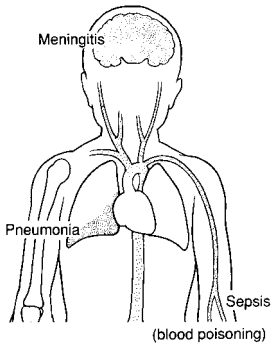
Anemia

A child with SCD has fragile, oxygen-carrying red blood cells, which causes them to have chronic anemia. They may easily fatigue and have less stamina and endurance. Due to their chronic anemia, the child may be slower to grow and develop. Children with SCD maintain a lower average height and weight than do unaffected children. Children with SCD are late in developing secondary sexual characteristics. The onset of puberty may be several years after their peer's onset.

Pain

Pain is a common complication of SCD. Pain occurs when the sickled cells clog up the blood vessels, preventing oxygen from getting to the tissues. It may or may not be accompanied by swelling or fever. Most often there are no physical signs that the child is having pain; therefore, the child should be taken seriously when pain is reported. Since the pain is not visible or detectable, we must rely on the child's report. Pain may occur *anywhere* in the body but is often reported or experienced in the arms, legs, abdomen, and back. School personnel should be informed of signs and symptoms so that adequate actions may be taken.

Infection

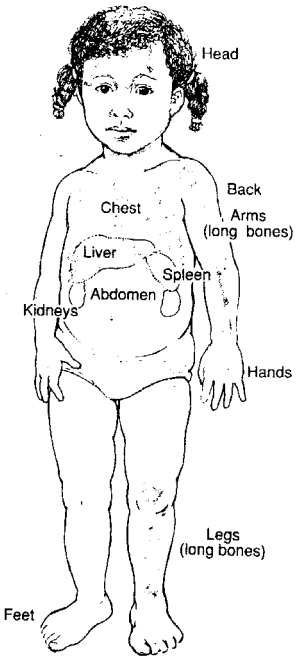


Children with SCD are at increased risk for developing infections. **Some types of infections in a child with SCD are life-threatening.** Some children with SCD take antibiotics daily to prevent them from getting infections. If a child with SCD shows any signs of having an infection, please notify their parents immediately.

Stroke

A small percentage of children with SCD have a stroke caused by sickled cells clogging the blood vessels in their head. Usually, the child recovers completely, but in some cases, intellectual, motor, and sensory impairments occur. If a child has a stroke, he or she is at great risk for having more strokes.

Children with SCD may have many other complications, but their descriptions exceed the scope of this resource.



SYMPTOMS

Signs/symptoms that need to be brought to the parent's or medical professional's attention are:

Pain

- Child complains of pain
- Child unwilling to use extremity
- Swelling
- Painful erection of the penis (priapism)

Stroke

- Seizures
- Paralysis
- Unsteady walk

- Slurred speech
- Changes in vision
- Weakness

Infection

- Fever 101 degrees or higher
- Rapid breathing
- Unusual sleepiness
- Coughing
- Irritability
- Paleness
- Difficulty in breathing

Other symptoms that should be reported:

- Rapid breathing or heartbeat
- Pain in left side of abdomen
- Headache
- Fainting
- Chest pain
- Congested cough

SCHOOL-RELATED ISSUES

Academic Performance

Children with SCD have several factors, which might interfere with school performance. Repeated absences due to their illness and frequent medical appointments, increased fatigue due to chronic anemia, and recurrent painful episodes may predispose them to poor performance.
Recent research provides conflicting information regarding how or if SCD affects overall cognitive functioning. However, studies do show that children with SCD tend to score lower on tests which assess motor skills, reading, and spelling. These deficits may be manifested as developmental immaturity and poor school performance, which may be symptoms of the illness rather than poor adjustment to a chronic illness. Mathematics may be affected because it builds on material previously learned. Attentional skills may tend to be less than those of their peers.
Children who have had a stroke have special educational needs that need to be assessed on an individual basis.
It is important to remember that children with SCD are a population at increased risk for learning difficulties. Neuropsychological and educational testing could be helpful in determining the child's strengths and weaknesses especially if the child has had small strokes. That information could assist school personnel in developing the best teaching strategy for that individual child.

Physical Activity

A child with SCD should be able to participate in physical activity when they are not having a painful episode or are ill. It is important for the child to participate as much as possible. Participating will allow them to reap the social benefits of group participation and promote overall physical fitness, strength, and motor control.

If a child cannot participate in the actual sport or activity, he/she could be the scorekeeper, squad leader, referee or could help set up the equipment. Sitting them out on the sidelines or bench can lower self esteem and cause friction in their relationships with their classmates.
It is VERY IMPORTANT to prevent dehydration in children with SCD. They need to take plenty of fluids before, during, and after an activity. They need to avoid becoming overheated or fatigued. Due to their chronic anemia, they may need more frequent rest times. Children with SCD also need to avoid being exposed to cold temperatures.
Excessive running or exercise and strenuous sports are not recommended for children with SCD.

Attendance

Painful episodes requiring at-home or hospital management are a common cause for children missing school. Because infections, illnesses, or emotional stress can precipitate a painful episode, students with SCD may be out of school for a longer duration than other students. Even when a child is healthy, children with SCD must see medical professionals often which may necessitate more days of missed school.
Extra effort on the parent's, school's, and child's behalf will be necessary for the child to keep up with school work. Assignments that the child can do at home and/or make-up work will enable him/her to keep up with the class.
It may be helpful to discuss the school's absentee policy with the parent to make them aware of the school's requirements. If the child has excessive absences which put them at risk of falling behind or failing, it is important to inform parents and medical professionals. This information may allow everyone involved to make some changes that would enable the child to catch up with their work and attend class on a more regular basis.
Children with SCD need to be encouraged to stay in school even if they have extended periods of absences. School offers socialization and peer interaction which are extremely important to a child's development. Homebound is sometimes recommended for children with SCD.

SPECIAL NEEDS AT SCHOOL

Medications

Children with SCD often experience painful episodes while at school. Children are often prescribed narcotics (ex. Tylenol with Codeine, Darvocet), anti-inflammatory medications (ex. Toradol, Advil) or steroids (Prednisone) to alleviate SCD pain. Tylenol is also given for pain or fever. It may be helpful for the child to have the medication on hand at school.
It is not always necessary, or in the child's best interest, to send him/her home if he/she is experiencing pain; however, parents should always be contacted. If it is possible for the child to take their medication and increase their fluid intake for awhile, they may be able to go back to class and continue their studies.

Fluids

It is **imperative** for children with SCD to maintain adequate fluid intake and to not get dehydrated. **If at all possible, it would benefit the child to be allowed to carry a water bottle with them at school or allow them to leave class to get water frequently**.

Bathroom Privileges

Special bathroom privileges are needed for SCD children due to the disease's effect on their kidneys. Additionally, the need for extra fluid causes them to urinate frequently. This frequent need to urinate can be embarrassing to the child and frustrating to the teacher; however, this is due to the disease and cannot be avoided. Children with SCD should be allowed to sit near the door so that he/she can leave as often as necessary to use the restroom.

Classmates

Peers of school-aged children are so important that the classmate may also need some information about SCD. It is common for children to tease or be unkind when there is a lack of understanding or fear about something of which they have little knowledge. It may be helpful for the teacher/counselor to assist the SCD child to plan a response when negative comments are made or when they are teased by classmates.
Giving the classmates information about the child's illness may eliminate fears or concerns and may foster acceptance and inclusion of the child with SCD. Information may only be given to the class after obtaining permission from the parents and the child. Some hospitals have nurses who can visit the school to educate the children about SCD and alleviate some fears or concerns they may have about the disease.
Listed below are some questions asked frequently by friends and classmates:

Q. Is Sickle Cell Disease contagious?
A. It is important to assure them that it is NOT contagious; it is inherited from their parents.

Q. Why are their eyes yellow? Do they have hepatitis?
A. No, they do not have hepatitis. Their eyes become yellow because of a substance released when their red blood cells break down.

Q Why do they keep a water bottle at their desk or get to leave class for water fountain and restroom breaks?
A Water helps veins to get bigger so that the sickle-shaped cells can flow through the veins easier and not get jammed together or cause as much pain. As a result of needing to drink so much water, and because SCD causes problems with their kidneys, they need to use the restroom more often.

Q. Why do children with sickle cell disease (SCD) look smaller and shorter and seem to be less physically developed than other kids their age?
A. Children with SCD have less oxygen-carrying cells functioning in their bodies. Oxygen is necessary for the production of energy. Less oxygen results in less energy for development. This makes it hard for them to grow and develop at a normal rate. They will develop the same way as other kids do, but they may take a little longer.

Q. Why do they miss school so much?
A. They have doctor appointments and periods of time when they are too sick or in too much pain to go to school.

SUMMARY

- Sickle cell pain is not visible to another person; it is important to rely on the child's report. Pain is an individual experience.
- It is imperative for children with SCD to maintain adequate hydration by drinking plenty of fluids and NOT get dehydrated.
- A child with SCD should participate in physical activities being sure to allow for adequate rest and fluid replacement. Strenuous exercise is not recommended.
- Children with SCD are a population at risk for learning difficulties. Neuropsychological and educational testing can be helpful in determining the child's strengths and weaknesses.
- What may appear to be poor motivation might be attributed to chronic fatigue or pain; therefore, it is important to get to know the child in order to make proper assessment.
- In order for a child with SCD to reach their greatest potential, it is necessary to have a collaborative effort between the child, parents, medical professionals, and school personnel.

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