



SICKLE CELL DISEASE:

INFORMATION FOR SCHOOL PERSONNEL

(Fourth Edition)



**Newborn Screening
and Genetic Services**

**Special Child Health and
Early Intervention Services**



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Introduction

This document is intended to be used as just one source of information regarding the care and treatment for students with sickle cell disease. It is meant to serve as a resource guide for school nurses and other personnel to alert them to the signs and symptoms of complications of sickle cell disease, and the steps to take should these complications occur. A glossary of terms is provided at the back of this document for personnel who are not familiar with the medical terminology in this document.

WHAT IS SICKLE CELL DISEASE?

Sickle Cell disease is a common and often life-threatening disease. It is an inherited hemolytic anemia (premature destruction of red blood cells) occurring in approximately 1 in every 400 African-American infants born in the United States each year. Individuals of Mediterranean, Arabian, Caribbean, South and Central American, and East Indian ancestry can also be affected.

Early identification can help prevent some of the serious medical problems associated with the disease. In New Jersey, universal newborn screening for Sickle Cell Disease began in 1990. Blood samples are drawn from newborns at hospitals and birth centers and submitted to the New Jersey Department of Health for laboratory analysis and reporting. Approximately 75 babies are identified each year with sickle cell disease in New Jersey.

Most of the medical care of sickle cell disease is focused upon the prevention and treatment of complications. Immediately after a newborn is identified to have sickle cell through this screening process, a referral is made to one of the Sickle Cell Treatment Centers located throughout the state of New Jersey.

Children with this disease can look forward to healthy, active and productive lives. Likewise, they can set their educational and career goals in accordance with their ambitions and innate abilities. Although a career in the military, or one that involves strenuous physical labor, may not be an option for some, there are people with sickle cell disease who have become doctors, lawyers, teachers, nurses, and other leaders in their communities.

Children with sickle cell disease are asymptomatic most of the time. However, the disease has some complications that may be severe and life threatening, with periods of crisis occurring suddenly and with little warning. These complications and crises might not happen to every child but you need to be aware of the warning signs.

Sickle Cell Trait and Disease Inheritance

Hemoglobin, a component of red blood cells, transports oxygen that is inhaled to the cells of the body. Each person has two genes for hemoglobin, one from each parent. In the person with sickle cell trait, one gene makes normal hemoglobin, known as hemoglobin A, and the other makes Hemoglobin S, or sickle hemoglobin. People with sickle cell trait **do not have** sickle cell disease and **will not develop** the disease during his or her lifetime.

The diagram below depicts the inheritance of sickle cell disease. Sickle cell disease results when both parents carrying sickle cell trait pass along the affected gene. There is a one in four, or 25 percent, chance that each parent will both pass along the affected gene resulting in sickle cell disease in the child. There are other inherited traits affecting the production of hemoglobin, most commonly thalassemia or hemoglobin-C trait. When inherited along with sickle cell trait, hemoglobin C and thalassemia result in forms of sickle cell disease. Likewise, if one parent passes along sickle cell trait and the other passes along thalassemia or hemoglobin C trait, the child will be born with sickle cell disease.

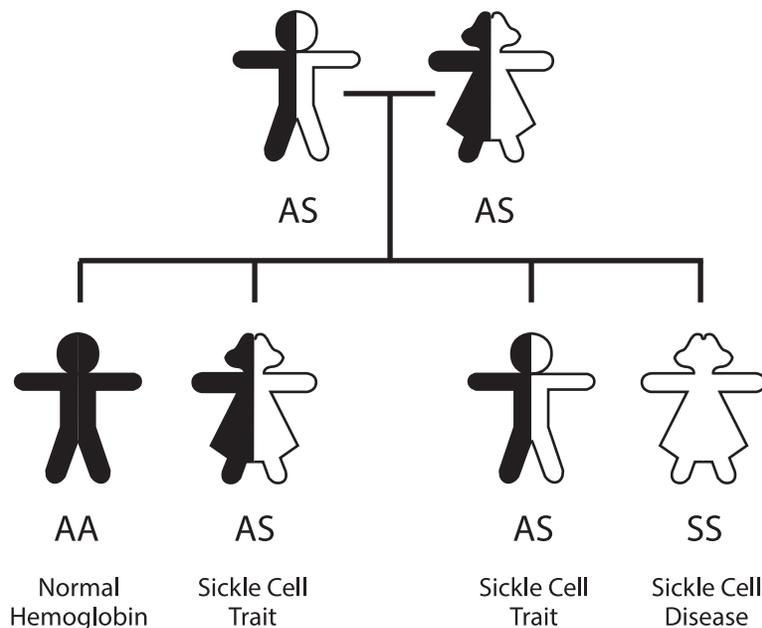


Figure 1. The inheritance pattern of sickle cell disease when each parent carries the trait.

MEDICAL EMERGENCIES

Immediately contact the child's family, and if necessary, the health care provider and/ or appropriate Sick Cell Treatment Center 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 child's family.

SYMPTOM	CALL 911	Call parent and refer to the sickle cell care team	Provide the child's parent or guardian with an update
FEVER	Lethargic, toxic in appearance, additional abnormal vital signs	Temperature 101 ° F or above or presence of shaking chills without fever	Low grade temperature
SWOLLEN SPLEEN	Lethargic, cold extremities, decreased peripheral pulses, abnormal vital signs	<ul style="list-style-type: none"> • Spleen is felt • Stomach pain • Paleness of lips, gums, nails • Fatigued 	
PAIN	Difficulty breathing, presence of fever, severe headache, neurological changes	Absence of relief from pain medications prescribed for school use, chest pain &/or cough, fever, nausea or vomiting, painful erection of the penis	Pain that is relieved by medications, supply of pain medications is running low or order is expired
HEADACHE OR WEAKNESS	Any neurological changes, hemiparesis, seizure, or severe or atypical headache always requires activation of 911 system	Non-severe headache in the absence of neurological symptoms that is unrelieved by pain medications	<p>Pain medication is helpful in relieving mild headache</p> <p>Unexpected changes in the child's learning and class room behavior</p>
LETHARGY	Extremely tired, difficult to wake up, unresponsive to commands	Responsive to commands but resistant. Too tired to participate in classroom activities after brief rest	Visit to nurse's office requiring a brief rest
DIFFICULTY BREATHING	Cyanosis, respiratory distress, use of accessory muscles, pulse oximetry below baseline with above symptoms	Chest pain with fever or increased work of breathing, chest pain unrelieved by pain medications, pulse oximetry below baseline without signs of respiratory distress, failure of bronchodilator (if ordered)	Use of nebulizers for reasons other than precautionary (i.e. physical education) with good response (if ordered), uncomplicated chest pain relieved with pain medications, if supply of bronchodilators is low or ordered expired

COMPLICATIONS OF SICKLE CELL DISEASE

Normal red blood cells are soft and flexible allowing the blood to flow easily through the vessels. In sickle cell disease, red blood cells can change to a rigid half-moon shape (sickled shape). When red blood cells sickle they are inflexible and obstruct small blood vessels. This causes a disruption to the flow of blood, and thus the delivery of oxygen. When the blood flow stops or slows suddenly, the problem is called painful crisis. The sickle-shaped red blood cells are fragile and break apart easily. The result is anemia, or a low hemoglobin level. Anemia can cause a child to become easily fatigued.

Sickle cell disease has many complications as a result of the sickling process described previously. Complications experienced vary from child to child. Complications of sickle cell disease can be categorized as acute or chronic. Acute complications often have a sudden onset, often without warning. Frequently acute complications need immediate medical attention. Chronic complications develop slowly over years. This section describes both acute and chronic complications.

ACUTE COMPLICATIONS

PAIN CRISIS

Pain crisis from sickle cell disease is the most common reason for seeking medical attention. Fever, infection, dehydration, exposure to extreme temperatures, overexertion, and/or unusual stress may trigger the pain. Sickle-shaped red blood cells occlude the venous system causing pain. Crisis may occur at any time and in any part of the body. Each person differs in terms of severity, duration, and frequency of pain episodes. The pain may be so mild that over-the-counter pain medicine is needed, or may be severe enough to require hospitalization. A child living with sickle cell disease is familiar with his or her own pain experience and will be your best resource should pain crisis occur in school. The child should be asked about location, intensity and accompanying symptoms. He or she may often be able to tell you if this pain is similar to previous experiences with sickle cell crisis to differentiate the cause of the pain. Pain scales can be very helpful in measuring the intensity. Often these are numeric scales but there are scales for younger children. The child's demeanor, use of the affected body part, depth and rate of respiration, pulse rate and blood pressure should also be observed as indicators of pain.

Be aware that due to their impaired immunity, children with sickle cell disease are at increased risk for osteomyelitis, or infection of the bone. Signs of osteomyelitis may include pain in the bone accompanied by fever and/or ill appearance. The area of pain may be swollen, red, and very tender to touch. The child with pain should be assessed for osteomyelitis.

• **SPECIAL CARE IN SCHOOL SETTING**

Treatment for pain crisis may include any or all of the following:

- ✓ Measure temperature to rule out accompanying infection, especially before giving medications that will lower the temperature and potentially mask a fever.
- ✓ Rest.
- ✓ Increased fluid intake.
- ✓ Analgesics as directed by the physician. Relaxation exercises to help relieve pain (if the child is familiar with these techniques).
- ✓ Ice and ice packs should **never** be applied to children with sickle cell disease as this will further exacerbate the sickling process.
- ✓ Assess area of pain for signs of osteomyelitis.

INFECTION/FEVER

Children with sickle cell disease are at risk for life-threatening infections. Thus, fever is always an emergency. Infection is the most common cause of morbidity and mortality in young children with sickle cell disease. The use of daily prophylactic penicillin begun shortly after birth has decreased, but not eradicated, this problem. Penicillin is often given until age 5 years. However, fever remains an emergency throughout the entire lifespan.

The child with a serious infection may not appear to be acutely ill in the early stages. Fever is the earliest sign of infection. Shaking chills and/or ill appearance in the absence of fever should be considered signs of infection. Therefore, it is important to measure the child's temperature and contact the family immediately if it is 101°F or higher. In the event of a fever and the family cannot be reached, a contingency plan for activating emergency services must be in place.

Classroom teachers should promptly send a child who does not look or feel well for evaluation by the school nurse. A temperature should be measured at each contact with the school nurse. The child's records should be "flagged" to alert substitute school nurses of the need to assess for fever.

• **SPECIAL CARE IN SCHOOL SETTING**

- ✓ **A fever of 101 degrees or higher in a patient with sickle cell disease is a medical emergency.**
- ✓ These patients must be seen in an emergency setting as soon as possible.
- ✓ **The child's family and physician should be contacted immediately.**
- ✓ Immunizations including Prevnar, Pneumovax, Menactra, HIB and the annual flu shot, must be kept up to date.

ACUTE CHEST SYNDROME

Acute chest syndrome is a life-threatening acute complication involving the lungs. It is the result of sickle shaped red blood cells clogging the vasculature of the lungs and causing the rapid spread of pulmonary infiltrates. The result can progress to overwhelming respiratory failure if emergency treatment is not implemented immediately. Antibiotics and blood transfusion must be given early in its course to reverse the progression of acute chest syndrome.

Symptoms and signs of acute chest syndrome include chest pain, tachypnea, use of accessory muscles/labored breathing, decreased breath sounds, falling pulse oximetry reading, and fever. Acute chest syndrome is often precipitated by pneumonia, fever, and pain crisis. Children with a history of acute chest syndrome are predisposed for recurrences. Parents and/or emergency services should be notified immediately if acute chest syndrome is suspected.

• SPECIAL CARE IN SCHOOL SETTING

- ✓ Assessment of child must always include a respiratory evaluation including auscultation of lungs.
- ✓ For those children who also have a diagnosis of asthma, maintain an up-to-date asthma action plan and administer medications according to that plan promptly in the event of chest pain and/or respiratory compromise.
- ✓ **A fever of 101 degrees or higher in a patient with sickle cell disease is a medical emergency, especially in the presence of cough or other respiratory symptoms.**
- ✓ These patients must be seen in an emergency setting as soon as possible.
- ✓ **The child's family and physician should be contacted immediately.**

SPLENIC SEQUESTRATION

The spleen is an organ in the upper left side of the abdominal cavity. It is normally tucked under the left rib cage. Splenic sequestration is an acute complication of sickle cell disease that occurs most commonly in children under 5 years of age, but can occur at any age. Sickle shaped red blood cells cause the sudden entrapment of a large amount of blood in the spleen and the spleen quickly becomes very large. As the spleen traps more blood the child can have signs and symptoms of severe anemia or, in very extreme instances, hypovolemic shock. Splenic sequestration is a medical emergency. It requires hospitalization and is treated with hydration, pain medication, and red blood cell transfusion. In some children, a splenectomy (removal of the spleen) is necessary.

The occurrence of splenic sequestration is difficult to predict. It comes on suddenly, and immediate action is required for the reversal of symptoms. For some children/young adults with sickle cell disease, the lower edge of the spleen may always be palpable and this is normal for that individual. Families are taught splenic palpation (feeling for the spleen) to help identify this complication at an early stage.

The school nurse should be comfortable with palpating the spleen and recognizing what is normal and abnormal. Splenic palpation should take place whenever a child with sickle cell has any of the following signs and symptoms of splenic sequestration:

- ✓ Enlarged spleen
- ✓ Sudden weakness
- ✓ Pallor (especially of lips, gums and nails)
- ✓ Abdominal pain
- ✓ Increase in abdominal girth
- ✓ Fever
- ✓ History of splenic sequestration

• ***SPECIAL CARE IN SCHOOL SETTING***

- ✓ **Should you observe these signs and symptoms, notify the child's family and advise them to contact the child's physician or sickle cell treatment center immediately. If there are signs of shock, activate emergency services (911).**
- ✓ Prompt recognition and treatment is necessary. Hospitalization and a blood transfusion may be required.

APLASTIC CRISIS

The red blood cells of people unaffected by sickle cell disease live approximately four months before they break down. For someone living with sickle cell disease, red blood cells live only a few weeks. Sometimes a viral infection can turn off the body's ability to make new red blood cells. This temporary shut off of production, in addition to the shortened life-span of sickled red blood cells, results in a sudden severe drop in hemoglobin. In other words, a person becomes more anemic than usual. If the anemia is severe enough, the child may need to be hospitalized. This acute complication is known as aplastic crisis.

Aplastic crisis is most often the result of infection with parvovirus. Parvovirus infection, also called Fifth Disease is a common childhood infection characterized by low fevers and a lacy rash on the arms. Often there is a facial rash known as the "slapped cheek rash" because it's characteristic red cheek pattern. The symptoms of Fifth Disease precede the aplastic crisis by a few days to a week. A child with aplastic crisis may have the following signs and symptoms:

- ✓ Sudden fatigue
- ✓ Pallor
- ✓ Lack of interest in play
- ✓ Shortness of breath
- ✓ Tachycardia (fast heart beat)

• ***SPECIAL CARE IN SCHOOL SETTING***

- ✓ Should you observe any of these signs and symptoms, notify the child's family immediately.
- ✓ Alert parents to any possible classroom exposures to known cases of Fifth Disease so that they child can be closely monitored.
- ✓ **Shortness of breath and tachycardia should be evaluated in an emergency setting as soon as possible.**

STROKE

Stroke is a life-threatening complication of sickle cell disease. It occurs when part of the brain does not get as much blood as it needs. This is caused by sickle cells blocking a blood vessel in the brain. The incidence of stroke in children with sickle cell disease is approximately 10%. Children diagnosed with HbSS or HbS-beta thalassemia-0 are at greatest risk. Once a child has experienced a stroke, he or she is at a significant risk for a subsequent stroke. To prevent this from occurring, these children will receive lifelong transfusions of red blood cells with the goal of limiting the amount of blood that can become sickle-shaped and block the vessels of the brain.

The child who has suffered a stroke will potentially experience life-long cognitive impairment requiring ongoing educational support. It is important that these children are assessed and supported at various times in their educational careers.

Any child with sickle cell experiencing one or more of these signs and symptoms should be considered a medical emergency:

- ✓ Aphasia,
- ✓ Hemiparesis,
- ✓ Vision loss or changes,
- ✓ Severe headache,
- ✓ Loss of consciousness,
- ✓ Seizure
- ✓ Any change in affect, orientation, behavior, or level of awareness.

• SPECIAL CARE IN SCHOOL SETTING

- ✓ Notify the child's parents or Sickle Cell Treatment Center if the above symptoms occur.
- ✓ **Activate emergency services (911).**
- ✓ These children need to be evaluated in an emergency setting as soon as possible. Rapid medical intervention may limit the devastating consequences of stroke.

PRIAPISM

Priapism (continued erection of the penis without sexual desire) occurs when sickled cells block blood circulation in the penis. Boys and young men who have experienced priapism in the past are likely to have a recurrent episode.

• SIGNS AND SYMPTOMS:

- ✓ Painful, prolonged swelling of the penis
- ✓ Sustained unwanted erection

• SPECIAL CARE IN SCHOOL SETTING

- ✓ Attempt to dispel embarrassment.
- ✓ Counsel adolescents regarding the importance of reporting signs and symptoms.
- ✓ Encourage child to empty bladder.
- ✓ Administer pain medication.
- ✓ Notify the family and physician IMMEDIATELY when symptoms are present.

CHRONIC COMPLICATIONS

RENAL CHANGES

Children with sickle cell disease urinate frequently due to an inability to concentrate urine. This inability to concentrate urine is the result of damage to the kidneys due to sickle cell disease. Children are also more prone to kidney infections. Nocturnal enuresis is often seen in these children beyond the age that would be expected.

• *SPECIAL CARE IN SCHOOL SETTING*

- ✓ The child should be allowed to use the bathroom frequently.
- ✓ Notify the family if child has hematuria (blood in the urine).
- ✓ The child needs to drink plenty of fluids throughout the day.
- ✓ The child may be concerned if there is an overnight field trip because of enuresis.
- ✓ If sleep is disturbed by enuresis, the child may exhibit fatigue.

CHOLELITHIASIS (GALLSTONES)

Adolescents and young adults often have problems with gallstones (cholelithiasis). This is due to excessive bilirubin, which is a product of increased breakdown of red blood cells. Excess bilirubin may collect in the gallbladder and form stones. The pain of gallstones is often intermittent and may be worse after eating. It comes on suddenly and is most likely felt in the right upper quadrant of the abdomen. Often the child will feel nauseous and may vomit. It is not uncommon for children to require that the gallbladder be removed surgically.

Gallstones can cause acute inflammation of the gallbladder or cause a blockage. This situation requires immediate attention. The signs and symptoms of inflammation or blockage may include the following: persistent abdominal pain, increased jaundice (yellow appearance of eyes, skin), vomiting, and possibly fever.

• *SPECIAL CARE IN SCHOOL SETTING*

- ✓ Notify the child's parents or Sickle Cell Treatment Center of any symptoms of gallstones.

IMPAIRED COGNITION

Children who have experienced stroke may or may not exhibit outward signs but should assume to have cognitive deficits until proven otherwise by neuropsychological testing. The child who has suffered a stroke secondary to sickle cell disease is at extremely high risk for another neurological event. For this reason, the child will be on a chronic transfusion regimen receiving a red blood cell transfusion every two to four weeks. The goal of transfusion therapy is to suppress the production of red cells that can sickle and obstruct the damaged vessels of the brain. Chronic transfusion therapy requires that a great deal of school be missed. Academic accommodation plans should take this into consideration.

Emerging research indicates that sickle cell disease, particularly the more severe types, may cause progressive loss of cognitive function. These changes are often very subtle. Students should be monitored closely and a child study evaluation arranged if there is a suspicion of academic concerns, even without a previous history. Neuropsychological testing should strongly be considered at school transitions particularly between elementary and high school, and then again before college.

AVASCULAR NECROSIS

When blood vessels supplying bone get blocked by sickle cells, the bone can become damaged. This condition, known as avascular necrosis, occurs most often in adolescents and adults with sickle cell disease. The joints most commonly affected are the ball-in-socket joints of the hips and shoulders. Avascular necrosis causes an arthritic type of pain in the affected joint that is persistent. If the hip is affected, it may cause a limp. Prolonged walking, sitting, or climbing of stairs may be painful. Physical therapy may help the symptoms. In severe cases, surgery to replace the joint may be needed. Accommodations may be made to protect the joints. An example may be the provision of an extra set of text books at home to avoid burden of carrying a backpack. The student may be allowed to leave early at the change of classes if walking quickly is difficult or use an available elevator.

• SPECIAL CARE IN SCHOOL SETTING

- ✓ Notify the child/young adult's family if these symptoms are present.
- ✓ Oral analgesics.
- ✓ Increased fluids
- ✓ Appropriate limitation of activities in physical education
- ✓ Rest

SICKLE CELL RETINOPATHY

Children with sickle cell disease may develop sickle cell retinopathy. If untreated, this can lead to blindness. This condition manifests slowly and rarely occurs before the age of ten years. Sickle cell retinopathy is more common in children with Hemoglobin SC disease.

• SPECIAL CARE IN SCHOOL SETTING

- ✓ **Notify the child's family immediately of any vision problems or trauma to the eye.**
- ✓ After the child reaches ten years of age, an ophthalmologist, who is familiar with sickle cell disease, should see him/her annually.
- ✓ Alert parents to the fact that, if the school provides vision screening, this does not take the place of an annual examination by an ophthalmologist.

SKIN PROBLEMS

Some patients develop poorly healing skin ulcers, especially around their ankles. This problem does not happen often and usually affects older adolescents and adults. These ulcers are often very hard to treat and care should be taken to prevent them. Wounds should be assessed and cleaned with an antiseptic soap.

• SPECIAL CARE IN SCHOOL SETTING

- ✓ Alert the parent/guardian if the child experiences a wound, especially on the lower part of the leg.
- ✓ Clean and dress the wound.
- ✓ Continue to assess the wound for adequate healing and alert the parent/guardian if there are concerns about poor healing or infection.

MEDICAL MANAGEMENT

Bone Marrow Transplant

Currently the cure for sickle cell disease is bone marrow transplant. Due to the complex nature of transplant and the high risk of complications, this treatment option is reserved for the most severe cases. Transplant requires a genetically matched donor. In the best case scenario, this donor is a sibling.

Children who undergo transplant will miss almost a great deal of school during the process and in the months required for recovery. The child will require homebound instruction during this period. Tutors should be made aware that the child is immunosuppressed and cancelling a session is necessary should the tutor feel ill. Before returning to school, a meeting should take place with the family, school personnel and the health care team to discuss accommodations and precautions necessary.

Red Blood Cell Transfusion

It is not uncommon for children with sickle cell disease to receive many blood transfusions. Typically, a transfusion is given to lessen or shorten an acute episode requiring hospitalization. Some children will be transfused regularly to avoid frequently recurring complications or avoid a stroke if the child is at increased risk. These children will miss a school day every 2-4 weeks for this procedure. Arrangements for making up that lost day should be made in advance.

Children on regular transfusions should feel well most of the time. If a child who has recently received a transfusion has complaints of worsening fatigue, lower back pain, hematuria (bloody urine), and is visibly jaundice, this may indicate a transfusion reaction is taking place. While rare, this type of delayed transfusion reaction is a medical emergency and the family and/or hematology treatment team should be notified immediately.

Children who receive a number of transfusions are at risk for a complication called iron overload. Each transfusion contains iron which the body holds on to in the organs. Over decades, this stored iron can cause irreversible organ damage, particularly to the heart and liver. Children with iron overload take a medication known as a chelator which removes iron from the body. Chelators may cause rashes, stomach upset and loose stools.

Hydroxyurea

Hydroxyurea is a form of chemotherapy that has been established as a treatment for moderate to severe sickle cell disease. Research has shown that hydroxyurea can significantly decrease the number of complications and hospitalizations. Hydroxyurea is given once a day. It is chemotherapy so the medication, as well as any bodily waste, should be handled while wearing gloves. Mild nausea is a possible side effect. Hydroxyurea may decrease the white blood cell count leading to increased risk of infection. A decreased platelet count can possibly result in bruising or bleeding.

Pain Medications

- **Mild pain** can often be successfully treated with non-opioid medications such as ibuprofen or acetaminophen. These medications are non-sedating and with the exception of mild stomach upset, do not have side effects. Both medications will lower fever. Therefore, the temperature should be checked prior to their administration to avoid masking a fever indicating there may be an infection. Because a child with mild discomfort can return to the classroom, it is important to make arrangements with the family for the school nurse to administer these medications in school.
- **Moderate to Severe Pain** is treated with the addition of opioid analgesics. The most commonly used opioids are codeine, morphine and oxycodone. Children with pain resulting from sickle cell disease may often continue to attend school. If the child is able, attendance should be encouraged. If opioid analgesics are taken, sedation is possible. Accommodations should be made for the child to rest in the nurse's office and return to school when feeling rested. School work may be modified to insure the concepts are learned but the volume of work is manageable during periods when opioid analgesics are required.
- **Nonmedical pain management** Self-hypnosis and relaxation exercises, massage, or the application of warm, never ice or cold, compresses may help decrease the use of narcotic analgesics and also provide the child with a sense of control.
- **Hydration** Fluid, preferably water, intake should be increased during painful episodes.

Antibiotics

Penicillin prophylaxis, given twice a day in a low dose, should be initiated by 3 months of age. Compliance is essential to prevent dangerous infection. Penicillin is stopped in most patients between three and five years of age. In patients experiencing previous infections with pneumococcal bacteria, or those who have undergone splenectomy, this prophylaxis may continue beyond age 5 years.

PSYCHOSOCIAL ISSUES

Taking care of a student with sickle cell disease not only involves managing the physical complications, but also includes resolving the psychological and social issues that are important to the student's well-being and continued development.

• PHYSICAL ACTIVITY

- ✓ Children should take an active part in any physical activity that interests them. Let them set their own pace.
- ✓ Aerobic exercises like running, swimming, walking, and biking are recommended. Just remember, the body will lose more fluid during exercise, so it is important to frequently replenish these fluids.
- ✓ Swimming in very cold water may trigger a painful crisis in some children. Therefore, check the water temperature before the child goes swimming.
- ✓ In the child with a spleen edge that is felt below the rib cage, contact sports should be avoided.

• NUTRITION AND DIET

- ✓ Eat a well-balanced diet appropriate for the student's age group. There is no special diet for children with sickle cell disease.
- ✓ Drink plenty of fluids, such as water and juice, to avoid dehydration
- ✓ Drink extra fluids during increased physical activity, when temperatures are warm, and during periods when the child is experiencing pain.

GROWTH AND DEVELOPMENT

Children with sickle cell disease may experience a delay with growth and development as compared to their peers. They may be shorter than their peers and puberty may be delayed. Self-esteem can be severely undermined by the teasing of classmates and/or by not being allowed to participate in group activities. Adults may treat the child as if he/she is younger because of his/her smaller size.

504 EDUCATION PLAN

Because of the disease and its treatment, it may be necessary for the child to be absent from school or be unable to concentrate on academic subjects. Implementation of a 504 plan can help children keep up with their class. Educators must be cautious, though, to not attribute true educational deficits to sickle cell disease. A child study team evaluation should be performed if a child is struggling to rule out learning impairments unrelated to sickle cell disease.

VOCATION COUNSELING

All young adults should be encouraged to set career goals. Guidance is especially important for those with sickle cell disease because there are certain careers that are not open to them. Military careers and jobs requiring heavy manual labor are not an option 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.

THE TEACHER

The teacher is a direct observer of the daily behavior, activities and functions of a child in the classroom. The teacher's observations should be included when assessing the child's ability to learn and progress at the appropriate pace.

Specific areas to assess:

- ✓ Behavior - related to learning and socialization. The teacher should concentrate on the child's ability to attend, achieve, and participate in academics and activities. The student should perceive that he/she is an integral and valuable member of the class and school.
- ✓ Physical Appearance.- the teacher should know the student well and recognize, often by assessing the child's body language, how he/she is feeling, both physically and emotionally.
- ✓ Medical - the teacher needs to have a comprehensive knowledge of the child's illness, symptoms and treatment plan. The teacher should also have a thorough awareness of the signs and symptoms that may be considered a medical emergency for a child with sickle cell disease.

THE SCHOOL NURSE

The school nurse plays a vital role in coordinating health services for students with sickle cell disease in the school setting. The Individualized Health Plan, or IHP, focuses on the specific health needs and problems of a student at a given time in their development and educational career. The IHP outlines the plan of health related services and programs required to support the student's safe and successful functioning in the school setting. Development of an IHP provides for effective and efficient delivery of health-related services that promote school success for the student.

Specific areas to assess:

- Previous history – type of sickle cell disease, primary healthcare provider, previous hospitalizations or emergency department visits.
- Current Status and Management – review home management routine including medications and fluid requirements, identify current problems, pain management scale, immunization requirements and current physical assessment.
- Self-care – areas the student manages alone, areas that require supervision. Assess age appropriate knowledge about sickle cell disease, knowledge of risk factors and health maintenance measures (diet, fluid requirements, rest, exercise, hygiene, reporting symptoms promptly). Review medical records and treatment plans from the healthcare provider.
- Psychosocial Status – student's strengths, family support system, friends, and social activities.
- Academic Issues – assess academic and attendance records, need for homebound or hospital instruction. Review the need for modified physical education, nursing assessment, medication and management of symptoms in the school, including off campus school activities. Serve as a health advocate for the child.

APPENDIX

GLOSSARY

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 which 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.

Chemotherapy - Medicines that are used to fight cancer.

Cholelithiasis - the presence of gallstones in the gallbladder.

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.

Dysfunction - unable to function normally.

Dyspnea - difficulty breathing.

Enuresis - the inability to control urination.

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

Genetic Centers - genetic specialists can arrange for genetic tests and can answer questions on how genetic conditions may be passed on in families.

Geneticist - a doctor who specializes in hereditary conditions.

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.

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.

Nephrotic Syndrome - a kidney disease.

Osteomyelitis - bone infection.

Pallor - paleness of skin.

Platelets - part of the blood that helps make blood clot.

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 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.

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

Splenic Sequestration - spleen overfills with blood.

Stroke - clogging of blood vessels to the brain.

Tachycardia - rapid heart beat

Tachypnea - fast rate of breathing.

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

Ulcers - sores on the skin.

Urination - passing water or peeing.

Venous - of the veins.

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

REFERENCE

National Heart, Lung, and Blood Institute (2014). Evidenced based management of sickle cell disease, expert panel report. Retrieved September 22, 2016 from <https://www.nhlbi.nih.gov/health-pro/guidelines/sickle-cell-disease-guidelines>