SICKLE CELL DISEASE:

INFORMATION FOR
SCHOOL PERSONNEL

Third Edition

Division of Family Health Services
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 school age students with sickle cell disease. It is meant to serve as a resource guide for school health nurses and other school personnel to alert them to the signs and symptoms of complications of the sickle cell diseases, and what should be done if they 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, non-contagious hemolytic anemia (premature destruction of red blood cells with the release of hemoglobin) 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 State Department of Health and Senior Services 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 involves prevention and treatment of complications. A team of specialists works cooperatively with each child's health care provider to improve access to primary and specialty care services through Sickle Cell Treatment Centers located throughout the state of New Jersey. A list of these centers can be found in the appendix at the end of this document (page 29).

Like other children, children with this disease can look forward to a healthy, active and productive life; and 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 should be urged to participate in school activities at an early age. They 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. Then the school, along with treatment centers and the child's health care provider, can assist the family in managing the child's health care needs.
**WARNING SIGNS**

Immediately contact the child's family, and if necessary, the health care provider and/or appropriate Sickle 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. The child may require emergency treatment at his/her health care provider’s office, sickle cell treatment center, or hospital emergency room.

<table>
<thead>
<tr>
<th>SIGNS</th>
<th>SYMPTOMS</th>
</tr>
</thead>
<tbody>
<tr>
<td>FEVER</td>
<td>101 degrees or above</td>
</tr>
<tr>
<td>IRRITABILITY</td>
<td>readily excited to impatience or anger</td>
</tr>
<tr>
<td>PALLOR</td>
<td>looks pale in face or tongue</td>
</tr>
<tr>
<td>BREATHING</td>
<td>dyspnea</td>
</tr>
<tr>
<td></td>
<td>tachypnea</td>
</tr>
<tr>
<td></td>
<td>stertorous breathing</td>
</tr>
<tr>
<td>HEADACHE</td>
<td>sudden or constant</td>
</tr>
<tr>
<td>HEARTBEAT</td>
<td>tachycardia</td>
</tr>
<tr>
<td></td>
<td>pounding</td>
</tr>
<tr>
<td>PAIN IN THESE AREAS</td>
<td>head chest</td>
</tr>
<tr>
<td></td>
<td>joints</td>
</tr>
<tr>
<td></td>
<td>abdomen</td>
</tr>
<tr>
<td></td>
<td>penis (with prolonged unexpected erection)</td>
</tr>
<tr>
<td>SWELLING</td>
<td>hands</td>
</tr>
<tr>
<td></td>
<td>feet</td>
</tr>
<tr>
<td></td>
<td>joints</td>
</tr>
<tr>
<td>HEMIPARESIS</td>
<td>on either side of the body</td>
</tr>
</tbody>
</table>
WHAT IS SICKLE CELL TRAIT?

Each person has a pair of genes for hemoglobin. Normal hemoglobin is referred to as Hemoglobin A. The letters (AA) are used to indicate that both hemoglobin genes are normal. In the child with sickle cell trait, one gene is normal and the other makes Hemoglobin S (Sickle). This combination of genes is referred to as (AS): (A) for the normal gene and (S) for the gene that makes Hemoglobin S.

If a child has only one gene that makes sickle hemoglobin, the child does not have sickle cell disease and will not develop the disease later in life. The condition is not contagious either. It is important, however, for the child to know that he or she has sickle cell trait. This information will be very important as part of genetic counseling to a person when he or she is planning to become sexually active. In each pregnancy with which both partners have sickle cell trait, there is a 25 percent chance that the offspring would be born with sickle cell disease.

Usually patients with sickle cell trait do not have any complications but hematuria, due to sickled blood cells in the kidney.

There are other hemoglobinopathies such as Thalassemia and Hemoglobin-C (AC) which can result in sickle cell disease when inherited along with sickle cell trait (AS). It is important to understand that people with other hemoglobinopathies such as Thalassemia and Hemoglobin-C will have negative results when screened specifically for sickle cell trait, but could have a child with a form of sickle cell disease.

For further information or any questions, contact a Sickle Cell Treatment Center (see page 29).
Sickle cell disease has many complications, some of which are severe and life threatening. Periods of crisis can occur suddenly and often with little warning. Complications and crises may not happen to every child but you need to be aware of the warning signs. This section describes many of those complications.

- **SIGNS AND SYMPTOMS** - details the early warning signs of each problem and
- **SPECIAL CARE IN THE SCHOOL SETTING** - describes some interventions that can be utilized by school personnel.

There are Sickle Cell Treatment Centers located throughout New Jersey with teams of specialists who work cooperatively with each child's health care provider to improve access to primary and specialty care services. School personnel, along with the Sickle Cell Treatment Centers and each student's health care provider, can assist the family in managing each child's health care needs.

**THE SICKLING PROCESS**

Sickle cell disease affects the hemoglobin part of red blood cells. Hemoglobin carries oxygen from the lungs to other parts of the body and gives blood its red color.

Normal red blood cells are round, soft, and flexible. They can squeeze through small blood vessels allowing the blood to flow easily. In sickle cell disease, red blood cells can change from round to half moon (or sickle shaped) when the hemoglobin gives off its oxygen. When red blood cells are shaped like a sickle, they are hard and inflexible and plug up small blood vessels. This causes the flow of blood and oxygen to be slowed down or stopped.

Pain, damage to parts of the body and anemia can result. When the blood flow stops or slows suddenly, the problem is called sickle cell crisis. The following chart (Sickling of Venous Blood) shows how some body organs and tissues may be affected by the plugging of blood vessels.
### SICKLING OF VENOUS BLOOD

<table>
<thead>
<tr>
<th>ORGAN/TISSUE INVOLVED</th>
<th>PROBLEMS CAUSED</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>KIDNEY</strong></td>
<td>* Enuresis</td>
</tr>
<tr>
<td></td>
<td>* Hematuria</td>
</tr>
<tr>
<td></td>
<td>* Nephrotic Syndrome</td>
</tr>
<tr>
<td></td>
<td>* Unconcentrated urine</td>
</tr>
<tr>
<td></td>
<td>* Urinary frequency</td>
</tr>
<tr>
<td><strong>SPLEEN</strong></td>
<td>* Increased risk for serious infections</td>
</tr>
<tr>
<td></td>
<td>* Splenic Sequestration</td>
</tr>
<tr>
<td></td>
<td>* Abdominal pain</td>
</tr>
<tr>
<td><strong>LUNGS</strong></td>
<td>* Pneumonia</td>
</tr>
<tr>
<td></td>
<td>* Acute Chest syndrome</td>
</tr>
<tr>
<td><strong>BONES</strong></td>
<td>* Infection</td>
</tr>
<tr>
<td></td>
<td>* Aseptic Necrosis</td>
</tr>
<tr>
<td><strong>BRAIN</strong></td>
<td>* Stroke</td>
</tr>
<tr>
<td></td>
<td>* Headache</td>
</tr>
<tr>
<td><strong>SKIN</strong></td>
<td>* Slow healing ulcers</td>
</tr>
<tr>
<td><strong>PENIS</strong></td>
<td>* Priapism</td>
</tr>
<tr>
<td><strong>EYES</strong></td>
<td>* Sickle Cell Retinopathy</td>
</tr>
<tr>
<td><strong>LIVER</strong></td>
<td>* Hepatomegaly</td>
</tr>
<tr>
<td></td>
<td>* Cholelithiasis</td>
</tr>
<tr>
<td></td>
<td>* Jaundice</td>
</tr>
</tbody>
</table>

* Not all these problems happen to everyone with Sickle Cell Disease. You need to know, however, that they can happen. **Notify the parents/caregivers immediately if you think their child has any of these problems.**
PAIN

Pain is the most common problem in sickle cell disease. Fever, infection, exposure to extreme cold, physical exhaustion, and/or unusual stress/anxiety may trigger the pain. Sickling in the venous system causes pain. It may occur at any time and in any part of the body. Each person differs in terms of severity, duration, and frequency of pain. The pain may be so mild that no analgesic (pain medicine) is needed, or may be severe enough to require hospitalization. Patients with sickle cell disease should be encouraged to take an active role in the management of their crises.

♦ SIGNS AND SYMPTOMS
  ✓ chest pain
  ✓ dyspnea
  ✓ fever 101 degrees or higher
  ✓ hemiparesis
  ✓ priapism
  ✓ nausea, vomiting
  ✓ severe abdominal pain
  ✓ bone pain and swelling

• SPECIAL CARE IN SCHOOL SETTING
  Treatment to relieve mild to moderate pain may include any or all of the following:
  ✓ Rest (bed-rest or a decrease in activity).
  ✓ Increased fluid intake with drinks such as caffeine-free soft drinks, juices, or bouillon.
  ✓ Analgesics as directed by the physician. There are slow-release preparations of morphine and codeine, which can be given every twelve hours, often obviating the need to give medication in school.
  ✓ Relaxation exercises to help relieve pain (if the child is familiar with these techniques).
  ✓ Avoid rapid immersion in cold water.
  ✓ Avoid excessive exposure to cold temperatures.

***Call the parent/caregiver and/or physician immediately if any of these symptoms are present. If the family is unavailable, call the child’s health care provider or the Sickle Cell Treatment Center immediately.
KIDNEY

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 repeated sickle cell crises. Children with sickle cell disease are also more prone to kidney infections.

◆ SIGNS AND SYMPTOMS
  ✓ dysuria
  ✓ hematuria
  ✓ more frequent urination than usual for this child
  ✓ enuresis remains for older children with sickle cell disease

◆ 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 get plenty of fluids throughout the day.
  ✓ The child may be concerned if there is an overnight trip because of enuresis.

FLUID REQUIREMENTS FOR CHILDREN WITH SICKLE CELL DISEASE

<table>
<thead>
<tr>
<th>CHILD’S WEIGHT</th>
<th>APPROXIMATE DAILY FLUID REQUIREMENTS</th>
</tr>
</thead>
<tbody>
<tr>
<td>25 lbs.</td>
<td>1½ to 1¾ qts.</td>
</tr>
<tr>
<td>50 lbs.</td>
<td>2 to 2½ qts.</td>
</tr>
<tr>
<td>75 lbs.</td>
<td>2¼ to 3 qts.</td>
</tr>
<tr>
<td>100 lbs.</td>
<td>2½ to 3½ qts.</td>
</tr>
</tbody>
</table>
**INFECTION/FEVER**

Infection is a life-threatening problem in sickle cell disease. Children with sickle cell disease have less resistance to infection and develop infections more easily and more frequently than other children. Children with sickle cell disease are typically on prophylactic (preventive) penicillin until five years of age. Infection is the most common cause of morbidity and mortality in young children with sickle cell disease. Prophylactic penicillin has decreased, but not eradicated, this problem.

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 (temperature of 101 degrees or higher) is the most common symptom of infection
  ✓ lethargy (unexplained tiredness)

♦ **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.**
  ✓ Up to date immunizations with Prevnar, Pneumovax, HIB.

**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. Dyspnea (difficulty breathing) and chest pain are typical symptoms. A transfusion is often necessary. The combined symptoms are called Acute Chest Syndrome, which can be life threatening.

♦ **SIGNS AND SYMPTOMS**
  ✓ chest pain ***
  ✓ cough
  ✓ dyspnea
  ✓ tachypnea (rapid breathing)
  ✓ fever

♦ **SPECIAL CARE IN SCHOOL SETTING**
  ✓ Encourage deep breathing exercises i.e. use of an incentive spirometer (machine that measures the amount of inhaled air) when recommended by child's physician (Sometimes pain may prevent the child from breathing deeply.)
  ✓ Encourage compliance with medications for asthma.
  ✓ Report any of the above signs and symptoms to the child's family or the Sickle Cell Treatment Center.

*** Recognize that chest pain is an emergency. Hospitalization of the child with acute chest syndrome is imperative.
SPLEEN

One of the spleen's functions in the body is to filter out bacteria from the bloodstream and help the body fight infection. Sickling in the spleen eventually causes fibrosis, impairing its ability to filter out bacteria. These children are therefore at a higher risk for developing infections.

Splenic sequestration is a possible complication of sickle cell disease that occurs most commonly in children under 5 years of age, but can occur at any age. It occurs when there is 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. The child can have signs and symptoms of a very low blood count (he/she becomes very anemic) or, in very extreme instances, shock. This is usually treated with hydration, analgesics (pain medication), and red 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 spleen may always be palpable and this is normal for that individual. Again, it is important to know what is normal for the child. Many families are taught splenic palpation (feeling for the spleen) to help identify this complication at an early stage.

♦ **SIGNS AND SYMPTOMS**
  ✓ sudden weakness
  ✓ pallor (especially of lips, gums and nails)
  ✓ abdominal pain
  ✓ increase in abdominal girth
  ✓ enlargement of the spleen
  ✓ fever

♦ **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. ***
  ✓ These patients need to be evaluated in an emergency setting as soon as possible.

***Prompt recognition and treatment is necessary. Hospitalization and a blood transfusion may be required.
ANEMIA

Children with sickle cell disease usually have anemia. They usually adjust and are asymptomatic (without symptoms). There are times, however, when a child has a substantial drop in the number of red blood cells. Failure to adequately replace the red blood cells can cause pallor, fatigue, and loss of appetite. An acute, (sudden) drop in the red cell count is referred to as an aplastic crisis. This is discussed in detail in the next section.

Normal red blood cells live for approximately four months before they break down. Regeneration normally occurs within four months. Sickled red blood cells live less than one month. The body is unable to produce sufficient red blood cells for replacement, resulting in anemia. When red blood cells break down, bilirubin (the orange-yellow pigment in bile) is produced. In sickle cell anemia, excess bilirubin causes jaundice (yellowing) which can be seen in the sclera (white portion) of the eyes, the color of the urine, and the color of the skin. These are often normal findings in a child with sickle cell disease.

♦ SIGNS AND SYMPTOMS
  ✓ fatigue
  ✓ poor appetite
  ✓ pallor
  ✓ jaundice that is greater than normal for the child

♦ SPECIAL CARE IN SCHOOL SETTING
  ✓ Should you observe any of these signs and symptoms, notify the child's family

APLASTIC CRISIS

An aplastic crisis 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 bone marrow slows down and much needed red blood cells are not produced. The life expectancy of a sickled red blood cell is 15 to 20 days instead of the typical four months for normal red blood cells.

♦ 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.
  ✓ Shortness of breath and tachycardia should be evaluated in an emergency setting as soon as possible***

*** Hospitalization and blood transfusion may be necessary.
BONES AND JOINTS

Osteomyelitis
Often the signs and symptoms of this infection are similar to those of sickle cell pain episodes in the joint or bone.

♦ SIGNS AND SYMPTOMS
  ✓ pain
  ✓ swelling
  ✓ erythematous (red) joint
  ✓ fever
  ✓ warmth

Painful Crisis
Almost all children with sickle cell disease will have a painful crisis at some point in their lives. The pain and swelling occur as the result of vessels being occluded (blocked) by sickle cells. Some children have them very often. A fever or infection, dehydration, exposure to extreme heat or cold, physical exhaustion, and unusual stress or anxiety may trigger the pain. However, most often there is no obvious reason why a child gets a painful crisis. The pain can happen in any part of the body.

♦ SIGNS AND SYMPTOMS OF PAIN CRISIS
  ✓ Bone pain
    ▪ pain
    ▪ swelling of the hands and feet in young children; long bones and back in older children
    ▪ fever
  ✓ Abdominal pain
    ▪ abdominal distention
    ▪ dyspnea

♦ SPECIAL CARE IN SCHOOL SETTING
  ✓ Fluids – It is recommended that the child be given juices rather than water so as to replace the electrolytes lost due to frequent urination.
  ✓ Rest.
  ✓ Analgesics (pain medicine) as recommended by the child’s treatment center.

CHOLELITHIASIS (GALLSTONES)
Adolescents and young adults often have problems with cholelithiasis. This is due to excess bilirubin (product of blood breakdown) caused by red blood cell destruction. Excess bilirubin may also collect in the gallbladder and form stones.

♦ SIGNS AND SYMPTOMS
  ✓ abdominal pain
  ✓ vomiting
  ✓ fatty food intolerance

♦ SPECIAL CARE IN SCHOOL SETTING
  ✓ Notify the child's parents or Sickle Cell Treatment Center. ***

*** Hospitalization and surgery may be necessary.

CENTRAL NERVOUS SYSTEM
Stroke is a serious complication of sickle cell disease. It occurs when part of the brain doesn't get as much blood as it needs. This is caused by sickle cells blocking a blood vessel in the brain. There is a lifetime incidence of approximately 10%. Once experienced, there is a tendency for recurrence.

- **SIGNS AND SYMPTOMS**
  - aphasia
  - hemiparesis
  - 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
  - speech trouble

(These symptoms do not necessarily indicate that a stroke has or will occur).

- **SPECIAL CARE IN SCHOOL SETTING**
  - Notify the child's parents or Sickle Cell Treatment Center if the above symptoms occur. ***
  - These patients need to be evaluated in an emergency setting as soon as possible.
  - There is now a screening test for children at risk of stroke called a transcranial Doppler. Patients of school age have this done annually.

*** If a stroke is diagnosed, the child/young adult should be hospitalized immediately for observation, testing, and treatment. Treatment is aimed at preventing a second stroke and this is achieved by giving transfusions of red blood cells on a chronic basis in order to maintain the percent of Hemoglobin S at less than 30%.

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.

**PRIAPISM**

Priapism (continued erection of the penis without sexual desire) occurs when sickle cells block blood circulation in the penis.

- **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.
  - Notify the family and physician IMMEDIATELY when symptoms are present. ***

*** Hospitalization may be required.
ASEPTIC NECROSIS

When blood vessels supplying bone get blocked by sickle cells, the bone can become damaged. When sickling occurs repeatedly in the veins supplying bone, damage may occur in these tissues.

♦ SIGNS AND SYMPTOMS
  ✓ Chronic hip, extremity or lower back pain which does not resolve
  ✓ Afebrile
  ✓ Occurs most commonly in the hip bone
  ✓ Persistent pain in groin or buttock
  ✓ Gait (manner of walking) disturbance

♦ 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
  ✓ Physical therapy may be necessary
  ✓ Surgery is necessary in severe cases

SICKLE CELL RETINOPIATHY

Children with sickle cell disease may develop sickle cell retinopathy, which can occasionally 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.

SKIN PROBLEMS

Some patients develop ulcers, especially around their ankles. This problem does not happen often and usually affects older children and adults.

♦ SIGNS AND SYMPTOMS
  ✓ Cut or wound that doesn't heal.
  ✓ A patch of dry itchy skin.

♦ SPECIAL CARE IN SCHOOL SETTING
  ✓ If the child has any ulcerations (skin sores), which are not healing normally, or look infected notify the parents.
  ✓ Keep the area clean.
  ✓ Put on a fresh bandage twice a day or as often as directed.
MEDICAL MANAGEMENT

There is currently no cure for sickle cell disease except for Bone Marrow Transplants, in select cases. Patients who have many complications may also be treated with a drug called, Hydroxyurea, that helps decrease sickling.

TRANSFUSION THERAPY

Transfusion therapy is generally used in the following circumstances:

♦ In patients with symptoms of
  o hypoxemia
  o severe anemia
  o acute chest syndrome

♦ In patients requiring a rapid decrease in Hemoglobin S (exchange transfusion) i.e.:
  o Severe infections
  o Stroke
  o Splenic sequestration
  o Severe Priapism
  o Acute Chest Syndrome
  o Refractory (unrelievable) pain crisis

♦ In preparation for surgery.

♦ In patients with certain chronic states every 2-4 weeks.
  o Minimum of 3-5 years after a stroke; may be life-long
  o Leg ulcers
  o During a complicated pregnancy
  o In severe recurrent vaso-occlusive crises

Children on chronic transfusion therapy run the risk of iron overload. When the ferritin levels rise above 2000 ng/ml., the patient is started on Desferoxamine, a chelating agent. The kidneys excrete Desferoxamine. This may cause the urine to turn a reddish color in some patients. The medication is usually given subcutaneously (under the skin) at home and intravenously while in the hospital.

A child on a chronic transfusion program will miss at least one day of school every three to four weeks. He or she may also be on a program of iron chelation with the drug Desferoxamine, to remove the iron that accumulates from these transfusions.

VASO-OCLUSION CRISIS (PAIN)

Any condition that tends to cause dehydration (loss of water) may precipitate sickling in the microvasculature (small blood vessels).

♦ Analgesics Mild pain in younger children can often be successfully treated with acetaminophen at regular doses. With more severe pain, the usual oral regimen is a combination of codeine and acetaminophen. Most pain episodes can be successfully treated at home with oral analgesics and increased oral hydration. Severe pain may require parenteral narcotic analgesics and IV hydration. Some patients may be treated with drugs that might prevent pain crisis, such as hydroxyurea.

♦ Nonmedical pain management Self-hypnosis and relaxation exercises may help decrease the use of narcotic analgesics and also provide the child with a sense of control.

♦ Hydration Fluid intake should be increased during painful episodes. Caffeine-free soft drinks, juices, and bouillon are recommended.
INFECTION/FEVER

Prevention or early aggressive treatment is very important. Infections will be treated with antibiotics.

♦ Penicillin prophylaxis (b.i.d. low dose penicillin) initiated by 3 month of age. Compliance is essential to prevent dangerous infection. Penicillin can be stopped in most patients by five years of age.

♦ With intercurrent infection, the physician may increase the dosage of penicillin and/or change the antibiotic.

Routine immunizations should be administered in accordance with New Jersey Health Services Guidelines, page 76, regarding Immunization compliance and documentation. It is especially important that patients with sickle cell receive Prevnar, Hib, and Hepatitis B vaccine. Administration of 23-valent pneumococcal polysaccharide vaccine should be given at 2 years of age, and repeated every three to five years until 10 years of age in all children with sickle cell disease. In a diagnosed child older than 10 years of age who has never received the vaccine, a one-time dose of 23-valent pneumococcal polysaccharide vaccine is recommended. This vaccine is given to children with sickle cell disease to help prevent infections caused by the pneumococcus.

PNEUMONIA/ACUTE CHEST SYNDROME

All patients with acute chest syndrome must be admitted to the hospital. Analgesics should be administered. Oxygen therapy is indicated for hypoxemia. Intravenous antibiotics and transfusions may also be necessary in treating this problem.

SPLENIC SEQUESTRATION

Treatment is directed toward prompt correction of hypovolemia with plasma expanders and particularly with blood transfusions. Splenectomy is considered if the child has had more than one episode.

APLASTIC CRISIS

Treatment is directed towards correcting the present problems. Transfusions should be given if needed. Hospitalization is not always required if the family continues follow-up visits.

STROKE

Management is aimed at early reversal of neurologic deficits and prevention of recurrence. Immediate treatment consists of intravenous hydration and transfusion to try to reverse or limit symptoms. Long-term management is aimed at prevention of a subsequent stroke.
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.

African Americans may feel singled out by a hereditary disease that affects them with more frequency than any hereditary disorder found in other racial and ethnic groups. Actually, sickle cell disease affects people of East Indian and Mediterranean descent also.

Lacking adequate information about the inheritance of the disease, parents may have feelings of helplessness and guilt that can often lead them to blame each other for the child's illness. Frequent crises may cause the parents to feel that they are being punished for something they did wrong. Because sickle cell disease is inherited, parents might feel a need to keep it a secret. Care and treatment may often be ignored.

♦ **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.

♦ **NUTRITION AND DIET**
  - Eat a well balanced diet 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 (loss of too much water).
  - Drink extra fluids during increased physical activity, when temperatures are warm, and during periods when the child has a fever.
  - Avoid caffeine drinks like regular coffee and colas. Caffeine can cause the blood vessels to constrict.
  - During a crisis, it is more important to drink fluids than it is to eat.
  - Take the vitamin folic acid daily, if advised by his/her physician.

♦ **SPECIAL CARE IN SCHOOL SETTING**
  - 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.
  - Educate the school community about the hemoglobinopathies.
SCHOOL-AGED CHILD

Children with sickle cell disease usually feel different. They are expected to develop along the same lines as children without the disease. However, some complications related to sickle cell disease influence their development. These factors may indicate to the child that he/she is different because of the sickle cell disease. The child may:

- Miss more school.
- Not be allowed to do things other children do because of physical limitations or overprotective parents and teachers.
- Need to go to the bathroom more frequently because they are required to drink more liquids especially during periods of increased exercises and heat.
- Not be able to spend the night at a friend’s house because of bedwetting.
- Be treated differently by parents, siblings, teachers, and others because “they’re sick”.
- May be small for their age. As they become adults, most children with sickle cell disease reach full size.
- Have delayed sexual maturation.

♦ SPECIAL CARE IN SCHOOL SETTING

- Help the child develop a sense of self worth. This teaches independent behavior (caring for self).
- Teach the child coping skills. Giving the child control, this helps the child to teach others.
- Discourage the child from blaming anyone for his/her disease.
- To increase self-esteem, emphasize those things about the child that he/she interprets as positive, not negative.
- Emphasize developing abilities rather than concentrating on the disadvantages of the disabilities.

ADOLESCENCE

Adolescence is a contradictory period of time during which young people are striving for independence, but are encouraged to remain dependent while completing their formal education. Some teens with sickle cell disease are afraid to grow up. Frequent absence from school may have held the teen back from normal grade advancement creating a longer period of dependence. Hospitalization further reduces the adolescent's independence, with their care being directed by adults and not by themselves. Also, during crises, regression to a more dependent behavior encourages those around him/her to treat the young adult like a child. This interaction heightens the child/adult contradiction, the conflict between dependency and independence. Teens with sickle cell disease may have a strong need to prove that they fit in.

♦ SPECIAL CARE IN SCHOOL SETTING

- Attempt to identify the source of the dependency:
  - Ongoing lifelong style of coping; or
  - Environmentally stimulated: has the school and/or family removed so much of the child’s control that he/she has turned to dependency as a means of coping?
  - Risk taking behavior.
- Do not criticize or openly acknowledge the dependent behavior.
- After careful explanation to the adolescent, set limits on the amount and type of dependent behavior that will be tolerated by the staff.
- Praise any independent behavior.
- Listen to what the teen has to say.
- Expect the best.
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 severely delayed. Self-esteem can be severely undermined by the teasing of classmates and/or by not being allowed to participate in group activities. Self-worth is promoted via peer acceptance. Adults may treat the child as if he/she is younger because of his/her smaller size.

Consequently, these children may either become very aggressive to compensate for their small size; or choose to associate with others like themselves, or with those they consider similar to themselves, to distance themselves from the teasing of others. Small size and delayed maturation are not the only physical concerns of the young adult with sickle cell disease. Many suffer from other physical characteristics that are a source of embarrassment such as: jaundice, dental deformities, and surgical scars.

♦ SPECIAL CARE IN SCHOOL SETTING
  o Provide avenues for the adolescent to express feelings. Teach assertiveness.
  o Be a good listener and accept what the adolescent verbalizes.
  o Assist the adolescent to understand his/her own ways of coping.
  o If the adolescent expresses anger or hostile feelings, remember not to take it personally. The adolescent may be handling his/her feelings the only way possible.
  o Give positive reinforcement for the adolescent's efforts to cope.

DEPRESSION

Adolescence is a period of transition from childhood to adulthood. Thoughts of the future may be present. Planning for the future may be stressful. 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 the young adult's ability to manage his/her disease and personal life. He/she may become dependent on drugs, develop somatic complaints, or seek miracle cures. This may impair his/her ability to concentrate and cause him/her to do poorly socially and academically.

♦ SPECIAL CARE IN THE SCHOOL SETTING
  o Focus on the young adult's feelings. Allow him/her to ventilate in ways that seem comfortable to him/her
  o Help explore ways to cope with feelings. Talk about alternate ways to express them
  o Give positive reinforcement to reality and realistic expectations
  o Maintain hope and share it with the young adult
  o Provide positive role models
  o Provide opportunities for vocational planning
Some young adults who have had frequent episodes of crises during childhood may feel that they are now less vulnerable to illness. Others may experience an increase in painful crises as they mature into adolescence and adulthood. The reason for this increase is not known, but it can result in fear on the part of the young adult and his/her parents. As they enter adolescence, they may not report obvious signs and symptoms of illness and instead choose to "wait and see if it goes away". They may become reluctant to assume responsibility for their own health care. The young adult wants to lead a "normal" life.

♦ **SPECIAL CARE IN SCHOOL SETTING**
  - Encourage the young adult to define and discuss his/her own needs. Refrain from forcing treatments.
  - Attempt to introduce realities slowly by beginning with the least threatening part of the reality.
  - Praise the young adult's efforts to recognize reality.
  - Share concerns about noncompliance with the young adult's parents.
  - Give control and responsibility to the young adult.

**EDUCATION**

Education must begin with parents, teachers, and the community. Children should be taught that knowledge is power, and with that power attitudes can be changed. To erase the stigma often and mistakenly associated with the disease, emphasis should be placed on building a positive self-esteem.

**Classification of Chronically Ill**

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. The child may fall behind at school and possibly fail the grade. This can be frustrating for the child. He may contemplate not trying in school because he feels that he will just fall behind again or fail. Being classified as chronically ill can help children to keep up with their class, since tutoring will be provided when appropriate. The school staff, the child study team, the child, and the family, the health care provider and/or treatment center need to cooperate and collaborate in resolving these problems.

**Sex Education**

We must be sensitive to the developing sexuality of young adults. Sex education and methods of birth control should be compatible with the disease. When young women choose to conceive, they need to know that with close medical supervision, they can have a healthy pregnancy with a successful outcome.

♦ **SPECIAL CARE IN SCHOOL SETTING**
  - Send home schoolwork for the child to do when he is feeling better, and suggest a tutor or instruction for the homebound as viable alternatives.
  - Teach the student to learn his/her own tolerance. Limitations can sometimes undermine the student's ability to develop a positive self-image. This especially refers to the tendency for these children to tire easily. Encourage all children to participate to their own level of tolerance in all activities.
  - Schools must develop ways to keep the child involved with peers, while adapting to his needs (i.e. adaptive physical education, individual education plan, etc.).
GENETIC COUNSELING

We need to emphasize the need for genetic counseling. The young adult should be aware that he/she could produce children who do not have sickle cell disease but will always have the trait, at a minimum. Genetic counseling should be provided prior to the person becoming sexually active and reinforced periodically.

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.

♦ SPECIAL CARE IN SCHOOL SETTING
  - Urge teen to stay in school and help him/her keep up his drive to do his/her best.
  - Help teen focus on what he/she can do, not what he/she can't do.
  - Some colleges have programs to help students with special needs.

SUPPORT GROUPS

Support groups are essential. Participation in support groups assist the student and family members to address the following issues:
  - guilt
  - balancing need to over-indulge
  - not being alone
  - learning how others coped
  - learning effective coping skills for greater control
  - being achievement oriented
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 - related to appropriate dress and cleanliness. 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 - related to the child's disease. 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.

Other areas to address:

- Referral to the child study team.
- Family Conference-ongoing to assess family's and student's adjustment to illness and school pressure.
- Learn child's special needs.
- Don't protect child too much, but don't ignore him/her.
- Expect the best.
An important component for the school community is counseling and community outreach. An African proverb says, “It takes an entire village to raise a child”. In keeping with that saying, “It takes an entire community to educate a child”. The school social worker has the expertise and knowledge to create the necessary community linkage.

The social worker should complete a psychosocial assessment on each child. The assessment must recognize that the activities, functions and behaviors of a child includes two major dimensions:

- Internal Dimensions - feelings, attitudes, thoughts, fantasies, memory, judgment, values and self image
- Social Dimensions - relationships with the environment, people and circumstances surrounding the child

**Specific Areas to Assess:**

- Family/significant others - Identify key individuals in the child's/young adult's life and the dynamics within the family structure, how parents and siblings relate to each other, ways in which affection is given and received among family members, etc.
- Traumatic/significant life events - i.e. household move, divorce, separation from friends, death of a relative or pet, death of another patient being treated at the center.
- General personality and behavior - mood, affect, attitude, responsiveness, and morale.
- Physical appearance - dress, cleanliness, and neatness.
- Social activities - hobbies and interests.
- Education - learning ability and attitude toward school.
- Family background - social/economic status.
- Sex - orientation, activity, and awareness.
- Awareness and understanding of illness - fears and questions.
- Health care costs

**Other Areas to Address:**

- Resource development.
- Community outreach - Referrals.
- Family conferences.
- Education groups - child and family.
THE SCHOOL NURSE

The school nurse plays a vital role in coordinating health services for students with sickle cell disease in the school setting. According to the American Nurses Association Standards of School Nursing Practice, school nurses are required to use the nursing process in the school setting.

Standard III, states:

“The nursing process includes Individualized Healthcare Plans (IHP) which are developed by the school nurse.”

The 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). Assess student’s self care skills, barriers to self-care, self-medications and treatment. Review medical records and treatment plans from the healthcare provider.

- Psychosocial Status – student’s strengths, family support system, friends, 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.

Other Areas to Address:

- Determine need for modifications in the school environment and the need for an Emergency Care Plan.

- Assess the need for an IEP.
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.

Dysuria - irritating or painful urination.

Enuresis - the inability to control urination.

Fibrosis - the spread of fiber like connective tissue over normal smooth muscle or other normal organ tissue.

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.

Hemoglobin SC Disease - a variant of sickle cell disease.

Hemoglobinopathy - abnormal hemoglobin.

Hemolytic - breakdown of red blood cells and release of hemoglobin.

Hepatomegaly - enlarged liver

Hyposthenuria - unconcentrated urine.

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 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.

**Sertoruous 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 an 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.

**Venous** - of the veins.

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


HYDROXYUREA REFERENCE


DOPPLER REFERENCE


INTERNET RESOURCE

The Internet is a very useful source of information about sickle cell disease. There are hundreds of web sites devoted to this topic. Many of these sites provide information about sickle cell disease including commonly asked questions, complications of the disease, treatments and potential cures. There is information about trans cranial doplers, hydroxyurea, and bone marrow transplant (see other sections of this manual). Some web sites will provide you with lists of organizations that have more information about sickle cell disease or can put you in contact with other persons who have sickle cell anemia. Please be sure to ask your physician or nurse which website(s) are most appropriate.

If you want to explore on your own, use one of the search engines (e.g. GOOGLE, YAHOO, AOL) and search the term: "Sickle Cell Anemia". You will come up with a number of matches that will start you on your search for topics related to sickle cell disease.
Comprehensive Sickle Cell/Hemoglobinopathies
Treatment Centers in New Jersey

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Children's Hospital of New Jersey
Newark Beth Israel Medical Center
Valerie Fund Children's Center
201 Lyons Avenue
Newark, NJ 07112-2094
Phone: (973) 926-7161
Fax: (973) 282-0395

The Children's Hospital at
St. Joseph's Hospital and Medical Center
703 Main Street
Paterson, NJ 07503-2691
Phone: (973) 754-3230
Fax: (973) 754-3331

University Hospital/UMDNJ
Pediatric Hematology/Oncology
UH Room F342
150 Bergen Street, Suite 5400
Newark, NJ 07103-2499
Phone: (973) 972-5106 or 972-0658
Fax: (973) 972-1340

Bristol-Meyers Squibb Children's Hospital
at Robert Wood Johnson University Hospital
Affiliated with UMDNJ/RWJ Medical School
Division of Pediatric Hematology/Oncology
195 Little Albany Street
New Brunswick, NJ 08903-0029
Phone: (732) 235-5437
Fax: (732) 235-6462

Children's Hospital of Philadelphia
New Jersey Section of Hematology/Oncology
Specialty Care Center
1012 Laurel Oak Road
Voorhees, NJ 08043
Phone: (856) 435-7502
Fax: (856) 627-2183 (office)

Genetic Centers in New Jersey

*SUPPORTED BY THE NEW JERSEY DEPARTMENT OF HEALTH AND SENIOR SERVICES

*Children's Hospital of New Jersey
Newark Beth Israel Medical Center
201 Lyons Avenue
Newark, NJ 07112
Phone: (973) 831-6020

*St. Joseph's Hospital and Medical Center
Section of Genetics
703 Main Street
Paterson, NJ 07503-2691
Phone: (973) 754-2727
Outreach Clinic: Fairfield

*Hackensack University Medical Center
Genetics Service
Don Imus Pediatric Center-Rm. 258
30 Prospect Avenue
Hackensack, NJ 07601-1991
Phone (201) 996-5264
Outreach Clinics: Hoboken, Parsippany

*UMDNJ/NJ Medical School
Center for Human & Molecular Genetics
90 Bergen Street, Suite 5400
Newark, NJ 07103-2499
Phone: (973) 972-3300
Outreach Clinics: Pompton Plains, West New York

*Saint Peter's University Hospital
Institute for Genetic Medicine
254 Easton Ave.
New Brunswick, NJ 08903
Phone: (732) 745-6678

*Cooper Hospital/University Medical Center
Division of Genetics
3 Cooper Plaza Suite 309
Camden, NJ 08103-1400
Phone: (856) 968-7255
Outreach Clinic: Children’s Regional Center at Voorhees