Sickle Cell Disease: Health Promotion and Maintenance and the Role of Primary Care Nurse Practitioners

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INTRODUCTION

Sickle cell disease (SCD) is an inherited autosomal recessive disease belonging to a group of conditions called hemoglobinopathies (Goldman & Bennett, 2000). This disorder was first described in 1910 by Dr. James B. Herrick as a “peculiar physical or chemical condition of the blood” (p. 517). Since 1910, much has been researched and published about the physiology, complications, treatments, and prevention of this disease (Fleming, 1989; Lombard, Xie, & Niihara, 2001; Olney, 1999; Simon, Lobo, & Jackson, 1999). However, there is a dearth of publications examining SCD health promotion, disease maintenance, and prevention from a primary care perspective. To this end, this article will focus on the role of primary care nurse practitioners (NPs) with regard to early identification of affected individuals, prevention of painful vaso-occlusive crises (VOCs), effective monitoring/screening, effective pain management and prophylaxis, and health education about this disease.

Complications from this disease (see Table 1) and frequent hospitalizations come with a societal cost. An estimated 75,000 Americans were hospitalized with SCD complications yearly during the years 1989 to 1993, with an average hospital stay of 6.1 days. In 1996, the average cost per hospitalization for sickle cell complications was $6,300, with a total annual cost of $475 million (Davis, Moore, & Gergen, 1997).

This disease primarily affects Black people of African descent as well as some people of Indian, Mediterranean, Italian, South and Central American, and Middle Eastern ancestry. It is estimated that 50,000 to 60,000 Americans have SCD. Some 8% to 10% of African American newborns have the trait; 1 of every 400 African American babies born has SCD. Due to SCD complications, life expectancy for affected people is approximately 42 years for men and 48 years for women (Dipiro et al., 1999; Goldman & Bennett, 2000). Because the patient population in America is diverse, NPs need to become knowledgeable about the different ethnic groups in which this disease is prominent, in order to target appropriate individuals for health promotion.

PSYCHOSOCIAL IMPLICATIONS

Several psychological and psychosocial problems associated with SCD have a major impact on the quality of the lives of affected individuals and their families. Self-esteem issues and concerns about body image due to delayed growth and growth abnormalities (small body size) are major psychosocial problems (Gentry, Varlik, & Dancer, 1997). Frequent leg ulcers from poor wound healing due to inadequate circulation also pose a major threat to body image (Dipiro et al., 1999; Thomas, Wilson-Barnett, & Goodhart, 1998). Research further suggests that problems with social relationships, peer acceptance, and isolation are other sources of psychosocial problems because affected individuals are restricted in their ability to interact with their healthy peers (Gentry et al.). The formation of significant rela-
In SCD (sometimes referred to as sickle cell anemia) both parents must have a sickle gene in order for the offspring to inherit the disorder. The condition is due to a single defective hemoglobin molecule; thus, the abnormal hemoglobin S (Hb-S) is produced instead of the normal Hb-A. The defective hemoglobin is due to the substitution of a single valine for glutamic acid at the sixth position in the β chain of the hemoglobin. In the presence of deoxygenation, the substitution changes the solubility of the hemoglobin molecule. Approximately 70% to 95% of the hemoglobin in the blood of individuals with SCD is abnormal; yet individuals with sickle cell trait (SCT), a benign condition, have less than 50% of the abnormal hemoglobin. Individuals with SCT are asymptomatic and experience no hematological complications (Dipiro et al., 1999; Rakel, 2002).

Genotypes of SCD include sickle cell–β-thalassemia, which comprises sickle cell–β⁺-thalassemia and sickle cell–β°-thalassemia. Patients with sickle cell–β⁺-thalassemia have approximately 3% to 25% Hb-A, but sickle cell–β°-thalassemia is marked by an absence of Hb-A. Sickle cell–β⁺-thalassemia has a benign clinical course compared to sickle cell–β°-thalassemia. Another genotype of SCD is Sickle C-disease (Hb SC disease), which is also marked by a generally milder clinical presentation. The main characteristics of SCD are chronic hemolytic anemia, organ infarction, and recurrent painful VOCs (Goldman & Bennett 2000; Rakel, 2002).

In SCD, the oxygen-carrying capacity of the red blood cells (RBCs) is altered, and their survival time is shortened from 120 days to 10 to 20 days. In the presence of hypoxia, the abnormal RBCs become rigid, sickled, or crescent in shape and clump in the vascular space, leading to (a) destruction of small blood vessels; (b) stasis in the vascular system, leading to impaired circulation; (c) increased blood viscosity and decreased perfusion to organs; and (d) occlusion of microcirculation leading to tissue hypoxia, infarction, and necrosis. The sequelae involve painful occlusive crises, organ infarcts, and complications from organ damage (Dipiro et al., 1999).

Patients often experience different types of SCD crises. These include hemolytic, sequestration, aplastic, and vaso-occlusive crises. In hemolytic crises, patients experience a sudden drop in hemoglobin and RBC levels due to excess hemolysis of RBCs; pain and fever may be present. In sequestration crises, the spleen and liver become extremely enlarged due to trapping of RBCs by the spleen. Sequestration causes a severe drop in hemoglobin and hematocrit levels, and hypotension, shock, and death may quickly ensue. In aplastic crises, patients experience severe anemia and decreased reticulocytes due to bone marrow failure. In VOCs, which are the most common crises and primary reason for frequent hospitalizations, patients experience severe pain in involved areas (Dipiro et al., 1999). The occlusion of blood vessels in the presence of hypoxia, infection, or dehydration causes VOCs (see Table 2). Any part of the body can be affected during a VOC, but the regions commonly affected include the musculoskeletal, cardiopulmonary, and abdominal areas. Laboratory changes that may occur during a VOC include leukocytosis, decreased blood levels of bicarbonate and pH, and increased serum fibrinogen. Repeated VOCs can lead to organ infarcts and damage, multisystem failure, and many clinical complications (Dipiro et al.; Goldman & Bennett, 2000).

**Early Identification of Symptoms**

Early detection of complications prevents frequent hospitalizations and prolongs lives because the complications and common infections seen in SCD, such as sepsis, bacterial meningitis, pneumonia, and osteomyelitis, may occur quickly and may be
### Table 1. Manifestations of Sickle Cell Disease Complications by Organ System

<table>
<thead>
<tr>
<th>Organ Involvement</th>
<th>Clinical Complications</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pulmonary</td>
<td>Called acute chest syndrome. Presenting symptoms: fever, dyspnea, chest pain, cough, tachypnea, wheezing, chills, and hemoptysis. Possible laboratory findings: leukocytosis, hypoxemia, and pulmonary infiltrate; caused by pulmonary embolism and infections.</td>
</tr>
<tr>
<td>Neurological</td>
<td>Cerebrovascular accidents due to brain infarction. Presenting symptoms: sudden weakness in extremities, aphasia, visual changes, and persistent headaches. Magnetic resonance imaging and transcranial Doppler ultrasonography can detect subclinical cerebral infarction.</td>
</tr>
<tr>
<td>Skin</td>
<td>Inadequate blood circulation. Presenting symptoms: slow healing chronic ulcers, especially in the lower extremities.</td>
</tr>
<tr>
<td>Cardiovascular</td>
<td>Chronic anemia associated with heart murmurs, dysrhythmias, heart enlargement, and possible heart failure. Presenting symptoms: tachycardia, exertional dyspnea, and palpitations.</td>
</tr>
<tr>
<td>Genitourinary</td>
<td>Sustained erections lasting hours or days. Presenting symptoms: severe pain in the penis (priapism) due to sickling of RBCs; can lead to impotence. Pregnant women are at risk for spontaneous abortions, preeclampsia, severe anemias and VOCs, infections, and death. Risks to the fetus include low birth weight, intrauterine growth retardation, and death.</td>
</tr>
<tr>
<td>Eyes</td>
<td>Proliferative retinopathy, venous microaneurysms, and other retina problems. Presenting symptoms: transient blindness and visual field defects.</td>
</tr>
<tr>
<td>Abdomen</td>
<td>Severe infarction of abdominal structures, splenomegaly, and splenic sequestration. Presenting symptoms: low-grade fever and abdominal pain, sudden drop in Hb and Hct, hypotension and shock with splenic sequestration.</td>
</tr>
<tr>
<td>Hepatobiliary</td>
<td>Chronic hemolysis resulting in increased serum bilirubin. Intrahepatic sickling may lead to hepatocellular swelling, necrosis, and pain. Presenting symptom: pain in right upper quadrant of abdomen.</td>
</tr>
<tr>
<td>Renal</td>
<td>Proximal and distal tubular, medullary, and glomerular dysfunction due to vaso-occlusion. Presenting symptoms: abnormalities in urinary concentration, hematuria, impaired clearance of potassium (hyperkalemia), hypertension, and proteinuria.</td>
</tr>
</tbody>
</table>

*Note.* RBCs = red blood cells; VOCs = vaso-occlusive crises. Hb = hemoglobin; Hct = hematocrit. Data from Dipiro et al., 1999; Goldman and Bennett, 2000; Quinn and Buchanan, 1999; Rakel, 2002; Wang et al., 2000.
severe (Wethers, 2000). Hence, detection of complications requires early identification of affected individuals, effective monitoring, effective treatment, and education for prevention.

**CLINICAL PRESENTATION**

SCD varies in its timing and clinical presentation. Understanding the timing and typical presenting symptoms is an essential component to timely and accurate diagnosis. Symptoms of SCD are usually delayed until about 3 to 4 months of age because the infant’s RBCs contain mainly fetal hemoglobin. Moderate to severe anemia is usually present by 6 to 9 months of age, and the first VOC often occurs by 6 to 12 months of age. In the first year of life, dactylitis (hand and foot swelling) and acute splenic sequestration are the most common clinical manifestations. Splenomegaly is commonly present after 6 months of age. After age 2, painful crises are the most common clinical presentation. Other physical presentations include height and weight abnormalities, protuberant abdomen, long and thin extremities, lumbar lordosis, tapered fingers, and barrel-shaped chest. Adolescents and adults with SCD almost always have a long-standing diagnosis; thus, clinical manifestations will be more related to disease complications and painful crises (Dipiro et al., 1999; Ignatavicius, Workman, & Mishler, 1999; Rakel, 2002).

**DIAGNOSTIC TESTING AND SCREENING**

Universal screening for all babies, regardless of ethnicity, has been proposed in order to initiate early medical intervention that can reduce morbidity and mortality. Currently over 40 U.S. states have adopted universal screening (Centers for Disease Control and Prevention, 1998). If universal screening is available and has been done in the hospital, the NP can easily confirm the diagnosis during well-baby checkups by reviewing medical records and can initiate an individualized plan of care for affected infants as young as 3 months old and for their families. If sickle cell screening is not performed at birth, evidence of the disease may be obtained from the history and physical examination.

If SCD is suspected in an infant not screened at birth, a peripheral blood smear is the initial diagnostic test. A positive blood smear would reveal sickled RBCs. Hemoglobin electrophoresis would confirm the diagnosis and would quantify the amount of the abnormal hemoglobin. Reticulocyte count would be elevated due to chronic hemolysis. The level of hemoglobin in the sickle cell patient can range between 5.5 and 9.5 g/dl, with an average of 7.5 g/dl. If the diagnosis of SCD is confirmed, the patient must be referred to a hematologist for further evaluation. Referral to other specialists (see Table 3) may be appropriate as well, depending on physical findings (Rakel, 2002).

In spite of referrals to specialists, NPs continue to play a crucial role in preventing VOCs, monitoring disease complications, maintaining these individuals’ health, and facilitating overall health promotion. For example, chest radiographs and measurements of arterial blood gases and pulmonary function are recommended biannually after age 5 to assess pulmonary compli-

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**Table 2. Sickle Cell Disease: Patient and Family Education**

<table>
<thead>
<tr>
<th>When to Seek Immediate Medical Care</th>
</tr>
</thead>
<tbody>
<tr>
<td>- Fevers or persistent low-grade fevers</td>
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<tr>
<td>- Pain unrelieved by prescribed oral analgesics</td>
</tr>
<tr>
<td>- Persistent abdominal pain and recurrent emesis</td>
</tr>
<tr>
<td>- Dyspnea, pain with breathing, hemoptysis, or feelings of tightness in the lungs</td>
</tr>
<tr>
<td>- Angina, exercise intolerance, or orthopnea</td>
</tr>
<tr>
<td>- Visual/speech changes, weakness/numbness in extremities, persistent headaches</td>
</tr>
<tr>
<td>- Sustained penile erections unrelieved by prescribed medication</td>
</tr>
<tr>
<td>- Increased lethargy, irritability, or pallor</td>
</tr>
</tbody>
</table>

**Factors That May Precipitate Painful Vaso-Occlusive Crises**

- Inadequate rest, emotional stress, and fatigue
- Vasoconstrictive drugs, smoking, and constrictive clothing
- Dehydration and strenuous physical exercise
- Extreme hot or cold temperatures, high altitudes, or unpressurized aircraft

Note. Data from Goldman and Bennett, 2000; Rakel, 2002; Rausch and Pollard, 1998.

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**Table 3. Sickle Cell Disease: Recommendations for Consultation**

- Fevers of 40°C or higher
- Toxic-appearing patients not on prophylactic penicillin with fever of 38.5°C or higher, positive cultures, and chest film
- Moderate to severe persistent pain in any body part
- Worsening respiratory symptoms and chest pain
- Worsening headaches
- Drop in hemoglobin level below 3g/dl from baseline
- Evidence of meningitis
- Jaundice and scleral icterus
- Inflammation in the extremities
- Initial and worsening hematuria and proteinuria
- Persistent leg ulcers
- Priapism
- Initial episodes of abnormal findings following ROS; objective findings including diagnostic tests

Note. ROS= review of systems. Data from Goldman & Bennett, 2000; Rakel, 2002; Wethers, 2000.
cations. To assess complications in the hips, magnetic resonance imaging is recommended every 3 to 5 years after age 10. Dental examinations are recommended yearly. Nutritional assessment and ophthalmologic examinations are done yearly after age 10. To assess hemologic, liver, and gallbladder function, a complete blood count and reticulocyte count are recommended quarterly, including an annual liver function test and hepatitis screening. Ultrasound of the abdomen is also recommended twice yearly after age 10. To assess kidney function, annual urinalysis as well as blood urea nitrogen, creatinine, and uric acid analysis are recommended. Stress echocardiogram and electrocardiogram are recommended twice yearly to assess cardiac functioning. Transcranial Doppler evaluations are initiated regularly to predict patients at risk for strokes. Referral to a social worker can be initiated to assess family coping and adjustment. Families can also be referred to the National Sickle Cell Foundation for support (Goldman & Bennett, 2000; Rausch & Pollard, 1998).

Female patients planning to become pregnant can be advised about genetic testing and counseling; DNA samples could be obtained via chorionic villus sampling at 8 to 10 weeks gestation to detect presence of the disease. In the second trimester, amniocentesis can also be done to detect SCD (Goldman & Bennett, 2000). Additionally, NPs have an essential role in monitoring growth, development, and child and family functioning.

EFFECTIVE MONITORING

Assessment: History Component (Subjective)

During the initial well-baby checkup, the NP should ascertain a family history and the presence of the disease. At the 3 or 4 month well-baby visit and thereafter, a review of systems (ROS) is crucial for detection of disease manifestations and for early initiation of the appropriate treatment regimen. Painful episodes, the usual initial presentation of the disease, commonly occur during the first 6 to 12 months of life; thus, evaluation of pain is crucial (Rakel, 2002). Information provided by the caregiver is critical in evaluating the infant's pain. When taking the history, the NP should obtain information from the infant's caregiver about the presence of pain (e.g., fussiness or persistent crying unrelied by medications and/or usual methods of consoling the infant).

Histories of fevers should be ascertained because infections, such as pneumonia, are sometimes the presenting evidence of SCD in infants. The NP should ask questions about abdominal pain (e.g., persistent crying and flexing of the legs), which may suggest the presence of infarction in abdominal structures. The caregiver should also be questioned regarding swelling and redness in the extremities, which may indicate dactylitis. Further, the NP should ask the caregiver about evidence of increased weakness, pallor, and fatigue. These conditions may suggest marked anemia due to splenic sequestration, which is one of the presenting symptoms in the first year of life. Questions about the infants’ nutrition and sleep patterns are also crucial in assessing growth and developmental needs (Dipiro et al., 1999; Rakel, 2002).

In young children, adolescents, and adults, an ROS is obtained in order to detect evidence of disease complications. To conduct an ROS, the NP asks the patient or caregiver about the presence of persistent headaches, convulsions, drowsiness, paralysis, aphasia, or visual disturbances, which may indicate neurological problems. The NP assesses pulmonary complications by asking questions about dyspnea, tachypnea, chest pain, hemoptysis, pain with breathing, and fevers. The patient is also asked questions about tachycardia, palpitations, exercise intolerance, fatigue, and orthopnea in order to assess cardiac involvement. Questions regarding abdominal pain and pain in the right upper quadrant and midepigastric area are aimed at detecting cholelithiasis/cholecystitis or infarction of abdominal structures, which are commonly seen in young children and adults. Repeated nausea and vomiting after eating may also suggest cholelithiasis or cholecystitis. To assess dietary intake, the NP obtains a 24-hr recall (Dipiro et al., 1999).

With regard to the genitourinary system, the presence, frequency, and duration of painful erections (priapism) are asked. Priapisms are commonly seen in ages 5 to 13 and 21 to 29. Five-year-old children have some awareness of their body parts because of increasing curiosity about their bodies; hence, it is developmentally appropriate to directly ask a 5-year-old boy with SCD if he experiences undesired painful erections in his penis or “private area” (Dixon & Stein, 2000). Female patients are asked about the presence of menstrual irregularities. The NP also asks patients about hematuria, which usually occurs as a result of hemolysis. Questions regarding leg ulcers, erythema, pain, and swelling in the joints are asked. Information about dental and visual problems is also obtained (Dipiro et al., 1999; Goldman & Bennett, 2000).

Assessment: Physical Component (Objective)

The primary goal during physical examination of infants is to detect evidence of disease manifestation. With children, adolescents, and adults, the goal is to assess the presence of disease complications. Follow-up assessments for infants less than 6 months old are recommended monthly and every 2 months for infants between 6 months and 1 year. For young children between ages 1 and 5, follow-up assessments are recommended every 3 months (Rausch & Pollard, 1998). With each visit, the height and weight are recorded to keep track of growth delays or significant changes. The NP examines the patient’s eyes for the presence of icterus and visual field defects. The retina is examined for evidence of vitreous hemorrhage, detachment, or occlusion, which are problems commonly seen in adults. The skin is examined for the presence jaundice and leg ulcers. Leg ulcers are commonly seen in individuals over age 10. Jaundice, may indicate hepatobiliary complications or hepatic crises, which are often seen in older patients (Dipiro et al., 1999; Goldman & Bennett, 2000).

The cardiopulmonary system is auscultated to detect early manifestations of complications. The abdomen is examined for the presence of splenomegaly, commonly present in the first 2 years of life. Evidence of cholelithiasis or cholecystitis (e.g., right upper quadrant and/or epigastric tenderness) is obtained during the abdominal examination. Musculoskeletal abnormalities (e.g., effusion, arthritic pain, or erythema) are also ascertained. Musculoskeletal problems, such as osteomyelitis and aseptic or avascular necrosis, are commonly experienced by these patients (Dipiro et al., 1999; Goldman & Bennett, 2000).
EFFECTIVE PROPHYLAXIS, PAIN MANAGEMENT, AND TREATMENT OPTIONS

Prophylaxis

Bacterial infections such as sepsis, meningitis, pneumonia, and osteomyelitis are common in adolescents and young children because of damage to the spleen. Although these infections are a major threat throughout the life span of these patients, they are the major causes of morbidity and mortality for children younger than 3 (Centers for Disease Control and Prevention, 2000). NPs may quickly and aggressively manage fevers of approximately 38.5°C in patients who do not appear toxic and who are receiving prophylactic penicillin. Treatments include broad-spectrum antibiotics, such as cephalosporins, after obtaining blood and urine cultures, chest radiography, and a complete blood count (Goldman & Bennett, 2000; Rakel, 2002; Wethers, 2000).

Hospitalization is recommended for individuals with a history of Streptococcus pneumoniae infection and fevers of 38.5°C or higher. Patients who live in an area with known antibiotic resistant to S. pneumoniae are also hospitalized with fevers of 38.5°C or higher. NPs can manage mild respiratory problems in the presence of normal chest film and oxygen saturation by using bronchodilators and incentive spirometry (Goldman & Bennett, 2000; Wethers, 2000). Table 3 outlines further indications for referral. In addition to the aforementioned treatments, immunizations and prophylactic antibiotics are recommended for preventing infections (see Table 4).

Effective Pain Management

Chronic pain interspersed with episodes of acute pain is one of the more difficult management challenges in patients with SCD. Because pain is a major characteristic of SCD, pain evaluation is essential during each patient encounter. It is important to remember that objective evidence of pain, such as increased blood pressure and tachypnea, may not be present in some patients with chronic sickle cell pain. In managing pain caused by VOCs, the NP must (a) establish an honest and mutually trusting relationship with the patient, (b) remain nonjudgmental about the patient’s pain, and (c) thoroughly assess the pain. Patients’ subjective account of pain along with pain scales, such as the visual analog and pain relief scales, may be used to quantify the severity of pain (Yale, Nagib, & Guthrie, 2000).

Painful episodes are the primary reason sickle cell patients seek medical care. Hence, using adequate hydration and analgesics to promptly and aggressively manage painful episodes is critical in decreasing suffering and preventing hospitalization. Milder painful episodes can be managed with adequate oral hydration, rest, warmth, and nonsteroidal anti-inflammatory drugs such as naproxen and indomethacin. A combination of codeine with acetaminophen or aspirin is another option. For optimal pain control, NPs must prescribe analgesics on a scheduled basis, such as every 4 to 6 hr, and avoid as-needed schedules. Patients with moderate to severe pain are generally hospitalized for treatment with parenteral narcotics, such as morphine or hydromorphone, and intravenous hydration (Dipiro et al., 1999; Reid, Charache, & Lubin, 1995).

Patients should also be educated about the effectiveness of biofeedback and cognitive behavioral therapies in pain management. Several studies suggest that these therapies can be effective, in combination with prescribed analgesics, in managing pain in SCD (Cozzi, Tryon, & Sedlacek, 1987; Thomas, 2000; Thomas et al., 1998; Thomas, Cpsychol, & Mphil, 2001).

Treatment Options

In order to increase patients’ understanding of their disease, NPs must educate them about the different treatment options and their side effects. Patients should be informed that bone marrow transplantation (BMT) is currently the only cure. However, only 1% to 2% of patients in America have successfully received BMT. Some of the major reasons include lack of a suitable sibling donor, financial issues, and graft versus host disease complications; thus, most patients are poor candidates for

![Table 4. Sickle Cell Disease: Prophylaxis Management and Recommended Immunizations](image-url)

<table>
<thead>
<tr>
<th>Drug</th>
<th>Dose</th>
<th>Frequency</th>
<th>Age</th>
</tr>
</thead>
<tbody>
<tr>
<td>Penicillin VK²</td>
<td>125 mg</td>
<td>Twice daily</td>
<td>2 months–3 years</td>
</tr>
<tr>
<td>Penicillin VK²</td>
<td>250 mg</td>
<td>Twice daily</td>
<td>3 years–5 or 6 years</td>
</tr>
<tr>
<td>Pneumococcal vaccine</td>
<td>Standard</td>
<td>Standard schedule</td>
<td>24 months and again after 5 years</td>
</tr>
<tr>
<td>OPV, DPT, MMR</td>
<td>Standard schedule</td>
<td>Standard schedule</td>
<td>Standard schedule</td>
</tr>
<tr>
<td>Varicella</td>
<td>Standard</td>
<td>Standard schedule</td>
<td>24 months and again after 5 years</td>
</tr>
<tr>
<td>Influenza vaccine</td>
<td>Yearly</td>
<td>Standard schedule</td>
<td>Standard schedule</td>
</tr>
<tr>
<td>H. Influenzae vaccine</td>
<td>Standard</td>
<td>Standard schedule</td>
<td>Standard schedule</td>
</tr>
<tr>
<td>Hepatitis B vaccine series</td>
<td>Standard</td>
<td>Standard schedule</td>
<td>Standard schedule</td>
</tr>
<tr>
<td>Hepatitis A (if indicated)</td>
<td>Standard</td>
<td>Standard schedule</td>
<td>Standard schedule</td>
</tr>
</tbody>
</table>

Note. OPV = oral poliovirus vaccine; DPT = diphtheria, pertussis, and tetanus; MMR = measles, mumps, rubella. Data from Dipiro et al., 1999; Rakel, 2002; Rausch & Pollard, 1998. *There is no determined time pertaining to how long prophylactic penicillin should be administered, but stopping at age 5 or 6 is recommended as safe.
Patients should also be educated about the use of blood transfusion therapies. Blood transfusions are frequently used to treat anemias, refractory VOCs and leg ulcers, persistent organ infarctions, cardiopulmonary complications, severe priapism, complicated pregnancies, and individuals at high risk for strokes. Major side effects of chronic transfusion include iron overload, alloimmunization, and infections. Individuals with serum ferritin blood levels over 2,000 ng/ml (iron overload) can be treated with deferoxamine chelation therapy, which assists in the excretion of excess iron. Chelation therapy, however, has been associated with increased cost, discomfort, and noncompliance in this patient population (Dipiro et al., 1999; Goldman & Bennett, 2000; Reed & Vichinsky, 1999).

Hydroxyurea drug therapy is another therapy for patients with SCD. Hydroxyurea, a known chemotherapeutic agent with an unknown mechanism of action, has been shown to decrease leukocyte, reticulocyte, and platelet counts and to increase hemoglobin, hematocrit, and mean corpuscular volume levels (Goldman & Bennett, 2000; Loukopoulos et al., 2000). Currently, there is no ideal hydroxyurea dosing regimen; however, research suggests a starting dose of 15 mg/kg/day, with gradual increments of 5 mg/kg/day while monitoring toxicity (Hoppe et al., 2000). In adults, a beginning dose of 500 mg/day is recommended, with gradual increments depending on toxicity (Steinberg, 1996).

The antisickling effect of hydroxyurea can decrease the frequency of VOCs, acute chest syndromes, and blood transfusions. The long-term risks for this therapy are still unknown, but myelotoxicity is a major side effect. Hydroxyurea is recommended to patients who experience impaired and refractory painful episodes and who would comply with frequent myelosuppression monitoring (Goldman & Bennett, 2000; Hoppe et al., 2000; Loukopoulos et al., 2000).

Sickle cell patients have a greater need for folic acid due to increased erythropoiesis. Thus, a 1 mg daily dose of folic acid is recommended in order to avoid decreased serum folate levels and megaloblastic anemia. A daily multivitamin supplement is also recommended (Dipiro et al., 1999; Rakel, 2002).

**Education for Preventing VOCs**

When the diagnosis of SCD is made, the entire family should be included in the plan of care and education; the health of individual family members may have a great impact on other family members and on family dynamics. Including all family members in the health education and plan may promote family normalcy, in spite of the effects of the disease on the family unit. Cohesive and strong family support has been documented as a major factor in effectively managing pain and in improving patients’ quality of life (Fleming, 1989; Yale et al., 2000).

Information for the patient and family about factors that may cause vaso-occlusion and precipitate VOCs and about when to seek emergency care is presented in Table 2. Individuals should also be educated about maintaining a well-balanced diet—including adequate calories, foods high in protein, folate, zinc, and iron—in order to promote growth and facilitate wound healing. A daily intake of 2 to 3 L of fluid is also recommended (Kasdan, 2000). Much more, the NP should continually educate patients and families about the importance of regular medical checkups and screening.

**Barriers in Decreasing the Severity of VOCs**

VOCs are the expected trajectory of this disease and, thus, cannot be fully prevented. However, the severity of the pain experienced during the crises can be lessened. Barriers to decreasing VOCs can be grouped into two categories: (a) patients’ coping styles and (b) health care providers’ perspectives and stereotyping regarding pain in SCD.

Patient’s coping styles, which are varied, subjective, and not entirely dependent on vital signs, can be confusing to health care providers. Research shows that patients who display passive and dependent styles of coping and negative thought patterns experience more painful episodes. These patients also exhibit increased psychological distress and frequent emergency room visits and hospitalizations. These coping styles are thought to contribute to and intensify pain and can be major barriers to effective pain management (Gil, Abrams, Phillips, & Keefe, 1989).

A lack of a comprehensive understanding of and knowledge about the nature of pain in SCD has been cited as a major barrier to effective pain management experienced by health care providers. The lack of a mutually trusting relationship between patients and health care providers has also been documented as another barrier in managing pain in SCD. Some health care providers view sickle cell patients as drug seeking, manipulative, and difficult because of their persistent demand for narcotics. Prescribing fewer narcotics than needed and inadequate pain assessment are other barriers faced by health care providers. These barriers result in suboptimal pain management, debilitating painful crises, and increased hospitalizations (Larsen, Neverett, & Larsen, 2001; Lombard et al., 2001; Marchiondo & Thompson, 1996).

In order to manage these barriers, the first essential element for the NP is increased knowledge about the nature of the pain in SCD, followed by prompt treatment of pain in a nonjudgmental manner. Encouraging patients to become involved in their own care may also decrease distress and dependency on health care providers and services, thus increasing patients’ sense of control over pain. Overcoming these barriers may decrease suffering, thereby preventing frequent hospitalizations and their psychosocial impact on quality of life.

**CONCLUSION**

SCD is a chronic disease affecting every organ of the body, thereby causing increased morbidity and mortality. To date there is no universal cure for this disease, although BMT has been effective in a small percentage of cases. The goal of treatment is, therefore, managing symptoms and decreasing the frequency of VOCs and disease complications. NPs are in key positions to decrease the high rate of morbidity and mortality associated with this disease.
by providing consistent and comprehensive primary care.

NPs must understand the importance of early identification of affected individuals, effective monitoring and screening, effective pain treatment, and prophylaxis. The unpredictable trajectory of SCD can lead to frustration, fear, helplessness, hopelessness, and emotional distress. In spite of the uncertainty of the course of this disease, NPs are in excellent positions to continually educate families about the necessity of frequent medical supervision and screening. Ineffective pain management is a major problem for patients with SCD. NPs can overcome this problem by initiating effective and prompt pain management in a nonjudgmental manner.

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Limited quantities of items remaining in stock include …

Denim Shirts, Adult T-Shirts, Kid's T-Shirts, Baseball Caps, Bumper Stickers, Computer Mouse Pads, License Plate Frames, Luggage Tags, Lunch Coolers, Tote Bags and Visors

To request a current 2003 Foundation Store Order Form, please contact the Foundation office located in Arizona. A 2003 Foundation Store Order Form will be faxed or mailed to you.

AANP Foundation Store
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