The *Critical Elements of Care* (CEC) considers care issues across the life span of the child. The intent of the document is to educate and support those caring for a child with sickle cell disease. The CEC is intended as a general aid to health care providers to assist in the recognition of symptoms, diagnosis and care management related to a specific diagnosis. The document provides a framework for a consistent approach to management of these children.

These guidelines were developed through a consensus process. The design team was multidisciplinary with statewide representation involving primary and tertiary care providers, family members and a representative from a Health Plan.

Content reviewed and updated 1/2012:
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This document is also available on the Center for Children with Special Needs website at www.cshcn.org.

**DISCLAIMER:** Individual variations in the condition of the patient, status of patient and family, and the response to treatment, as well as other circumstances, mean that the optimal treatment outcome for some patients may be obtained from practices other than those recommended in this document. This consensus-based document is not intended to replace sound clinical judgment or individualized consultation with the responsible provider regarding patient care needs.

S.B., age 6, describing her sickle cell pain
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## Sickle Cell Disease

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I. OVERVIEW OF SICKLE CELL DISEASE

Definition of Sickle Cell Disease
Sickle cell disease comprises a group of genetic disorders characterized by the inheritance of sickle hemoglobin (Hb S) from both parents, or Hb S from one parent and a gene for an abnormal hemoglobin or β-thalassemia from the other parent. The presence of Hb S can cause red blood cells to change from their usual biconcave disc shape to a crescent or sickle shape during de-oxygenation. Upon re-oxygenation, the red cell initially resumes a normal configuration, but after repeated cycles of “sickling and un-sickling,” the erythrocyte becomes damaged permanently and may remain sickled or may hemolyze. This hemolysis is responsible for the anemia that is the hallmark of sickle cell disease.

Acute and chronic tissue injury can occur when blood flow through the vessels is obstructed due to the abnormalities in the sickled red cells. Complications may include painful episodes involving soft tissues and bones, acute chest syndrome, priapism, cerebral vascular accidents, and both splenic and renal dysfunction. Historically, common causes of mortality among children with sickle cell disease included bacterial infections, splenic sequestration crisis and acute chest syndrome.

Sickle cell disease affects 70,000 to 100,000 Americans, primarily those of African heritage, but also those of Mediterranean, Caribbean, South and Central American, Arabian or East Indian ancestry. It is estimated that eight percent of the African American population carries the sickle cell trait, and approximately one African American child in every 375 is affected by sickle cell disease. Thus, it is the most common inherited blood disorder, and among the most prevalent of genetic diseases in the United States.

Psychosocial Aspects of Sickle Cell Disease
Sickle cell disease is life-altering for most families. Learning to accept, cope and respond to this chronic illness requires that the practitioner and family work together. Cooperation occurs best in an environment where the family feels comfortable, safe and un-judged. The practitioner sets a tone for the relationship. That tone should encourage the family to view the practitioner as a resource, confidante and advocate.

When working with children and families affected by sickle cell disease, it is important to develop a comprehensive approach that encompasses psychosocial issues. Working to understand the issues faced by many of these families will help improve relationships and ensure a positive outcome.

The Status of African Americans
In the U.S., sickle cell disease is primarily a disorder of African Americans. Disproportionate numbers of African Americans face economic challenges of housing, employment and daily living, and often encounter barriers to health care access. The challenge of overcoming discrimination and racism are daily realities for many families. In addition, patients and families often do not feel accepted or welcomed in many health care settings, which can significantly interfere with a child with a chronic disease receiving optimal medical care.

Although women are the head of many households, family structures vary. Raising children as a single parent is challenging – particularly in the areas of economic support, childcare and respite time for the parent. As we have become a more mobile society, single parents often face a lack of family support and experience general feelings of isolation. Extended families may include both biological family members and those who are not biologically related but who fill family roles. It is not unusual to have large numbers of “family” who care for a child and take various levels of responsibility for that child. In some cases, extended families can be overwhelming for
Parents may need support in articulating their needs in this setting and in particular, their need for privacy.

Generally, African Americans have strong spiritual beliefs that may be historical and cultural. Some families may be active participants in a church congregation and find great support or assistance from their church family. Others, while having beliefs, may not participate in any organized religious group. Still other African Americans are Muslim or Buddhist. It is important to respect these beliefs. Insensitivity or infringement upon a family’s belief system can create a rift between practitioner and family.

**Effects of Physical Appearance**

Children with sickle cell disease may display physical manifestations of their illness. As a result of short stature, low muscle mass or jaundiced eyes and nailbeds, ridicule by peers and others is possible. This is particularly common in children 8 to 12 years of age. Children and their parents should be prepared to use coping strategies to help them in these situations. Gaining knowledge and understanding of their illness is one such strategy. Education of schools and peers can also be helpful.

**School Attendance and Adjustment**

Some children with sickle cell disease are frequently absent from school. These absences may be the result of a painful episode, hospitalization, outpatient visits and procedures or other illnesses. Frequent absences from school may result in incomplete class work and incomplete development of social skills. Students can feel disenfranchised from classroom activities and classmates.

There are a variety of responses these students may have, but the extremes of withdrawal or disruptive behavior are particularly troublesome for school personnel or families. Withdrawal may manifest in a lack of participation in classroom activities or with classmates, daydreaming, a lack of enthusiasm in the process of learning, or opposition to attending school as evidenced by verbalization or behavior. Disruptive behavior may be displayed through choices in dress or problems in interacting with other children.

These behaviors may indicate that a child is feeling overwhelmed by schoolwork, and they may not know how to ask for assistance. They may not be able to catch up on missed assignments and may not feel a sense of belonging in the classroom. This can lead to intense feelings regarding relationships at school. In most cases the child will not be able to clearly state their feelings, so they may need assistance in defining the problems. This may include testing by a neuropsychologist experienced in working with children affected by sickle cell to determine if there is an organic basis for impaired school performance. A counselor or social worker may also be helpful in working with the school system.

We encourage families to contact the school each year and to provide information about sickle cell disease to teachers, coaches, and school nurses. There may be other community professionals or resources to help families with this task. Addressing the needs of sickle cell patients, such as adequate fluid intake, frequent restroom visits, working with the child during pain episodes to decrease pain while avoiding excessive absences, and careful review of academic performance, enables the school system to become an ally of the family. School accommodations, covered by federal and state laws, should be pursued as needed.

**Physical Activities**

Physical exhaustion can precipitate a painful episode in children with sickle cell disease. While it is important for children with sickle cell to participate in physical activities at school, this often occurs without the necessary supportive measures to prevent difficulties. The educational process for affected children is to ensure adequate knowledge about their disease. When affected children request fluids or petition for modified physical activity they are often seen as problem students who want special treatment. On the contrary, as children grow to understand the precipitating factors that affect their illness, the fact that they begin to advocate on their own behalf should be viewed as a positive development.

However, balancing between disease-appropriate behaviors and avoiding a negative label is difficult for children. It is imperative for parents to be involved.
each year in their child’s classroom, and that they explain to teachers and administrators the special needs of their child.

As children get older, some may experience an increase in desire to compete in sports. This can result from peer or family pressure. The desire to “fit in” or “be like others” is very important for children aged 8 to 12 years. It may not be possible for some children to participate in contact sports, particularly strenuous sports, due to problems with easy fatigue or enlarged spleens. The result may be teasing by peers for not being able to participate. The child may look for other ways to prove themselves, or may participate in activities that are medically risky. At this age, children need activities that help build their self-esteem and improve understanding about their illness.

Effects of Frequent Hospitalizations

Small children who are hospitalized should be encouraged to bring special toys, like stuffed animals to provide comfort when familiar faces are not around. Similarly, a favorite blanket or pillow can be soothing while sleeping away from home. If possible, consults with pain management teams and child life specialists can provide strategies to reduce the trauma of painful procedures (see Pain Management). This is important for children who may experience frequent and prolonged hospitalizations.

Some children require frequent hospitalizations as a result of painful episodes, infections or transfusion protocols. Long hospitalizations can cause boredom, especially if the facility does not have an orientation toward children’s activities. If a child is having problems with other children as a result of their illness, it is likely that these behaviors will continue during hospitalization.

Consulting with families about home strategies for modifying unwanted behavior should provide some support for hospital staff. Alternatively, it is important to recognize that some parents may not have adequate strategies. In this case, it is important that a child life specialist, social worker or other professional be consulted as a resource for families and staff. It is essential to assure patients have support and advocates. This can be from family, community or friends.

Children should be encouraged to bring schoolwork to the hospital. Some facilities may have volunteers who can assist them, or paid staff members who fulfill this academic role. The school system may also provide tutors for students under certain conditions.

Children should be encouraged to phone and text friends and family members in an effort to stay connected to life outside the hospital. These strategies allow the child to stay focused on regular activities rather than focused on their illness. Living with a chronic illness can result in a general apathy about life, which can lead to sadness or depression.

If frequent admissions have been necessary, adolescents and their families will know the hospital system well. Many will develop expectations as to how an admission should go, and what interactions with staff will be like. In addition, they will know the flaws of the system as well, which can create tense moments for staff, patients and their families. For practitioners, it may be difficult to be confronted about staffing, equipment or the lack of communication between medical staff and families. Families may not know the best ways to communicate their concerns, so it may be necessary to help them define the problem. Some problems, like personality conflicts between specific staff members and families may not be easily remedied by the practitioner, but validating the experience and providing suggestions on how to handle situations can help reduce stress. Many hospital system problems do not have simple answers, although some families insist otherwise.

Mortality and Sickle Cell Disease

For families, the sickle cell diagnosis raises concerns about the affected child’s life span. It is important to talk openly about this fear with families and their children. With improvements in medical care, and parents’ involvement in learning about and teaching their children about the illness, 95% of children will live beyond age 18. The possibility of death should be addressed routinely with encouragement, emphasizing the importance of good care at home and creating a positive attitude toward life in spite of the chronic illness. Despite this, families and children should be reminded that having sickle cell should not be used as a reason to not pursue secondary education, have a career, and have a family and children.
The Basic Tenets of the Hemoglobinopathy Follow-Up Program

- Every child with sickle cell disease should have a source of primary medical care.
- Well-child care should follow the normal guidelines of the American Academy of Pediatrics. Hematology care is not a substitute for well-child care. The primary care provider should become familiar with the Management and Therapy of Sickle Cell Diseases publication from the U.S. Department of Health and Human Services (see References on page 50).
- A protocol for access to emergency care should be established early on.
- Every child should have regular consultation with a physician who has expertise in the sickling disorders. Some primary physicians with special interest and skill in the sickling diseases may act both as primary physicians and consultants.
- Children with major sickle complications (stroke, acute chest syndrome, renal or cardiac disease) should be evaluated by a tertiary care consultant familiar with treating these disorders.
- Positive sickle hemoglobinopathy screening results should be rechecked with a second newborn screen. Confirmatory testing should then be done after 1 year of age, when Hb F levels have normalized.
- When clinically significant hemoglobinopathies are confirmed, the primary care provider should refer to consultative care. Consultative care should be established in the first two months of life.
- Positive sickle hemoglobinopathy screening should lead to early prophylaxis of infection and anticipatory family education about the risks to a child with a sickling disease.
- The family should have access to 24-hour-a-day medical services through the primary physician or their on-call arrangements. Sickle cell specialists and tertiary level consultation should be available 24 hours a day to physicians.
- To ensure access to care, a social worker should be available to assist the family in identifying financial and other resources, and to connect to other state agencies.
- Genetic counseling services should be available to all families of children with hemoglobinopathies.
- Data on all newborn hemoglobinopathy screens should be centrally maintained so that clinicians can identify a child's hemoglobin status without rescreening.
- Communication should be maintained between those at all levels of care.
- Normal patterns of medical confidentiality and information exchange should be maintained.
### Diagnostic Testing for the Common Sickle Cell Syndromes

<table>
<thead>
<tr>
<th>Syndrome</th>
<th>Phenotype</th>
<th>Genotype</th>
<th>Neonatal Screening</th>
<th>MCV*</th>
<th>Hb A2 (%)**</th>
<th>Hb A (%)**</th>
<th>Hb S (%)**</th>
<th>Hb F (%)**</th>
<th>Hb C (%)**</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sickle Cell Disease (Hb SS)</td>
<td>Hemolysis and anemia by age 6-12 months</td>
<td>S-S</td>
<td>FS</td>
<td>Normal</td>
<td>&lt;3.5</td>
<td>0</td>
<td>80-95</td>
<td>2-25</td>
<td>0</td>
</tr>
<tr>
<td>Sickled β°-Thalassemia (1)</td>
<td>Hemolysis and anemia by age 6-12 months</td>
<td>S-B°</td>
<td>FS</td>
<td>Decreased</td>
<td>3.5-7</td>
<td>0</td>
<td>80-92</td>
<td>2-15</td>
<td>0</td>
</tr>
<tr>
<td>Sickle-C Disease (Hb SC)</td>
<td>Milder hemolysis and anemia</td>
<td>S-C</td>
<td>FSC</td>
<td>Normal or decreased</td>
<td>NA</td>
<td>0</td>
<td>45-50</td>
<td>1-5</td>
<td>45-50</td>
</tr>
<tr>
<td>Sickled β+-Thalassemia (1)</td>
<td>Milder hemolysis and anemia</td>
<td>S-B+</td>
<td>FSA or FS (3)</td>
<td>Normal or decreased</td>
<td>3.5-6</td>
<td>5-30</td>
<td>65-90</td>
<td>&lt;2</td>
<td>0</td>
</tr>
<tr>
<td>Sickle Cell Trait</td>
<td></td>
<td>AS</td>
<td>FAS</td>
<td>Normal</td>
<td>&lt;3.5</td>
<td>50-60</td>
<td>35-45</td>
<td>&lt;2</td>
<td>0</td>
</tr>
<tr>
<td>Normal</td>
<td></td>
<td>AA</td>
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<td>Normal</td>
<td>&lt;3.5</td>
<td>95-98</td>
<td>0</td>
<td>&lt;2</td>
<td>0</td>
</tr>
</tbody>
</table>

* By age 2 years.

** These are typical findings, but due to the large variability observed in sickle hemoglobinopathies actual values may fall outside of these ranges.

1. **β°** indicates a thalassemia mutation with absent production of **β**-globin; **β+** indicates a thalassemia mutation with reduced (but not absent) production of **β**-globin.

2. Hemoglobins reported in order of quantity (e.g. FSA = F>S>A); F, fetal hemoglobin; S, sickle hemoglobin; C, hemoglobin C; A, hemoglobin A. All abnormal results, including FAS, require confirmation with second newborn screen and Hb electrophoresis and confirmation testing at age 1 year.

3. The quantity of Hb A at birth is sometimes insufficient for detection.

4. Hb F levels in rare cases of Hb SS may be high enough to cause confusion with Hb S-Pancellular Hereditary Persistence of Fetal Hemoglobin (S-HPFH), a more benign disorder with less severe anemia and vaso-occlusion. In such cases, family studies and laboratory tests to evaluate the distribution of Hb F among red cells may be helpful.

5. The quantity of Hb A2 cannot be measured in presence of Hb C.

III. GUIDELINES FOR CARE OF CHILDREN WITH SICKLE CELL DISEASE

Definition of Levels of Care
This care plan assumes three levels of care for children with sickle cell disease:
1. The primary care physician;
2. A multidisciplinary program skilled in the nuances of sickle cell disease; and
3. Tertiary care for management of unusual or major complications.

Where skills and resources are appropriate, one medical site may provide several levels of care simultaneously. Whenever possible, the regular well-child care and immunizations should be managed by the primary physician, and disease-specific activities managed at the multidisciplinary program. The recommended timing and substance of visits will be described, but will vary with the needs of the patient, family and skills of the primary care provider. In general, infants should have monthly health care visits through the first six months, which can be alternated between primary and comprehensive sites, followed by visits every three to six months through 6 years of age.

These are guidelines, not standards. Their intent and the desired quality of care may be met by programs other than those described below.

The comprehensive program visits described below define counseling and teaching needs for age-specific sickle disease risks. This counseling may occur during the course of the normal primary provider visits listed if the primary caretaker is skilled in the problems of sickle diseases. Alternatively, the counseling and teaching goals may be met by outreach or in-home service providers such as public health nurses skilled in sickling diseases or tertiary program nurse clinicians. However, it is desirable for the child to visit the comprehensive program by 4 to 6 weeks of age and at least annually to establish the rapport and trust needed in case of major complications, and to keep abreast of new trends in the evaluation and treatment of sickle disease.

Visits listed with the primary care provider correspond to the current American Academy of Pediatrics well-child guidelines.

Refer to pages 18-24 for the three levels of the comprehensive care plan for children with sickle cell disease.

Clinic Requirements
Most of the care for sickle cell patients occurs in an outpatient setting. Comprehensive outpatient management has been shown to reduce morbidity, lessen the frequency of complications, lessen psychological burdens, and reduce the rate of hospitalization.

Primary Care Requirements
Primary caretakers should be familiar with and capable of providing the level of care outlined in The Management of Sickle Cell Disease.

Primary providers should help facilitate patients’ follow-up with the comprehensive sickle clinic.

Secondary Care Requirements
A. Diagnosis
- Ability to obtain and interpret results of screening and definitive tests for hemoglobinopathies.
- Ability to provide genetic counseling to affected families.
- Provide information about newborn screening program.

B. Ambulatory Care
- Provide general information about sickle cell diseases.
- Ability to follow guidelines for routine ambulatory care, as outlined in Management and Therapy of Sickle Cell Diseases.
- Access to educational materials to reinforce counseling.
- Participation of physicians, nurse practitioners and/or physician assistants with expertise in care of sickle cell patients.
Critical Elements of Care: Sickle Cell Disease

C. Complications

- Health care staff with experience and resources capable of identifying early signs of, and providing initial treatment for acute and chronic complications of sickle cell disease (such as: organ damage to include stroke, acute chest syndrome, splenic sequestration crises, sepsis, hand-foot syndrome, painful episodes, priapism, leg ulcers, avascular necrosis, sickle glomerulopathy, retinopathy, pulmonary hypertension, growth issues, and sickle lung disease).
- Proximity of tertiary level inpatient services, including surgical and medical services capable of providing initial care and stabilization for the above complications.
- Understanding the unique risks of surgery and anesthesia associated with sickling diseases.
- Availability to appropriately matched blood products.
- Availability of specialized pain management services, as well as availability of referral services for drug addictions.
- Access to academic and vocational counseling services.

D. Adolescent and Adult Care

- Transition strategy for patients transferring from pediatric care to adult care services.
- Birth control counseling and management.
- Genetic counseling
- Reproductive counseling and expertise in managing sickle cell patients through pregnancy and delivery.
- Understanding of the natural history of sickle cell disease and the development of approaches to monitor patients for chronic organ failure.

E. Access and Availability

- Patient access to expert physician/medical staff available 24 hours a day. Staff must be knowledgeable in sickle hemoglobinopathies and capable of inpatient management.

Comprehensive Sickle Cell Clinic: Tertiary Care

A. Diagnosis

- Physician level genetic counseling services.
- Availability of pain management team for design of individualized pain treatment protocols and for application of coping techniques for chronic pain.
- Neuropsychologist with expertise in recognition of neurocognitive deficits common to sickle cell disease.
- Availability of neuro-imaging technology (e.g. MRI/MRA and angiography) for delineation of neurologic abnormalities encountered in sickle cell disease.
- Availability of trans-cranial doppler and specialists trained in assessing patients with sickle cell anemia to screen for the risk of stroke.
- Availability of diagnostic testing for delineation of complications of sickle cell disease (e.g. pulmonary function testing, DEXA or bone density scan, abdominal US, echocardiography).
- Access to radiologists with experience differentiating sickle complications from other concerns.
Critical Elements of Care: Sickle Cell Disease

1. **Access to MRI based quantitative assessment of iron overload (e.g. T2*, Ferriscan or SQUID).**

   **B. Ambulatory Care**
   All components of secondary care, plus:
   - Social work and nutrition should have experience with sickle cell and have time dedicated to the clinic.

   **C. Complications**
   All components of secondary care, plus:
   - Clinician available to provide or directly access definitive care for acute and chronic complications of sickling diseases.
   - Participation in a tertiary care inpatient center capable of providing definitive medical and surgical care for complications of sickling diseases.
   - Ability to design and maintain patients on chronic transfusion programs and iron chelation therapy, as well as understand and monitor for the complications of iron overload and chelation therapy.
   - Familiarity with recent advances and ongoing experimental therapy in sickling diseases.
   - Involvement in clinical trials designed to improve the quality of life and care provided to sickle cell disease patients.
   - Access to a blood bank that performs extended red cell phenotyping and provides similarly matched blood products.

   **D. Adolescent and Adult Care**
   All components of secondary care, plus:
   - Sickle cell experience.

   **E. Access and Availability**
   Same as secondary level.

**Age-Specific Activities**

**2- to 4-Week Check by PRIMARY CARE PROVIDER**
- Conduct usual 2-week, well-child care.
- Review results of state newborn metabolic screen, which includes hemoglobinopathy screening results.
- Check if Hepatitis B vaccine given at birth. If not, begin series.

**When Presumptive Positive**

**Hemoglobinopathy Screen Becomes Available to PRIMARY PHYSICIAN**
- Discuss usual expectations of well-child care and practice arrangements, including after-hours coverage. It is important to encourage parents to maintain as normal a lifestyle as possible for children with sickle cell disease.
- No immediate confirmatory testing is necessary if the state lab has received two independent specimens as per standard policy for all newborns.
- Testing, including quantitation of hemoglobin types and for thalassemia, should be performed at 1 year of age after consultation or referral to a pediatric hematologist (a current listing is provided with the newborn screening program notification letter in Washington state).
- Begin Penicillin prophylaxis with Penicillin VK 125 mg BID orally to prevent pneumococcal sepsis.
- Provide prescription for folic acid supplements, 0.1 mg QD. Folate is consumed at increased rates in hemolytic anemias. It may be difficult finding liquid formulations; if preferred, please contact a pediatric hematologist.
- Emphasize the importance of observing for fever. The family should be taught to take a rectal temperature and appropriate use of antipyretics (e.g. avoiding antipyretics until the child has been evaluated for fever a health care provider). They should be taught to call the primary care provider immediately if fever develops.
• Emphasize the importance of fluid hydration.

• Make referral to your regional genetic counselor for assistance. A list of counselors with expertise in hemoglobinopathies is provided with the notification letter from the newborn screening program.

• Refer to WIC program for nutrition assistance (if eligible).

• Contact the County Health Department Children with Special Health Care Needs Program to have a public health nurse assigned.

6-Week Check by COMPREHENSIVE HEMOGLOBINOPATHY CARE PROGRAM ("COMPREHENSIVE PROGRAM")

• Discuss the identified hemoglobinopathy with the family. Answer further questions. Briefly discuss genetic basis, and if not already done, refer for genetic counseling.

• Highlight the following problems:

  Fever: Parents should check the child for fever if he or she is acting ill (demonstrate taking a rectal temperature). The family should be instructed to call the child’s physician or a tertiary care center if fever develops. Overwhelming sepsis should be discussed as well as its normal evaluation and management. The emergent risk of sepsis should be discussed and the need for immediate medical evaluation emphasized.

  Antibiotic Prophylaxis: Should be started by 4 to 6 weeks of age in patients with SS and Sβ0 Thalassemia. Use Penicillin 125 mg BID until age 3 years, and 250 mg BID from age 3 to age 6 years (Gaston et al., 1986). Some comprehensive hemoglobinopathy programs recommend continued prophylactic treatment throughout life, however, a randomized prospective trial for older patients without surgical splenectomy or prior pneumococcal sepsis has demonstrated no benefit (Falletta et al., 1995). Sepsis risk in sickle genotypes other than HbSS (e.g. SC, Sβ+ Thalassemia) is lower and penicillin for these patients may not be indicated. Erythromycin (20 mg/kg divided into two daily doses) may be used in cases of penicillin allergy.

Splenic Sequestration Crisis: Instruct the family in recognition of splenic sequestration crisis and examination of the spleen. To learn about the exam and their child’s normal splenic size, they should practice this daily when the child is quiet. In cases of irritability, pallor, increasing abdominal girth and tenderness or respiratory distress, they should know to examine the spleen and, if enlarged, seek care at once.

Other Medical Providers: Discuss the importance of identifying the child’s sickle disease diagnosis with other medical providers.

• Initiate social work evaluation. Include discussion of family structure, strengths, coping mechanisms and financial resources. Discuss normal reactions to chronic illness in one’s child. Provide information about the parent support group. Where appropriate, refer for financial support for medical care. Where available, refer to a care coordination program.

• Confirm that second hepatitis B vaccine was given.

• If appropriate and not yet done, refer to WIC or alternate nutrition counseling.

• Coordinate nurse review care plan with family.

• If appropriate, confirm public health nurse referral.

• Begin teaching awareness about coping with common problems associated with children with chronic illnesses.

2-Month Check by the PRIMARY CARE PROVIDER

• Perform routine well-child care and physical exam, and demonstrate spleen exam. Reinforce home palpation of spleen.

• Reaffirm antibiotic prophylaxis and review emergency care arrangements.

• Reinforce teaching about the significance and management of fever. Discuss use of liberal fluids and of antipyretics in illness.

• Review folate therapy.

• Give standard 2-month immunizations.
Critical Elements of Care: Sickle Cell Disease

III. GUIDELINES FOR CARE OF CHILDREN WITH SICKLE CELL DISEASE

3-Month Check by COMPREHENSIVE PROGRAM/Teaching Goals for Age

- Perform physical exam.
- Reinforce earlier teaching.
- Highlight:
  **Pain Episodes, Sickle Dactylitis:** Discuss how "colic" or fussiness may be symptoms of pain. Discuss administration of liberal oral fluids and appropriate outpatient pain medications. If pain is not relieved by fluids, rest, and oral analgesics, the child should be medically evaluated. Make available resources for coping with pain.

  **Causes of Sickling:** Discuss inciting causes or triggers of sickling. Include the kidney's limited ability to conserve water and consequent need for liberal fluid intake. Discuss fluids appropriate for maintaining hydration in illness or hot weather. Discuss the effects of cold, infections and tiring.

  - Social work update.
  - Coordinating nurse review care plan with family.
  - Review strategies to maximize health care access and introduce the patient and family to the Emergency Room, and reinforce strategies for positive interactions.

4-Month Check by PRIMARY CARE PROVIDER

- Perform routine well-child care.
- Give standard 4-month immunizations.
- Reinforce teaching about fever, splenic size, fluids, antibiotics, folic acid and pain therapy.
- Introduce coping strategies for blood draws and other invasive procedures.

5-Month Check by COMPREHENSIVE PROGRAM/Teaching Goals for Age

- Perform physical exam.
- Reinforce earlier teaching.
- Initiate dietary/nutrition counseling. Discuss the fact that good nutrition is important for the child's health but will not correct sickle diseases. Growth should be followed at each visit. Enroll in WIC if appropriate.
- Increase folic acid dose to 0.25 mg QD.
- Highlight:
  **Acute Chest Syndrome:** Discuss how respiratory distress or chest pain may signal problems and call for immediate medical evaluation. Normally, chest X-ray, CBC, retic and oximetry would be done. Antibiotics and oxygen should be administered, and transfusion may be provided in acute chest syndrome. Consider including antibiotic coverage for chlamydia and mycoplasma infection. Discuss the importance of expanding lungs to avoid atelectasis and recruit collapsed regions of lung. This is done with age-appropriate approaches.

  **Neurologic Complications:** Discuss neurologic complications of sickle cell disease. The family should be taught to look for and seek help if seizures, severe headache, weakness, paralysis/paresis, vertigo, visual changes or loss of speech occur. Emergent medical evaluation for CVA should be performed; if fever is present, the possibility of meningitis should be considered. An exchange transfusion is indicated for stroke. The tertiary care program should be contacted for advice.

  **Nurse:** Review care plan with family.

6-Month Check by PRIMARY CARE PROVIDER

- Perform routine well-child care.
- Reinforce previous teaching.
- Give standard 6-month immunizations.

8- to 9-Month Check by COMPREHENSIVE/PRIMARY CARE PROGRAM/Teaching Goals for Age

- Review and discuss prior teaching.
- Physical exam.
- Social service re-evaluation.
- Nurse review care plan with family.
- Influenza booster (initial two-dose vaccine during early first winter).
Note that the 8- to 9-month visit (and subsequent tri-monthly visits through 6 years of age) may either be performed as a single primary care visit, or separately as a primary care and comprehensive care visit, according to the expertise and comfort of the primary care provider.

11- to 12-Month Check by COMPREHENSIVE/PRIMARY CARE PROGRAM/Teaching Goals for Age
- History and PE.
- Labs: CBC, diff, retic, plt, BUN, Cr, Bili, Alk P, LDH, ALT, Iron Studies (other than FEP, ZPP), UA.
- Hemoglobin quantitation and thalassemia screen; electrophoresis or HPLC to quantitate hemoglobins (HbS, A, A2, F, C) and inclusion body or BCB prep. Should be done in an approved diagnostic laboratory.
- Tuberculin test, if indicated.
- Increase folic acid dose to 0.4 to 0.5 mg QD.
- Perform blood typing, and include sickle cell extended RBC matching panel (at a minimum RhD, Cc, Ee and Kell). Inform blood bank patient has sickle cell and should always receive blood with this extended matching.
- Introduce priapism.
- Confirm that genetic counseling occurred, and review.
- Nutrition counseling.
- Nurse review care plan with family.
- Routine immunizations/updates
- Distribute pain questionnaire.

17- to 18-Month Check by COMPREHENSIVE/PRIMARY CARE PROGRAM/Teaching Goals for Age
- Routine well-child care.
- Routine immunizations/ updates
- Review past teaching and examination.
- Nurse review care plan with family.
- Distribute pain questionnaire.

21-Month Check by COMPREHENSIVE/PRIMARY CARE PROGRAM/Teaching Goals for Age
- Review past teaching and examination.
- Social service case review.
- Discuss hyposthenuria and enuresis.
- Nurse review care plan with family
- Discuss Transcranial Doppler Study to identify children at increased risk for stroke (SS and Sßo patients)

24-Month Check by PRIMARY CARE PROVIDER
- Routine well-child care, review previous teaching.
- Pneumovax™ (PPV23), meningococcal, other routine immunizations/updates
- Increase folic acid to 0.8 to 1 mg QD.
- Discuss oral hygiene.

2 1/2-Year Check by COMPREHENSIVE PROGRAM/Teaching Goals for age (Annually on the half-year)
- Review need and importance of yearly studies.
- Review past teaching, PCN prophylaxis and exam.
- CBC, diff, plt, retic, BUN, Cr, Alk P, AST, Bili. LDH, Iron Studies
- Transcranial Doppler Study at 2 years of age and then yearly for patients with SS or Sßo-thalassemia, and some patients with Sß+ thalassemia (should be done at a tertiary care facility by personnel trained to study patients with hemoglobinopathies).
• Introduce concepts of incentive spirometry for lung expansion when sick or during pain episodes. Discuss age appropriate substitutes for incentive spirometry.
• Evaluate for asthma.
• Review status of new potential treatments and interventions.
• Annually in the fall, give booster influenza vaccine.
• Social service PRN.
• Nurse review care plan with family.
• Routine immunizations.
• If frequently transfused, please refer to guidelines for age 7 1/2 and older (below).

3- and 4-Year Check by PRIMARY CARE PROVIDER
• Routine well-child care.
• BP, UA with all subsequent annual visits.
• Ensure penicillin dose of 250 mg BID.
• Refer for routine dental care.
• Age four: Begin routine hearing and vision screening.
• Assess pain status, counsel family on pain management prevention and treatment.
• Begin coping strategy teaching with child.
• Assess and teach self-care skills.
• Developmental assessment.

5-Year Check by PRIMARY CARE PROVIDER
• Routine well-child care.
• Routine immunizations.

5 1/2- and 6 1/2-Year Check by COMPREHENSIVE PROGRAM/Teaching Goals for Age
• Review past teaching and examination.
• CBC, diff, plt, retic, BUN, Cr, Alk P, ALT, Bili, LDH, iron studies, UA.

Annual Check by PRIMARY CARE PROVIDER
• Routine well-child care.
• Pneumovax™ one-time booster five years after initial dose. Menactra booster every five years.
• Routine immunizations.
• Discontinue penicillin prophylaxis at age 6 years (children with a history of sepsis should continue on penicillin prophylaxis for life).
• Review yearly studies.

Annually from age 7 1/2 to 13 years on the Half-Year Check by COMPREHENSIVE PROGRAM/Teaching Goals for Age
• Review past teaching and examination.
• Discuss leg ulcers, priapism, delays in sexual maturation, sexual activity, smoking/drugs, activities and career goals as developmentally appropriate.
• Monitor/counsel on pain management.
• Monitor school progress and educational intervention as needed.
• Social service and nutritional evaluation as needed.
• Nurse review care plan with family.
• Review status of new potential treatments and interventions.
• Assess and teach self-care skills.
• Review yearly studies.
• Abdominal ultrasound for gall bladder stones, as needed for symptoms, and every other year routinely.
• Neuropsychologic evaluation q 2 to 3 years.
• Screen for depression and discuss coping strategies to provide mental health services.
• Pulmonary function tests, CXR, O2 saturation, TCD, ophthalmology and dental evaluations yearly.
• EKG every other year.
• Echocardiogram, including documentation of tricuspid regurgitation jet velocity for all patients with a history of decreasing exercise tolerance/activity, multiple pneumonias, progressive restrictive lung disease. Timing of re-evaluation depends on results and clinical progression.
• Repeat meningococcal immunization q 5 years.
• Chronic transfusion programs, if needed, will usually be managed by tertiary care programs. Transfusion-dependent children are at risk of iron toxicity to the liver, heart, pancreas and pituitary gland. Ferritin, Fe, TIBC, as well as percent HbS are followed closely. At least annually, hepatic and renal function should be tested. Annual 24-hour Holter monitoring may be appropriate. Clinical and serologic pituitary function testing, including gonadotropins, can be used to monitor pituitary function. Quantitative assessment of organ iron accumulation is required, preferably non-invasively with specific MRI sequences (T2* or Ferriscan), or SQUID. Liver biopsy to assess for portal fibrosis and chronic hepatitis may be needed if progressive liver damage is suspected. HIV and hepatitis serologies should be done yearly.

Annually from 14 to 18 years:

ADOLESCENCE ISSUES
• Review past teaching and examination.
• Discuss leg ulcers, priapism, potential delays in sexual maturation, sexual activity, smoking/drugs, activities and career goals as developmentally appropriate.
• Genetic counseling directed toward patient early adolescence.
• Monitor/counsel on pain management.
• Monitor school progress and educational intervention as needed.
• Social service and nutritional evaluation as needed.
• Nurse review care plan with family.
• Assess and teach self-care skills. Distinguish self-care from transition.
• Begin to develop a plan for transition to adult care.
• Discuss birth control options.
• Review yearly studies.
• Neuropsychologic evaluation q 2 to 3 years.
• Screen for depression.
• Abdominal ultrasound for gall bladder stones, as needed for symptoms, and every other year routinely.
• Pulmonary function tests, CXR, O2 saturation, TCD, ophthalmology and dental evaluations yearly.
• EKG every other year.
• Echocardiogram including documentation of tricuspid regurgitation jet velocity for all patients with a history of decreasing exercise tolerance/activity, multiple pneumonias, progressive restrictive lung disease. Timing of re-evaluation depends on results and clinical progression.
• Repeat meningococcal immunization q 5 years.
### Care Recommendations for SS and Other Sickle Syndromes
*Disease specific recommendations are noted by *(see footnotes)*

#### INFANCY Sickle and Primary Care Visits

<table>
<thead>
<tr>
<th>AGE</th>
<th>PCP 2- to 4-week</th>
<th>SICKLE 6-week</th>
<th>PCP 2-month</th>
</tr>
</thead>
<tbody>
<tr>
<td>History/PE</td>
<td>X</td>
<td>X</td>
<td>X</td>
</tr>
<tr>
<td>Immunizations</td>
<td>Hep B if needed</td>
<td></td>
<td>All routine immunizations, flu for household</td>
</tr>
<tr>
<td>Medications</td>
<td>PCN 125 mg PO BID, Folate 0.1 mg PO daily</td>
<td>Verify PCN 125 mg PO BID, Folate 0.1 mg PO daily</td>
<td>PCN 125 mg PO BID, Folic 0.1 PO daily</td>
</tr>
<tr>
<td>Labs/ Diagnostics</td>
<td>2nd NBS if needed</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Education</td>
<td>WCC, 24-hour access to care</td>
<td>Basic pathophysiology of sickle cell and natural history, improved survival</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Review NBS results Fever, hydration</td>
<td>Identify family’s preferred learning methods Inheritance/genetics Fever/sepsis/rectal temps Splenic sequestration Chronic illness awareness Discuss plan of care and emergency access Assess language preference</td>
<td></td>
</tr>
<tr>
<td>Referrals</td>
<td>Sickle care, Public Health Nurse (PHN), WIC, genetic counselor</td>
<td>SW, verify PHN, WIC, GC</td>
<td></td>
</tr>
</tbody>
</table>
### Care Recommendations for SS and Other Sickle Syndromes

#### INFANCY Sickle and Primary Care Visits

<table>
<thead>
<tr>
<th>AGE</th>
<th>History/PE</th>
<th>Immunizations</th>
<th>Medications</th>
<th>Labs/Diagnostics</th>
<th>Education</th>
<th>Referrals</th>
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<tbody>
<tr>
<td>PCP</td>
<td>X</td>
<td></td>
<td>PCN 125 mg PO, Folate 0.1 mg PO daily</td>
<td>Pathophysiology of sickling, dactylitis, pain episodes, avoidance of temperature extremes</td>
<td>Review of care, visit schedule, review health care access, what to expect with ER and inpatient hospitalizations</td>
<td>Verify WIC, parent support, other PRN</td>
</tr>
<tr>
<td>3-month PCP</td>
<td>Routine</td>
<td></td>
<td>PCN 125 mg PO BID, Folate 0.25 mg PO daily</td>
<td>Cord blood, pain, dactylitis, assessing pain in babies</td>
<td>Review respiratory monitoring, introduce neurological complications</td>
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<tr>
<td>4-month PCP</td>
<td>Routine, flu</td>
<td></td>
<td>PCN 125 mg PO BID, Folate 0.25 mg PO daily</td>
<td>Acute chest, nutrition, diet, fever, resp, spleen, pain</td>
<td>Fever, respiratory symptoms for pain, review dactylitis, develop pain plan, review temperature triggers for pain</td>
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<tr>
<td>5-month PCP</td>
<td>X</td>
<td></td>
<td>PCN 125 mg PO BID, Folate 0.25 mg PO daily</td>
<td>Fever, resp, spleen, pain</td>
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<tr>
<td>6-month PCP</td>
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<td>PCN 125 mg PO BID, Folate 0.25 mg PO daily</td>
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<td>Fever, resp, spleen, pain</td>
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<td>7-month PCP</td>
<td>X</td>
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<td>PCN 125 mg PO BID, Folate 0.25 mg PO daily</td>
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<td>Fever, resp, spleen, pain</td>
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<tr>
<td>9-month PCP</td>
<td>X</td>
<td></td>
<td>PCN 125 mg PO BID, Folate 0.25 mg PO daily</td>
<td>Fever, resp, spleen, pain</td>
<td>Fever, resp, spleen, pain</td>
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<tr>
<td>12-month PCP</td>
<td>Routine</td>
<td></td>
<td>PCN 125 mg PO BID, Folate 0.25 mg PO daily</td>
<td>Fever, resp, spleen, pain</td>
<td>Fever, resp, spleen, pain</td>
<td></td>
</tr>
</tbody>
</table>

#### Comments

- **Critical Elements of Care:**
  - Sickle Cell Disease

- **Care Recommendations for SS and Other Sickle Syndromes**
  - **INFANCY Sickle and Primary Care Visits**
    - **History/PE**
    - **Immunizations**
    - **Medications**
    - **Labs/Diagnostics**
    - **Education**
    - **Referrals**
## Care Recommendations for SS and Other Sickle Syndromes
### TODDLER Sickle and Primary Care Visits

<table>
<thead>
<tr>
<th>AGE</th>
<th>SICKLE 13-month</th>
<th>PCP 15-month</th>
<th>PCP 18-month</th>
<th>SICKLE 19-month</th>
<th>PCP 2-year</th>
<th>SICKLE 2-year</th>
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<tbody>
<tr>
<td>History/PE</td>
<td>X</td>
<td>X</td>
<td>X</td>
<td>X</td>
<td>X</td>
<td>X</td>
</tr>
<tr>
<td>Immunizations</td>
<td></td>
<td>Routine</td>
<td>Routine</td>
<td></td>
<td>PPV 23, Menactra</td>
<td></td>
</tr>
<tr>
<td>Medications</td>
<td>Increase Folate to 0.4-0.5 mg PO daily</td>
<td>PCN 125 mg PO BID, Folate 0.4-0.5 mg PO daily</td>
<td>PCN 125 mg PO BID, Folate 0.4-0.5 mg PO daily</td>
<td>PCN 125 mg PO BID, Folate 0.4-0.5 mg PO daily</td>
<td>Increase Folate to 0.8-1 mg/d PO daily</td>
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</tr>
<tr>
<td>Labs/ Diagnostics</td>
<td>Confirmatory labs*, annual labs, extended red cell phenotyping</td>
<td></td>
<td></td>
<td></td>
<td>Annual labs TCD*</td>
<td></td>
</tr>
<tr>
<td>Education</td>
<td>Review diagnosis, medications, genetic counseling and family planning</td>
<td>Routine WCC, nutrition and fluids, Fever, resp, spleen, pain</td>
<td>Routine WCC, nutrition and fluids, Fever, resp, spleen, pain</td>
<td>Create personal care plan for hospital use, Introduce neurological risk and TCD* with next visit, Introduce hyposthenuria and enuresis, Review developmental behavior vs. pain</td>
<td>Routine WCC, nutrition and fluids, Dental referral and oral hygiene, Fever, resp, spleen, pain</td>
<td>Discuss neurological and TCD* results, Review labs and pathophys, Introduce additional pain triggers, Introduce priapism, Ensure dental care</td>
</tr>
<tr>
<td>Referrals</td>
<td></td>
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</table>
## Care Recommendations for SS and Other Sickle Syndromes

### YEARLY Sickle Visits (Primary Care Visits continue per individual clinic protocol)

<table>
<thead>
<tr>
<th>AGE</th>
<th>3 YEARS</th>
<th>4 YEARS</th>
<th>5 YEARS</th>
<th>6 YEARS</th>
</tr>
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<tbody>
<tr>
<td><strong>History/PE</strong></td>
<td>X</td>
<td>X</td>
<td>X</td>
<td>X</td>
</tr>
<tr>
<td><strong>Immunizations</strong></td>
<td>As needed</td>
<td></td>
<td>Menactra booster 3 years after first dose if given between 2-6 1/2. If no Hib given previously, child should receive 1st dose.</td>
<td></td>
</tr>
<tr>
<td><strong>Medications</strong></td>
<td>Folate 0.8-1 mg/d PO Increase PCN to 250 mg BID</td>
<td>Folate 0.8-1 mg/d PO PCN 250 mg BID</td>
<td>Folate 0.8-1 mg/d PO PCN 250 mg BID</td>
<td>Folate 0.8-1 mg/d PO Discontinue PCN at age 6 years</td>
</tr>
<tr>
<td><strong>Labs/Diagnostics</strong></td>
<td>Annual labs TCD* Neuropsych (every 2-3 years)</td>
<td>Annual labs TCD*</td>
<td>Annual labs TCD*</td>
<td>Annual labs TCD*</td>
</tr>
<tr>
<td><strong>Education</strong></td>
<td>Distraction and stories with pain management Promote social opportunities, preschool Promote decision-making by offering choices</td>
<td>Start imagery work Promote activity/hobby as a long-term pain reliever (those involved cope with pain better) Promote decision-making by offering choices Have child help with simple chores (clean-up)</td>
<td>School readiness and accommodations, plan Early self-care (hydration) Encourage descriptions of pain Encourage monthly counseling for coping, stress Promote activity/hobby as a long-term pain reliever (those involved cope better with pain) Promote decision-making by offering choices Have child help with simple chores (clean-up)</td>
<td>Assess and build on child’s disease understanding Self-care: hydration, initial pain management (non-meds), warmth YMCA, boys/girls club, art, drama, music Promote decision-making, natural consequences allow child to experience consequences</td>
</tr>
<tr>
<td><strong>Referrals</strong></td>
<td></td>
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</tbody>
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*Note: TCD* may refer to Transcranial Doppler, a non-invasive test to measure blood flow in the brain.*
## Care Recommendations for SS and Other Sickle Syndromes

### YEARLY Sickle Visits

<table>
<thead>
<tr>
<th>AGE</th>
<th>7 YEARS</th>
<th>8 YEARS</th>
<th>9 YEARS</th>
<th>10 YEARS</th>
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<tr>
<td>History/PE</td>
<td>X</td>
<td>X</td>
<td>X</td>
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<tr>
<td>Immunizations</td>
<td>PPV23 booster 5 years after first dose</td>
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<tr>
<td>Medications</td>
<td>Folate 0.8-1 mg PO daily</td>
<td>Folate 0.8-1 mg PO daily</td>
<td>Folate 0.8-1 mg PO daily</td>
<td>Folate 0.8-1 mg PO daily</td>
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<td>Labs/Diagnostics</td>
<td>Annual labs Yearly studies</td>
<td>Annual labs Yearly studies Neuropsych</td>
<td>Annual labs Yearly studies</td>
<td>Annual labs Yearly studies</td>
</tr>
<tr>
<td>Education</td>
<td>Assess and build on child's disease understanding Self-care: hydration, initial pain management (non-meds), warmth Encourage social activities such as camps, school trips, outings, or sleep overs.</td>
<td>Assess and build on child's disease understanding Self-care: hydration, initial pain management (non-meds), warmth Sickle camp or other camp/social experience for children with special needs</td>
<td>Long-term interests Teasing, social opportunities Promote involvement in hobbies and extracurricular activities.</td>
<td>Explore teasing or bullying at school. Discuss advocating for self at school and other arena. Promote involvement in extracurricular activities and hobbies. Promote social gatherings with other families with sickle cell</td>
</tr>
<tr>
<td>Referrals</td>
<td></td>
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</tr>
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</table>
### Care Recommendations for SS and Other Sickle Syndromes

#### YEARLY Sickle Visits

<table>
<thead>
<tr>
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<th>12 YEARS</th>
<th>13 YEARS</th>
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<tr>
<td>Immunizations</td>
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<td></td>
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<tr>
<td>Medications</td>
<td>Folate 0.8-1 mg PO daily</td>
<td>Folate 0.8-1 mg PO daily</td>
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<tr>
<td></td>
<td>Neuropsych</td>
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</tr>
<tr>
<td>Education</td>
<td>Assess and build on child's disease understanding</td>
<td>Assess and build on child's disease understanding and self-care</td>
<td>Assess and build on child's disease understanding and self-care</td>
<td>Assess and build on child's disease understanding and self-care</td>
</tr>
<tr>
<td></td>
<td>Self-care: hydration, initial pain management, learn about meds</td>
<td>Asset-building with child: determine and build on strengths</td>
<td>Ensure youth knows emergency plan and pain plan</td>
<td>Begin to conduct at least part of visit without parent</td>
</tr>
<tr>
<td></td>
<td>Triggers</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Teach child to start keeping records of labs, studies, visits</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Referrals</td>
<td></td>
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## Care Recommendations for SS and Other Sickle Syndromes

### YEARLY Sickle Visits

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<th>16 YEARS</th>
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<tr>
<td>Education</td>
<td>Assess and build on child’s disease understanding and self-care</td>
<td>Assess and build on child’s disease understanding and self-care</td>
<td>Assess and build on child’s disease understanding and self-care</td>
</tr>
<tr>
<td></td>
<td>Discuss family planning, birth control, genetics</td>
<td>Encourage youth to start making own appointments, tracking progress, and managing medications with parental help</td>
<td>Introduce to adult hematology, tour adult facility, arrange adult primary care provider</td>
</tr>
<tr>
<td></td>
<td>Discuss drugs and alcohol especially in relation to sickle cell as well as interactions with opiates.</td>
<td>Discuss drugs and alcohol especially in relation to sickle cell as well as interactions with opiates.</td>
<td>Discuss drugs and alcohol especially in relation to sickle cell as well as interactions with opiates.</td>
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<tr>
<td>Referrals</td>
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</table>
Pain Related to Sickle Cell Disease

Pain is the hallmark of sickle cell disease. The pain associated with sickle cell is complex in that it can be acute, recurring, chronic or a mixture of these. Unlike other causes of pain, there may be no biomarkers or physical indicators for the clinician to use to evaluate pain, and over time patients may adapt to the pain and objective findings such as elevations in heart rate or blood pressure are not always observed.

Trust in the patient’s report and eliciting a good description of pain are therefore critical components in the evaluation of sickle pain, and in the differentiation between sickle pain and other etiologies of pain. Management of pain must be individualized to each patient and plans of care should be created that work best for each person. Treatment should, however, always be multimodal incorporating the alleviation of triggers in addition to non-pharmacological and pharmacological approaches.

**Severity:** Varies from mild to extremely intense.

**Character:** Deep, aching, tiring, fatiguing, relentless. Described as “body chewing,” “body biting” or “bone breaking.”

**Developmental Aspects:** Can occur as early as 4 to 9 months of age when fetal hemoglobin levels are diminished.

**Region:** Can occur in any part of the body and may involve single or multiple body parts. Common complaints:
- Extremity pain
- Abdominal pain
- Back pain

Pain due to swelling in hands and feet from dactylitis typically occurs in children under 3 years of age.

**Frequency:** Sickle cell pain forms a continuum from acute to chronic:
- 30% never or rarely have pain
- 50% have few episodes
- 20% have frequent, severe episodes (6% of patients account for 30% of all painful episodes)

Precipitating Factors:
- Infection
- Hypoxemia
- Dehydration
- Fatigue
- Exposure to cold, changes in weather
- Strenuous exercise
- Sleep apnea

General Principles of Pain Management

A number of general principles can be applied to the management of pain in sickle cell disease.

A. Pain must be viewed within a chronic disease continuum: Promotion of wellness and development while also consistently addressing pain is necessary.

B. Health care professionals have the accountability/responsibility for using a proactive, not a reactive approach. Multiple interventions and approaches should be integrated in the management of pain, not simply medication alone.

C. Emphasize the value of a system-wide approach

1. Effective pain management is contingent on involvement by administration, managers, practitioners and family members.
2. Role of child and family:
   a. To expect that pain be treated/integrated into a plan of treatment
   b. To participate in designing and modifying plan, informing providers of personal belief system that impacts care choices.
   c. To obtain education and support
3. Role of administration, managers, and practitioners:
   a. Pain relief is a quality assurance/continuous quality improvement issue for children with chronic illness. Care effectiveness must be evaluated.
b. Develop standards of care/clinical guidelines for common pain problems such as:
   Emergency room treatment of sickle cell pain episode, home management procedures, and developing multiple healthy coping strategies.

D. Adequate assessment is the cornerstone of therapy
   1. Pain assessment should be developmentally appropriate and a routine part of the inpatient and outpatient care of children with these chronic diseases.
   2. The child's complaints of pain should be believed. Verbal self-report is primary and cannot be disputed.

E. Assess and develop a plan of care with the first episode of pain
   1. Online “Pain profiles” that are accessible or transferable, regardless of site of care.
   2. Summarizes pain history and details the pain care plan based on child and family input and past experiences. Plan should include both non-pharmacologic and pharmacologic details. Plan is modified and updated on a real-time basis.
   3. Life records: Eliminates the need for repeated questioning of child/parents(s), particularly as they enter different hospital areas (ER, clinic, inpatient, OR).
   4. A pain problem list should be instituted so that pain stemming from the disease and its treatment can be isolated and treated appropriately.
   5. Hand-held records: Empowers child and family.

F. Guidelines for clinical care
   1. Avoid the use of the term pain “crisis” as this can contribute to a sense of anxiety. A more appropriate term is “pain episode.”
   2. Please refer to the management of sickle cell pain algorithms and charts in this document.
   3. General principles of pharmacologic management:
      a. Severe pain is an emergency and must be treated accordingly.

b. Use a stepwise approach to pharmacologic therapy that includes: initial therapy with NSAIDs, add low potency short acting opioids if necessary, change to higher potency short acting opioids if needed, add long acting opioids and adjuvant therapies as needed.

c. Assessment and re-assessment must be ongoing throughout the course of pain treatment.

d. Be certain that adequate analgesics are given to allow nighttime sleep.

e. In the majority of cases, oral routes of analgesia are effective and should be used.

f. Scheduled administration to prevent anticipated return of pain is appropriate, unless pain is truly episodic and unpredictable.

g. Avoid noxious routes of administration (e.g. I.M. injections) since children will often deny pain due to a fear of needles.

h. Addiction is rare. Fear of addiction should not restrict adequate opioid administration.

i. Do not use placebos.

j. Involve the child and his/her family in the treatment, and respect personal preferences and cultural diversity.

k. If dose reduction is indicated, it should be done slowly to avoid precipitating severe pain withdrawal.

l. Side effects should be anticipated and treated.

m. The goal of therapy should be adequate analgesia to allow increased function as determined by the patient, family and staff.

n. Although there are guidelines for starting doses, there is no maximum dose for opioids. The right dose is the dose that is adequate to relieve the pain without undue toxicity.

o. Assess often for respiratory compromise, as hypoxemia may contribute to episodes of acute chest syndrome. Incentive spirometry while on opiates.
4. Complimentary non-pharmacologic strategies—developmental approaches:

<table>
<thead>
<tr>
<th>Ages</th>
<th>Explanations</th>
<th>Distractions</th>
<th>Containment</th>
<th>Physical</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Infants</strong></td>
<td>Caregiver teaching</td>
<td>Music/mobiles, soothing talk, soft or a novel voice, calm demeanor, oral-motor stimulation (pacifiers, non-nutritive sucking)</td>
<td>Holding/cuddling/swaddling, positioning, pacifier</td>
<td>Massage (applicability/efficacy being determined)</td>
</tr>
<tr>
<td><strong>Toddlers/Preschoolers</strong></td>
<td>Pop-up books, magic circle/magic game, puppets, kaleidoscopes, counting ABCs, music-sing-along songs, squeezing on koosh ball</td>
<td>Pinwheel, blowing bubbles, “meow-woof” breathing, party blowers</td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>School-Age/Adolescents</strong></td>
<td>Modeling/desensitization: Explanations to child and family</td>
<td>(younger): Pop-up books; counting ABCs, puppets, kaleidoscopes, music with I-Pod, DVD player, video games</td>
<td>Imagery (older): Pain switch, familiar images with stories, biofeedback</td>
<td></td>
</tr>
</tbody>
</table>

**Breathing/relaxation:**
- “Go limp as a ragdoll,” or “You’re blowing hurt away,” or ask the child to yawn, choo-choo like a train

**Imagery:** Stories-use images familiar to the child

**Explanations:** Before procedure, provide concrete and brief explanations to caregiver and child; during procedure, provide sensory information and emphasize informational affective aspects of the experience; after procedure, use therapeutic play

**Physical:** Massage, heat, acupuncture, acupressure, Transcutaneous Electrical Nerve Stimulation (TENS)
### Common Pain States Associated with Sickle Cell Disease

<table>
<thead>
<tr>
<th>Pain States</th>
<th>Clinical Signs &amp; Symptoms</th>
<th>Signs &amp; Underlying Cause</th>
<th>Special Features &amp; Considerations</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Acute Painful Event</strong></td>
<td>• Sudden onset</td>
<td>• Vaso-occlusion</td>
<td>• Unpredictable, recurrent</td>
</tr>
<tr>
<td></td>
<td>• Pain in any or all parts of the body</td>
<td>• Endothelial damage</td>
<td>• Great variability</td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Inflammation</td>
<td>• All ages</td>
</tr>
<tr>
<td><strong>Acute Hand-Foot Syndrome</strong></td>
<td>• Painful dorsal swelling of hands and feet</td>
<td>• Symmetrical infarcts of metacarpal and metatarsal bones due to obstruction of developing blood vessels</td>
<td>• More common in childhood</td>
</tr>
<tr>
<td>(Dactylitis)</td>
<td></td>
<td></td>
<td>• Often first manifestation of disease (occurs as early as 6 months of age)</td>
</tr>
<tr>
<td><strong>Acute Inflammation of Joints</strong></td>
<td>• Painful, swollen joints</td>
<td>• Vaso-occlusion/injury</td>
<td>• May accompany dactylitis</td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Inflammation</td>
<td>• Acute flare-ups as isolated events</td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Infected joints</td>
<td>• Septic flare-ups are rare but may occur</td>
</tr>
<tr>
<td><strong>Acute Chest Syndrome</strong></td>
<td>• Chest pain, particularly rib and substernal area</td>
<td>• Pulmonary infiltrate</td>
<td>• May require transfusion and can be fatal</td>
</tr>
<tr>
<td></td>
<td>• Chest pain posteriorly (upper back)</td>
<td>• May be associated with infarction, infection or hemorrhage, or any combination of these</td>
<td>• Common cause of mortality in children and adults</td>
</tr>
<tr>
<td></td>
<td>• Fever, tachypnea, and/or hypoxia</td>
<td>• Unilateral pain (splinting from atelectasis)</td>
<td></td>
</tr>
<tr>
<td><strong>Splenic Sequestration</strong></td>
<td>• Left upper-quadrant pain</td>
<td>• Blood trapped in the spleen</td>
<td>• Can be catastrophic in children, with possibility of circulatory collapse</td>
</tr>
<tr>
<td></td>
<td>• Marked pallor</td>
<td></td>
<td>• Insidious onset in adults</td>
</tr>
<tr>
<td></td>
<td>• Sudden decrease in hemoglobin concentration</td>
<td></td>
<td>• Occurs in older children and adults with HbSC and sickle β-thalasemia</td>
</tr>
<tr>
<td></td>
<td>• Enlarged spleen</td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Intrahepatic Sickling or Hepatic Sequestration</strong></td>
<td>• Right upper-quadrant pain</td>
<td>• Blood pooling in the liver</td>
<td>• Occurs more commonly in adults</td>
</tr>
<tr>
<td></td>
<td>• Sudden decrease in hemoglobin</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>• Enlarged liver</td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Abdominal and Intra-abdominal Pain</strong></td>
<td>• Jaundice</td>
<td>• Cholelithiasis</td>
<td>• Can be initial manifestation of acute chest syndrome</td>
</tr>
<tr>
<td></td>
<td>• Diffuse abdominal pain</td>
<td>• Gastritis</td>
<td>• Involve surgery if severe symptoms</td>
</tr>
<tr>
<td></td>
<td>• Enlarged spleen</td>
<td>• Constipation secondary to opioid therapy</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Splenic infarction</td>
<td></td>
</tr>
<tr>
<td><strong>Priapism</strong></td>
<td>• Painful erection</td>
<td>• Sickling in sinusoids of penis</td>
<td>• May be chronic or stuttering (intermittent)</td>
</tr>
<tr>
<td><strong>Avascular Necrosis of Femur or Humerus</strong></td>
<td>• Prolonged, constant bone pain</td>
<td>• Associated with bone infarction, sickle arthritis</td>
<td>• Physical therapy may be useful for reducing pain and maintaining function</td>
</tr>
<tr>
<td></td>
<td>• Shoulder pain</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>• Knee pain</td>
<td>• Physical therapy may be useful for reducing pain and maintaining function</td>
<td></td>
</tr>
<tr>
<td></td>
<td>• Hip pain</td>
<td>• Associated with bone infarction, sickle arthritis</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Physical therapy may be useful for reducing pain and maintaining function</td>
<td></td>
</tr>
<tr>
<td><strong>Chronic Neuropathic Pain</strong></td>
<td>• Pain in back, lower extremities, other sites</td>
<td>• Older adults: disc disease, infections</td>
<td>• Must be considered in patients with a decreased response to opioids</td>
</tr>
<tr>
<td></td>
<td>• Spontaneous</td>
<td>• Collapsed vertebrae</td>
<td>• Physical therapy</td>
</tr>
<tr>
<td></td>
<td>• Lancinating</td>
<td>• Iron overload neuropathy</td>
<td>• Consider medications for neuropathic pain such as gabapentin</td>
</tr>
<tr>
<td></td>
<td>• Burning</td>
<td></td>
<td>• Treatment modalities may require days or weeks before taking effect</td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Creates chronic pain state</td>
<td></td>
</tr>
</tbody>
</table>

Adapted from APS guidelines
Pain Assessment Tools

There are a variety of tools available to measure pain severity and functional impact. The following are a variety of tools to choose from. The practitioner should use what is most appropriate for the patient and situation.

Assessment Tool 1: The Oucher

*Which part of the scale should be used?*

If children can count to 100, they can use the numerical scale; if not, they should use the photographic scale.

*How does one use the Oucher?*

A. Let children practice using the Oucher.
   1. Ask them to recall times they hurt in the past.
   2. Have them describe these episodes to you and then rate them on the Oucher.

B. Collect data and convert to scores.
   1. After re-explaining the scale, ask, “How much hurt do you have right now?”
   2. If the child uses the numerical scale, the number they give is the Oucher score; if the child uses the photographic scale, the picture they select is converted to the appropriate predetermined score shown on the Oucher (0, 20, 40, 60, 80 or 100).

Assessment Tool 2: Pain Intensity Number Scale

*Children Developmentally Later School-Age and Adolescent*

Instructions:

1. “I need to know how much pain you have because I can’t feel your pain. I want you to use a scale so you can tell me how much pain you have right now.”

2. “The numbers between 0 and 10 represent all the pain a person could have. Zero means no pain and 10 means pain as bad as it could be. You can use any number between 0 and 10 to let me know how much you have right now.”

3. “Give your pain a number between 0 and 10 so I will know the intensity of the pain you feel now.”

4. Record the pain intensity on the nursing flow sheet as 0/10, 1/10, 2/10, etc.


5. For younger children, substitute the FACES pain scale for number scale.

Pain Intensity FACES Scale
Assessment Tool 3: Work Graphic Rating Scale

*Children Developmentally Later School-Age and Adolescent*

Instructions:
1. Place a straight up-and-down mark on this line to show how much pain you have.

<table>
<thead>
<tr>
<th>No pain</th>
<th>Little pain</th>
<th>Medium pain</th>
<th>Large pain</th>
<th>Worst possible pain</th>
</tr>
</thead>
</table>

2. Record the pain intensity on the nursing flow sheet as “none,” “little,” “medium,” “large” or “worst possible.”

**Assessment Tool 4: Functional Assessment**

Record functional assessment of pain on flow sheet, for example:
- “Unable to sit up or walk”
- “Able to eat”
- “Able to do self care”
Sickle Cell Pain Assessment

Determine type of pain (onset, duration, frequency)

Acute or chronic?

Acute

Brief or persistent?

Brief

Determine characteristics, location, and intensity based on self-report

Persistent

Determine characteristics, location, and intensity based on self-report and observation

Determine related symptoms

Determine probable cause(s)

Related to SCD?

NO

Conduct complete work-up to determine etiology of pain

YES

Treatment History
- Frequency of painful episodes in previous year
- Number of ED visits in past year
- Frequency and duration of hospitalizations
- Pain medication history
- Current medication regime

Physical Factors
- Blood pressure
- Heart rate
- Respiration
- Oxygen saturation level
- Chest/abdomen
- Pain sites
- Tenderness
- Warmth
- Swelling
- Lab/X-ray data

Demographic/Psychosocial Factors
- Age
- Gender
- Developmental level
- Family factors
- Cultural factors
- Adaptation to SCD
- Coping styles
- Cognitive abilities
- Mood
- Level of distress

Dimensions of Pain
- Intensity of 0-10 scale:
  - Mild (0-3)
  - Moderate (4-6)
  - Severe (7-10)
- Characteristics:
  - Location
  - Quality
  - Precipitating factors

Impact of Pain on Functioning
- Self-care
- School/work
- Social activities
- Relationships
- Parent ability (adults)

Summarize, assimilate and prioritize profile

Identify appropriate interventions based on comprehensive assessment

Adapted from APS guidelines
Treatment Flowchart

Identify appropriate intervention based on comprehensive assessment

**PHARMACOLOGICAL**
- Acetaminophen or NSAIDs
- Opioids
- Adjuvants

**BEHAVIORAL**
- Relaxation
- Deep breathing
- Behavior modification
- Biofeedback
- Exercise

**PSYCHOLOGICAL**
- Cognitive therapies
- Hypnotherapy
- Imagery
- Distraction
- Social support

**PHYSICAL**
- Hydration
- Heat
- Massage
- Hydrotherapy
- Ultrasound
- Acupuncture/Acupressure
- Physical therapy

Identify patient/family educational needs

Formulate treatment plan

Adapted from APS guidelines
Management of an Episode of Acute Pain in Sickle Cell Disease

**Arrival at emergency department**

Severe acute pain

Assess for common acute pain states associated with sickle cell disease (SCD) (See table on page 31)

Determine type of pain (onset, duration, frequency)

Typical pain?

NO ▼

Determine related symptoms (Infections, complications, other comorbidities/precipitating factors)

Related to SCD?

YES ▲

NO ▼

Determine pain characteristics (intensity, location and quality based on self-report)

Obtain treatment history (home meds, acute pain, hospital rx, meds past 24 hours, out of home meds?)

Examine pertinent physical factors (defer more extensive history and physical until patient is comfortable)

Summarize assessment profile and select treatment (based on characteristics of episode and prior treatment history and physical findings)

Administer a parenteral or oral non-steroidal anti-inflammatory unless contra-indicted

Currently on chronic opioid therapy?

NO ▼

Start IV loading dose of short-acting opioid. Refer to patient specific plan if available. In not, use the following defaults:

- Adults/children > 50 kg body weight:
  - Morphine 0.1-0.15 mg/kg IV
  - Hydromorphone 0.015-0.020 mg/kg IV

- Adults/children > 50 kg body weight:
  - Morphine 5-10 mg IV
  - Hydromorphone 0.75 mg-1.5 mg IV

YES ▲

Select medication and loading dose based on overall assessment and prior treatment history

(Note: Patient/family often know what medication and dosage have been effective in the past)
Management of an Episode of Acute Pain in Sickle Cell Disease  

**Time elapsed from admission to emergency department**

**15-20 min.**

Administer by IV (if sufficient venous access) or subcutaneous route (if insufficient venous access)

Assess degree of relief q 15-30 min.

Moderate pain relief?

- YES ▼
  - Side effects tolerable?
    - YES ▼
      - Begin around-the-clock dosing
    - NO ▼
      - Moderate pain relief?
        - NO ▼
          - Continue IV analgesic at initial dose used (or fraction thereof) or per pain plan
        - YES ▼
          - Use adjuvant medications in combination to enhance the efficacy side-effect ratio
          - Monitor effectiveness of pain control

- NO ▼
  - Make disposition
    - Treatment effective?
      - NO ▼
        - Complications?
          - YES ▼
            - Admit to hospital
          - NO ▼
            - Can be maintained at home with oral medications?
              - YES ▼
                - Send home with prescribed level of medication (PO) to maintain adequate pain relief
              - NO ▼
                - Refer back to clinician managing SCD

  - NO ▼
    - Reassess often
    - Titrate to relief
    - Maintain relief

**30 min.**

Breakthrough pain?

- YES ▼
  - Provide rescue dosing

- NO ▼
  - Treatment effective?
    - NO ▼
      - Complications?
        - YES ▼
          - Admit to hospital
        - NO ▼
          - Can be maintained at home with oral medications?
            - YES ▼
              - Send home with prescribed level of medication (PO) to maintain adequate pain relief
            - NO ▼
              - Refer back to clinician managing SCD

- YES ▼
  - Maintain relief

**2-8 hours**

Refer back to clinician managing SCD

These are the American Pain Society guidelines. Individual institutions, including Seattle Children’s Hospital, may have differing practices.

Adapted from APS guidelines
# Vaso-Occlusive Pain

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>Monitoring</th>
<th>Diagnostic (if not previously obtained)</th>
<th>Fluids, Nutrition, General Care</th>
<th>Medications/Treatments</th>
<th>Discharge Criteria</th>
</tr>
</thead>
<tbody>
<tr>
<td>Vaso-occlusive pain in a child with sickle cell disease</td>
<td>1. Vital signs q 4 hr 2. Record I + O, daily weight 3. Continuous pulse ox. if any respiratory symptoms present, or if on parenteral opiates 4. Consider CR monitor</td>
<td>1. CBC, diff., plt count and retic count initially (compare with patient’s baseline data); consider a hold tube for the blood center (for later type and cross) if severe anemia suspected or transfusion anticipated 2. CXR: low threshold if cough or any respiratory signs or symptoms are present, or develop after admission; encourage incentive spirometry prior to CXR 3. Blood culture if &gt;38.2°C; urinalysis, urine culture and other cultures (e.g. CSF) as indicated 4. Consider diagnostic tests to evaluate possible non-sickle causes of pain (e.g. abdominal ultrasound, liver function tests for RUQ to R/O cholelithiasis and cholecystitis)</td>
<td>1. IV + PO 1.25 x maintenance. Increased fluids only if patient is dehydrated and/or insensible losses are increased (e.g. persistent fever); avoid excessive fluids, which may worsen respiratory status 2. Avoid IV fluid bolus unless clinically dehydrated or clinically indicated (not for pain alone) 3. Incentive spirometry−10 breaths q 2 hr. from 0800-2200 and while awake. 4. Encourage ambulation and activity</td>
<td>1. Follow patient-specific care plan if available; if not, follow generic steps below 2. Offer heat pads, imagery, relaxation methods or other comfort measures as adjunct to pharmaceuticals. 3. A parenteral or oral non-steroidal anti-inflammatory agent if no contraindication (i.e. gastritis, ulcer or renal impairment) 4. If no established pain plan: Morphine sulfate 0.1 mg/kg/dose IV q 2 hr. or 0.01-0.1 mg/kg/hr. continuous infusion or via PCA (doses above 0.1 mg/kg/hr. may be required but should be used with caution); alternative analgesics may be used in individual cases* 5. Reassess pain control at least twice daily and after every intervention; analgesics may be weaned as tolerated by decreasing dose, not by prolonging interval between doses; discuss analgesic changes with patient/family 6. Start oral opiates as soon as tolerated from a gastrointestinal standpoint, even if requiring IV opiates 7. Consider pain team consultation 8. Ceftriaxone 75mg/kg q 24 hr if febrile (prophylactic penicillin may be discontinued while on broad-spectrum antibiotics) 9. Continue prophylactic folic acid, if applicable 10. O₂ by nasal cannula as needed to keep O₂ saturation &gt; 93% 11. Colace or laxative to prevent narcotic-induced constipation 12. See other Clinical Care Paths for acute chest syndrome, acute anemia crisis, stroke, priapism, if present 13. Avoid use of ice or cold packs</td>
<td>1. Taking oral fluids well and able to take all PO meds (e.g. prophylactic penicillin) if applicable 2. Adequate pain relief on oral analgesics 3. Afebrile &gt;24 hr and negative cultures &gt;24 hours if applicable 4. Resolution of any pulmonary symptoms or documentation of adequate oxygenation on room air</td>
</tr>
</tbody>
</table>

* These are the American Pain Society guidelines. Individual institutions, including Seattle Children’s Hospital, may have differing practices.

Modified from Mountain States Regional Genetic Services Network, 1996
**Sedation Scale and Indications for Action**

<table>
<thead>
<tr>
<th>Sedation Scale</th>
<th>Action Required</th>
</tr>
</thead>
</table>
| 5  Sleeping, easily aroused | None  
Consider monitoring O₂ saturation |
| 1  Awake and alert | None  
Consider monitoring O₂ saturation |
| 2  Occasionally drowsy, easy to arouse | None  
Consider monitoring O₂ saturation |
| 3  Frequently drowsy, arousable, drifts off to sleep during conversations | Physically stimulate  
Decrease or discontinue the opioid  
Monitor O₂ saturation |
| 4  Somnolent, minimal or no response to stimuli | Discontinue opioid and consider titration of naloxone  
Monitor O₂ saturation |


Note: The above scale and recommendations are not appropriate for patients who are terminally ill, have developed a tolerance to respiratory depression, or in whom sedation is not caused by opioids.
# NSAIDs Dosing Data

<table>
<thead>
<tr>
<th>Medication</th>
<th>Usual Dose for Adults and Children ≤50 kg Body Weight</th>
<th>Usual Dose for Children(^1) and Adults(^2) ≤50 kg Body Weight</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Acetaminophen and Over-the-Counter NSAIDs</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Acetaminophen(^3)</td>
<td>650 mg q 4 hr</td>
<td>10-15 mg/kg q 4 hr (oral)</td>
</tr>
<tr>
<td></td>
<td>975-1,000 mg q 6 hr</td>
<td>15-20 mg/kg q 4 hr (rectal)</td>
</tr>
<tr>
<td>Ibuprofen</td>
<td>400-800 mg q 6 hr</td>
<td>10 mg/kg q 6-8 hr</td>
</tr>
<tr>
<td>Ketoprofen (Orudis)</td>
<td>25-75 mg q 6-8 hr</td>
<td>0.5 mg/kg q 6 hr</td>
</tr>
<tr>
<td>Naproxen (Naprosyn)</td>
<td>500 mg initially, then 250 mg q 6-8 hr</td>
<td>5-7 mg/kg q 8-12 hr</td>
</tr>
<tr>
<td>Naproxen sodium (Anaprox)</td>
<td>550 mg initially, then 275 mg q 6-8 hr</td>
<td></td>
</tr>
<tr>
<td><strong>Parenteral NSAIDs</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Ketorolac tromethamine(^5,6) (Toradol)</td>
<td>30 mg initially, then 15-30 mg q 6 hr, parenteral dose not to exceed 5 days or 120 mg/day</td>
<td>0.5 mg/kg q 8 hr intravenously(^5,6)</td>
</tr>
</tbody>
</table>

\(^1\) Only medications that are FDA-approved as analgesics for children are included.

\(^2\) Acetaminophen and NSAID dosages for adults weighing less than 50 kg should be adjusted for weight.

\(^3\) Acetaminophen lacks the peripheral anti-inflammatory and antiplatelet activities of other NSAIDs.

\(^4\) Ibuprofen is not FDA-approved for use in children as an over-the-counter medication. It has FDA approval for use in children as a prescription medication for fever; however, some clinicians have had experience in prescribing ibuprofen for pain relief in children.

\(^5\) For short-term use only.

\(^6\) Has the same GI toxicities as oral NSAIDs. Safety and efficacy not established for use in children.

Modified from “Guidelines for the Management of Acute and Chronic Pain in Sickle Cell Disease,” American Pain Society, August 1999. These are the American Pain Society guidelines. Individual institutions, including Seattle Children’s Hospital, may have differing practices.
**Opioid Dosing Table**  
*(For patients without established pain plan)*

<table>
<thead>
<tr>
<th>Opioid Agonist</th>
<th>Approx Equianalgesic Oral Dose</th>
<th>Approx Equianalgesic Parenteral Dose</th>
<th>Recommended Starting Dose (Adults &gt; 50 kg) Oral</th>
<th>Recommended Starting Dose (Adults &gt; 50 kg) Parenteral</th>
<th>Recommended Starting Dose (Children, adults &lt; 50 kg)</th>
<th>Recommended Starting Dose (Children, adults &lt; 50 kg) Parenteral</th>
</tr>
</thead>
<tbody>
<tr>
<td>Morphine&lt;sup&gt;2&lt;/sup&gt;</td>
<td>30 mg q 3-4 hr (around-the-clock dosing); 60 mg q 3-4 hr (single dose or intermittent dosing)</td>
<td>10 mg q 3-4 hr</td>
<td>10-30 mg q 3-4 hr</td>
<td>5-10 mg q 2-4 hr</td>
<td>0.3 mg/kg q 3-4 hr</td>
<td>0.1 mg/kg q 3-4 hr</td>
</tr>
<tr>
<td>Codeine&lt;sup&gt;3&lt;/sup&gt;</td>
<td>200 mg q 3-4 hr</td>
<td></td>
<td>15-60 mg q 3-4 hr</td>
<td>Not recommended</td>
<td>0.5-1 mg/kg q 3-4 hr</td>
<td>Not recommended</td>
</tr>
<tr>
<td>Hydromorphone&lt;sup&gt;2&lt;/sup&gt; (Dilaudid)</td>
<td>7.5 mg q 3-4 hr</td>
<td>1.5 mg q 3-4 hr</td>
<td>6 mg q 3-4 hr</td>
<td>1.5 mg q 3-4 hr</td>
<td>0.06 mg/kg q 3-4 hr</td>
<td>0.015 mg/kg q 3-4 hr</td>
</tr>
<tr>
<td>Oxycodone</td>
<td>30 mg q 3-4 hr</td>
<td>Not available</td>
<td>10 mg q 3-4 hr</td>
<td>Not available</td>
<td>0.15-0.2 mg/kg q 3-4 hr</td>
<td>Not available</td>
</tr>
</tbody>
</table>

**Opioid Agonist – Antagonist and Partial Antagonist**

| Nalbuphine (Nubain) | Not available | Not available | Not available | Not available | Not available | 0.1 mg/kg q 3-6 hr |

Note: Tables vary in the suggested doses that are equianalgesic to morphine. Clinical response is the criterion that must be applied for each patient; Titration to clinical response is necessary. Because there is not complete cross-tolerance among these drugs, it is usually necessary to use a lower than equianalgesic dose when changing drugs and to retitrate to response.

Caution: Recommended doses do not apply to patients with renal or hepatic insufficiency or other conditions affecting drug metabolism and kinetics.

<sup>1</sup> Caution: Doses listed for patients with body weight less than 50 kg cannot be used as initial starting doses in babies less than 6 months of age. Consult *Clinical Practice Guideline for Acute Pain Management: Operative or Medical Procedures and Trauma section on management of pain in neonates for recommendations*.

<sup>2</sup> For morphine (hydromorphone and oxymorphone), rectal administration is an alternate route for patients unable to take oral medications, but equianalgesic doses may differ from oral and parenteral doses because of pharmokinetic differences.

<sup>3</sup> Caution: Codeine doses above 65 mg often are not appropriate due to diminishing incremental analgesia with increasing doses, but continually increasing constipation and other side effects.

Caution: Doses of aspirin, ibuprofen and acetaminophen in combination opioid/NSAID preparations should be avoided to prevent inadvertent toxicity from the non-opiate component.

These are the American Pain Society guidelines. Individual institutions, including Seattle Children’s Hospital, may have differing practices.
Critical Elements of Care: Sickle Cell Disease

**Anemia Algorithm**

**Signs/Symptoms**
1. Increased pallor
2. Lethargy
3. Poor appetite
4. Shortness of breath
5. Jaundice

**Evaluation**
1. PE
2. CBC, retic, stool, guaiac
3. Measure spleen size
4. Consider CXR
5. UA

Compare labs to patient’s baseline; fetal Hb and HU can increase MCV, alpha and β thalassemia can decrease MCV

**Hct < 18**
- Transfuse unless clinical reasons not to

**Hct ≥ 18**

-↑ MCV
-↓ Retic
-↓ HCT

-Consider folate deficiency, check folate, Vit B 12 levels
-Replete as indicated

-↓ Erythropoiesis

-↓ Retic below baseline

-↓ Retic below baseline

-↓ Retic below baseline +/− ↓ platelets

-Consider serial Hct/Retic
-Assess need for transfusion

-↓ Hct

-Consider serial monitoring Hct/
-Retic
-Consider need for transfusion

-↓ Hct

-↑ MCV
-↓ Retic
-↓ HCT

-Consider iron deficiency
-Evaluate Fe profile TIBC, Fe, % saturation

-↑ MCV
-↓ HCT

- ↑ Retic
- ↑ MCV
- ↓ HCT

- ↑ Retic
- ↓ MCV
- ↓ HCT

**Hct > 6% drop**
- MCV at baseline
- Retic at baseline or increased

- ↑ Hct/Ret

- ↓ Retic below baseline

- Aplastic crisis

- ↓ HCT

- ↑ MCV
- ↓ Retic
- ↓ HCT

**Hct > 6% drop**
- MCV at baseline
- Retic at baseline or increased

- ↑ Hct/Ret

- ↓ Retic below baseline

- Aplastic crisis

- ↓ HCT

- ↑ MCV
- ↓ Retic
- ↓ HCT

**Spleen size increased**
- +/− ↓ platelets

- ↑ Retic
- ↓ MCV
- ↓ HCT

- Consider serial monitoring Hct/
-Retic
- Consider need for transfusion

- ↑ Retic
- ↓ MCV
- ↓ HCT

**Spleen size increased**
- +/− ↓ platelets

- ↑ Retic
- ↓ MCV
- ↓ HCT

- Consider serial monitoring Hct/
-Retic
- Consider need for transfusion

- ↑ Retic
- ↓ MCV
- ↓ HCT

**Spleen size increased**
- +/− ↓ platelets

- ↑ Retic
- ↓ MCV
- ↓ HCT

- Consider serial monitoring Hct/
-Retic
- Consider need for transfusion
Fever and Sepsis Algorithm

**Signs/Symptoms**
1. Temp 38.3°C (101.0°F) or greater, or a temp greater than 38.0°C (100.4°F) for more than 8 hours
2. Poor appetite
3. Fussy
4. Lethargic

**Work-up**
1. Physical exam, vital signs, evidence of systemic or localized infection, cardiopulmonary assessment, spleen size and neurologic exam
2. Blood culture
3. CBC, retic count, platelets
4. Low threshold for a chest X-ray
5. Culture other bodily fluids as clinically indicated

**A. Obvious infection; or**

**B. Ill-appearing; or**

**C. WBC > 30,000 or < 5,000**

**D. T > 39°C (102.2°F)**

**E. Age < 6 mos. with HbSS or Sβ0-thalassemia**

**Concerns about family ability to follow up**

**Hospitalization for IV antibiotics-broad spectrum**

**Treat 48 hours pending (-) culture**

**If culture (+), treat as per organism**

**If afebrile and cultures are negative for 24 hrs, consider discharge after 2nd dose of ceftriaxone, resume oral penicillin if applicable, and follow up if fever returns**

**If persistently febrile with T < 39.0°C (102°F), clinically well and cultures remain (-) after 48 hrs, consider discharge on PO antibiotics until afebrile for 24 hrs with follow-up evaluation every 3-4 days**

**If culture (-), well-appearing, T > 38.3°C (101°F) and T < 39°C (102.2°F)**

**Consider PO ABX with follow-up every 3-4 days**

**If culture (+), ill-appearing, or T > 39°C (102.2°F)**

**Hospitalization**

**If culture (-) and T < 38.3°C (101°F)**

**Discontinue ABX if no obvious infection; resume PCN, prophylaxis if appropriate**

**A. Not Ill-appearing**

**B. Lab evaluation lacks high-risk features**

**Parenteral (IM or IV) ceftriaxone 75 mg/kg (2 gm maximum)**

**Repeat vital signs and assessment 30 mins after parenteral ceftriaxone. If stable, discharge to home; follow up in 18-24 hrs for second dose of ceftriaxone.**

**Repeat vital signs**

**At re-evaluation 18-24 hours after the initial ceftriaxone dose**

**V. ALGORITHMS AND COMPLICATION-SPECIFIC GUIDELINES**
### Acute Chest Syndrome

**Definition:** A new infiltrate on CXR in a patient with sickle cell disease.

**Diagnostic Monitoring (if not previously obtained):**
1. CBC, diff., platelet count and reticulocyte count initially and daily until improving (compare with patient’s baseline values).
2. CXR. Repeat for clinical deterioration looking for progression. May need serial CXRs.
3. Type and cross match (minor-antigen-matched if available, sickle negative, leukocyte depleted RBC) to have blood available. Obtain red cell extended phenotyping for sickle cell patients if not done previously (at a minimum type for RhD, Cc, Ee and Kell).
4. Blood cultures if T > 38.2°C or history of recent fever.
5. Capillary or arterial blood gas and assessment by PICU team for severe illness.

**Fluids, Nutrition, General Care:**
1. Maintain “euvolemia.” IV + PO 1-1.25 x maintenance. More fluid is appropriate only if patient is dehydrated or if insensible losses are increased (e.g. persistent fever).
2. Incentive spirometry x 10 breaths q 2 hr. during the day (0800-2200), if awake at night, and prior to all CXRs.
3. Encourage ambulation, activity.

**Medications/Treatments:**
1. Oxygen to maintain O2 saturation > 93%.
2. Ceftriaxone 75mg/kg Q 24 hr. IV. (Prophylactic penicillin may be discontinued while on broad-spectrum antibiotics.)
3. Azithromycin 10mg/kg PO x1, then 5 mg/kg PO QD days 2-5, or other macrolide antibiotic.
4. Chest physical therapy if consolidation is present.
5. Follow patients specific pain plan. If not available consider morphine 0.1 mg/kg IV q 2 hr. or 0.01 - 0.1 mg/kg/hr. continuous infusion or PCA for severe pain. Alternative analgesics (but not demerol) may be used in individual cases. Adequate pain relief is essential to avoid splinting improve respiratory dynamics that worsens respiratory status.
6. Consider round the clock bronchodilators, especially if patient has history of wheezing or asthma. Some patients benefit even if not clinically obstructed.
7. Consider use of BiPAP or CPAP if not improving with routine measures.
8. Consider red cell transfusion for progressive respiratory decline despite incentive spirometry and #1-6 above. Pain control and incentive spirometry are commonly under-utilized. Transfuse initially if severely ill.
   a. Simple transfusion to a HCT of 30% (no clear benefit to exchange transfusion).
   b. Exchange transfusion for patients with progressive disease and a Hct > 27% or lack of improvement > 36 hrs post simple transfusion. Target a Hct of 30% and Hb S or Hb S + C < 30 %. (May require transfer to ICU for erythrocytapheresis). Remove femoral or central venous catheters as soon as possible after exchange transfusion to reduce risk of thrombosis.
9. See other Clinical Care Paths for acute anemic crisis, stroke, priapism, if present.
10. Continue prophylactic folic acid, if applicable.

**Discharge Criteria:**
1. Off O2.
2. Afebrile > 24 hr and negative cultures for 24 hours (if applicable).
3. Good oral intake, able to take all oral medications including antibiotics.
4. Adequate pain relief (if needed) with oral analgesics.
5. Discharge instruction completed regarding home use of incentive spirometry while on opiates.
6. Follow-up plans coordinated with hematology service. CXR to establish new baseline in 2-3 months.

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Modified from Mountain States Regional Genetic Services Network, 1996
### Stroke or Acute Neurological Event

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>Monitoring</th>
<th>Diagnostic (if not previously obtained)</th>
<th>Fluids, Nutrition, General Care</th>
<th>Medications/Treatments</th>
<th>Discharge Criteria</th>
</tr>
</thead>
<tbody>
<tr>
<td>Stroke or acute neurologic event in child with sickle cell disease</td>
<td>1. ICU admission first 24 hr and until stable</td>
<td>1. CBC, diff, platelet count, reticulocyte count initially (compare with patient’s baseline data)</td>
<td>1. IV + po 1-1.25 x maintenance</td>
<td>1. Oxygen to maintain $O_2$ saturation &gt; 93%</td>
<td>1. Clinically and neurologically stable &gt; 24 hr after transfusions</td>
</tr>
<tr>
<td></td>
<td>2. Vital signs, neuro checks q 2 hr</td>
<td>2. Coagulation profile; consider evaluation for hypercoagulability</td>
<td></td>
<td>2. Rx seizures if present</td>
<td>2. Afebrile &gt; 24 hr</td>
</tr>
<tr>
<td></td>
<td>3. CR monitor</td>
<td>3. Blood and urine cultures if febrile</td>
<td></td>
<td>3. Rx increased intracranial pressure if present</td>
<td>3. Hematology and physical therapy follow-up organized</td>
</tr>
<tr>
<td></td>
<td>4. Continuous pulse ox</td>
<td>4. Type and cross-match (minor-antigen-matched if available (at a minimum RhD, Cc, Ee and Kell), sickle-negative, leukocyte-depleted RBC) for exchange transfusion (consider erythrocytapheresis)</td>
<td></td>
<td>4. Broad-spectrum IV antibiotics if febrile</td>
<td></td>
</tr>
<tr>
<td></td>
<td>5. Record I + O, daily weight</td>
<td>5. Electrolytes initially and daily until stable</td>
<td></td>
<td>5. If applicable and not on broad-spectrum antibiotics, continue prophylactic penicillin</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>6. Emergent CT scan or MRI/MRA to exclude intracranial hemorrhage, MRI/MRA when stable if not obtained initially</td>
<td></td>
<td>6. Exchange transfusion or erythrocytapheresis to a Hct of 30% and Hb S ≤ 30%; remove femoral or central venous catheter as soon as possible to reduce risk of thrombosis</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>7. Consider CSF culture if febrile and no contraindication present</td>
<td></td>
<td>7. If exchange transfusion cannot be done acutely a simple transfusion with RBC to a Hct of 30% may be done but should be followed by and exchange transfusion when available.</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>8. Consider hypercoagulation/thrombosis evaluation</td>
<td></td>
<td>8. Quantitative HbS after transfusion or at discharge</td>
<td></td>
</tr>
</tbody>
</table>
# Priapism

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>Monitoring</th>
<th>Diagnostic (if not previously obtained)</th>
<th>Fluids, Nutrition, General Care</th>
<th>Medications/ Treatments</th>
<th>Discharge Criteria</th>
</tr>
</thead>
</table>
| Stuttering priapism: under 2 hrs at a time, may be recurrent | 1. Vital signs q 2-4 hr  
2. Record I+ O, daily weight  
3. CR monitor and continuous pulse ox if receiving parenteral narcotics | 1. CBC, diff, platelet count, reticulocyte count initially (compare with patient’s baseline data)  
2. Urinalysis and urine culture  
3. Blood culture if febrile; consider other cultures (e.g. CSF) as indicated | 1. IV fluids: 10 cc/kg over 1 hr, then IV + po = 1.25 - 1.5 x maintenance  
2. Encourage ambulation and voiding  
3. Offer warm packs or hot shower/bath if available  
4. Never use ice or cold packs  
5. Incentive spirometry - 10 breaths q 2 hr 0800-2200 and when awake if on parenteral narcotics  
6. NPO if surgical procedure planned  
7. Observe for severe headache or neurologic signs or symptoms (some suggest increased risk of stroke) | 1. Urology referral urgently if priapism persists more than 2 hrs  
2. Consider pseudoephedrine < 2 yr 4 mg/kg/day split q 6 hr po; 2-5 yr 15 mg q 6 hr po; 6-12 yr 30 mg q 6 hr po, >12 yr 60 mg po q 6 hr  
3. Strongly consider arranging aspiration and irrigation with epinephrine (1:1000,000) after 2 hrs. Notify urology within 2 hrs of onset of priapism with goal of performing procedure before 6 hours. All attempts should be made to do this within 12 hrs of onset. This can be done with local and a benzodiazepine, conscious sedation or general. Repeat procedures may be required.  
4. Pain control  
   a. Ibuprofen 10 mg/kg po q 6 hr or other anti-inflammatory agent if no gastritis, ulcer or renal impairment present  
   b. Morphine 0.05-0.1 mg/kg IV q 2 hr or 0.01-0.1 mg/kg/hr continuous infusion or PCA pump (max total dose) for severe pain  
5. Broad-spectrum IV antibiotics if febrile  
6. If applicable, continue prophylactic penicillin (if not on broad-spectrum antibiotics) and folic acid  
7. O₂ by nasal cannula if needed to keep pulse ox >93%.  
8. See other Clinical Care Paths for acute chest syndrome, acute anemic crisis, stroke, if present | 1. Priapism resolving (complete detumescence may take 1-2 wks)  
2. Taking oral fluids well and able to take po medications (e.g. prophylactic penicillin) if applicable  
3. Adequate pain relief on oral analgesics  
4. Afebrile ≥ 24 hr  
5. Adequate oxygenation on room air |
| Persistent: over 2 hrs continuously          |                                                                            |                                        |                                 |                                                                                         |                                                                                                                                                   |

Modified from Mountain States Regional Genetic Services Network, 1996
## General Anesthesia and Surgery

<table>
<thead>
<tr>
<th>Pre-Op Evaluation</th>
<th>Pre-Op Transfusion and Pulmonary Care</th>
<th>Intraoperative</th>
<th>Post-Operatives</th>
</tr>
</thead>
</table>
| • Baseline CXR, pulse ox CBC, retic, U/A | • Patient typically admitted the day before for transfusion and hydration while NPO | • Minimum 50% $O_2$ with anesthetic agent | • $O_2$ by nasal cannula at 2 L/min and continuous pulse ox even if $O_2$ saturations are high. Continue O/N and assess the next day. Maintain saturations >93%.
| • Consider pulmonary function tests and/or ECHO for patients with prior history of acute chest syndrome, suspicion of chronic lung disease or decreased exercise performance | • Simple transfusion targeting a Hct of 30-33% should be strongly considered for all children with Hb SS or Sβ0-thalassemia prior to any procedure requiring general anesthesia | • Avoid hypoxia (continuous pulse ox), hypercarbia, or hyperventilation | • Document $O_2$ saturations on room air intermittently to screen for increasing $O_2$ need.
| • Coordination of peri-operative plan with Hematology, Surgery and Anesthesia | • Surgery without pre-op transfusion in children with HbS/S and Sβ+-thalassemia may be considered in selected cases non- or minimally-invasive procedures (e.g. PE tubes or MRI/MRA). Note: tonsillectomy and/or adenoidectomy is not considered a minor procedure. Recommendations for patients with Hb S/C and Sβ+-thalassemia vary. In general, transfusion is not required for smaller procedures such as tonsillectomy and/or adenoidectomy, but transfusion is required for abdominal surgery. Due to a high baseline HCT, these patients often require exchange transfusion. | • Avoid tourniquets | • Encourage early ambulation, activity.
| | • Use antigen-matched if available, sickle-negative, leukocyte-depleted PRBC (at a minimum RhD, Cc, Ee and Kell) | | • IV + po 1-1.25 x maintenance. Avoid excessive hydration, which may precipitate acute chest syndrome.
| | • Practice incentive spirometry or developmentally appropriate substitute (e.g. bubbles) | • If history of obstructive disease, start steroid inhaler 3 days before and scheduled albuterol the night before surgery | • Strict adherence incentive spirometry: 10 breaths q 2 hr while awake. Use of pain medication before this may be useful.
| | • If history of obstructive disease, start steroid inhaler 3 days before and scheduled albuterol the night before surgery | • Avoid hypoxia (continuous pulse ox), hypercarbia, or hyperventilation | | • IV hydration 1-1.25 x maintenance while NPO before procedure. Hold while receiving blood transfusion.

 Modified from Mountain States Regional Genetic Services Network, 1996
References and Resources

General References


VI. REFERENCES AND RESOURCES


Critical Elements of Care: Sickle Cell Disease

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RESOURCES

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Texas Department of Health, Newborn Screening Program. Information on sickle cell trait, sickle β-thalassemia, hemoglobin C disease, pain in children and various complications. www.dshs.state.tx.us/newborn/sickle.shtm


Things to Know About Sickle Cell Trait. Texas State Department of Health. www.dshs.state.tx.us/newborn/sctrait.shtm

Newborn Screening Information


Regional and National Foundations

American Sickle Cell Anemia Association. www.ascaa.org


Northwest Sickle Cell Collaborative. Resources, information, support and community events. For families, kids and health care professionals. www.nwsicklecell.org/

Parent to Parent (P2P). Support programs of Washington State that provides emotional support and information to families of children with special needs and/or disabilities. www.arcwa.org/parent_to_parent.htm

Sickle Cell Disease Association of America, Inc. www.sicklecelldisease.org

Starbright Foundation. Starlight Children’s Foundation has been dedicated to improving the quality of life for children with chronic and life-threatening illnesses and life-altering injuries by providing entertainment, education and family activities that help them cope with the pain, fear and isolation of prolonged illness. www.starbright.org/Default.aspx?id=2147483750&terms=sickle+cell