

# Sickle Cell Disease

---

*CRITICAL ELEMENTS  
OF CARE*

---

**Produced by  
The Center for Children with Special Health Needs  
Children's Hospital and Regional Medical Center, Seattle, WA**

Fourth Edition, 6/2006

---

The *Critical Elements of Care* (CEC) consider 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 to assist the Primary Care Provider 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 state-wide representation involving primary and tertiary care providers, family members and a representative from a Health Plan.

Original Consensus Team:

B.J. Bryson, MSW

Kyna Byerly

Phyllis Coolen

Carol Flanagan, PNP

Kathy Mullin, RN

Daniel Niebrugge, MD

Maureen Pomietto, CNS

Jean Popalisky, RN, MN

Belinda Sandidge

Terri Stewart, PHN

Stephen Sulzbacher, Ph.D.

George Tanbara, MD

Mark Walters, MD

Content reviewed and updated 6/06:

M. Bender, MD, Ph.D.

Ruth White, MN, ARNP

Trinna E. Bloomquist, BSN

This document is also available on the Center for Children with Special Needs website at:

[www.cshcn.org](http://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 judgement or individualized consultation with the responsible provider regarding patient care needs.

---

# TABLE OF CONTENTS

## SICKLE CELL DISEASE CRITICAL ELEMENTS OF CARE

### I. OVERVIEW

- Definition of Sickle Cell Disease . . . . . 1
- Diagnostic Testing for the Common Sickle Cell Syndromes . . . . . 2

### II. BASIC TENETS OF HEMOGLOBINOPATHY FOLLOW-UP

- Hemoglobinopathy Follow-Up Program . . . . . 3

### III. GUIDELINES FOR CARE OF CHILDREN WITH SICKLE CELL

- Definition of Levels of Care . . . . . 4
- Clinic Requirements . . . . . 5
- Age-Specific Activities . . . . . 7
- Sickle Cell Disease Flow Sheet . . . . . 14

### IV. GUIDELINES FOR PAIN MANAGEMENT

- Pain Related to Sickle Cell Disease . . . . . 17
- General Principles of Pain Management . . . . . 17
- Complication-Specific Guidelines: Vaso-Occlusive Pain . . . . . 20
- Pain Episode Algorithm . . . . . 21
- ER Management: Pain Assessment . . . . . 22
- Guide to Determination of Schedule for Maintenance Analgesia in the Hospital . . . . . 23
- Pain Assessment Tools:
  - Assessment Tool 1: The Oucher . . . . . 24
  - Assessment Tool 2: Pain Intensity Number Scale . . . . . 24
  - Assessment Tool 3: Work Graphic Rating Scale . . . . . 24
- Pain Management References:
  - Table 1: Research Dosage Guidelines, Dosing Data for NSAIDS . . . . . 25
  - Table 2: Research Dosage Guidelines, Dosing Data for Opioid Analgesics . . . . . 26

---

# TABLE OF CONTENTS (cont.)

## SICKLE CELL DISEASE CRITICAL ELEMENTS OF CARE

### V. APPENDICES

I. Psychosocial Aspects of Sickle Cell Disease . . . . .	27
II. Sickle Cell Disease Algorithms and Complication-Specific Guidelines	
• Anemia Algorithm . . . . .	29
• Sepsis Algorithm . . . . .	30
• Acute Chest Syndrome . . . . .	31
• Stroke or Acute Neurologic Event . . . . .	32
• Priapism . . . . .	33
• General Anesthesia and Surgery . . . . .	34
III. References and Resources	
• Patient Education Materials Sources . . . . .	35
• Counseling References for Parents of Newborns . . . . .	35
• General References . . . . .	36



## Diagnostic Testing for the Common Sickle Cell Syndromes\*

Syndrome	Genotype	Neonatal Screening <sup>2</sup>	Hb A (%)	Hb S (%)	Hb F (%)	Hb A <sub>2</sub> (%)	Hb C (%)	Solubility Test <sup>6</sup>
Sickle cell anemia (HbSS)	S-S	FS	0	80-95	2-25 <sup>4</sup>	<3.50	0	Pos
Sickle $\beta^0$ - thalassemia <sup>1</sup>	S- $\beta^0$	FS	0	80-92	2-15	3.5-7.0	0	Pos
Sickle-hemoglobin C disease (Hb SC)	S-C	FSC	0	45-50	1-5	NA <sup>5</sup>	45-50	Pos
Sickle $\beta^+$ - thalassemia <sup>1</sup>	S- $\beta^+$	FSA or FS <sup>3</sup>	5-30	65-90	2-10	3.5-6.0	0	Pos
Sickle cell trait	A-S	FAS	50-60	35-45	<2	<3.5	0	Pos
Normal	A-A	FA	95-98	0	<2	<3.5	0	Neg

<sup>1</sup>  $\beta^0$  indicates thalassemia mutation with absent production of  $\beta$ -globin;  $\beta^+$  indicates thalassemia mutation with reduced (but not absent) production of  $\beta$ -globin.

<sup>2</sup> 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 Hb electrophoresis.

<sup>3</sup> Quantity of Hb A at birth sometimes insufficient for detection.

<sup>4</sup> 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 benign disorder not usually associated with significant anemia or vaso-occlusion. In such cases, family studies and laboratory tests to evaluate the distribution of Hb F among red cells may be helpful.

<sup>5</sup> Quantity of Hb A<sub>2</sub> cannot be measured in presence of Hb C.

<sup>6</sup> Note that this test does not distinguish sickle cell trait from sickle cell disease syndromes. High fetal hemoglobin level (as in HPFH) can lead to false negatives. False negative results occur during infancy in all sickle syndromes.

\* Modified with permission from Lane PA: Sickle cell disease. *Pediatr Clin North Am* 1996; 43:639-64.

---

# II. BASIC TENETS OF HEMOGLOBINOPATHY FOLLOW-UP

## The Basic Tenets of the Hemoglobinopathy Follow-Up Program are as Follows:

- Every child with sickle cell disease should have a source of primary medical care.
  - Wherever possible, well-child care should follow the normal guidelines of the American Academy of Pediatrics. 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 34).
  - 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 lead to immediate definitive testing by the primary care physician through qualified diagnostic laboratories.
  - When clinically significant hemoglobinopathies are confirmed, the primary care provider should consider referral to consultative care. Consultative care should be established in the first 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 her/his on-call arrangements. Tertiary level consultation should be available 24 hours a day to physicians.
  - To ensure access to care, other state agencies should assist the family in identifying financial and other resources.

- 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.

---

# 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-six months through six 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 four to six 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 treatment of sickle disease.

• Visits listed with the primary care provider  
• correspond to the current American Academy of  
• Pediatrics well-child guidelines, with the addition of  
• visits at 15 months, four years, and annually (instead of  
• semiannually) from age five years.

• Refer to pages 7-12 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 psychologic 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*.

### 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 versed in care of sickle cell patients.
- Participation of nursing staff with expertise in sickle cell issues. Nursing staff must have the skill and time available to provide educational support, perform phone triage, coordinate delivery of services with social services, and provide regular family outreach to ensure that families consistently receive care.
- Availability of vaccines specific to the infection risks of sickling diseases.
- Availability of social services to coordinate delivery of health care services and provide basic counseling.
- Access to nutrition services.
- Access to dental care with referral ability to those experienced in issues of infection and anesthesia specific to sickling diseases.
- Knowledge of community and family support resources for families of children with sickling diseases.

#### C. Complications

- Health care staff with experience and resources capable of identifying early signs of and providing initial treatment for acute and chronic 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, and sickle lung disease.
- Proximity of secondary 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 associated with sickling diseases.
- Availability of specialized pain management services, as well as availability of referral services for drug addictions.
- Transition strategy for patients transferring from pediatric care to adult care services.
- Access to academic and vocational counseling services.

#### D. Adolescent and Adult Care

- Birth control counseling and management.
- Reproductive counseling and expertise in managing sickle cell patients through pregnancy and delivery.
- Understanding of the natural history of sickle cell anemia and development of approaches to monitor patients for chronic organ failure.

E. Access and Availability

- Patient access to expert physician 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 (MRI/angiography, SPECT, etc.) 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.

B. Ambulatory Care

- Same as secondary care, but social work and nutrition should have time dedicated to the clinic.

C. Complications

- Same as secondary care, but should be able 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.

D. Adolescent and Adult Care

- Same as secondary level.

E. Access and Availability

- Same as secondary level.

## Two-to-Four-Week Check by PRIMARY CARE PROVIDER

- Conduct usual two-week, well-child care.
- Review results of second 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 after consultation or referral to a pediatric hematologist (a current listing is provided with the newborn screening program notification letter).
- 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. They should be taught to call the primary physician 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.

## Six-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.** The 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 four to six weeks of age in patients with SS and S $\beta^0$  Thalassemia—Penicillin 125 mg BID until age three years, and 250 mg BID from age three to age six 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, S $\beta^+$  Thalassemia) is lower and penicillin for these patients may not be indicated. Erythromycin (20mg/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.
- Administer second hepatitis B vaccine.
- 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.

### **Two-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 DTAP, IPV, approved H. influenza conjugate vaccine (HIB), Hep B #1 or 2, and pneumococcal conjugate vaccine (PCV).

### **Three-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 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 and tiring.

- 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.
- 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.

### **Four-Month Check by PRIMARY CARE PROVIDER**

- Perform routine well-child care.
- Give DTAP, IPV, approved H. influenza conjugate vaccine (HIB), Hep B #2 or 3, and pneumococcal conjugate vaccine (PCV).
- Reinforce teaching about fever, splenic size, fluids, antibiotics, folic acid and pain therapy.
- Introduce coping strategies for blood draws and other invasive procedures.

### **Five-Month Check by COMPREHENSIVE PROGRAM/Teaching Goals for Age**

- Perform physical exam.
- Reinforce earlier teaching.
- 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 blood gases or oximetry would be done. Oxygen should be administered, and simple or exchange transfusion provided in acute chest syndrome. Until infection is ruled out, empiric antibiotic therapy is usually warranted. Consider including antibiotic coverage for chlamydia and mycoplasma infection.

**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. 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.

- Collect CBC, diff, platelets and retic count. The child's normal levels should be established by serial testing.

### **Six-Month Check by PRIMARY CARE PROVIDER**

- Perform routine well-child care.
- Reinforce previous teaching.
- Give DTAP, third Hep B vaccine if not done, and HIB and pneumococcal conjugate vaccine (PCV).
- Adjust folic acid dose to 0.25 mg QD.

### **Eight-to-Nine-Month Check by COMPREHENSIVE/PRIMARY CARE PROGRAM/ Teaching Goals for Age**

- Review and discuss prior teaching.
- Physical exam.
- CBC, diff, plt and retic.
- Social service re-evaluation.
- Nurse review care plan with family.
- Influenza booster (initial two-dose vaccine during early first winter).

Note that the eight- to nine-month visit (and subsequent tri-monthly visits through six 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.
- Quantitate hemoglobins; e.g., HbS, A, A<sub>2</sub>, F, C, evaluate for thalassemia in an approved diagnostic laboratory.
- Tuberculin test.
- Adjust folic acid dose to 0.5 mg QD.
- Perform blood typing, including all minor blood groups.
- Introduce priapism.
- Confirm that genetic counseling occurred, and review.
- Nutrition counseling.
- Nurse review care plan with family.
- IPV, MMR
- Annually in the fall, give booster influenza vaccine.

### **14-to-15 Month Check by COMPREHENSIVE/PRIMARY CARE PROGRAM/Teaching Goals for Age**

- Routine well-child care.
- Review past teaching and examination.
- Social service case review.
- HIB, PCV #4, DTAP
- Nurse review care plan with family.

### **17-to-18 Month Check by COMPREHENSIVE/PRIMARY CARE PROGRAM/Teaching Goals for Age**

- Routine well-child care.
- Varicella (optional after age 12 months)
- Review past teaching and examination.
- Nurse review care plan with family.
- Distribue 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 Sβ° patients)

### **24-Month Check by PRIMARY CARE PROVIDER**

- Routine well-child care, review previous teaching.
- Pneumovax™, meningococcal, Hepatitis A (optional)
- Adjust penicillin dose to 250 mg BID.
- Adjust folic acid to 1mg QD.
- CBC, diff, plt, retic, BUN, Cr, Bili, Alk Phos, ALT, Iron Studies.
- Consider discussion of 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 S $\beta^{\circ}$ -thalassemia, and some patients with S $\beta^{+}$  thalassemia (should be done at a tertiary care facility by personnel trained to study patients with hemoglobinopathies).
- 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.
- Review status of new potential treatments and interventions.
- Hep A #2

### **3-and-4-Year Check by PRIMARY CARE PROVIDER**

- Routine well-child care.
- BP, UA with all subsequent annual visits.
- 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.
- Pneumovax<sup>TM</sup> booster 1-2 years after initial vaccination.
- Assess and teach self-care skills.
- Developmental assessment.

### **5-Year Check by PRIMARY CARE PROVIDER**

- Routine well-child care.
- DTAP, IPV, Pneumovax

### **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, UA.
- Social service PRN.
- Nurse review care plan with family.
- Promote self-care, reinforce coping strategies.
- Initiate school outreach and provide schools with resources about sickle cell disease.
- Continue Transcranial Doppler Study yearly for patients with SS or S $\beta^{\circ}$ -thalassemia and some patients with S $\beta^{+}$  thalassemia (should be done at a tertiary care facility by personnel trained to study patients with hemoglobinopathies).
- Review status of new potential treatments and interventions.
- Assess and teach self-care skills.
- Developmental and neuropsychologic assessment.

### **Annual Check by PRIMARY CARE PROVIDER**

- Routine well-child care.
- Pneumovax™ every three to five years.
- dT booster age 15 years.
- MMR booster after age five years.
- Discontinue penicillin prophylaxis at age six 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.
- Abdominal ultrasound for gall bladder stones, as needed for symptoms, and every other year routinely.
- 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.
- Neuropsychologic evaluation q 2-3 years.
- Screen for depression.
- Pulmonary function tests, CXR, O2 saturation, TCD, ophthalmology and dental evaluations yearly.
- EKG every other year.
- ECHOCardiogram including documentation of tricuspid regurgitation jet velocity yearly for all patients with a history of multiple pneumonias, acute chest syndrome or restrictive lung disease.
- Repeat pneumococcal and meningococcal immunization q 3-5 years.

Chronic transfusion programs 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. Liver biopsy to assess for portal fibrosis, chronic hepatitis, and iron content on a regular basis may be indicated. 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.
- Abdominal ultrasound for gall bladder stones, as needed for symptoms, and every other year routinely.
- Review yearly studies.
- 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
- Begin to develop a plan for transition to adult care.
- Discuss birth control options.
- Neuropsychologic evaluation q 2-3 years.
- Screen for depression.
- Pulmonary function tests, CXR, O2 saturation, TCD, ophthalmology and dental evaluations yearly.
- EKG every other year.
- ECHOCardiogram including documentation of tricuspid regurgitation jet velocity every two years and yearly for all patients with a history of multiple pneumonias, acute chest syndrome or restrictive lung disease.
- Repeat pneumococcal and meningococcal immunization q 3-5 years.

Name: \_\_\_\_\_ Diagnosis: \_\_\_\_\_ Birth Date: \_\_\_\_\_  
 Phone: \_\_\_\_\_ Hospital Phone: \_\_\_\_\_  
 Primary Care Provider: \_\_\_\_\_ Comprehensive Care Clinic/Provider: \_\_\_\_\_

Date	Visit	Topic	Medications	Labs	Immunizations
_____	2-4wks./primary care (following second state newborn screen)	<input type="checkbox"/> review screening results <input type="checkbox"/> review well-child care/24-hr. access <input type="checkbox"/> referrals: PHN, WIC, genetic counselor <input type="checkbox"/> emphasize fever and hydration issues	<input type="checkbox"/> start PCNVK 125mg BID (SS & Sβ <sup>0</sup> thalassemia) <input type="checkbox"/> start folate: 0.1 mg/d	<input type="checkbox"/> diagnostic specimen	<input type="checkbox"/> HepB. vaccine
_____	4-6wks./compr. care	<input type="checkbox"/> review fever, risk of sepsis <input type="checkbox"/> review splenic sequestration <input type="checkbox"/> SW eval./RN review of care plan <input type="checkbox"/> verify PHN, WIC and genetic counselor referrals <input type="checkbox"/> begin chronic illness awareness			<input type="checkbox"/> HepB. vaccine
_____	2mos./prim. care	<input type="checkbox"/> routine WCC/PE <input type="checkbox"/> reinforce PCN proph., Folate admin., fever, fluids <input type="checkbox"/> review splenic sequest. crisis			<input type="checkbox"/> Hib, DTAP, IPV, HepB. #1 or 2, PCV.
_____	3mos./compr. care	<input type="checkbox"/> PE, review fever, fluids, PCN, splenic sequest. <input type="checkbox"/> introduce mechanism of sickling, dactylitis, pain episodes, avoidance of temperature extremes <input type="checkbox"/> dietary/nutrition counseling <input type="checkbox"/> SW update/RN review of care plan <input type="checkbox"/> review health care access, interaction with ER			<input type="checkbox"/> Influenza for household**
_____	4mos./prim. care	<input type="checkbox"/> PE, WCC <input type="checkbox"/> reinforce fever, fluids, other issues <input type="checkbox"/> PCN, splenic sequest., etc. <input type="checkbox"/> introduce acute chest syndrome			<input type="checkbox"/> Hib, DTAP, IPV, HepB #2 or 3, PCV
_____	6mos./compr. care	<input type="checkbox"/> PE, WCC <input type="checkbox"/> reinforce previous teaching <input type="checkbox"/> Inform parents of resources to store cord blood cells from newborn full siblings of patients <input type="checkbox"/> introduce neurologic complications	<input type="checkbox"/> ↑ folate to 0.25mg Qd		<input type="checkbox"/> DTAP, Hib, PCV <input type="checkbox"/> HepB vaccine* if not completed
_____	8-9mos./prim. care	<input type="checkbox"/> review previous teaching <input type="checkbox"/> PE <input type="checkbox"/> SW update/RN review of care plan			<input type="checkbox"/> Influenza**
_____	11-12mos./compr. care	<input type="checkbox"/> PE, WCC <input type="checkbox"/> review previous teaching <input type="checkbox"/> confirm genetic counseling occurred <input type="checkbox"/> nutrition counseling/RN review	<input type="checkbox"/> ↑ folate to 0.50 mg Qd	<input type="checkbox"/> CBC, diff., plt, retic, BUN, CR <input type="checkbox"/> Bili, Alk, phos, ALT, iron studies, LDH <input type="checkbox"/> quant. HbA, A <sub>2</sub> , F, S, C, thalassemia screen <input type="checkbox"/> TB test <input type="checkbox"/> UA	<input type="checkbox"/> IPV***, MMR

\* If unable to follow this schedule, or if the mother of the infant is HepB SAg+, please consult current approved guidelines.  
 \*\* Influenza vaccination must be addressed early winter of each year. For children under 6mo, household members should be vaccinated. Children over 6mo of age should receive the two-dose influenza vaccine the first time, then one yearly after.  
 \*\*\* Optional

Name: \_\_\_\_\_ Diagnosis: \_\_\_\_\_

<b>Date</b>	<b>Visit</b>	<b>Topic</b>	<b>Medications</b>	<b>Labs</b>	<b>Immunizations</b>
_____	14-15mos./prim.care	<input type="checkbox"/> PE, WCC, review previous teaching <input type="checkbox"/> SW review/RN review			<input type="checkbox"/> Hib, PCV#4, DTAP
_____	17-18mos./compr.care	<input type="checkbox"/> PE, WCC, review previous teaching <input type="checkbox"/> RN review <input type="checkbox"/> distribute pain questionnaire <input type="checkbox"/> initiate development of individualized treatment plan			<input type="checkbox"/> Varicella***
_____	21mos./prim.care	<input type="checkbox"/> PE, review past teaching <input type="checkbox"/> SW review/RN care review <input type="checkbox"/> discuss hyposthenuria/enuresis			
_____	24mos./compr.care	<input type="checkbox"/> PE, WCC <input type="checkbox"/> consider discussion of oral hygiene <input type="checkbox"/> introduce priapism	<input type="checkbox"/> ↑ folate to 1mg Qd	<input type="checkbox"/> CBC, diff., plt, retic, BUN, Cr, Bili, Alk Phos, AST	<input type="checkbox"/> pneumovax™ <input type="checkbox"/> meningococcal vaccine <input type="checkbox"/> HepA #1***

\*\*\*Optional.

III. Guidelines for Care of Children with Sickle Cell Disease: *Sickle Cell Disease Flow Sheet*

Name: \_\_\_\_\_ Diagnosis: \_\_\_\_\_

Date	Visit	Topic
------	-------	-------

For all patients review status of new potential treatments and interventions and inform families of resources to store cord blood cells from newborn full siblings of patients.

- |       |   |  |
|-------|---|--|
| _____ | every 3 mos. through age 6 years/compr. care or patients with Hb SS, Sβ <sup>0</sup> -thalassemia | <input type="checkbox"/> review previous teaching, including chest syndrome<br><input type="checkbox"/> PE, BP*, vision/hearing screen*<br><input type="checkbox"/> RN review of care plan   |
| _____ | every 6 mos. in patients with SC, Sβ <sup>+</sup> thalassemia                                     | <input type="checkbox"/> SW follow-up as needed<br><input type="checkbox"/> review PCN prophylaxis, neurologic event teaching<br><input type="checkbox"/> begin coping strategy teaching w/ child  |
| _____ | every 6 mos. after age 6 years/compr. care with SS, Sβ <sup>0</sup> thalassemia                   | <input type="checkbox"/> review previous teaching<br><input type="checkbox"/> PE, BP, vision/hearing screen<br><input type="checkbox"/> RN review of care plan   |
| _____ | yearly in patients with SC, Sβ <sup>+</sup> thalassemia   | <input type="checkbox"/> SW follow-up as needed<br><input type="checkbox"/> promote self-care, reinforce coping strategies   |
| _____ | yearly visit/prim. care   | <input type="checkbox"/> PE, routine well-child care   |
| _____ | 3 years/compr. care/prim. care  | <input type="checkbox"/> PE, routine well-child care<br><input type="checkbox"/> refer for routine dental care<br><input type="checkbox"/> RBC pitscore annually in patients with SC, Sβ <sup>+</sup> Thalassemia<br><input type="checkbox"/> increase PCN + 250mg BID at age 3; discontinue PCN prophylaxis at age 6  |
| _____ | 5 years/prim. care/compr. care  | <input type="checkbox"/> PE, routine well-child care<br><input type="checkbox"/> school outreach<br><input type="checkbox"/> quantitate HbF by Hgb electrophoresis   |
| _____ | pre-adolescence/adolescence/compr. care   | <input type="checkbox"/> review previous teaching<br><input type="checkbox"/> PE, BP, vision/hearing screen<br><input type="checkbox"/> discuss leg ulcers, priapism, delays in sexual maturation, sexual activity, smoking, drugs, activities, career goals as developmentally appropriate<br><input type="checkbox"/> refer for genetic counseling directed toward patient in early adolescence<br><input type="checkbox"/> initiate discussions on options and process of transitioning to an adult medical facility<br><input type="checkbox"/> discuss and screen for depression yearly |

**Yearly Labs and Studies**

- Yearly: cbc, diff., retic, plt, BUN, Cr, Alk P, SGPT (ALT), Bili, LDH, and Iron
- UA annually
- Yearly after age 2: in patients with SS and Sβ-thalassemia begin Transcranial Doppler Studies
- Yearly from 2-3 through 13 years: neuropsychologic evaluation
- Yearly after age 7 in patients with Hb SS, Sβ<sup>0</sup> thalassemia: begin yearly CXR, Oximetry or ABG, EKG
- Yearly after age 7 in patients with Sβ<sup>0</sup> thalassemia, Hb, SS, and SC: begin yearly ophthalmology exam
- Yearly from 7<sup>1/2</sup> - 13 years: screen for depression, pulmonary function tests including DICO and pleythsmography, ophthomology and dental evaluation, ECHOcardiogram including documentation of tricuspid regurgitation jet velocity yearly for all patients with a history of multiple pneumonias, acute chest syndrome or restrictive lung disease
- U/S in alternating years to assess for gallstones in all patients
- other\*\*

**Immunizations**

- |   |   |
|---|---|
| <input type="checkbox"/> annual influenza booster in the fall                       | <input type="checkbox"/> Dt booster age 15  |
| <input type="checkbox"/> pneumovax™ booster 1-2 yrs. following initial immunization | <input type="checkbox"/> MMR booster after age 5  |
| <input type="checkbox"/> HepA #2 6 months after 1st dose                            | <input type="checkbox"/> repeat pneumococcal and meningococcal immunization q 3-5 years |
| <input type="checkbox"/> DPT, IPV, pneumovax™                                       |   |

\*Check BP beginning age three years; vision/hearing screen beginning age four years.

\*\*Children on transfusion programs should have ferritin, Fe, TIBC; %Hb S monitored; annually, 24-hour Holter monitor; pituitary function testing; liver and renal functions should be tested. Consider liver biopsy to regularly assess hepatic iron content and adequacy of chelation therapy.

---

# IV. GUIDELINES FOR PAIN MANAGEMENT

## Pain Related to Sickle Cell Disease

**Severity:** Varies from mild to extremely intense and described as “body chewing,” “body biting.”

**Character:** Deep, aching, tiring, fatiguing

**Developmental Aspects:** Can occur as early as four to nine 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. Pain due to swelling in hands and feet from dactylitis typically occurs in children under three years. Common complaints:

- extremity pain
- abdominal pain
- back pain

**Frequency:** Sickle cell pain forms a continuum from acute to chronic

- 30 percent never or rarely have pain
- 50 percent have few episodes
- 20 percent have frequent, severe episodes (6 percent of patients account for 30 percent of all painful episodes)

**Precipitating Factors:**

- infection
- hypoxemia
- dehydration
- fatigue
- exposure to cold
- 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 Consistently Addressing Pain
- B. Health Care Professionals Have the Accountability/Responsibility for Using a Proactive, Not a Reactive Approach.

### C. Emphasize the Value of a System-Wide Approach

1. Effective pain management is contingent on involvement by administration, manager and practitioners.
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
  - c. To obtain education and support
3. Pain relief is a quality assurance/continuous quality improvement issue for children with chronic illness. Care effectiveness must be evaluated.
4. 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 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 at the Beginning

1. Computerized profiles — “recipe cards”: Gathers pain history and then child, parent(s) and health care team develop plan on the computer. This plan is modified and updated on a real time basis.
2. Life records: Eliminates the need for repeated questioning of child/parents(s), particularly as they enter different hospital areas (ER, clinic, inpatient, OR).
3. A pain problem list should be instituted so that pain stemming from the disease and its treatment can be isolated and treated appropriately.

- 4. Hand-held records: Empowers child and family.

F. Guidelines for Clinical Judgment

- 1. World Health Organization Guidelines (WHO): Basic foundation for pharmacologic management. (See Figure 1 on page 18).
- 2. General Principles of Pharmacologic Management
  - a. Severe pain is an emergency and must be treated accordingly.
  - b. Use the WHO step-wise approach to pharmacologic management.
  - 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 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.
  - o. In general, avoid continuous infusion narcotics; assess often for

respiratory compromise, as hypoxemia may contribute to episodes of acute chest syndrome. Incentive spirometry while on continuous infusions.

- 3. Complimentary Nonpharmacologic Strategies—Developmental Approaches

**Infants:**

- Explanations: Caregiver teaching
- Distractions: Music/mobiles, soothing talk, soft or a novel voice, calm demeanor, oral-motor stimulation (pacifiers, non-nutritive sucking)
- Containment: Holding/cuddling/swaddling, positioning, pacifier
- Physical: Massage (applicability/efficacy being determined)

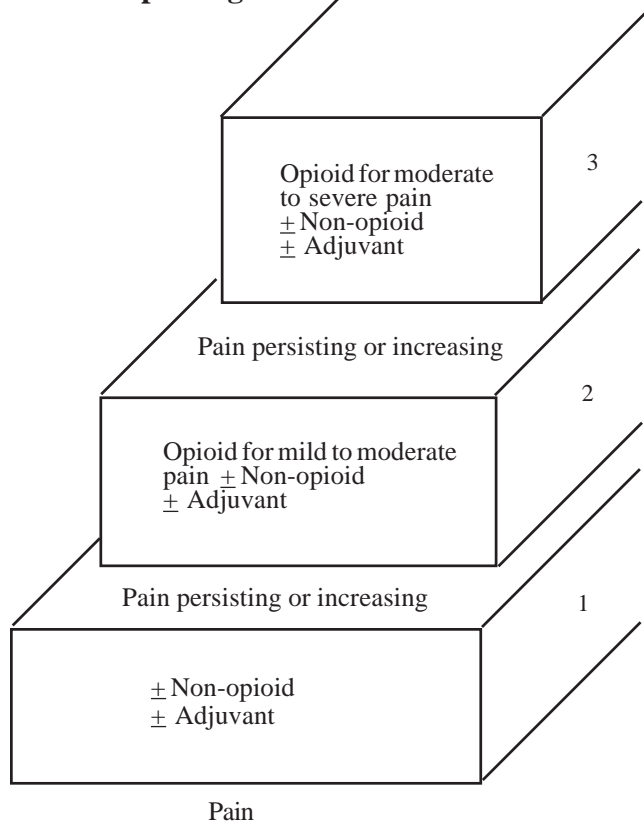
**Toddlers/Preschoolers:**

- Distraction: Pop-up books, magic circle/magic game, puppets, kaleidoscopes, counting ABCs, music-sing-along songs, squeezing on koosh ball
- Distraction with breathing: Pinwheel, blowing bubbles, “meow-woof” breathing, party blowers
- Breathing/relaxation: “Go limp as a rag doll” 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/cold, acupuncture, acupressure, Transcutaneous Electrical Nerve Stimulation (TENS)

**School-Age/Adolescents:**

- Explanations to child and family.
- Modeling/desensitization: Explanations to child and family.
- Distraction (younger): Pop-up books; counting ABC’s, puppets, kaleidoscopes, music with walkman tape player
- Imagery (older): Pain switch familiar images with stories, biofeedback

**Figure 1. The WHO (1986)  
Three-Step Analgesic Ladder**

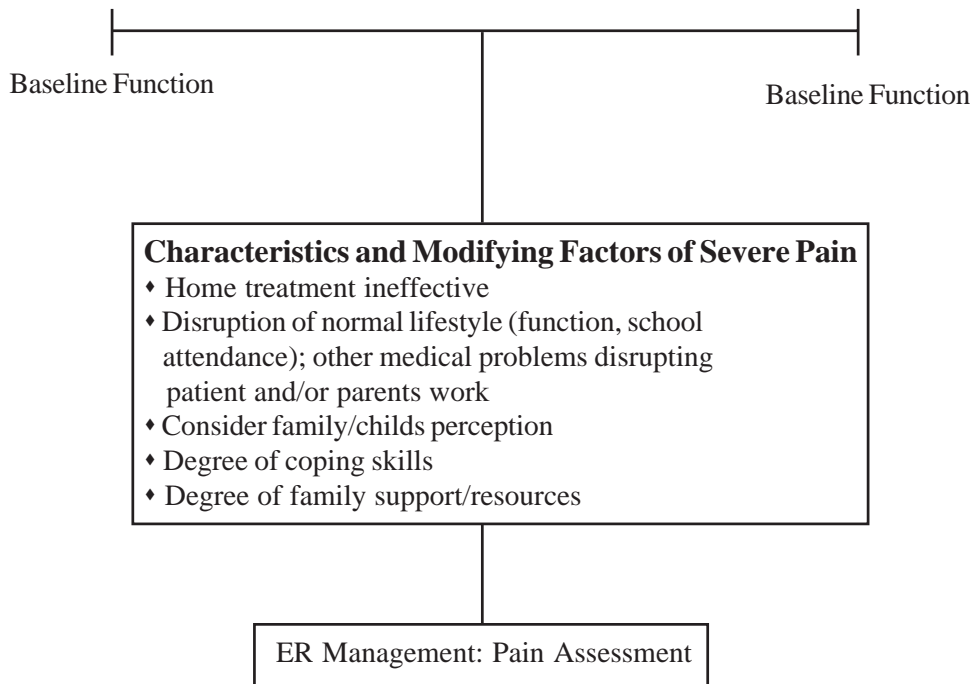
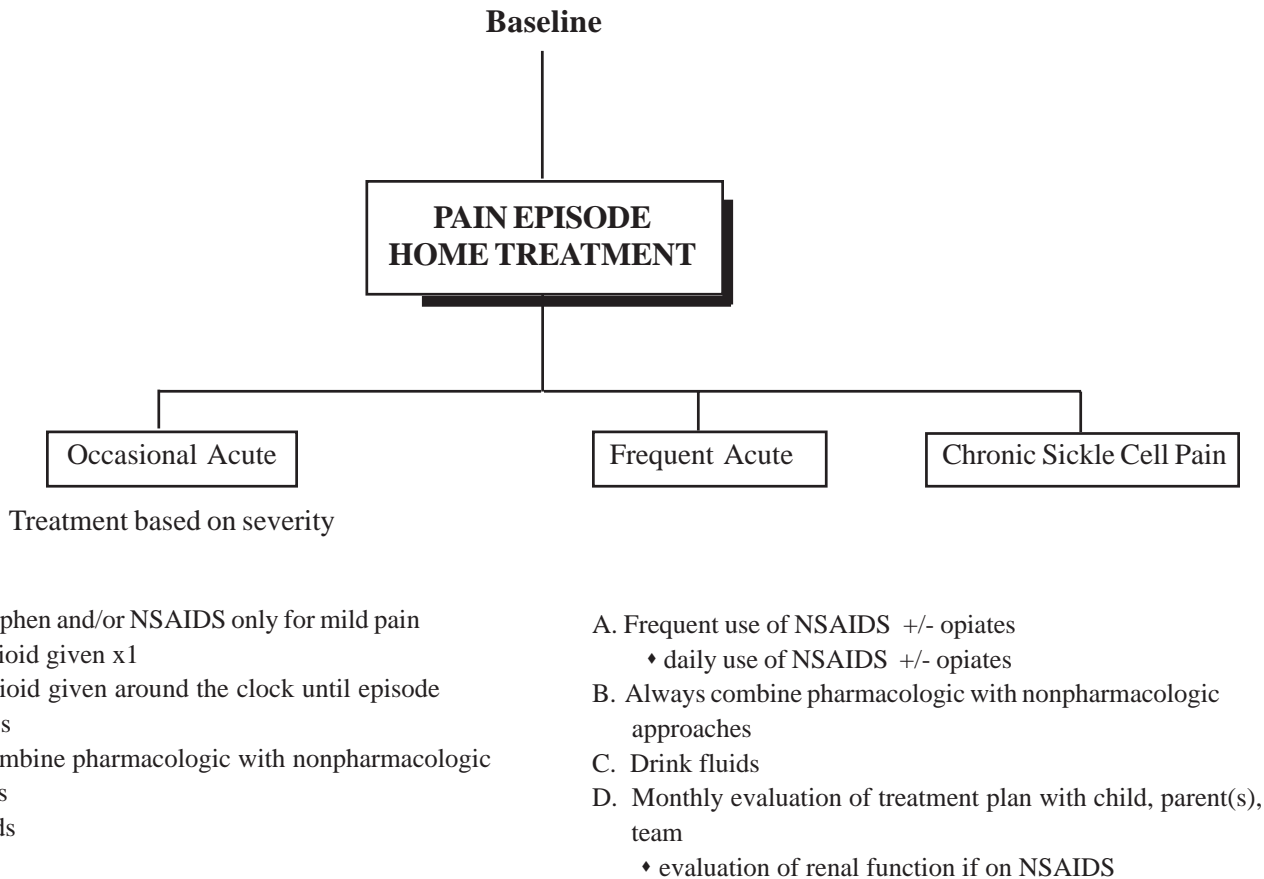


## VASO-OCCLUSIVE PAIN

Diagnosis	Monitoring	Diagnostics (if not previously obtained)	Fluids, General Care	Medication/Treatment	Discharge Criteria
<p>Vaso-occlusive pain in child with sickle cell disease.</p>	<ol style="list-style-type: none"> <li>1. Vital signs q 4 hr.</li> <li>2. Record I + O, daily weight.</li> <li>3. Continuous pulse ox. if any respiratory symptoms present, or if on parenteral narcotics.</li> <li>4. Consider CR monitor.</li> </ol>	<ol style="list-style-type: none"> <li>1. CBC, diff., plt count and retic count initially. (Compare with patient's baseline data.)</li> <li>2. CXR if cough, chest pain or any respiratory symptoms present or develop after admission.</li> <li>3. Blood culture, urinalysis, urine culture and other cultures (e.g. CSF) if febrile.</li> <li>4. Consider renal (BUN, creat) and liver (fractioned bili, ALT) function tests for very severe pain or any evidence of encephalopathy (R/O acute multi-organ failure syndrome).</li> <li>5. Consider abdominal ultrasound, liver function tests for RUQ, epigastric pain (R/O cholelithiasis, cholecystitis).</li> <li>6. Type and cross match for RBC (minor-antigen-matched if available, leukocyte-depleted, sickle-negative), if Hb is 1-2 gm/dl or more below baseline, and/or if evidence of acute chest syndrome (see acute chest syndrome protocol), or cardiovascular compromise present.</li> </ol>	<ol style="list-style-type: none"> <li>1. IV + po 1-1.5 x maintenance. Increased fluids may be needed if patient is dehydrated and/or insensible losses are increased (e.g. persistent fever). Avoid excessive fluids, which may precipitate or exacerbate acute chest syndrome.</li> <li>2. Avoid routine bolusing of IV fluids unless clinically dehydrated or otherwise clinically indicated.</li> <li>3. Incentive spirometry—10 breaths q 2 hr. when awake.</li> <li>4. Encourage ambulation and activity.</li> </ol>	<ol style="list-style-type: none"> <li>1. Cefuroxime 50 mg/kg IV q 8 hr. if febrile. (Prophylactic penicillin may be discontinued while on broad-spectrum antibiotics.)</li> <li>2. Continue prophylactic folic acid, if applicable.</li> <li>3. Ibuprofen 10 mg/kg po q 8 hr. or other anti-inflammatory agent if no contraindication (i.e. gastritis, ulcer or renal impairment).</li> <li>4. Morphine sulfate 0.05 - 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 occasionally be required but should be used with extreme caution). Alternative analgesics may be used in individual cases. In most cases, prn analgesic orders are not appropriate.</li> <li>5. Encourage incentive spirometry for all patients, but it is essential for patients with history of acute chest or on continuous infusions.</li> <li>6. Start oral opiates as soon as tolerated from a gastrointestinal standpoint, even if requiring IV opiates.</li> <li>7. O<sub>2</sub> by nasal cannula as needed to keep pulse ox <math>\geq</math>92% or <math>\geq</math> patients baseline value. Avoid excessive or unnecessary O<sub>2</sub>, which may suppress the reticulocyte count and exacerbate anemia.</li> <li>8. Consider pain team consultation.</li> <li>9. Offer heating pads or other comfort measures previously used by patient.</li> <li>10. Consider colace or laxative for narcotic-induced constipation.</li> <li>11. See other Clinical Care Paths for acute chest syndrome, acute anemia crisis, stroke, priapism, if present.</li> <li>12. Reassess pain control at least twice daily. Analgesics may be weaned as tolerated by decreasing dose, not by prolonging interval between doses. Discuss analgesic changes with patient/family.</li> </ol>	<ol style="list-style-type: none"> <li>1. Taking oral fluids well and able to take po medications (e.g. prophylactic penicillin) if applicable.</li> <li>2. Adequate pain relief on oral analgesics.</li> <li>3. Afebrile &gt;24 hr.</li> <li>4. Resolution of any pulmonary symptoms or documentation of adequate oxygenation on room air.</li> </ol>

Modified from Mountain States Regional Genetic Services Network, 1996

## PAIN EPISODE ALGORITHM



## ER MANAGEMENT: PAIN ASSESSMENT

- ♦ Region/site
- ♦ Character
- ♦ Intensity
- ♦ Quality
- ♦ Precipitating factors
- \* Determine if pain is sickle cell related (not all pain is sickle cell pain)
- ♦ What relieves/what aggravates
- ♦ Child self report/pain score
- ♦ Parent report
- ♦ Recent medication use
- ♦ Past therapy that helped

### ER Management: Immediate Treatment

- ♦ Hydration (PO or IV depending on status)
- ♦ Non-steroidal anti-inflammatory agent, or non-opioid analgesic (PO or IV depending on status)
- ♦ Administer opioid (PO or IV depending on status)
- ♦ Use established pain treatment plans

Response to initial doses of IV opioid in ER:

GOOD

- ♦ Discharge on non-opioid analgesics (Tyl, Ibu)
- ♦ Oral opioid around the clock for 24 hrs. or treatment plan

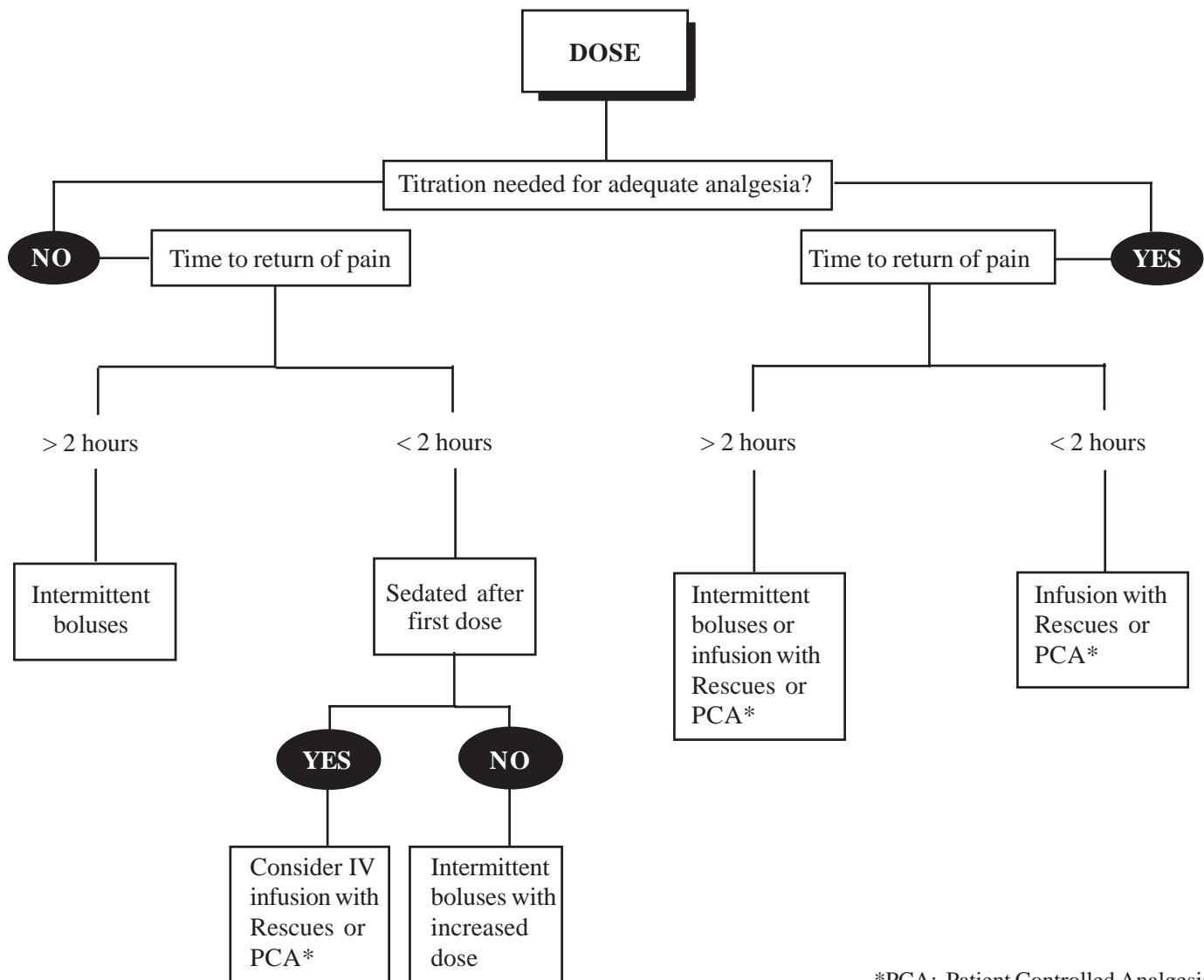
NONE or INADEQUATE

- ♦ Dose titration
- ♦ Admit to holding area or inpatient unit (see next chart)

## GUIDE TO DETERMINATION OF SCHEDULE FOR MAINTENANCE ANALGESIA IN THE HOSPITAL

(Shapiro, 1992)

(Note- This is a generic plan. Individual patient plans may vary.)



\*PCA: Patient Controlled Analgesia

### Methods for Administering Opioid Analgesia in the Hospital

- Intermittent IV doses around the clock
- IV infusion caution: infusions can result in respiratory compromise
- IV infusion with intermittent IV doses offered every two hours
- Sustained release or immediate acting agents given PO around the clock, with intermittent IV doses offered every two hours
- PCA with basal infusion
- Sustained release or immediate acting agents given PO around the clock, with PCA without basal infusion

## **THE OUCHER**

### **Assessment Tool 1**

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. Ask them to recall times they hurt in the past. 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 he/she gives is the Oucher score; if the child uses the photographic scale, the picture he/she selects is converted to the appropriate predetermined score shown on the oucher (0, 20, 40, 60, 80 or 100).

## **PAIN INTENSITY NUMBER SCALE**

### **Assessment Tool 2**

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.  
(Wilke, D.J., et. al. *The Hospice Journal*, 6(1), 1-13. *Essentials of Pain Management: A Nursing Handbook*. Optioncare: Seattle, WA.)

## **WORK GRAPHIC RATING SCALE**

### **Assessment Tool 3**

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.

---

no pain                      little pain                      medium pain                      large pain                      worst possible pain

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

**Research Dosage Guidelines**  
**TABLE 1**  
**Dosing Data for NSAIDS**

<b>Oral NSAIDS</b>	<b>Usual Adult Dose</b>	<b>Usual Pediatric Dose<sup>1</sup></b>	<b>Comments</b>
Acetaminophen	650-975 mg q 4 hr.	10-15 mg/kg q 4 hr.	Acetaminophen lacks the peripheral anti-inflammatory activity of other NSAIDS.
Aspirin	650-975 mg q 4 hr.	10-15 mg/kg q 4 hr. <sup>2</sup>	The standard against which other NSAIDS are compared. Inhibits platelet aggregation; may cause post-operative bleeding.
Cholinemagnesium Trisalicylate (Trilisate)	1000-1500 mg bid	25 mg/kg bid	May have minimal antiplatelet activity; also available as oral liquid.
Diflunisal (Dolobid)	1000 mg initial dose, followed by 500mg q 12 hr.		
Etodolac (Lodine)	200-400 mg q 4-6 hr.		
Fenoprofen calcium (Nalfon)	200 mg q 4-6 hr.		
Ibuprofen (Motrin, others)	400 mg q 4-6 hr.	10 mg/kg q 6-8 hr.	Available as several brand names and as generic; also available as oral suspension.
Ketoprofen (Orudis)	25-75 mg q 6-8 hr.		
Magnesium salicylate	650 mg q hr.		Many brands and generic forms available.
Meclofenamate sodium (Meclomen)	50 mg q 4-6 hr.		
Mefenamic acid (Ponstel)	250 mg q 6 hr.		
Naproxen (Naprosyn)	500 mg initial dose followed by 250 mg q 6-8 hr.	5 mg/kg q 12 hr.	Also available as oral liquid.
Naproxen sodium (Anaprox)	550 mg initial dose followed by 275 mg q 6-8 hr.		
Salsalate (Disalcid, others)	500 mg q 4 hr.		May have minimal antiplatelet activity.
Sodium salicylate	325-650 mg q 3-4 hr.		Available in generic form from several distributors.
<b>Parenteral NSAID</b>			
Ketorolac	30 or 60 mg IM or IV initial dose followed by 15 or 30 mg q 6 hr. Oral dose following IM or IV dosage: 10mg q 6-8 hr.		Intravascular and intramuscular dose not to exceed 3 days.

Note: Only the above NSAIDS have FDA approval for use as simple analgesics, but clinical experience has been gained with other drugs as well.

<sup>1</sup> Drug recommendations are limited to NSAIDS when pediatric dosing experience is available.

<sup>2</sup> Contraindicated in presence of fever or other evidence of viral illness.

Source: *Quick Reference Guide for Clinicians, Acute Pain Management in Infants, Children and Adolescents*: U.S. Department of Health and Human Services; Public Health Service, Agency for Health Care Policy, 1972.

**Research Dosage Guidelines**  
**TABLE 2**  
**Dosing Data for Opioid Analgesics**

<b>Opioid Agonist</b>	<b>Approx. Equianalgesic Oral Dose</b>	<b>Approx. Equianalgesic Parenteral Dose</b>	<b>Recommended Starting Dose (Adults &gt;50kg body wt.) Oral</b>	<b>Recommended Starting Dose (Adults &gt;50kg body wt.) Parenteral</b>	<b>Recommended Starting Dose (Children, adults &lt;50 kg body wt.)<sup>1</sup> Oral</b>	<b>Recommended Starting Dose (Children, adults &lt;50 kg body wt.)<sup>1</sup> Parenteral</b>
Morphine <sup>2</sup>	30 mg q 3-4 hr. (around-the-clock dosing) 60 mg q 3-4 hr. (single dose or intermittent dosing)	10 mg q 3-4 hr.	30 mg q 3-4 hr.	10 mg q 304 hr.	0.3 mg/kg q 3-4 hr.	0.1 mg/kg q 3-4 hr
Codeine <sup>3</sup>	130 mg q 3-4 hr.	75 mg q 3-4 hr.	60 mg q 3-4 hr.	60 mg q 2 hr. (intramuscular subcutaneous)	1 mg/kg q 3-4 hr. <sup>4</sup>	Not recommended
Hydromorphone <sup>2</sup> (Dilaudid)	7.5 mg q 3-4 hr.	1.5 mg q 3-4 hr.	6 mg q 3-4 hr.	1.5 mg q 3-4 hr.	0.06 mg/kg q 3-4 hr.	0.015 mg/kg q 3-4 hr.
Hydrocodone (in Lorcet, Lortab, Vicodin, others)	30 mg q 3-4 hr.	Not available	10 mg q 3-4 hr.	Not available	0.2 mg/kg q 3-4 hr. <sup>4</sup>	Not available
Levorphanol (Levo-Dromoran)	4 mg q 6-8 hr.	2 mg q 6-8 hr.	4 mg q 6-8 hr.	2 mg q 6-8 hr.	0.04 mg/kg q 6-8 hr.	0.02 mg/kg q 6-8 hr.
Meperidine (Demerol)	300 mg q 2-3 hr.	100 mg q 3 hr.	Not recommended	100 mg q 3 hr.	Not recommended	0.75 mg/kg q 2-3 hr.
Methadone (Dolophine, others)	20 mg q 6-8 hr.	10 mg q 6-8 hr.	20 mg q 6-8 hr.	10 mg q 6-8 hr.	0.2 mg/kg q 6-8 hr.	0.1 mg/kg q 6-8 hr.
Oxycodone (Roxicodone, also in Percocet, Percodan, Tylox, others)	30 mg q 304 hr.	Not available	10 mg q 3-4 hr.	Not available	0.2 mg/kg q 3-4 hr. <sup>4</sup>	Not available
Oxymorphone (Numorphan)	Not available	1 mg q 3-4 hr.	Not available	1 mg q 3-4 hr.	Not recommended	Not recommended
<b>Opioid – Antagonist and Partial Antagonist</b>						
Buprenorphine (Buprenex)	Not available	0.3-0.4 mg q 6-8 hr.	Not available	0.4 mg q 6-8 hr.	Not available	0.004 mg/kg q 6-8 hr.
Butorphanol (Stadol)	Not available	2 mg q 3-4 hr.	Not available	2 mg q 3-4 hr.	Not available	Not recommended
Nalbuphine (Nubain)	Not available	10 mg q 3-4 hr.	Not available	10 mg q 3-4 hr.	Not available	0.1 mg/kg q 3-4 hr.
Pentazocine (Talwin, others)	150 mg q 3-4 hr.	60 mg q 3-4 hr.	50 mg q 4-6 hr.	Not recommended	Not recommended	Not recommended

Note: Published 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 the *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 pharmacokinetic differences.

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

<sup>4</sup> Caution: Doses of aspirin and acetaminophen in combination opioid/NSAID preparations must also be adjusted to the patient's body weight.

## 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 must occur in an environment where the family feels comfortable. 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.

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, child care 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 loneliness. 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 the parents. 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 have migrated to religious groups like Muslim or Buddhist faiths. 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 8-12 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.

## 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 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, day dreaming, 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 school work, and s/he may not know how to ask for assistance. S/he 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 her/his feelings, so s/he 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 dealing with the school system.

We encourage families to contact the school each year and to provide information about sickle cell disease to teachers 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 and careful review of academic performance, enables the school system to become an ally of the family.

## Physical Activities

Physical exhaustion can precipitate a painful episode in children with sickle cell disease. However, children may be expected to participate in physical activities at school without 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 behavior 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 8-12 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. If possible, consults with pain management personnel 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.

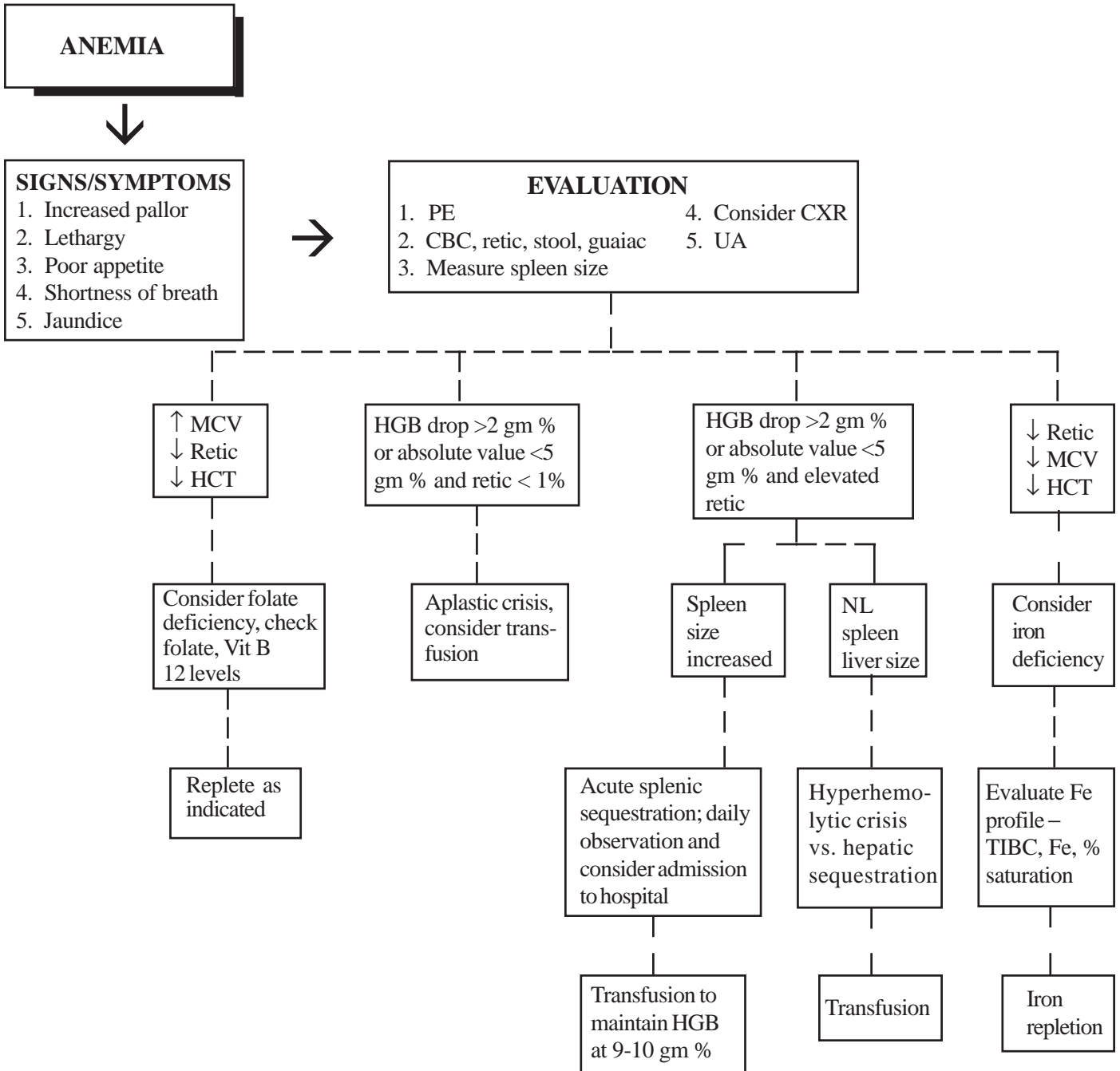
Children should be encouraged to bring school work to the hospital. Some facilities may have volunteers who can assist them, or paid staff members who fulfill this academic role. The school system may also provide tutors for students under certain conditions. Children should be encouraged to telephone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

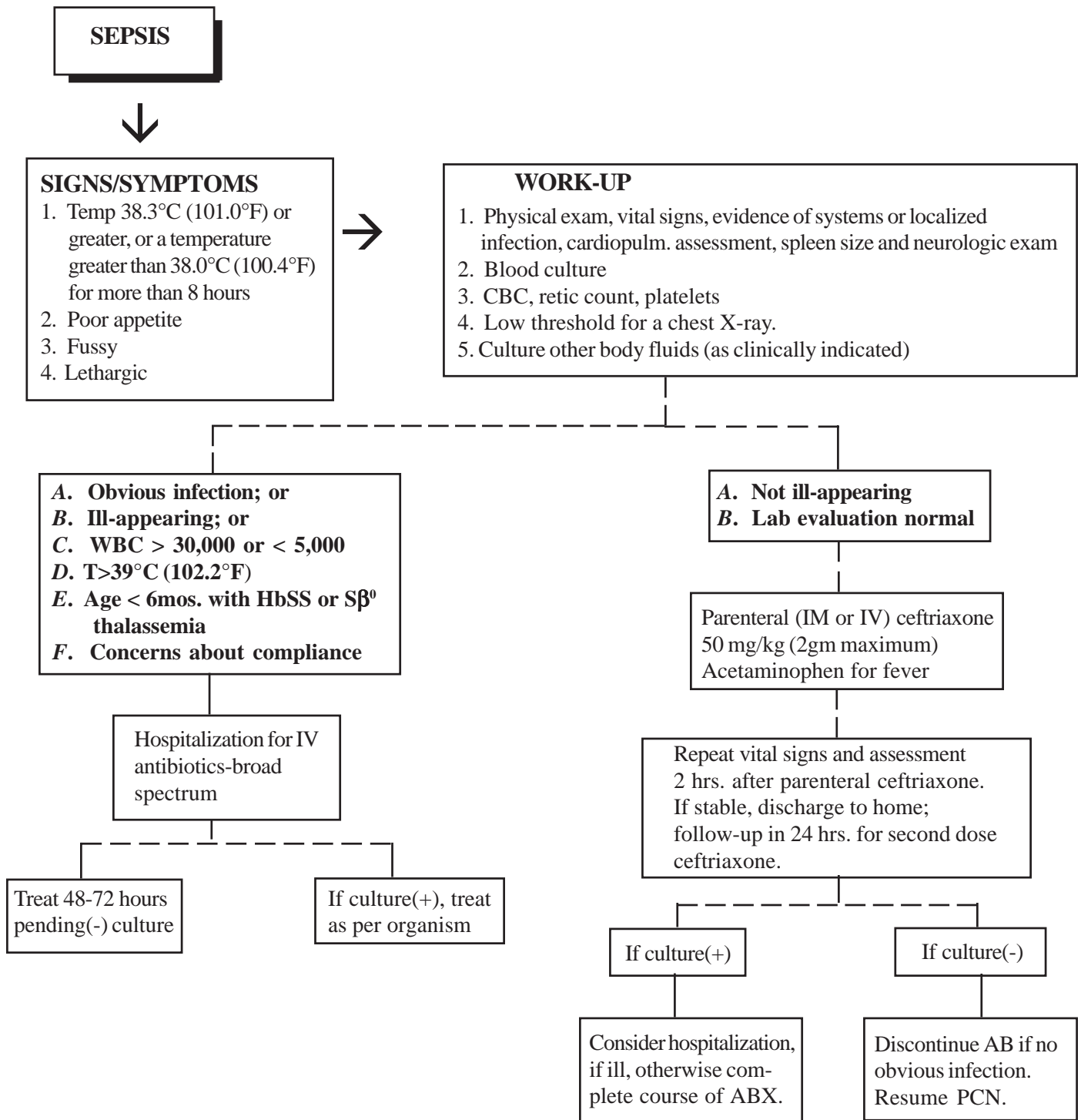
## Mortality and Sickle Cell Disease

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 percent 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.

## ANEMIA ALGORITHM



## SEPSIS ALGORITHM



## ACUTE CHEST SYNDROME

Diagnosis	Monitoring	Diagnostics (if not previously obtained)	Fluids, Nutrition, General Care	Medications/Treatments	Discharge Criteria
<p>Acute Chest Syndrome in child with sickle cell disease.</p> <p><u>Definition:</u> Any acute illness associated with lower respiratory symptoms, hypoxemia or new infiltrate on CXR.</p>	<ol style="list-style-type: none"> <li>1. Vital signs q 2-4 hr.</li> <li>2. Continuous pulse ox.</li> <li>3. Consider CR monitor.</li> <li>4. Record I + 0, daily weight.</li> </ol>	<ol style="list-style-type: none"> <li>1. CBC, diff., platelet count and reticulocyte count initially and daily until improving (compare with patient’s baseline values).</li> <li>2. CXR initially, repeat for clinical deterioration - may need serial CXRs looking for progression.</li> <li>3. Consider:                             <ol style="list-style-type: none"> <li>a) type and cross match (minor-antigen-matched if available, sickle-negative, leukocyte-depleted RBC) for severe illness or if Hb &gt; 1 gm/dl below baseline.</li> <li>b) blood cultures if febrile or history of recent fever.</li> <li>c) ABG for severe illness.</li> <li>d) renal (BUN, creat) and liver (fractioned bili, ALT) function tests for severe illness or if diffuse encephalopathy present (R/O acute multiorgan failure syndrome).</li> </ol> </li> </ol>	<ol style="list-style-type: none"> <li>1. Maintain “euvoemia.” 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).</li> <li>2. Incentive spirometry x 10 breaths q 2 hr. when awake.</li> <li>3. Encourage ambulation, activity.</li> </ol>	<ol style="list-style-type: none"> <li>1. Oxygen to maintain O<sub>2</sub> saturation ≥ 94% or ≥ baseline value.</li> <li>2. Acetaminophen 15 mg/kg po q 4 hr. or prn T &gt;38.0°C.</li> <li>3. Ibuprofen 10 mg/kg po q 8 hr. or other anti-inflammatory agent if no contraindication present (i.e. gastritis, ulcer, renal impairment).</li> <li>4. Morphine 0.05 - 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 <u>not</u> demerol) may be used in individual cases.</li> <li>5. Cefuroxime 50 mg/kg q 8 hr. IV. (Prophylactic penicillin may be discontinued while on broad-spectrum antibiotics.)</li> <li>6. Erythromycin 10 mg/kg q 6 hr. po, Clarithromycin 15 mg/kg split q 12 hr. po or other macrolide antibiotic.</li> <li>7. Continue prophylactic folic acid, if applicable.</li> <li>8. Consider bronchodilators, especially if patient has history of restrictive airway disease.</li> <li>9. Consider red cell transfusion:                             <ol style="list-style-type: none"> <li>a) simple transfusion for moderately severe illness, especially if Hb &gt; 1 gm/dl below baseline (do not transfuse acutely to Hb &gt; 10 gm/dl, Hct &gt; 30%).</li> <li>b) partial exchange transfusion to Hb 10 gm/dl and Hb S or Hb S + C (patient’s RBC) ≤ 30 % for severe or rapidly progressive disease. (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.</li> </ol> </li> <li>10. See other Clinical Care Paths for acute anemic crisis, stroke, priapism, if present.</li> </ol>	<ol style="list-style-type: none"> <li>1. Off O<sub>2</sub>.</li> <li>2. Afebrile ≥ 24 hr.</li> <li>3. Good oral intake, including oral antibiotics.</li> <li>4. Adequate pain relief (if needed) with oral analgesics.</li> <li>5. Follow-up plans coordinated with hematology service.</li> </ol>

Modified from Mountain States Regional Genetic Services Network, 1996

## STROKE OR ACUTE NEUROLOGIC EVENT

Diagnosis	Monitoring	Diagnostics (if not previously obtained)	Fluids, Nutrition, General Care	Medications/Treatments	Discharge Criteria
Stroke or acute neurologic event in child with sickle cell disease.	<ol style="list-style-type: none"> <li>1. ICU admission first 24 hr. and until stable.</li> <li>2. Vital signs, neuro checks q 2 hr.</li> <li>3. CR monitor.</li> <li>4. Continuous pulse ox.</li> <li>5. Record I + 0, daily weight.</li> </ol>	<ol style="list-style-type: none"> <li>1. CBC, diff., platelet count, and reticulocyte count initially (compare with patient's baseline data).</li> <li>2. Coagulation profile. Consider evaluation for hypercoagulability.</li> <li>3. Blood and urine cultures if febrile.</li> <li>4. Type and cross match (minor-antigen-matched if available, sickle-negative, leukocyte-depleted RBC) for partial exchange transfusion (erythrocytapheresis).</li> <li>5. Electrolytes initially and daily until stable.</li> <li>6. CT scan or MRI to exclude intracranial hemorrhage.</li> <li>7. Consider CSF culture if febrile and no contraindication present.</li> </ol>	<ol style="list-style-type: none"> <li>1. IV + po 1-1.5 x maintenance.</li> </ol>	<ol style="list-style-type: none"> <li>1. Rx seizures if present.</li> <li>2. Rx increased intracranial pressure if present.</li> <li>3. Broad-spectrum IV antibiotics if febrile.</li> <li>4. If applicable and not on broad-spectrum antibiotics, continue prophylactic penicillin.</li> <li>5. Partial exchange transfusion or erythrocytapheresis to Hb 10 gm/dl and Hb S (patient's RBC) <math>\leq</math> 30% (may consider for erythrocytapheresis). Remove femoral or central venous catheter as soon as possible after exchange transfusion to reduce risk of thrombosis.</li> <li>6. Simple transfusion with RBC to Hb approximately 10 gm/dl may be considered as an alternative to partial exchange transfusion for stable patients with Hb &lt; 6-7 gm/dl (do <u>not</u> transfuse acutely to Hb &gt; 10 gm/dl, Hct &gt; 30 %).</li> <li>7. Hemoglobin electrophoresis after partial exchange transfusion or at discharge.</li> </ol>	<ol style="list-style-type: none"> <li>1. Clinically and neurologically stable &gt; 24 hr. after transfusions.</li> <li>2. Afebrile &gt; 24 hr.</li> <li>3. Hematology and physical therapy follow-up organized.</li> </ol>

Modified from Mountain States Regional Genetic Services Network, 1996

## PRIAPISM

Diagnosis	Monitoring	Diagnostics (if not previously obtained)	Fluids, General Care	Medication/Treatment	Discharge Criteria
<p>Priapism in child with sickle cell disease.</p> <p>Notify urology if priapism persists for greater than 2 hours.</p>	<ol style="list-style-type: none"> <li>1. Vital signs 1 2-4 hr.</li> <li>2. Record 1 + O, daily weight.</li> <li>3. CR monitor and continuous pulse ox. if receiving parenteral narcotics.</li> </ol>	<ol style="list-style-type: none"> <li>1. CBC, diff., platelet count, and reticulocyte count initially (compare with patient's baseline data).</li> <li>2. Type and cross match (minor-antigen-matched if available, sickle-negative, leukocyte-depleted RBC) for partial exchange transfusion.</li> <li>3. Urinalysis and urine culture.</li> <li>4. Blood culture if febrile. Consider other cultures (e.g. CSF).</li> </ol>	<ol style="list-style-type: none"> <li>1. IV fluids - 10 cc/kg over 1 hr., then IV + po = 1.5 x maintenance.</li> <li>2. Encourage ambulation.</li> <li>3. Incentive spirometry—10 breaths q 2 hr. when awake if on parenteral narcotics.</li> </ol>	<ol style="list-style-type: none"> <li>1. Broad-spectrum in antibiotics IV if febrile.</li> <li>2. If applicable, continue prophylactic penicillin (if not on broad-spectrum antibiotics) and folic acid.</li> <li>3. Mild to moderately severe pain—acetaminophen with codeine ( 1 mg/kg) po q 4 hr.</li> <li>4. Ibuprofen 10 mg/kg po q 8 hr. or other anti-inflammatory agent if no gastritis, ulcer or renal impairment present.</li> <li>5. 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.</li> <li>6. Consider prostatic massage. Strongly consider drainage and irrigation with epinephrine (1:1,000,000) under local anesthetic per urology. Notify urology within 2 hours with the goal of performing the procedure within 4 hours of onset. All attempts should be made to do this within 12 hours of onset.</li> <li>7. Pseudoephedrine &lt; 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.</li> <li><b>8. <u>Never</u> use ice or cold packs.</b></li> <li>9. O<sub>2</sub> by nasal cannula if needed to keep pulse ox ≥ 92% or ≥ patient's baseline value. Avoid excessive or unnecessary O<sub>2</sub>, which may suppress the reticulocyte count and exacerbate anemia.</li> <li>10. Reassess pain control at least twice daily. Analgesics may be weaned as tolerated by decreasing dose, <u>not</u> by prolonging interval between doses.</li> <li>11. Transfusion if no evidence of detumescence within 12 hr.                         <ol style="list-style-type: none"> <li>a) Partial exchange or erythrocytapheresis to Hb 10 gm/dl and Hb S (patient's RBC) ≤ 30%.</li> <li>b) May consider simple transfusion as alternative to partial exchange transfusion if Hb &lt; 6-7 gm/dl (do <u>not</u> transfuse acutely to Hb &gt; 10 gm/dl, hct &gt; 30%).</li> </ol> </li> <li>12. Surgical drainage (i.e. Winter shunt) is usually indicated if priapism persists for ≥ 24 hrs., unresponsive to supportive care and transfusions.</li> <li>13. Observe for severe headache or neurologic signs or symptoms. (Ischemic stroke may occur 1-10 days after onset of priapism).</li> <li>14. See other Clinical Care Paths for acute chest syndrome, acute anemic crisis, stroke, if present.</li> </ol>	<ol style="list-style-type: none"> <li>1. Priapism resolving (complete detumescence may take 1-2 weeks).</li> <li>2. Taking oral fluids well and able to take po medications (e.g. prophylactic penicillin) if applicable.</li> <li>3. Adequate pain relief on oral analgesics.</li> <li>4. Afebrile ≥ 24 hr.</li> <li>5. Resolution of any pulmonary symptoms or documentation of adequate oxygenation on room air.</li> </ol>

Modified from Mountain States Regional Genetic Services Network, 1996

## GENERAL ANESTHESIA AND SURGERY

Pre-Op Evaluation	Pre-Op Transfusion	Day Prior to Surgery	Intraoperative	Post-Operative
<ul style="list-style-type: none"> <li>• Baseline CXR, pulse ox.</li> <li>• CBC, retic, U/A.</li> <li>• Consider pulmonary function tests for patients with prior history of acute chest syndrome or with suspicion of chronic lung disease.</li> </ul>	<ul style="list-style-type: none"> <li>• Simple transfusion should be strongly considered for all children with Hb SS or Sβ<sup>0</sup>-thalassemia prior to any procedure requiring general anesthesia.</li> <li>• Use minor-antigen-matched if available, sickle-negative, leukocyte-depleted RBC. Simple transfusion: RBC's to increase Hb to 10.0 gm/dl.</li> <li>• Surgery without pre-op transfusion in children with Hb SS and Sβ<sup>0</sup>-thalassemia may be considered in selected cases for <u>minor</u> procedures (e.g. PE tubes) with brief anesthetics. (Tonsillectomy and/or adenoidectomy is <u>not</u> considered a minor procedure). Recommendations for patients with Hb SC or Sβ<sup>+</sup>-thalassemia vary. In general, transfusion is not required for smaller procedures such as tonsillectomy and/or adenoidectomy, but transfusion is required for abdominal surgery. Do to a high baseline HCT these patients often require partial exchange transfusion.</li> </ul>	<ul style="list-style-type: none"> <li>• CBC, retic.</li> <li>• IV hydration (1 - 1.5 x maintenance) ≥ 12 hr. before procedure.</li> </ul>	<ul style="list-style-type: none"> <li>• Minimum 50% O<sub>2</sub> with anesthetic agent.</li> <li>• Avoid hypoxia (continuous pulse ox.), hypercarbia, or hyperventilation.</li> <li>• Avoid tourniquets.</li> </ul>	<ul style="list-style-type: none"> <li>• O<sub>2</sub> by nasal cannula @ 2L (even if pulse ox. ≥ 99%) until side effects of anesthesia have worn off and patient is ambulatory.</li> <li>• Continuous pulse oximetry for ≥ 24 hr. to ensure that 2L O<sub>2</sub> is sufficient to keep saturation &gt; 95%.</li> <li>• Encourage early ambulation, activity.</li> <li>• IV + po 1 - 1.5 x maintenance. Avoid excessive hydration, which may precipitate acute chest syndrome.</li> <li>• Incentive spirometry- 10 breaths q 2 hr. while awake.</li> </ul>

Modified from Mountain States Regional Genetic Services Network, 1996

## References and Resources

### PATIENT EDUCATION MATERIALS AND RESOURCES

*The Family Connection — Sickle Cell Trait* (English, French, Spanish); *The Family Connection — Hemoglobin C Trait* (English, French, Spanish); *Newborn Screening for Your Baby's Health* (English, Spanish) Sickle Cell Anemia, New York State Department of Health, Newborn Screening Program, Wadsworth Center for Laboratories and Research, P.O. Box 509, Albany, NY 12201-0509; phone: 518-473-7552.

*Help* (resource book listing sources of care for patients with sickle cell disease in the United States, Puerto Rico and the Virgin Islands); *Sickle Cell Disease: How to help your child to take it in stride — A Parent/Teacher Guide Viewpoints*. Also available: brochures on sickle cell trait, anemia and other topics, home study kit, games and a video on parenting. National Association for Sickle Cell Disease, 3345 Wilshire Blvd., Suite 1106, Los Angeles, CA 90010-1880; phone: 1-800-421-8453.

*All You Ever Wanted to Know about Sickle Cell Trait*. State of California Genetic Disease Center, phone: 510-412-1542, leave message with desired number of copies and contact information. First copy is free, additional copies are \$.30 each.

*The Infant and Young Child with Sickle Cell Anemia* (a guide for parents, in English, Spanish); *Pneumococcal Infection and Penicillin*; *So Your Baby Has the Sickle Cell Trait* (English, Spanish). Also available: brochures on sickle cell trait, sickle beta-thalassemia, hemoglobin C disease, pain in children and various complications. Texas Department of Health, Newborn Screening Program, 1100 West 49th St., Austin, TX 78756-3199; [www.dshs.state.tx.us/newborn/sickle.shtm](http://www.dshs.state.tx.us/newborn/sickle.shtm)

*Sickle Cell Anemia — What Is It?* Cincinnati Children's Sickle Cell Center, Cincinnati Children's Hospital Medical Center, 3333 Burnet Ave., Cincinnati, OH 45229; phone: 1-800-344-2462, ext. 4541.

*Thalassemia Information Sheet; Sickle Cell Anemia Public Health Information Sheet*. March of Dimes, Birth Defects Foundation, 1275 Mamaroneck Ave., White Plains, NY 10605; [www.marchofdimes.com](http://www.marchofdimes.com)

Note: Additional materials may be available from your own state or local health department, sickle cell agency or community agency.

### COUNSELING REFERENCES FOR PARENTS OF NEWBORNS WITH SICKLE CELL DISEASE AND TRAIT

American Society of Human Genetics Membership Directory. <http://genetics.faseb.org/cgi-bin/ASHG-Search> A searchable database of American Society of Human Genetics members, and their contact information.

Cantor A.B., Miller M.C. III, Larisey L, et al. (1979). A study of media effectiveness for sickle cell anemia education in a rural community. *J Natl Med Assoc*, 71, 1055-1057.

Grossman L.K., Holtzman N.A., Charney E., et al. (1985). Neonatal screening and genetic counseling for sickle cell trait. *Am J Dis Child*, 139, 241-244.

Loader S., Sutera C.J., Walden M., et al. (1991). Prenatal screening for hemoglobinopathies; Evaluation of counseling. *Am J Hum Genet*, 48, 447-451.

Miller D.R. (1979). Pitfalls of newborn screening for sickle cell anemia. *Am J Dis Child*, 133, 1235-1236.

Neal-Cooper F., Scott R.B. (1988). Genetic counseling in sickle cell anemia: Experiences with couples at risk. *Public Health Rep*, 103, 174-178.

Rowley P.T., Mack L. Jr., Lawrence F. (1984). Screening and genetic counseling for beta-thalassemia trait in a population unselected for interest: Comparison of three counseling methods. *Am J Human Genet*, 36, 677-689.

### GENERAL REFERENCES

Adams R.J., McKie VC, Hsu L et.al (1998). Prevention of a first stroke by transfusions in children with sickle cell anemia and abnormal results on transcranial Doppler ultrasonography. *N Engl J Med*, 339, 5-11.

Adams T., McKie V., Nichols F., et al. (1992). The use of transcranial ultrasonography to predict stroke in sickle cell disease. *N Engl J Med*, 326, 605-610.

Armstrong P.J. & Bersten A. (1986). Normeperidine toxicity. *Anesthesia and Analgesia*, 65, 536-538.

Angling D.L., Siegel J.D., Pacini D.L. (1984). Effect of penicillin prophylaxis on nasopharyngeal colonization with streptococcus pneumonia in children with sickle cell anemia. *J Pediatr*, 104, 18-22.

Bainbrige R., Higgs D.R., Maude G.H., et al. (1985). Clinical presentation of homozygous sickle cell disease. *J Pediatr*, 106, 881-885.

Baum F.K., Dunn D.T., Maude G.H., Serjeant G.R. (1987). The

### III. Appendix: References and Resources

- painful crisis of homozygous sickle cell disease. A study of the risk factors. Arch Intern Med, 47, 1231-1234.
- Bakshi S.S., Grover R., Cabezon E., et al. (1991). Febrile episodes in children with sickle cell disease treated on an ambulatory basis. J Assoc Acad Minor Phys, 2, 80-83.
- Belgrave F.Z., Molock S.D. (1991). The role of depression in hospital admissions and emergency treatment of patients with sickle cell disease. J Natl Med Assoc, 83, 777-781.
- Berde C.B. In Max, M.B., Portenoy R.K., & Laska E.M. (eds.). (1991). Advances in pain research and therapy: The design of analgesic clinical trials. Pediatric Analgesic Trials, 18, 445-455.
- Brown A.K., Miller S.T., Agatista P. (1989). Care of infants with sickle cell disease. Pediatrics, 83 (5 Pt 20), 897-900.
- Buchanan G.R., Schiffman G. (1980). Antibody responses of polyvalent pneumococcal vaccine in infants with sickle cell anemia. J Pediatr, 96, 264-266.
- Buchanan G.R., Siegel J.D., Smith S.S., et al. (1982). Oral penicillin prophylaxis in children with impaired splenic function: A study of compliance. Pediatrics, 70, 926-930.
- Buchanan G.R., Smith S.J. (1986). Pneumococcal septimicemia despite pneumococcal vaccine and prescription of penicillin prophylaxis in children with sickle cell anemia. Am J Dis Child, 140, 428-432.
- Burghardt-Fitzgerald, D. (1989). Pain behavior contracts: Effective management of adolescents in sickle cell crisis. Journal of Pediatric Nursing, 4, 320 - 324.
- Centers for Disease Control. (1989). Risk associated with human parvo-virus B19 infection. MMWR 38, 81-97.
- Charache, S. (1991). Hydroxyurea as treatment for sickle cell anemia. Hematology/Oncology Clinics of North America, 5, 571-583.
- Chudwin D.S., Wara D.W., Matthay K.K. (1983). Increased serum opsonic activity and antibody concentration in patients with sickle cell disease after pneumococcal polysaccharide immunization. J Pediatr, 102, 51-54.
- Committee on Infectious Disease. (1990). Parvovirus, erythema infectiosum and pregnancy. Pediatr, 85, 131.
- Consensus Conference. (1987). Newborn screening for sickle cell disease and other hemoglobinopathies. JAMA, 258, 1205-1209.
- Cummins D., Heuschkel R., Davies S.C. (1991). Penicillin prophylaxis in children with sickle cell disease in Brent study of physician's knowledge, patient compliance. BMJ, 302, 989-990.
- Davies S.C., Brozovic M. (1989). The presentation, management and prophylaxis of sickle cell disease. Blood Rev, 3, 29.
- Day S., Brunson G., Wang W. (1992). A successful education program for parents of infants with newly diagnosed sickle cell disease. J Pediatr Nurs, 7, 52-57.
- DeCeulaer K., McMullen K.W., Maude G.H., et al. (1985). Pneumonia in young children with homozygous sickle cell disease: Risk and clinical features. Eur J Pediatr, 144, 255-258.
- Eckman J.R., Walco G.A., Erickson C.J. & Blankson V. (1994). Non-pharmacologic management of sickle syndromes. Proceedings of the 19th Annual Meeting of the National Sickle Disease Program, 53.
- Edmond A.M. (1985). Acute splenic sequestration in homozygous sickle cell disease: Natural history and management. J Pediatr, 107, 201.
- Enwonwu C.O. (1988). Nutritional support in sickle cell anemia: theoretical considerations. J Natl Med Assoc, 80, 139-144.
- Flanagan C. (1980). Home management of sickle cell anemia. Pediatr Nurs, 6, A-D.
- Falletta J.M, Woods G.M., Verter J.I., Buchanan G.R., Pegelow C.H., Iyer R.V., Miller S.T., Holbrook C.T., Kinney T.R., Vichinsky E., et al. (1995). Discontinuing penicillin prophylaxis in children with sickle cell anemia. Prophylactic penicillin study II. J Pediatr, 127, 685-690.
- Foley M.K., Inturrisi E.C. (1987). Analgesic drug therapy in cancer pain: Principles and practice. Med Clin North Am, 71, 207-232.
- Gaston M.H., Verter J.L., Woods G., et al. (1986). Prophylaxis with oral penicillin in children with sickle cell anemia: A randomized trial. N Engl J Med, 314, 1593-1599.
- Gigliotti F., Feldman S., Wang W.C. (1986). Serologic follow-up of children with sickle cell disease immunized with Haemophilus influenza type B conjugate vaccine during early infancy. J Pediatr, 118, 917-919.
- Gil K., Williams D., Thompson R. & Kinney T. (1991). Sickle cell disease in children and adolescents: The relation of child and parent pain coping strategies to adjustment. Journal of Pediatric Psychology, 16, 643 - 663.
- Givner L.B., Luddy R.E., Schwartz A.D. (1981). Etiology of osteomyelitis in patients with major sickle hemoglobinopathies. J Pediatr, 99, 411-413.
- Graff-Lonnevig V., Hedlin G., Lindfors A. (1988). Penicillin allergy: a rare pediatric condition? Arch Dis Child, 63, 1342-1346.
- Griffin T.C., McIntire D. and Buchanan G.R. (1994). High-dose intravenous methylprednisolone therapy for pain in children and adolescents with sickle cell disease. N Engl J Med, 330, 733-737.
- Grover R., Withers D.L. (1990). Management of acute splenic sequestration crisis in sickle cell disease. J Assoc Acad Minority Phys, 1, 167.
- Grover R., Shahidi S., Fisher B., et al. (1983). Current sickle cell screening program in New York City. Am J Public Health, 73, 249-253.

- Hales K., Barriere S. (1979). Polyvalent pneumococcal vaccine: a review. Am J Hosp Pharm, 36, 773-777.
- Hatton C.S.R., et al. (1985). Hepatic sequestration in sickle cell anemia. Brit Med J, 290, 744.
- International Rheumatic Fever Study Group. (1991). Allergic reaction to long-term benzathine penicillin prophylaxis for rheumatic fever. Lancet, 337, 1308-1310.
- Inturrisi C.E., Foley K.M. (1984). Narcotic analgesics in the management of pain. In M. Kuhar, G. Pasternack (Eds.), Analgesics: Neurochemical, behavioral and clinical perspectives. New York: Raven Press.
- Jackson S.M., Sporrer K.A., Agner S.A., Laver J.H. and Abboud M.R. (1994). Pain in children and adolescents with sickle cell anemia: a prospective study utilizing self reporting. Proceedings of the 19th Annual Meeting of the National Sickle Cell Disease Program, New York City, 86.
- Jenkins M.E., Scott R.B., Vaird R.L. (1960). Studies in sickle cell anemia: Sudden death during sickle cell anemia crisis in young children. J Pediatr, 56, 30.
- John A.B., Ramlal A., Jackson H., et al. (1984). Prevention of pneumococcal infection in children with homozygous sickle cell disease. BMJ, 288, 1567-1570.
- Kaplan E.C., Berrios X., Speth J., et al. (1989). Pharmacokinetics of benzathine penicillin serum levels during the 29 days after IM injection of 1.2 IU. J Pediatr, 115, 146-150.
- Karko R.F., Foley K.M., Grabinsky P.Y., et al. (1983). Central nervous system excitatory effects of meperidine in cancer patients. Ann Neurol, 13, 180-185.
- Kim H.C., Alavi A., Russel M., et al. (1989). Differentiation of bone and bone marrow in infants from osteomyelitis in sickle cell diseases. Clin Nucl Med, 141, 249-254.
- Kinney T.R., et al. (1990). Long-term management of splenic sequestration in children with sickle cell disease. J Pediatr, 117, 194.
- Kuisel L., Shearer J., Jackson S.M., et al. (1994). Home pain management in adolescents with severe sickle cell disease. Proceedings of the 19th Annual Meeting of the National Sickle Cell Disease Program, 104.
- Leikin S.L., Gallagher D., Kinney T.R. (1989). Mortality in children and adolescents with sickle cell disease. Pediatrics, 84, 500-508.
- Listernick R., Frisone L., Silverman B.L. (1992). Delayed diagnosis of infection with abnormal neonatal screens. JAMA, 267, 1095-1099.
- Lobel J.S., Cameron B.F., Johnson E., et al. (1989). Value of screening umbilical cord blood for hemoglobinopathy. Pediatrics, 83(5 Pt 2), 823-826.
- Marcinak J.F., Frank A.L., Labotka R.L., et al. (1991). Haemophilus influenza type B vaccine in children with sickle cell disease: Antibody persistence after vaccination at age one and one half to six years. Pediatr Infect Dis J, 10, 157-159.
- McIntosh S., Rooks Y., Richey A.K., et al. (1980). Fever in young children with sickle cell disease. J Pediatr, 96, 199-204.
- Miller S.T., Stilerman T.V., Rao S.P., et al. (1990). Newborn screening for sickle cell disease: When is an infant lost to follow-up? Am J Dis Child, 144, 1343-1345.
- Ndumbe P.M., Nyouma E. (1990). Transmission of hepatitis B virus by blood transfusion in Yaounde, Cameroon. BMJ, 301, 523-524.
- Nussbaum R.L., Powell C., Graham H.L., et al. (1984). Newborn screening for sickling hemoglobinopathies. Houston, 1976 to 1980. Am J Dis Child, 138, 44-48.
- O'Brien R.T., McIntosh S., Aspnes G.T., et al. (1976). Prospective study of sickle cell anemia in infancy. J Pediatr, 89, 205-210.
- Ohene-Frempong K. (1991). Stroke in sickle cell disease: Demographic, clinical and therapeutic considerations. Semin Hematol, 28, 213-219.
- Overturf G.D. (1982). Pneumococcal vaccination: Update and evaluation. Prog Clin Biol Res, 98, 59-71.
- Overturf G.D., Powars D., Baraff L.J. (1977). Bacterial meningitis and septicemia in sickle cell disease. Am J Dis Child, 131, 784-787.
- Pavlikis S.G., Prohovnik I., Piomelli S., et al. (1989). Neurologic complications of sickle cell disease [Retraction of Barness LA]. Adv Pediatr, 36, 247-276.
- Pattison Jr., et al. (1981). Parvovirus infections and hypoplastic crisis in sickle cell anemia. Lancet, 1, 644.
- Pearson H.A. (1986). Sickle cell disease: Diagnosis and management in infants and children. Pediatr Rev, 9, 121.
- Pearson H.A., Cobb W.T. (1964). Folic acid studies in sickle cell anemia. J Lab Clin Med, 64, 913-921.
- Pegelow C.H., Armstrong F.D., Light S., et al. (1991). Experience with the use of prophylactic penicillin in children with sickle cell anemia. J Pediatr, 118, 736-738.
- Pegelow C. (1992). Survey of pain management therapy provided for children with sickle cell disease. Clinical Pediatrics, 31, 211-214.
- Platt O. (1994). Easing the suffering caused by sickle cell disease. New Engl J Med, 330, 783-784.
- Platt A., Beyer J., Kinney T., & West M. (1995). Recommendations for assessing sickle cell pain. Proceedings of the 19th Annual Meeting of the National Sickle Cell Disease Program, 145.
- Porter C. & Villarruel A. (1992). Socialization and caring for hospitalized African-American and Mexican-American children. Issues in Comprehensive Pediatric Nursing, 14, 1-16.

### III. Appendix: References and Resources

- Porter J, Jick II. (1980). Addiction is rare in patients treated with narcotics. New Engl J Med, 302, 123.
- Powars D.R. (1975). Natural history of sickle cell disease: First 10 years. Semin Hematol, 12, 267-285.
- Powars D.R., Overturf G, Weiss J., et al. (1981). Pneumococcal septicemia in children with sickle cell anemia: Changing trend of survival. JAMA, 245, 1839-1842.
- Rabb, et al. (1983). A trial of folate supplementation in children with homozygous sickle cell disease. Br J Haematol, 54, 589.
- Rao S., Pang. (1982). Transfusion therapy for subacute splenic sequestration in sickle cell anemia. Blood, 60 (suppl), 489.
- Rodgers G.P., Dover G.J., Noguchi C.T., et al. (1990). Hematologic responses of patients with sickle cell disease to hydroxyurea. New Engl J Med, 322, 1037-1045.
- Rogers D.W., et al. (1978). Early deaths in Jamaican children with sickle cell disease. Br J Med, 1, 1515.
- Rogers Z.R., Morrison R.A., Vedro D.A., et al. (1990). Outpatient management of febrile illness in infants and young children with sickle cell anemia. J Pediatr, 117, 736-739.
- Rowley P.T., Huntzinger D.J. (1983). Newborn sickle cell screening: Benefits and burdens realized. Am J Dis Child, 137, 341-345.
- St. Jude Children's Research Hospital, Department of Hematology-Oncology: Chesney P.J., Day S.W., Eguiguren J.M., Fairclough D.L., Flynn P.M., Harris S., Rodman J.H., Smith R., Wang W.C., & Williams J.A. (1993). A randomized study of outpatient treatment with deftriaxone for selected febrile children with sickle cell disease. New Engl J Med, 329, 472-476.
- Sandler D.P., Smith J.C., Weinberg C.R., et al. (1989). Analgesic use and chronic renal disease. New Engl J Med, 320, 123-138.
- Schechter N., Berrien F. & Katz S. (1988). The use of patient-controlled analgesia in adolescents with sickle cell pain crisis: A preliminary report. Journal of Pain and Symptom Management, 3, 1029-1045.
- Schubert T.T. (1986). Hepatobiliary system in sickle disease. Gastroenterology, 90, 2013.
- Seeler R.A., Metzger W., Mufson A. (1972). *Ciplococcus pneumoniae* infections in children with sickle cell anemia. Am J Dis Child, 123, 8-10.
- Seeler R.A., Shwiaki M.Z. (1972). Acute splenic sequestration crisis (ASSC) in young children with sickle cell anemia: Clinical observations in 20 episodes in 14 children. Clin Pediatr, 11, 701-704.
- Serjeant G.R. (1985). Treatment of sickle cell disease in early childhood in Jamaica. Am J Pediatr Hematol Oncol, 7, 235-239.
- Shapiro B. (1989). The management of pain in sickle cell disease. Pediatric Clinics of North America, 36, 1029-1045.
- Shaprior B. (1991). Sickle cell disease-related pain. IASP Newsletter, January-February, 2-4.
- Sharpe J., Brown R., Thompson N., Buchanan I. and Eckman J. (1994). Predictors of coping with pain in mothers and their children with sickle cell disease. Proceedings of the 19th Annual Meeting of the National Sickle Cell Disease Program, 174.
- Sheehy T.W. (1970). Sickle cell hepatopathy. So Med J, 70, 533.
- Sills R. (1987). Splenic function: Physiology and splenic hypofunction. Crit Rev Oncol Hematol, 7, 1-36.
- Smith J. (1993). Management of sickle cell disease: Progress during the past 10 years. American Journal of Pediatric Hematology/Oncology, 5, 360 - 365.
- Spirer A. (1980). The role of the spleen in immunity and infection. Adv Pediatr, 27, 55-88.
- Thurn J. (1988). Human parvovirus B19: Historical and clinical review. Rev Inf Dis, 10, 1005-1011.
- Topley J.M., Rogers D.W., Stevens M.C., et al. (1981). Acute splenic sequestration and hypersplenism in the first five years in homozygous sickle cell disease. Arch Dis Child, 56, 765-769.
- Topley J.M., et al. (1981). Acute splenic sequestration crisis in your children with sickle cell anemia. Arch Dis Child, 56, 765.
- University of Texas Southwestern Medical Center, Department of Pediatrics: Buchanan G.R., Morrison R.A., Rogers Z.R. & Vedro D.A., Dallas TX 75235-9063. (1990). Outpatient management of febrile illness in infants and young children with sickle cell anemia. Journal of Pediatrics, 117, 736-739.
- Vermlyen C., Cornu G, Philippe M., et al. (1991). Bone marrow transplantation in sickle cell anemia. Arch Dis Child, 66, 1195-1198.
- Vichinsky E., Hurst D., Earles A., et al. (1988). Newborn screening for sickle cell disease: Effect on mortality. Pediatrics, 81, 749-755.
- Vishinsky E., Lubin B.H. (1987). Suggested guidelines for the treatment of children with sickle cell anemia. Hematol Oncol Clin North Am, 1, 483-501.
- Vichinsky E., et al. (1981). The diagnosis of iron deficiency anemia in sickle cell disease. Blood, 58, 963.
- Warren N.S., Carter T.P., Humbert J.R., et al. (1982). Newborn screening for hemoglobinopathies in New York state: Experience of physicians and parents of affected children. J Pediatr, 100, 373-377.
- Webb D.K., Serjeant G.R. (1989). Systemic salmonella infection in sickle cell anemia. Ann Trop Pediatr, 9, 169-172.

White P.F. (1988). Use of patient-controlled analgesia for management of acute pain. *JAMA*, 259, 243-247.

Whitten C.F. (1989). Newborn screening for sickle cell disease and other hemoglobinopathies: Perspective from the National Association for Sickle Cell Disease. *Pediatrics*, 83 (5 Pt 2), 906-7.

Wright L., Brown A., Davidson-Mundt A. (1992). Newborn screening: The miracle and the challenge. *J Pediatr Nurs*, 7, 26-42.

Zarkowsky H.S., Gallagher D., Gill F.M., et al. "Bacteremia in sickle hemoglobinopathies." *J Pediatr* 1986.

---

### GENERAL REFERENCES

National Heart Lung and Blood Institute. (2002). The management of Sickle Cell Disease. Publication #02-2117. [www.nhlbi.nih.gov/health/prof/blood/sickle/sc\\_mngt.pdf](http://www.nhlbi.nih.gov/health/prof/blood/sickle/sc_mngt.pdf)

**Funded by the Washington State Department of Health  
Children with Special Health Care Needs Program**



**Item #002**

