

# Minnesota Sickle Cell Disease Emergency Guidelines

9/1/05

- Triage the patient to a room and assign staff as rapidly as possible
- Ask patient if he or she has a care plan for sickle cell disease and consult this plan, if available**
- Assess vital signs: blood pressure, heart rate, respiratory rate, oxygen saturation, temperature
- Administer oxygen for O<sub>2</sub> saturation <90% or more than 3% below patient's baseline status, if known

## Pain Management

- Obtain history of pain, self-assessment of pain, and treatments attempted at home
- Ask about previous hospitalizations, treatments, and medication allergies or reactions
- Administer IV fluids (NS). In adults, central access is preferred to leg access. (Pediatric patients with normal renal/cardiac function: 20cc/kg IV NS; adult patients with normal renal/cardiac function: 1-2 liters NS; smaller volumes and extra caution for patients with known cardiac/renal disease)
- Initiate pain treatment. **Do not use Demerol/meperidine.** Opt for subcutaneous or IV infusion of morphine or hydromorphone/Dilaudid. Avoid intramuscular injection and do not under-dose. PCA administration for ongoing pain control is an option. Example starting doses listed on the back of this checklist. Consider NSAIDs and other adjunct therapies
- Assess pain relief every 5-10 minutes and modify dosing until pain is controlled
- Encourage use of distraction techniques (walking, etc.) and supportive cares (massage by support person, warm packs)

## Patients Presenting with Other Complications of Sickle Cell Disease

- Consider labs including CBC with differential, platelet count, reticulocyte count, and other labs as indicated (including pregnancy test). If transfusion might be needed as inpatient, order type and screen (most likely in chest crisis, neurologic symptoms)
  - Infection (fever >38.5, other signs of infection or sepsis)**
    - Obtain a blood culture and other tests as clinically indicated
    - Obtain a chest X-ray in patients less than 5-years-old
    - Initiate broad-spectrum antibiotic therapy with ceftriaxone (plus vancomycin if toxic). Do not wait for result of lab tests
  - Respiratory symptoms (chest pain, shortness of breath, tenderness, breathing problems)**
    - Obtain chest X-ray and administer oxygen if indicated by O<sub>2</sub> saturation
    - Treatment should include antibiotics and consideration of transfusion
  - Acute neurologic symptoms (including painless limp, acute headache, seizures)**
    - CT, MRI/A as indicated
    - Consult hematology/neurology
  - Abdominal symptoms (distension, "silent" abdomen, pain, anemia, weakness, or pallor)**
    - Check spleen and liver size. Enlargement, especially coupled with falling hematocrit and increased reticulocyte count may indicate sequestration crisis. Type and cross for suspected sequestration
    - Isotonic IV fluids
    - If gallbladder disease is suspected, consider abdominal ultrasound and chemistries
  - Priapism**
    - IV hydration, subcutaneous terbutaline. Consult urology if not resolved within 4-6 hours
    - Avoid transfusion
- Obtain OB consult for pregnant patients

## Pregnant Patients

**Admission/Transfer Criteria** (designate situations in which patients should be considered for admission or transfer to a hospital knowledgeable in sickle cell disease): pain not controlled within 4-6 hours; patient returns to ER within 48 hours due to treatment failure; patient is pregnant; significant acidosis or hypoxia from baseline; infant less than 1 year with fever > 38.5 or significant infection is suspected; acute neurologic symptoms; splenic or hepatic syndromes, cholecystitis; evidence of acute chest syndrome (pulmonary infiltrate on x-ray, decrease of hemoglobin more than 1g from baseline, patient requires oxygen, chest pain); urology consult not immediately available for priapism lasting more than 4 hours

## Emergency Room Discharge Criteria

*If patient will be discharged, consider patient's resources in achieving required follow-up. Consider admission for patients unable to achieve follow-up as outpatient.*

- Pain under control and patient able to manage at home
- Stable cardio-respiratory status
- Plan in place for timely outpatient follow-up
- Stable hemoglobin (sickle cell patients are uniformly anemic at baseline)

## Hematology Specialists Willing to Provide Physician Consultation

### Children's Hospitals and Clinics of Minnesota

Pediatrics: Stephen Nelson, MD; Jane Hennessy (Minneapolis) (612) 813-5940  
 Pediatrics: David Slomiany, MD; Kristin Moquist (St. Paul) (651) 220-6732

### Minnesota Oncology/Hematology

Adults: M. Obinna Nwaneri, MD (612) 863-8585

### Hennepin County Medical Center

Adults: Douglas Rausch, MD (612) 873-6369  
 After hours call (612) 873-2639

### University of Minnesota

Pediatrics: Joseph Neglia, MD (612) 626-2778  
 Adults: Mark Reding, MD (612) 273-3000

## Usual Starting Dose for Moderate-to-Severe Pain Medication for Sickle Cell Disease

Table 1

Usual Starting Doses of Opioid Analgesics in Opioid-Naive Adults and Children ≤50 kg Body Weight

	Oral	Parenteral
Morphine	0.3 mg/kg every 3-4 h	0.1-0.15 mg/kg every 2-4 h
Hydromorphone (Dilaudid)	0.06-0.08 mg/kg every 3-4 h	0.015-0.020 mg/kg every 3-4 h

Table 2

Usual Starting Doses of Opioid Analgesics in Opioid-Naive Adults and Children ≥50 kg Body Weight

	Oral	Parenteral
Morphine	10-30 mg every 3-4 h	5-10 mg every 2-4 h
Hydromorphone (Dilaudid)	7.5 mg every 3-4 h	1.5 mg every 3-4 h

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

NOTE: Published tables vary in the suggested doses that are equianalgesic to morphine. Titration to clinical responses is necessary because there is not complete cross-tolerance among these drugs, a lower than equianalgesic dose is usually necessary when changing drugs; re-titrating to clinical response is also necessary.

SOURCE: The Management of Sickle Cell Disease. NATIONAL INSTITUTES OF HEALTH/National Heart, Lung, and Blood Institute Division of Blood Diseases and Resources. NIH PUBLICATION NO. 02-2117 REVISED JUNE 2002 (FOURTH EDITION)

## Notes

This guideline is intended to provide the minimum considerations necessary in the evaluation and treatment of all patients with sickle cell disease and does not address needs of specific patients. For individual patients, the Minnesota Hemoglobinopathy Collaborative also endorses the use of individualized care plans and tools such as the Emergency Information form available from the American Academy of Pediatrics and the College of Emergency Physicians at: <http://www.aap.org/advocacy/emergprep.htm>

This guideline was developed through a collaborative effort between the Minnesota Department of Health and the Minnesota Hemoglobinopathy Collaborative.

