

OUTPATIENT EVALUATION AND MANAGEMENT OF PAIN WITH SICKLE CELL DISEASE (Jan 2009)

1. Rapid triage on arrival. Sickle cell pain can be excruciating and requires urgent treatment.
2. **History:**
 - Location, duration, character and intensity of pain. Is pain similar to previous sickle pain?
 - Consider etiologies other than sickle vasoocclusion (e.g. delayed hemolytic transfusion reaction, cholecystitis, appendicitis, trauma)
 - Associated symptoms - especially fever, respiratory symptoms, evidence of dehydration, increased jaundice, or dark urine
 - Ask about dose, timing, and efficacy of analgesics already used for this episode
 - Previous experience with analgesics (efficacy, side effects, allergies). What does patient/family feel best alleviates pain?
3. **Physical Exam:** Complete with emphasis on:
 - vital signs with BP (**see Fever Guidelines if T>38.3°C**), hydration status, and pulse ox (compare with patient's baseline)
 - degree of pallor and cardiopulmonary status. Use O₂ for pulse ox ≤92% or < baseline & look for signs of respiratory illness
 - evidence of localized or systemic infection
 - spleen size (compare with baseline exam)
 - penis (priapism)
 - bones & joints (dactylitis, osteonecrosis, osteomyelitis)
 - neurologic exam (stroke)
4. **Diagnostics:**
 - CBC, diff, platelet, and reticulocyte count (compare with patient's baseline values)
 - Urinalysis and urine culture for abdominal or flank pain
 - Blood cultures if febrile (**see Fever Guidelines**)
 - Type, screen, and Xmatch for extreme pallor (Hgb <6 or 25% below baseline), respiratory symptoms, or acute splenic enlargement (see inpatient pain guidelines). Request leukocyte-depleted, C, E, Kell-compatible (requires minor antigen phenotype) and sickle-negative RBC. In absence of alloantibodies, urgent transfusion should not be delayed by search for minor-antigen matched units.
 - CXR if fever, chest pain, tachypnea, or other lower respiratory symptoms or signs present (Acute Chest Syndrome)
 - Consider abdominal ultrasound and liver function tests for RUQ, epigastric pain (R/O cholelithiasis/cholecystitis)
 - Consider pelvic exam for adolescent female with lower abdominal pain
5. **Treatment:** Base choice and dose of analgesics on intensity of pain, analgesics already used, prior experience of patient with efficacy and side effects, and patient preference. Selected patients may be treated initially with oral analgesics such as ibuprofen 10 mg/kg, acetaminophen with codeine 1 mg/kg, oxycodone 0.2 mg/kg, or immediate release oral morphine 0.3-0.5mg/kg. Most patients who present with pain have failed oral analgesics and/or are experiencing severe pain. Strongly consider use of both opioid and NSAID. Never use a placebo. Use age-appropriate pain intensity rating to assess intensity of pain and monitor efficacy of treatment. Refer to individual patient care plan if available.
 - Opioid options:
 - **Morphine** 0.1-0.15mg/kg IV. Reassess pain q 15-30 min. Patients with severe pain may require repeated doses of morphine 0.02-0.05 mg/kg IV q 15-30 min to achieve pain relief. Subsequent doses IV q 2 hr.
 - Other opioids, such as hydromorphone (**Dilaudid**) 0.015-0.02 mg/kg IV, Nalbuphine (Nubain) 0.1-0.3 mg/kg IV, may be appropriate in some cases. Avoid meperidine (Demerol) in part because of the risk of seizures with repeated doses.
 - NSAID options, if no contraindication (i.e. gastritis, ulcer, coagulopathy, dehydration, renal impairment)
 - **Ketorolac** (Toradol) 0.5 mg/kg (30 mg max dose) IV q 6 hr. For p.o. consider **Ibuprofen** 10 mg/kg po q 6-8 h.
 - **IV fluids:** Consider bolus 10 cc/kg NS over 1 hr, then **D₅ 1/2 NS @ 1 1/2 maintenance** rate. Excessive fluids may precipitate or exacerbate acute chest syndrome and should be avoided unless patient is dehydrated, hypotensive, or has poor perfusion.
 - Monitor **pulse ox**. Use O₂ by nasal cannula or face mask if needed to keep O₂ saturation ≥ 92% or ≥ patient's baseline value, if baseline >92%. The etiology of a supplemental O₂ requirement should be investigated.
 - Prompt administration of parenteral antibiotics if febrile (**see Fever Guidelines**).
6. Discuss the **option of hospitalization** with patient & family. Contact on call hematologist to review management & disposition.
 - If adequate pain relief with one or two doses of opioid, consider giving oral analgesics as trial of outpatient therapy. Call (352) 392-5633 for appt or to speak with hematologist (8a-5p).
 - Consider hospitalization for around-the-clock parenteral analgesics if pain inadequately relieved or if more than one or two doses of parenteral opioid required.

These guidelines do not indicate an exclusive course of treatment or serve as a standard of care. Variations based on a physician's best medical judgement may be appropriate in individual cases.