

Outpatient Evaluation & Management of Pain **in the Child with Sickle Cell Disease**

1. Rapid triage on arrival. Sickle cell pain can be excruciating and requires urgent treatment.
2. History:
 - Nature, location, duration, and severity of pain – is pain similar to previous sickle pain?
 - Consider etiologies other than sickle vasoocclusion (e.g. cholecystitis, appendicitis, trauma)
 - Associated symptoms - especially fever, respiratory symptoms, or evidence of dehydration
 - 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 (see Fever Care Path if $T > 38.5^{\circ}\text{C}$), hydration status, and pulse ox (compare with patient's baseline)
 - degree of pallor and cardiopulmonary status. Use supplemental oxygen for pulse ox less than baseline or signs of respiratory illness.
 - evidence of localized or systemic infection
 - spleen size (compare with baseline exam)
 - penis (R/O priapism)
 - bones and joints (dactylitis, osteomyelitis)
 - neurologic exam
4. Lab:
 - 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 Care Path)
 - Type, screen, and crossmatch for extreme pallor, respiratory or neurologic symptoms, or acute splenic enlargement. Request leukocyte-depleted and, if available, 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
 - 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 severity 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 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. Use age-appropriate pain scale 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.
 - Nalbuphine (Nubain) 0.2-0.3 mg/kg IV. Subsequent doses IV q 3 hr. Do not use nalbuphine for patients receiving chronic opioids (e.g., MS contin, oxycontin, fentanyl patch).
 - Other opioids, such as hydromorphone (Dilaudid) 0.015-0.02 mg/kg IV, may be appropriate in individual 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 maximum dose) IV q 6 hr. Do not use higher loading dose.
 - Ibuprofen 10 mg/kg po q 6-8 h.
 - IV fluids: 10 cc/kg NS over 1 hr, then D₅ ¼ NS @ 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.
6. Discuss option of hospitalization with patient and family. Contact on call pediatric hematologist to discuss management and disposition.
- If adequate pain relief with one or two doses of opioid, consider giving oral analgesics as trial of outpatient therapy.
 - Consider hospitalization for around-the-clock parenteral analgesics if pain inadequately relieved or if more than one or two doses of parenteral opioid required.