Sickle Cell Pain Pathway: ED Management

March 2026

Children's Healthcare of Atlanta

Pain Management (for those without a pain plan in EPIC)

The primary management of a Vaso-Occlusive Crisis (VOC) is analgesics guided by patient report of pain severity
No biomarker or imaging studies can validate pain or assess its severity

Triage

Rapid triage on arrival (ESI Level 2)

- Sickle cell pain can be excruciating and requires urgent treatment
- Review medical record for individualized Emergency pain plan – if one is identified please follow that plan
- If patient does not have an individualized pain plan - begin ED Caregiver Initiated Protocol (see Sickle Cell Pain CIP 5.15)

Initial Evaluation and Monitoring

History

- Location, duration, frequency, character and intensity of pain. Is pain similar to previous sickle cell pain?
- Consider etiologies other than sickle vaso-occlusion (delayed hemolytic transfusion reaction, cholecystitis, appendicitis, trauma)
- Associated symptoms (fever, respiratory symptoms, evidence of dehydration, increased jaundice, or dark urine)
- Dose, timing, and efficacy of analgesics already used for this episode
- Previous experience with analgesics (efficacy, side effects, allergies). What does patient/family feels best alleviates pain?
- Non-pharmacological and psychological strategies currently being used to manage pain
- Recent ED visit or hospitalization within 72 hours
- Transfusion in the last 4 weeks

Complete Exam with emphasis on

- Vital signs with BP, temperature >38.3°C (see Sickle Cell Fever in ED Guideline), hydration status, and pulse ox (compare with patient's baseline Sp02)
- Location of pain
- Degree of pallor and cardiopulmonary status. Use supplemental oxygen for Sp02 below baseline or signs of respiratory distress
- Evidence of localized or systemic infection
- Spleen size (compare with baseline exam)
- Penis (priapism)
- Bones and joints (dactylitis, osteomyelitis)
- Neurologic exam (see Pedatric Stroke <18 yrs Imaging Guideline)

Consider

- Osteomyelitis or fracture with bone pain accompanied by swelling, fever, point tenderness, redness, and increased warmth
- Splenic or liver sequestration, appendicitis, cholecysitis, UTI, PID, acute chest syndrome with mild to severe abdominal pain
- Possible concurrent conditions: CNS event (stroke), priapism, aplastic crisis, fever/sepsis, acute chest syndrome (ACS), acute sequestration crisis, delayed hemolytic transfusion reaction (if received transfusion in last 4 weeks, get type and screen)
- Consider opioid withdrawal on 72 hours returns and discuss with hematology)

Diagnostics

Initial Labs

- CBC with Diff and Reticulocyte Count (compare with baseline)
- CMP — Risk of elevated liver function or high creatinine, receiving pain treatment with ketorolac and acetaminophen

Consider

- Chest X-ray — IF: History of acute chest syndrome, tachypnea, cough, or respiratory symptoms, chest, back, or abdominal pain
  Note: The CXR shall be interpreted prior to discharge by either Radiology (if available), ED, or clinic attending
- Blood Culture — IF: Febrile: temperature >38.3°C (see Sickle Cell Fever in ED Guideline)
- Urine Culture and U/A — IF: Febrile, dysuria, suprapubic pain, unexplained abdominal pain
  Ideally, the urine sample should be obtained prior to (or within 1 hour) after administration of IV antibiotics
- CSF — IF: Altered mental status, seizures, neurologic symptoms
- Type and Cross Match — IF: Hgb <6 gm/dl or 20% or more below baseline and/or if evidence of acute chest syndrome, acute splenic enlargement, or cardiovascular compromise is present; or transfusion in last 4 weeks
  - Request leukocyte-depleted and if available, C, E, Kell-compatible (req. minor antigen phenotype) and sickle-negative RBC
  - In absence of alloantibodies, urgent transfusion should not be delayed by search for minor-antigen matched units
- Abdominal Ultrasound and Liver Function Tests — IF: Consider for RUQ, epigastric, or severe abdominal pain or marked jaundice (R/O cholelithiasis)
- HCG, Chlamydia, N. gonorrhea — IF: If clinical suspicion for pregnancy or sexually transmitted disease in post-pubertal patient
- HIV testing — IF: 13 years or older, and not tested in last year

ED Caregiver Initiated Protocol Medications (5.15)

Medication	Dosage	Max Dose	Notes
Intranasal Fentanyl	2 mcg/kg IN *Give INF BERFORE IV placement and independent of IV morphine	100 mcg	Rapid pain management prior to IV access; Divide dose equally between each nare (Not to exceed 1 ml per nare); Contraindicated if current URI or nasal infection/obstruction, concern for stroke, altered conciousness, any form of head injury, parental/guardian refusal
Hydrocodone/ Acetaminophen	0.2 mg/kg PO x1	10 mg Hydrocodone	If patient has not received pain medication or acetaminophen in last 4 hours
Ketorolac (Toradol)	0.5 mg/kg IV x1	30 mg	If pain greater than 5; Do not administer if known allergy/sensitivity or if patient has had ibuprofen in last 6 hours or known renal dysfunction

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IV Opiods			
Morphine Sulfate	0.1-0.15 mg/kg IV (First Dose), then repeat every 30-60 minutes until adequate relief	10 mg	Reassess pain at peak onset of medication. Patients with severe pain may require repeated doses to achieve pain relief
Hydromorphone	0.015-0.02 mg/kg IV (First Dose), then repeat every 30-60 minutes until adequate relief	2 mg	Reassess pain at peak onset of medication. Patients with severe pain may require repeated doses to achieve pain relief
Nalbuphine	0.3 mg/kg IV (First Dose), then repeat 0.2 mg/kg IV 1-2 hrs until adequate relief	20 mg	Do not use Nalbuphine for patients receiving chronic opioids (e.g. MS Contin, Oxycontin, or Fentanyl patch)
NSAIDs			
Ketorolac (Toradol)	0.5 mg/kg IV x1	30 mg	Initial dose should be given 6 hours after last dose of ibuprofen. Do not administer if known allergy/sensitivity
Ibuprofen	10 mg/kg PO x1	800 mg	Alternative NSAID when IV Toradol not available or oral route preferred
Other Medictions			
Hydrocodone/ Acetaminophen	0.2 mg/kg PO x1	10 mg Hydrocodone	If patient has not received pain medication or acetaminophen in the last 4 hours

- Strongly consider use of both opioids and NSAIDs
- Base choice, dose, and schedule (Bolus ATC or PCA) of analgesics in part on analgesics already used and prior patient experience and preference. Refer to individual patient care plan if available
- Never use a placebo
- Use age appropriate pain intensity rating to assess intensity of pain and monitor efficacy of treatment

General Care (review personal pain plan if present)

- Reassess pain at peak onset of medication per policy with pulse oximetry, at minimum every 15-30 minutes
- Reassess vital signs with pulse oximetry every 60 minutes and consider CR monitor
- Encourage oral hydration
- For well- hydrated or mild/moderately dehydrated patients with normal BP and perfusion: Administer hypotonic IV fluids (D5 1/4 NS) with combined IV and/or oral fluid rate at 1 to 1.5x maintenance
- For severely dehydrated or hypotensive patients: Administer lactated ringers bolus 20 ml/kg, then D5 ¼ NS at 1.5x maintenance rate

Notify ED Attending if fluid bolus is clinically indicated. Caution: Excessive fluids may precipitate or exacerbate acute chest syndrome.

Respiratory

- Monitor pulse oximetry
- Determine baseline oxygenation (Sp02)
  - If baseline Sp02 is ≥ 95%, keep Sp02 at ≥ 95%, use supplemental oxygen as needed
  - If baseline Sp02 is <95%, use supplemental oxygen as needed to keep at baseline
- The etiology of a new or increasing supplemental 02 requirement should be investigated; obtain chest xray

Consult

- Identify patient's primary Hematology campus (AMBH, Hughes Spalding, or Scottish Rite)
- Contact the primary campus' on-call pediatric Hematologist after initial evaluation and labs resulted to discuss management and disposition

Admission Criteria

- Discuss hospitalization with patient and family and ability to maintain pain relief as outpatient
- Consider hospitalization if pain inadequately relieved after 2 hours of appropriate IV treatment including 2 or more doses of an IV opioid
- If adequate pain relief with 1 or 2 doses of IV opioid, consider giving oral analgesics as trial of outpatient therapy
- Consider hospitalization if patient/family is uncomfortable with discharge or feels additional doses of parenteral analgesics will be needed
- Avoid lapses of analgesic treatment prior to transfer to inpatient. Initiate holding or admission orders that include scheduled IV opioids and rescue dosing instructions

Discharge

Discharge Criteria

- Pain resolved or adequately controlled 60 minutes after last IV opioid dose
- Patient and family comfortable with managing pain on home medications
- Taking adequate oral liquids
- Safe home environment and Hematology follow-up discussed

Discharge Instructions and Outpatient Management

- Discharge instructions should indicate duration of scheduled narcotics and any changes to home pain management
- Follow-up for uncomplicated VOC may concur with routine Hematology care for the patient
- Arrange Hematology follow up in 1-2 weeks in cases of any of the following:
  - History of frequent ED visits
  - Daily opioid use
  - Poor compliance with outpatient care (no clinic visit in past 6 months)
  - Other symptoms or abnormal lab values of concern

SCD Clinic Numbers: AMBH: 404.785.1200 | Hughes Spalding: 404.785.9800