Pediatric Sickle Cell Disease Pain Management in the Emergency Department

The following information is intended as a guideline for the acute management of children with vaso-occlusive crisis (VOC) associated with sickle cell disease (SS, SC, S beta- thalassemia). Management of your patient may require a more individualized approach.

MD/APP/RN Team Assessment

Vitals, H&P (ESI TRIAGE LEVEL 2 at minimum): Vital signs with blood pressure, temperature >38.3 C/ 101 F (see Sickle Cell Fever Pathway), hydration status, and pulse ox; Degree of pallor/ cardiopulmonary status; Spleen size; Penis (priaprism); Bones/joints (dactylitis, osteomyelitis); Neurologic exam; Allergies
Assess and document pain:
(**Mild pain: 1-4, Moderate pain: 5-7, Severe pain: 8-10, Different pain than standard: treat level of pain accordingly and consider alternate diagnosis**)
Recent pain medication, dose, time of last dose
Goal for initial treatment of pain: Pain meds within 30 mins of triage. Consider IN fentanyl if difficult access anticipated/encountered for mod/severe.
Individualized Care Plan: Look in EPIC for individualized care plan if one exists for the patient.

Laboratory studies: **CBC+differential and reticulocyte count for moderate/severe pain**

Additional interventions and diagnostic testing to be considered:
IV Fluids: ***Attempt oral hydration first. If unable to tolerate PO initiate D5 1/2 NS at maintenance or 1.5 X maintenance; AVOID BOLUSING UNLESS REQUIRING ACTIVE RESUSCITATION FOR SIGNIFICANT HEMODYNAMIC INSTABILITY***
Type and Screen: Pale, persistent tachycardia, ill-appearing; suspected splenic sequestration; Acute chest syndrome; Focal neurologic findings; Hgb <6 g/dl or 20% or more below baseline; Reticulocyte count <1%
Chest xray: cough, parental report of difficulty breathing or respiratory sx, chest pain, new hypoxemia, or clinical suspicion for pneumonia/acute chest syndrome
Blood and urine cultures: If febrile >38.3C (see Sickle Cell Disease Fever Management Pathway)
Urine/Serum HCG: Any female ≥12 years or <12 with menarche

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Mild Pain
Ibuprofen + Oxycodone
Patient Comfortable at 30 mins x 2
Discharge (See Discharge Criteria and instructions)
Persistent Pain

Moderate/Severe Pain or Pain refractory to home pain management
(If no IV access, place IV or consider IN/subQ route)

Initial Bundle
IV (Morphine OR Hydromorphone) + IV Ketorolac
Pain significantly improved
No significant improvement at 15 minute pain and sedation reassessment

IV Opioid Dose #2
Maintain or consider escalation of initial dose by 25%
Pain significantly improved
No significant improvement at 15 minute pain and sedation reassessment

IV Opioid Dose #3
Maintain or consider escalation of Dose #2 by 25%
Pain significantly improved
Reassess pain and sedation at 15 mins

Persistent Significant Pain
IV Opioid #4
Give same dose as Dose #3 and order PCA according to Hematology recs

Admission Criteria
- Pain inadequately relieved after 2 hours of appropriate IV treatment including 2 or more doses of an IV opioid
- Family uncomfortable with discharge or feels additional doses of parenteral analgesics will be needed
- Other sequelae of HbSS including but not exclusive to: focal neurologic findings, Acute chest syndrome, Splenic sequestration, Severe anemia

Patient comfortable
PO Opioid
Reassess pain and sedation in 1 hr
Patient remains comfortable

Discharge Criteria
Patients with pain relief:
- After 1-3 doses of IV analgesia
- Continued pain relief at least 60 mins after PO analgesia
- Absence of other complications of HbSS, tolerating PO, and safe home environment and Hematology f/u

Table 1. Pain Medications

Medication	Route	Admin	Dose	Comment
Fentanyl	IN	Use with atomizer Onset 5-10 mins	1.5 mcg/kg/dose Max 100 mcg/dose	Initial pain management prior to IV access
Ketorolac (Toradol)	IV	IV Push 2-3 min	0.5 mg/kg/dose Max 15 mg/dose	Initial dose should be given 4 hours after last dose of Ibuprofen
Morphine	IV	Slow IV push	0.1-0.15 mg/kg/dose Max 8 mg/dose	
Hydromorphone (Dilaudid)	IV	Slow IV push	0.015-0.02 mg/kg/dose Max 2 mg/dose	
Oxycodone	PO		<6 yo: 0.15 mg/kg/dose up to 2.5 mg; 6-12 yo: 0.2 mg/kg/dose up to 5 mg; >12 yo: 0.2 mg/kg/dose up to 10 mg	
Hydrocodone/ Acetaminophen	PO		0.2 mg/kg Max 10 mg Hydrocodone	If patient has not received acetaminophen in the last 4 hours
Ibuprofen	PO		10 mg/kg	alternative NSAID when IV Toradol not available or oral route preferred

Discharge Instructions
- Duration of scheduled narcotics and any changes to home pain management
- Hematology follow-up
- Important Sickle Cell Disease Clinic Numbers: Emergent question or consult at all hours **984-974-1000** and ask for pediatric hematologist on call; Routine questions from 8a-4p call office at **919-966-0178**