ED GUIDELINE

Sickle Cell Pain Crisis Management

< 21 years – Initial ED Management

Children's MINNESOTA

Aim: To reduce delays in analgesia administration to better manage pain and improve outcomes in patients presenting with Sickle Cell Pain Crisis.

0 MIN (Patient in room)

20 MIN

40 MIN

1 Hr

2-4 Hrs

Prehospital

- Hematologist calls ED to discuss anticipated pain plan
- **Rapid start orders placed by ED (ED Sickle Cell/Pain Rapid Start)**
- Mpls campus preferred

Patient arrives to ED

RN initial assessment (Triage and Rooming)

- Pivot/Triage Nurse will check for Pre-Registration account, activating appropriate account if available.
- Patient to be assigned ESI 2 (yellow) and room immediately, if possible; VS and start O2 for sats <92%. Notify provider.
- During rooming, or in triage if room not available, document last pain med administration at home.
- Preliminary assessment of mild/moderate/severe pain - document pain score 0-10 (Note 3)
  - Check for pre-arrival orders (*ED Sickle Cell/Pain Rapid Start*). If appropriate, offer analgesia through the *"ED Protocol for Pain and Fever"*
  - If pain score ≥4/10, refer to *"Sickle Cell Disease Pain Crisis w/wo Fever: Line Placement and Labs"* for prompt IV placement
  - If no *ED Sickle Cell/Pain Rapid Start* orders present, quickly discuss pain severity level with provider ASAP to expedite opioid orders recommended in next step
  - Offer heat pack, warm blanket, and avoid cold
- Give oral ondansetron if any nausea or vomiting

INCLUSION CRITERIA

- Patient with known Sickle Cell Disease
- Pain consistent with Sickle Cell Pain Crisis (Notes 1 and 2).

EXCLUSION CRITERIA

- Priapism
- Trauma
- Atypical pain symptoms: severe abdominal pain, concern for stroke, severe HA, altered mental status, new seizure, focal neurologic finding.

Moderate/Severe Pain (Note 3)*

- Intranasal fentanyl 1-2 mcg/kg (max 100 mcg) if ≥ 3 yrs old
- IV Morphine 0.1 mg/kg (max 6 mg) (or per patient established pain plan)
- IV Ketorolac 0.5 mg/kg (max 15 mg) if no NSAID in last 6 hours
- Labs: CBC, BMP, Retic count, LDH, Total/Direct Bili
- IV fluids at maintenance rate unless dehydrated and needs bolus
- **IV access delayed beyond 30 min?** Consider Intranasal (IN) Fentanyl, Intramuscular (IM) Ketorolac, Oral oxycodone (Note 4 for dosing).

***If available, review individualized pain plan for med substitutions or guidance**

Mild Pain (Note 3)*

- Oral Ibuprofen 10 mg/kg (Max 600 mg) if no NSAID in last 6 hours
- Oral oxycodone 0.05 - 0.1 mg/kg (max 10 mg) (or per patient established pain plan)

Please consider accompanying SCD related conditions: fever, acute chest syndrome, acute multi-organ failure, acute splenic or hepatic sequestration, delayed hemolytic transfusion crisis, renal infarct, cerebral infarct/stroke

Nurse to reassess pain every 30 min

If not improving, nurse to ask provider to order opioid (50% original dose or per patient established pain plan), ensure NSAID, acetaminophen, and comfort measures are maximized

Nurse to reassess pain every 60 min

Determine disposition after 3-5 cycles (2-4 hours)

Admission: Admit to Minneapolis

- ED provider: place IV morphine PRN dose (0.05 mg/kg, max 3 mg) in ED transition of care orders (or per pain plan)
- Hematologist: preps admit orders including **STAT** Patient-Controlled-Analgesia (PCA) to be started on arrival to floor (Note 5)
- RN: Continue q30 min pain assessment and discuss with provider if not improving

Discharge:

- Patient required ≤3 doses of opioid and last dose was oral opioid with sustained pain control. Home plan feasible for family.
- Lab evaluation reassuring
- Absence of other complications of Sickle Cell Disease (Note 6)
- Reviewed with Hematologist on call
- Discharge education and follow-up completed (Note 7)

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NOTES:

Note 1. Signs/Symptoms consistent with Sickle Cell Pain crisis includes extremity pain, back pain, abdomen or chest wall pain often without overlying skin or joint changes.

1. Reported pain remains the gold standard for pain assessment
2. Assess vital signs and lab values (CBC, retic, T/D bili, LDH, BMP)
3. Differential diagnosis: Skin/Soft Tissue Infection, osteomyelitis, vascular necrosis, trauma, gout, arthritis, DVT/PE, MI, stroke, migraine, meningitis, stroke, acute cholecystitis, pelvic inflammatory disease, constipation. Please evaluate and exclude etiology from standard of care (i.e. abdominal pain appendicitis etc.)
4. Please consider accompanying SCD related conditions: Fever, acute chest syndrome, acute multi-organ failure, acute splenic or hepatic sequestration, delayed hemolytic transfusion crisis, renal infarct, stroke. If concern for stroke, then excluded from this guideline and move to ED suspected stroke guideline, consulting heme STAT and other consultants per stroke guideline.

Note 2. Possible triggers of Sickle Pain include Infections, dehydration, stress, menses, alcohol, rapid temperature change, prior steroid course (rebound pain).

Note 3. Mild vs Moderate vs Severe pain episodes:
Reported pain remains gold standard for pain assessment. Utilize pain scores to help assess pain level. Because each patient has a unique experience for each pain scale number, also assess how patient would rate their score of mild (generally 1-3), moderate (generally 4-6), or severe pain (generally 7-10).

Note 4. Dosing of pain medication:

1. Acetaminophen 15 mg/kg (Max 650 mg) by mouth
2. Intranasal (IN) Fentanyl 1-2 mcg/kg (Max 100 mcg) if ≥ 3 yrs old. Can repeat every 1 hour
3. Ibuprofen 10 mg/kg (Max 600 mg) by mouth
4. Ketorolac 0.5 mg/kg (Max 15 mg) IM or IV; must be >= 6 hours from last NSAID dose (Oral ketorolac is **not** on formulary)
5. Oxycodone 0.05 - 0.1 mg/kg (Max 10 mg) by mouth
6. Morphine 0.1 mg/kg (max 6 mg) IV

Note 5. Patient-Controlled Analgesia (PCA) Pumps

1. Hematology preps STAT PCA order which will be initiated once admitted. Simultaneously, provider will place order for naloxone drip at 2 mcg/kg/h. As needed naloxone bolus dose for oversedation will automatically order within the EMR secondary to the PCA order.
2. PCA Medications and Dosing
    - If patient has not had PCA previously, then start morphine PCA. Otherwise, per pain plan or last PCA used during hospital stay
    - Morphine PCA: morphine 0.015 mg/kg/h continuous infusion (max 0.75 mg/h), 0.015 mg/kg demand dose (max 0.75 mg), 10-minute lockout, maximum hourly dose = 0.075 mg/kg/h (max 3.75 mg/h)
    - All patients on opioid PCA should have naloxone drip starting at 2 mcg/kg/h
    - Side effects (treatment options) include:
        - Pruritus (diphenhydramine, naloxone drip)
        - Nausea/vomiting (ondansetron)
        - Constipation (polyethylene glycol, senna, bisacodyl, docusate)
        - Respiratory depression (naloxone 0.01 mg/kg/dose (Max 0.4mg) IV or IN to alleviate sedation – not reverse analgesia)

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Note 6. Complications of sickle cell that would inhibit discharge:

1. Respiratory symptoms/hypoxia concerning for Acute Chest Syndrome
2. Focal neurologic symptoms or unevaluated headache
3. Significant marrow suppression (Hgb below baseline without adequate reticulocytosis)
4. Evidence of splenic sequestration
5. Priapism
6. Acute infectious concerns requiring admission

Note 7. Patient discharge education and follow-up:

1. Continue oral ibuprofen and acetaminophen every 6 hours for 2-3 days after opioid discontinued. Advise family of timing for next doses
2. Ensure family has 12 doses of oral Oxycodone (3 days of q6 hours or 4x/day dosing) at time of d/c unless otherwise directed by pain plan. Advise family of timing for next doses.
3. Review non-pharmacologic pain management options
    - Deep Breathing, Progressive Muscle Relaxation, Imagery, Self-Hypnosis, Aromatherapy, Acupressure, Distraction, Massage, Application of Heat, Guided Imagery
4. Ask family to call the following day to give an update on symptoms; patient can also call sooner if worsening (if Mon-Fri, patient should call heme clinic; if Sat-Sun or overnight, patient should call the on-call provider). Hematologist will also notify Heme RN as well to reach out during weekdays.

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