ED GUIDELINE

Sickle Cell Vaso-Occlusive Pain Episode 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 vaso-occlusive pain episode (VOE).

0 MIN (Patient arrival)

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.
- Triage as ESI 2 (yellow) and room immediately, if possible; VS and start O2 for sats <92%. Start continuous saturation monitoring. Notify provider and child life specialist
- 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 Pain or Fever*"
  - 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
  - If pain score ≥4/10, refer to "*ED Protocol Sickle Cell Crisis IV Placement*" for prompt IV placement with child life assistance as needed
  - Offer heat pack, warm blanket and avoid cold
- Give oral ondansetron if any nausea or vomiting

INCLUSION CRITERIA

- Patient with known Sickle Cell Disease (SCD)
- Pain consistent with Sickle Cell vaso-occlusive episode (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 (Pain score ≥ 4, Note 3)*

- Medications (in order of priority; Note 4)
  - Intranasal fentanyl 1-2 mcg/kg (max 100 mcg)
  - 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
  - Oral (or IV) acetaminophen 15 mg/kg (Max 650 mg) if none in last 4 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 (Pain score 1-3) Note 3)*

- Oral Ibuprofen 10 mg/kg (Max 600 mg) if no NSAID in last 6 hours
- Oral acetaminophen 15 mg/kg (Max 650 mg) if none in last 4 hours
- Oral oxycodone 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

- Go to page 2 for PCA initiation in the emergency department if the patient is in Minneapolis. If patient is in St. Paul, continue to re-dose opioids per guideline while awaiting transport.
- ED provider orders ongoing opioid doses as needed before PCA initiation.

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)

ED GUIDELINE

Sickle Cell Vaso-Occlusive Pain Episode Management

< 21 years - PCA Initiation for Minneapolis ED

Children's MINNESOTA

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INCLUSION CRITERIA
- Patient with known Sickle Cell Disease
- Pain consistent with Sickle Cell vaso-occlusive episode (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.

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

2-4 Hrs

Patient in Minneapolis and will be admitted for sickle cell disease vaso-occlusive pain episode. ED physician calls hematologist to discuss admission.

ED PROVIDER:
- Place ED transition of care orders
- Discuss admission with patient. Explain that PCA will be started in Emergency Department (unless room ready for patient immediately upstairs)
- Provide ongoing pain management until PCA started (continue q30 min pain assessment). Ensure pain control adequate before starting PCA.

ADMITTING HEMATOLOGIST*:
Add *Hematology Sickle Cell Admission Orderset*: **Sign** PCA order phase and **pend** Phase I and II (Note 5)
- Order naloxone drip and naloxone PRN in PCA phase
- Note: Acetaminophen/ketorolac scheduled q6 hours and bowel regimen can be ordered in the PCA phase, but these will not be verified/started until on the floor unless specifically asking the nurse to do so.

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

NURSING:
PCA to be started in the Emergency Department:*
- RN: Initiate patient on PCA and naloxone drip. If pain not under control before PCA is started, reach out to ED physician for STAT pain control optimization before PCA.
- RN to reach out to:
  - Hematology if questions/concerns regarding pain while on PCA or PCA questions
  - ED attending if other concerns or needing immediate assistance

*If room is ready on 7th floor and patient can be moved up immediately, move patient to 7th floor and then initiate PCA on floor. Otherwise, don't delay PCA initiation while awaiting transfer.

Nurse can follow PCA Step Up instructions in order. Notify hematology if concerns. If applicable, optimize acetaminophen/ketorolac re-administration through ED physician.

Nurse to reassess pain per PCA orders

Patient moved to 7th floor Minneapolis when room is ready and admitted to hematology.
- Hematologist signs phase I and II of admission orders.
- ED and floor nurse perform PCA double check on patient's arrival to floor

Sickle Cell Vaso-Occlusive Pain Episode Management

< 21 years – Notes

Aim: To reduce delays in analgesia administration to better manage pain and improve outcomes in patients presenting with Sickle Cell vaso-occlusive pain episode (VOE).

NOTES:

Note 1. Signs/Symptoms consistent with Sickle Cell vaso-occlusive episode 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. Intranasal (IN) Fentanyl 1-2 mcg/kg (Max 100 mcg). Can repeat every 1 hour. Goal to utilize before or during IV placement. If patient gets IV placed quickly, it is ok to give both IN Fentanyl and IV morphine at the same time in patients that are NOT opioid naïve, which is typically patients with sickle cell disease in a vaso-occlusive episode.
2. Morphine 0.1 mg/kg (max 6 mg) IV
3. Acetaminophen 15 mg/kg (Max 650 mg) by mouth
4. Ibuprofen 10 mg/kg (Max 600 mg) by mouth
5. 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)
6. Oxycodone 0.05 - 0.1 mg/kg (Max 10 mg) by mouth

Note 5. Patient-Controlled Analgesia (PCA) Pumps

1. Hematology places signed orders for STAT PCA and 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. PCA will be initiated in the Emergency Department by ED nurses.
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 (ambulation as tolerated, increased fluid intake, 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. Re-dose as necessary, no minimum time required between doses given rapid onset of action.

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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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REFERENCES:

1. Brandow A, et al, American Society of Hematology 2020 guidelines for sickle cell disease: management of acute and chronic pain. *Blood Adv* (2020) 4 (12): 2656–2701 https://doi.org/10.1182/bloodadvances.2020001851
2. Children's Hospital of Philadelphia, Umana J, et al., Emergency Department Clinical Pathway for the Evaluation/Treatment of Children with Sickle Cell Disease and Pain. Posted: July 2010
   Revised: March 2024. Available from Sickle Cell Disease with Pain Clinical Pathway — Emergency Department | Children's Hospital of Philadelphia (chop.edu)
3. Seattle Children's Hospital, Bender, M., Burns, B., Crandall, C., Dassler, A., Herrman, A., Hrachovec, J., Lo, M., Reichert, E., Staples, S., Thomas, A., Migita, D., 2024 January. Sickle Cell ED Vaso-occlusive Pain Pathway. Available from CSW Sickle Cell - ED Vaso-occlusive Pain Pathway (seattlechildrens.org)
4. New England Pediatric Sickle Cell Consortium: Management of Acute Pain in Pediatric Patients with Sickle Cell Disease (Vaso-Occlusive Episodes). Available from Microsoft Word - PAIN CPG 2-09.doc (nepscc.org)