# *CLINICAL PATHWAY:*
# Sickle Cell: Management of Acute Pain Crisis

THIS PATHWAY SERVES AS A GUIDE AND DOES NOT REPLACE CLINICAL JUDGMENT.

**Inclusion Criteria:** ≥7 years old with Sickle Cell Disease during an acute painful crisis that failed outpatient home management
**Exclusion Criteria:** Patients with Sickle Cell Disease that are <7 years old, have neurological symptoms, or concern for acute chest. If febrile, please refer to **Sickle Cell Disease with Fever Pathway**.

**[1]History should include:**
- Similarity to previous sickle cell pain
- Analgesics used for this episode
- Allergies

**[1]Physical Exam should include:**
- Vitals, O2 sat
- Hydration status
- Pallor
- Potential infection
- Spleen size
- Penis (priapism)
- Neurological
- Other potential etiology of pain other than Sickle Cell related pain crisis

**Emergency Room Initial Management:**

*Goal of care: initial assessment and first dose of opioid given within 1 hour of presentation.*

Level of Care: ESI 2

- Obtain thorough history and comprehensive physical examination[1]
- Obtain IV access (see **Venous Access – Emergency Department Care Pathway**). If unable to obtain IV access, notify attending to consider alternate opioid route (intranasal, PO, subQ)
- Labs and imaging: obtain CBC w/diff and reticulocyte count; if chest pain, consider CXR
- Review most recent creatinine and do not administer Ketorolac if above normal range
- ***Review Pain Plan in "Letters" section of Care Navigator***

**Emergency Room Pain Management:**

- Ketorolac IV 0.5 mg/kg/dose (max 15 mg/dose) x1 dose AND/OR Acetaminophen PO/IV 15 mg/kg/dose (max 1 g/dose) x1 dose
- If patient failed PO opioid at home: give IV opioid per Pain Plan
- If patient does not have Pain Plan: morphine IV 0.1 mg/kg/dose (max 5 mg/dose) or morphine PO 0.3 mg/kg/dose (max 15 mg/dose)at attending's discretion
- If unable to obtain IV access, consider intranasal fentanyl 1-2 mcg/kg/dose
- IVFs at the discretion of the provider and/or patient with signs of dehydration
- Reassess 30 minutes after opioid dose:
  - If insufficient improvement in 30 minutes, give 2nd dose of opioid PO/IV per Pain Plan. If no Pain Plan, repeat IV morphine 0.05 mg/kg/dose (max 2.5 mg/dose) *OR* PO morphine 0.15 mg/kg/dose (max 7.5 mg/dose) at attending's discretion
  - If insufficient improvement in 30 minutes after 2nd dose of opioid, give 3rd dose PO/IV per Pain Plan. If no Pain Plan, IV morphine 0.05 mg/kg/dose (max 2.5 mg/dose) *OR* PO morphine 0.15 mg/kg/dose (max 7.5 mg/dose) at attending's discretion
- See "Adjuvant Medications" below for GI, pruritus, and nausea management

**Disposition:**

Call on-call Heme/Onc attending to notify of discharge or admission

- **ED Discharge criteria:** Pain relief after 1-3 doses of IV opioids and no other complications of sickle cell disease
- **Admission criteria:** Pain insufficiently controlled after 3 opioid doses
  - Admit to Inpatient Services (Heme/Onc)
  - Consider initiating inpatient pain plan below prior to transfer to avoid delays in analgesic administration

## Opioids

**Order opioids ASAP upon admission. Review Pain Plan in "Letters" section of Care Navigator.**

*If home regimen includes PO long-acting opioid (eg, Methadone, oxyContin, or MS contin):*
- Continue home long-acting regimen per Pain Plan
- AND add bolus-only PCA (no continuous). For Connecticut Children's Employees, please refer to **Connecticut Children's PCA Policy**.
- **HOLD** PO immediate release opioid

*If patient not on long-acting opioid:*
- Schedule their immediate release opioid ATC as per Pain Plan
- AND add bolus only PCA (no continuous). **Connecticut Children's PCA Policy**.
- Do not start a long-acting opioid

Considerations:
- *If opioid naïve,* do not use continuous PCA infusion or ATC PO opioids
- *If ≤7 yrs old:* consider intermittent IV opioids PRN or authorized agent controlled analgesia (AACA)
- *If >7 yrs old:* consider PCA
- *If not tolerating PO:* may utilize PCA + continuous infusion. **Connecticut Children's PCA Policy**.

Continued monitoring per primary team with goal of transitioning to PO pain regimen within 3 days if clinically appropriate

## Non-Opioid Pain Management

- Ketorolac IV: 0.5 mg/kg/dose (max 15 mg/dose) q6hrs up to 20 doses
  - Max: no more than 20 doses in 30 day period
  - At, or before, 20th dose: change to PO Ibuprofen: 10 mg/kg/dose (max 600 mg/dose) q6hr ATC

AND

- Acetaminophen IV/PO: 15 mg/kg/dose (max 1000 mg/dose or 4 g/day) q6hr ATC for 2-3 days

## Adjuvant Medications

- Offer Lidoderm patches for regional pain in patients ≥6 years of age (12 hours on, 12 hours off) q24 hours [may take several days to reach full effect]

**Further Considerations:**

*If insufficient pain control within the first 24 hours, may consider adding the following to the opioid + non-opioid plan:*
- Ketamine 2 mcg/kg/min, escalating by 2 mcg/kg/min every 8-12 hours as needed (max 6 mcg/kg/min)

*If patient has isolated limb pain, may consider regional anesthesia.*

**Other Considerations:**

**GI:**
- Famotidine PO while on NSAIDS: 0.5-1 mg/kg/day divided daily or BID (max 40 mg daily)
- Start bowel regimen while on opioids with goal of one stool per day:
  - Miralax 8.5 g-17 g daily
  - +/- Senna: 1-2 tabs BID (Colace not recommended)

**Pruritus:**

*If on long-acting opioids or PCA with continuous opioid infusion, consider the following:*
- Low dose naloxone infusion: 0.25 mcg/kg/hr
- Nalbuphine IV PRN:
  - <50 kg: 0.1 mg/kg/dose (max 2.5 mg/dose) q6hr PRN
  - ≥ 50 kg: 2.5-5 mg/dose (max 5 mg/dose) q6hr PRN
- Ondansetron PO/IV: 0.15 mg/kg/dose q8hr PRN (max 8 mg/dose)
- Diphenhydramine is contraindicated for opioid-induced pruritus

**Nausea:**
- Ondansetron PO/IV: 0.15 mg/kg/dose q8hr PRN (max 8 mg/dose)

## Adjuvant Therapies

- Encourage functional plan (**Appendix A**)
- Notify Sickle Cell Social Worker
- If patient previously used TENS, encourage use

**Consult:**
- Massage therapy – obtain consent on admission.
- Integrative Medicine
- Child Life
- Case Management

**Considerations:**
- Pain Team consult if pain not improved after 24 hours, or with hx of chronic pain
- Psych consult if existing relationship with psychology or presents with emotional/behavioral issues (if seen by Hem/Onc psych, please note in consult comments)
- PT +/- TENS if inpatient >24 hrs with little/no improvement

**Discharge Criteria and Instructions:**
Pain well-controlled on PO Pain Plan, return to baseline functionality

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## Appendix A: Functional Plan for Patients with Sickle Cell

1.) Regulate sleep/wake cycle - Lights on/blinds open 0900, lights off/electronics off 2200.
2.) Changing for bed into "night clothes" and getting dressed in clothes in AM (if able).
3.) Daily or every other day shower.
4.) Complete activities of daily living (ADL's) independently as tolerated.
5.) Out of bed (OOB) for meals/during meal times if not eating. As admission progresses, OOB more than exclusively for meals (after day one or two) with a rest break in bed in the morning and in the afternoon for up to 1 hour only.
6.) Participation in floor activities. Out of bed, preferably in play room rather than bed side, for special Child Life events, Hole in the Wall Gang Camp activities and art/play projects.
7.) For frequent flyers: school work.
8.) Walks around med/surg unit per PT and/or Primary Team

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