Clinical Pathway for Evaluation/Treatment of Adults with Sickle Cell Disease and Pain (ED)

Time elapsed from arrival

Triage

ED Team Assessment

Goals	• Administer pain medication within 60 mins of patient's arrival to the ED with crisis pain • Reassess pain 30 minutes after IV administration • Provide additional doses of opioids for persistent pain in a timely way
Considerations	• Use **oral diphenhydramine** for opioid induced pruritis. – Avoid IV diphenhydramine due to increased sedation • Use IV ondansetron for opioid induced nausea • Use non-pharmacologic measures such as heating pads and lidocaine patches for local pain relief • Use IV fluids as clinically indicated for dehydration/ intravascular volume depletion • **Caution:** Corticosteroids may exacerbate crisis
Vascular Access	• Place IV, ultrasound-guided as needed • If delayed due to poor venous access, consider SubQ opioids for pain control
Initial Work-Up	History and Physical Exam **Labs:** • STAT CBC w/diff, retic count, CMP • Consider UPT in all post-menarchal females • Urine and blood cultures if febrile • Type and Cross should be drawn for: – Focal neuro deficits – Concern for acute chest syndrome (ACS) – Hgb < 6 g/dL or Hgb drop >2 g/dL from baseline » Call hematology service if acute anemia is present – **Be mindful that blood products may be delayed due to antibodies and early typing may be warranted** **Imaging:** • Chest x-ray – Fever, respiratory symptoms, hypoxia, or otherwise indicated – **Consult hematology service if chest infiltrate is present to discuss admission as decompensation can occur with acute chest syndrome**

40 min

Management of Moderate/Severe Pain

Initial Bundle	15 mg IV Ketorolac* **and** Morphine 6 mg OR (0.1 mg/kg) **OR** Hydromorphone 1 mg OR (0.01 mg/kg) *Do NOT use if patient has nephropathy/CKD
Repeat IV Opioid Dosing	**Increase IV opioid dose by 50% if no improvement in pain levels after 30 mins** **Maximum recommended dose:** Morphine 8 mg Hydromorphone 1.5 mg
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60 min

Reassess in 30 minutes

90 min

Reassess in 30 minutes

120 min

150 min

180 min

Determine Disposition, Hematology Consult

Persistent pain

Admit, consult Hematology

IV Opioid #4

- Start PCA if available (preferred) or every 2 hr scheduled opioid
- Schedule ketorolac 15 mg IV every 6 hours if normal renal function
- Hourly pain assessment
- Continue non-pharmacologic interventions

Suggested PCA starting doses

Morphine (IV)
2 mg q15 min with 32 mg 4-hour lockout

Hydromorphone (IV)
0.5 mg q15 min with 16 mg 4-hour lockout

Fentanyl (IV)
50 mCg q15 min with 800 mg 4-hour lockout

Pain controlled, absence of other complications of sickle cell disease

Discharge

Educational Media

Managing Sickle Cell Disease in the ED, ACEP

PEM Episode 9: Sickle Cell Disease

Evidence

ASH 2020 Guidelines for SCD:
Management of Acute and Chronic Pain

Clinical Pearls

Patients may have learned distraction techniques for pain coping on assessment which can include:

- **Sleep**
- **Television**
- **Phone use**
- **Headphones/music**
- **Apps/games**
- **Comfort measures**

Most patients have received some form of IV pain medication since childhood. We can trust they know which medication works best for them.

If a patient suggests a medication that worked well for them in the past, this is self-advocacy. Please do not misconstrue this behavior as drug seeking.

Please refer to individualized treatment plan that patient has been advised to carry

Re-evaluating pain every 30 minutes expedites disposition to either discharge or admission!

Discharge considerations:

- Pain rating does not have to be 0/10, however, provider should discuss with patient if they can feel like pain can be managed at home or if admission is needed.
- Encourage follow-up with outpatient hematologist within 7-10 days
- If sending in a 3-5 day supply of opioid medication, pick a 24 hour pharmacy if patient is seen after hours
- Advise patient to alert pain medication prescriber that a script was given in the ER if they are on an outpatient pain mgmt contract

References:

1. Brandow AM, Carroll CP, Creary S, et al. American Society of Hematology 2020 guidelines for sickle cell disease: management of acute and chronic pain. Blood Advances. 2020;4(12):2656-2701. doi:https://doi.org/10.1182/bloodadvances.2020001851
2. Philadelphia TCH of. Sickle Cell Disease with Pain Clinical Pathway Emergency Department. www.chop.edu. Published August 12, 2014. https://www.chop.edu/clinical-pathway/sickle-cell-disease-with-pain-clinical-pathway

Disclaimer: Innovative Hematology, Inc. ("IHI") developed the Sickle Cell Disease Emergency Department ("SCD ED") Clinical Pathway as an evidence-based tool and reference for Emergency Department providers treating patients with sickle cell pain. Clinical Pathways provide best practices and are intended to assist health care providers in clinical decision making. The SCD ED Clinical Pathway is based upon publicly available medical evidence and consensus of health care providers at IHI. The SCD ED Clinical Pathway is not intended to substitute for the independent professional judgement of the treating health care provider as there may be individual variation among patients.

The health care provider treating the patient assumes all risks associated with clinical decision making. IHI assumes no responsibility for any adverse consequences or omission that may arise from the use or reliance on the SC ED Clinical Pathway. IHI reviews Clinical Pathways periodically, however new evidence or developments may not be represented.

If you have any questions/comments, please contact our team 317-871-0000.

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