University Hospital Emergency Department Clinical Practice Guideline
Management of Sickle Cell Anemia Vasoocclusive Crisis (VOC)

VOC is the hallmark acute complication for persons with sickle cell disease (SCD) and manifests as acute, severe pain. Best practice suggests rapid triage, care area placement, and administration of analgesics

The Emergency Severity Index (ESI) Version 4 triage system suggests that patients with VOC be triaged as **ESI level 2**, a very high priority, and rapid evaluation and treatment should be facilitated

- MD/RN rapid assessment and treatment may include, but is not limited to: IV access, cardiac monitor, pulse oximetry, CBC with differential, reticulocyte count, CMP
    - Pain assessment
    - Analgesics
    - Physical exam
    - Specific etiology of crisis

Goals of ED management:

- Administration of analgesia within 30 minutes of patient arrival
- Assessment for etiology of pain, concomitant disease, complication of VOC
- Appropriate titration of analgesics to adequately control pain
- Frequent assessment and documentation of patient's pain by bedside nurse
- Use of adjuncts for pain management, e.g., distraction (e.g. music, movies) as available

Pain management is best guided by **patient report** of pain severity along with clinical judgment

- Biomarkers or imaging cannot specifically validate pain or assess severity
    - Documentation of sickle cell anemia should be obtained
- Guidelines supplemented with clinical judgment should be utilized in selecting drug, dose, and route of administration
- If opioids are felt necessary, the recommendation is to start low and titrate slowly, as needed
- Goal is to safely decrease pain score by at least 2 points on a numerical rating scale; goal is not necessarily to obtain complete pain relief.

Pain management typically includes use of opioids for analgesia for patients with severe pain

- Addition of NSAIDs reduces pain and decreases hospital length of stay
- Refer to attached algorithms for recommended management of adults and pediatrics with VOC

Supportive care should be patient specific

- Consider maintenance rate of IV hydration in euvolemic patients who are unable to drink fluids
- Consider supplemental oxygen in patients with an oxygen saturation <95%

Careful consideration should be given to discharge medications

- NSAIDs (ibuprofen or naproxen) should be utilized primarily for outpatient pain control
- Many patients are on home opioids, often at high doses. These patients should be referred to their primary care, sickle cell, or pain management provider for medication.
- The NJ PDMP should be reviewed for consistency with patient report

Disposition decision should be made within **2-4 hours**

References:

- National Heart, Lung, and Blood Institute. Evidence-based management of sickle cell disease: expert panel report, 2014. September 2014. http://www.nhlbi.nih.gov/ (last accessed 29 November 2016)

Management of Sickle Cell Anemia VOC in Adults

- Assess pain score and home opioid and non-opioid pain medications
- NSAID (choose one)
  - **Ketorolac 15 mg IV** Q4-6 hours (max 15 mg per dose, 90 mg in 24 hours)
    - Pediatrics: **Ketorolac 0.5 mg/kg IV** (max 15 mg per dose)
  - **Ibuprofen 200-400 PO mg** Q4-6 hours (max 3200 mg in 24 hours)
    - Pediatrics: **Ibuprofen 10 mg/kg PO** (max 400 mg per dose)
- Non-pharmacologic therapy: heat pack or topical 2% lidocaine jelly on joints
- Consider adjuvant therapy
  - **Ondansetron PO/IV** for nausea or vomiting
  - **Diphenhydramine PO** once if there is itching associated with opioid therapy
    - Not to be administered with each dose
    - Diphenhydramine not to be administered IV for this indication

Opioid Naïve

Mild-moderate pain:
- NSAIDs typically adequate for treatment

Severe pain:
- If able to tolerate PO: Administer **morphine IR 15 mg PO**
  - Pediatrics: **morphine solution 0.2 mg/kg PO** (max 10mg)
- Alternative: Administer **intranasal fentanyl 1-2 mcg/kg** (maximum of 1mL administered per nostril)
- If unable to tolerate/fail PO: Administer **morphine 0.1 mg/kg IV** (max dose 10 mg)

Reassess pain within **30 minutes**

If pain is not controlled, place patient on capnography AND repeat initial dose of **morphine IV***

If pain is adequately controlled, consider transition to PO and discharge (with NSAIDs)
If pain is not controlled, patient should be admitted or placed in observation

Opioid Tolerant

- Assess patient home dose of opioids
- If able to tolerate PO: Administer **PO morphine IR** based on home dose

Patient Home PO Analgesic	Patient Home PO Analgesic Dose	**Recommended PO Morphine IR Dose**
Oxycodone	5 mg	15 mg
	10 mg	30 mg
Morphine	15 mg	30 mg
	30 mg	45 mg
Hydromorphone	2 mg	15 mg
	4 mg	30 mg

- Alternative: Administer **intranasal fentanyl 1-2 mcg/kg** (maximum of 1mL administered per nostril)
- If unable to tolerate/failed PO: Administer **morphine 0.2 mg/kg IV** (max dose 16 mg)

Reassess pain within **30 minutes**

- If pain is not controlled, place patient on capnography AND administer **morphine 0.2 mg/kg IV** (max dose 16 mg)

Reassess pain within **30 minutes**

If pain is not controlled, administer **ketamine 0.3 mg/kg IV*** over 15 minutes

If pain is adequately controlled, consider transition to PO and discharge (with NSAIDs)
If pain is not controlled, patient should be admitted or placed in observation

***Morphine IV**
- Administer as slow IV push
- Administer subcutaneously if no IV access
- Consider lower initial dose in elderly or obese patients

+Ketamine IV
- 0.3 mg/kg is the pain management dose
- Administer as slow infusion over 15 minutes
- Higher doses can lead to dissociation
- Please counsel your patient on effects of ketamine including delirium, sense of unease, etc.
- Avoid use in patients with history of psychosis

Developed by ED Guideline Committee, December 2017 (Latest revision – January 2019)