Sickle Cell Suspected VOE Clinical Pathway

September 2021

Outcomes/Goals	**1.** Create an efficient, team-oriented approach for patients with sickle cell disease presenting with possible VOE to achieve rapid evaluation, treatment, and disposition **2.** Manage pain to a level acceptable to the patient and in accordance with OHSU Evidence-Based Practice Guideline
Inclusion Criteria	1. Patients with SCD of any age still followed by OHSU Pediatric Hem/Onc or non-OHSU patients with SCD <20 yo presenting with pain
Exclusion Criteria	1. Pts presenting with T≥38.3°C (refer to ***sickle cell with fever*** pathway)
NURSE Documentation	Document location of pain, symptoms associated with pain, and treatments that have worked and not worked. Document evidence of shock/decompensation, mental, respiratory and circulatory status. Document presence of fever. Document presence of central line or port and any history of line infections. Medications, allergies, vital signs, height and weight per Peds ED NPEOC.
INTERVENTIONS Initiate on arrival	ESI II Place on continuous cardiac and pulse ox monitor. Apply Oxygen for SpO2<92% and/or patient comfort. PIV or access central line/port per CLABSI prevention Bundle policy; policy # HC-NSG-259-POL and HC-NSG-260-PRO Consider NS fluid bolus if signs or symptoms of dehydration or hypovolemia **Offer intranasal fentanyl 2 µg/kg x1 dose** **(For patients ≥10 kg; max single dose 100 µg)**
DIAGNOSTICS	CBC, with Diff, Reticulocyte Count, CMP, Type and screen, LDH total, Bili direct Chest X-ray, if c/o chest pain, SOB, demonstrating increased WOB, or O2 need HCG in females ≥ 12 or younger if menarche
PHYSICIAN (LIP)	
Documentation	Assess, document pain, document recent pain medication, dose, time of last dose Allergies to any medication Order pain medication immediately (see below)
Fluids Bolus	Consider NS 20 mL/kg bolus if signs or symptoms of dehydration or hypovolemia (no proven therapeutic benefit)
Blood Products	*Consult Pediatric Hem/Onc prior to transfusion to determine number of units to transfuse and whether an exchange transfusion is indicated*
Medication **Pain**	IN fentanyl PO NSAID, PO oxycodone (mild/moderate pain) *if not tried at home* IV ketorolac if normal renal function, IV opioid (moderate/severe pain) **(See pain meds panel on flowsheet page)**
ADMISSION	Consult to Pediatric Hematology Oncology
*High Risk versus Low Risk Considerations	**Low Risk:** • Well-appearing with Stable vital signs • Tolerating good po • No concern for acute chest syndrome or sequestration crisis • Tolerating po pain medication • No new hypoxemia and chest x-ray without infiltrate • Reticulocyte count at baseline • Patient and family comfortable with plan **High Risk**: • Toxic/ill appearing • Oxygen requirement • Multiple recent ED visits for pain

Suspected Vaso-Occlusive Episode (VOE)

Immediate Action

- Place on continuous cardiac and pulse ox monitor.
- Offer and administer **IN fentanyl** (for pts ≥ 10 kg, 2 mcg/kg; 100 mcg max)
- Determine pain level, last home dose of NSAID, and order appropriate NSAIDs and PO/IV opioids
- PIV placement
- CBC, reticulocyte count, HCG, BMP if getting NSAIDS
- Give 20 ml/kg NS bolus IF signs of dehydration or hypovolemia (DO NOT OVER-RESUSCITATE—can precipitate ACS, worsen aplastic crisis, cause cerebral edema with CVA)

Fever > 38.3 or 101?

YES

Fever Management·

- All pediatric patients with SCD are considered functionally asplenic and can devolve to severe sepsis quickly
- Go to ***sickle cell with fever*** pathway

NO

Headache or Neuro findings?

YES

Evaluate for Stroke:

- Careful neurologic exam
- Emergently consult Peds Neuro, Peds PHO
- CVA in SCD requires emergent RBC apheresis through a dialysis catheter

NO

Chest pain, pain "typical" for VOE?

YES

Evaluate for Acute Chest Syndrome:

- Splinting from pain can lead to atelectasis and vasoocclusive damage in lungs causing ACS
- Diagnostic Criteria:
  - new pulmonary infiltrate
  - *PLUS*
  - ANY of the following: fever, cough, hypoxia, tachypnea
- If suspicion for ACS, obtain CXR PA & lateral
- If concern for ACS:
  - Encourage incentive spirometry
  - Supplemental oxygen if SpO2<92%
  - Empiric broad-spectrum abx (see ***sickle cell with fever*** pathway)
  - Consult PHO

NO

Patient anemic relative to baseline?

YES

Anemia Management in SCD:

- If anemic and retic count inappropriately low/normal, consult hematology for possible aplastic crisis
- If anemia below baseline, consult hematology before considering transfusion

NO

Treat pain and review possible causes:

- Possible causes include infection, cold, stress, extreme exercise, dehydration, alcohol, menses, pregnancy, exacerbation of other medical issues, idiopathic
- Treat pain according to VOE PAIN MANAGEMENT algorithm below

VOE PAIN MANAGEMENT

Mild pain (0-4) OR no trial of PO analgesics at home prior to ED

Ibuprofen + Oxycodone

Reassess after 30-60 mins

Patient comfortable

Persistent pain

Moderate/severe pain (5/10-10/10) and failed PO analgesic trial

IV Bundle
- IV hydromorphone OR IV morphine
- IV ketorolac*

Reassess after 15-30 mins

Patient comfortable

Persistent pain

IV Opioid #2
- Initial dose + 25%

Reassess after 15-30 mins

Patient comfortable

Persistent pain

IV Opioid #3
- Initial dose + 50%

Reassess after 15-30 mins

Patient comfortable

Persistent pain

Admit

Oral Opioid

Discharge home with enough pain meds to get them to next PHO clinic (usually Thursday)

Follow personal pain plan as outlined in PHO clinic note. If no personal pain plan, consider starting medication dosing as follows:

PO Meds

Oxycodone
- < 6 yrs: 0.15 mg/kg up to 2.5 mg
- 6-12 yrs: 0.2 mg/kg up to 5 mg
- > 12 yrs: 0.2 mg/kg up to 10 mg

Ibuprofen
- 10 mg/kg

IV Meds

Ketorolac*
- 0.5 mg/kg, 30 mg max

Morphine
- 0.1-0.15 mg/kg, 8 mg max

Hydromorphone
- 0.015-0.02 mg/kg, 2 mg max

*Ketorolac
- Ketorolac may be given within 1 hour of last PO NSAID dose as escalation of therapy if no hx of renal insufficiency
- If patient has significant dehydration/volume depletion, consider NS bolus prior to administration
- It can be given w/o HCG results unless the patient is in the 3rd trimester of pregnancy, Class D

Sickle Cell Suspected VOE Rationale and Data

Goals of Clinical Pathway

1. Create an efficient team-oriented approach for patients with sickle cell disease presenting with possible VOE to achieve rapid evaluation, treatment, and disposition
2. Manage pain to a level acceptable to the patient

Data Considerations	Interventions	Rationale
Assessing pain	Patient report	Pain related to sickle cell disease is intense, and may be precipitated by cold, dehydration, stress, alcohol, exercise, and illness, among other triggers. It may be a manifestation of vaso-occlusive episodes but also may mask other complications, such as acute chest syndrome, osteomyelitis, sepsis, stroke, and the like. The best method of assessing pain is the patient's report. There is no persuasive evidence that physical exam findings (e.g. tachycardia, hypertension) or laboratory tests (e.g. decrease in hemoglobin) correlate with the degree of pain a patient is experiencing.
Analgesics	NSAIDs, Opioids	National guidelines recommend rapidly initiating analgesic therapy, ideally within 30 minutes of triage or within 60 minutes of registration. For children with severe pain due to SCD and a VOE, initiate treatment with parenteral opioids. For children with SCD and a VOE associated with mild to moderate pain, use NSAIDs if they have been successful in the absence of contraindications. Opioids are also frequently required.
Fluid repletion	Normal saline bolus	Patients with SCD with VOE are frequently volume deplete from decreased oral intake, increased insensible losses, and reduced urinary concentrating ability. Fluid repletion may therefore be of some benefit. However, there is no good evidence suggesting any particular volume, rate, or type of fluid in alleviating pain; furthermore, over-resuscitation with fluid can contribute to the development of acute chest syndrome, worsen cerebral edema in stroke, and cause hemodilution in aplastic crisis. In light of this, fluid should be used judiciously in patients experiencing a VOE.
Anemia	Blood transfusion	Absolute indications for blood transfusion include aplastic crisis, splenic sequestration, CVA, and rapidly progressing ACS. SCD patients with anemia relative to baseline may benefit from transfusion; however, they are at high risk for alloimmunization and iron overload so risks must be balanced. Therefore, PHO should be consulted PRIOR to RBC transfusion.
Lab studies	CBC, CMP, retic count, T&S	CBC, CMP, reticulocyte count may be helpful to assess precipitants of VOE, including infections, aplastic crisis, splenic sequestration, cholecystitis, and so on. Though with an uncomplicated vaso-occlusive episode obtaining these tests routinely does not frequently alter management, the results may suggest alternative diagnoses or elucidate the etiology of VOE in a minority of cases.
Monitoring	Cardiac and pulse ox monitoring	Patients being administered high doses of opioid medications are at risk of respiratory depression and for safety purposes should be maintained on continuous cardiac and pulse oximetry monitoring.

References:

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