1.0 Introduction

The cause of vaso-occlusive crisis (VOC) is believed to be ischemic tissue injury from the obstruction of blood flow by sickled erythrocytes. Reduced blood flow causes hypoxia and acidosis. This further increases the sickling process, leading to further hypoxia and acidosis—a cycle that eventually leads to ischemic tissue injury. Each VOC varies in intensity and duration. Infection, fever, acidosis, hypoxia, dehydration, sleep apnea, and exposure to extremes of heat and cold can precipitate crises. Often, no cause is identified.

Painful VOC is the most frequent complication of Sickle Cell Disease. Common sites of pain include bone (extremities, dactylitis or hand/foot syndrome, back) and abdominal pain. Bone pain, the most common type of VOC, may or may not be accompanied by swelling, low-grade fever, redness, and warmth. It may be symmetrical, asymmetrical, or migratory. Dactylitis is a common presentation in infants and toddlers; back and abdominal pain are more common in older children. Abdominal pain in children with sickle cell disease is usually a simple VOC, but other diagnoses may present similarly (splenic sequestration, liver sequestration, appendicitis, pancreatitis, biliary colic and cholecystitis, urinary tract infection, pelvic inflammatory disease, etc.) and should be ruled out. In addition, pneumonia and chest crisis may present as, or accompany abdominal pain. During a severe painful crisis, a patient may also develop an acute chest syndrome, or a CNS event.

Pain should be treated early and aggressively. No laboratory features are pathognomonic of VOC; diagnosis is based strictly on the history and physical examination. When treating a painful crisis, the Healthcare Provider needs to be aware that concurrent illnesses such as an acute sequestration, priapism, aplastic episode, or fever/sepsis (see other protocols) may also occur, which must be dealt with concurrently.

This clinical practice guideline has been developed for the management of sickle cell patients with an acute painful episode who present to the emergency department and/or inpatient units.
2.0 Clinical Practice Recommendations for Management of Vaso-occlusive Crisis

Child with Sickle Cell Disease (SCD) presents to ED with acute painful episodes (vaso-occlusive crisis):

**ED Initial Assessment and Management:**
- Place a child with vaso-occlusive crisis (VOC) or SCD fever or higher immediately into a team. Administrator of Pain Management Team (CDE, CDE or RN) as needed by type of crisis.
- Venous access must be obtained within 15 minutes of triage by a qualified provider. Refer to Sickle Cell Fever Guidelines.
- Place child in the head of the bed in the Trendelenburg position; refer to Sickle Cell Fever Guidelines.
- Pain management should be considered as first line of treatment for VMC.
- Pain assessment and initial treatment ideally within one hour of registration (or 30 minutes from transfer).
- PCP should be informed as early as possible (based on CT signs), and conduct a brief history and physical and order laboratory tests if needed. Include blood cultures, CBC, platelet count, electrolytes, glucose, creatinine, and urinalysis. Refer to Electrolyte Guidelines.
- Initiate Pain Management Plan and order pain medications as needed. Refer to Pain Management Guidelines.
- Pain management involves non-pharmacological interventions such as heating pads, massage, warm baths, and other interventions. 
- The Child Life Specialist can recommend structured activity. Imaging and diagnostic tests should be performed as needed.
- Consult Hematology service for patients with the following:
  - Presence of jaundice
  - Neurologic exam
  - Associated symptoms
  - Hemoglobin less than 8
  - Presumed acute splenic sequestration
  - Acute chest syndrome (refer to Sickle Cell Fever Guidelines). 
  - Presence of fever, chills, or other symptoms (refer to Acute chest Syndrome Guidelines).
- Pain management
- If child has painful episode without fever or fever: review pain order set and conduct a brief history and physical while obtaining laboratory tests as indicated.
- Initiate VOC order set in Epic. Refer to Sickle Cell Fever Guidelines.
- Place child in the head of the bed with continuous oxygen saturation monitoring with continuous opioid infusion.
- Refer to Acute chest Syndrome Guidelines.
- Consider physical or psychological interventions such as heating pads, massage, warm baths, and other interventions.
- On-site consultation of the Child Life Specialist can recommend structured activity. Imaging and diagnostic tests should be performed as needed.
- Consult Hematology service for patients with the following:
  - Presence of jaundice
  - Neurologic exam
  - Associated symptoms
  - Hemoglobin less than 8
  - Presumed acute splenic sequestration
  - Acute chest syndrome (refer to Sickle Cell Fever Guidelines). 
- Pain management
- If child has painful episode with fever or fever: review pain order set and conduct a brief history and physical while obtaining laboratory tests as indicated.
- Place child in the head of the bed with continuous oxygen saturation monitoring with continuous opioid infusion.
- Refer to Acute chest Syndrome Guidelines.
- Consider physical or psychological interventions such as heating pads, massage, warm baths, and other interventions.
- On-site consultation of the Child Life Specialist can recommend structured activity. Imaging and diagnostic tests should be performed as needed.
- Consult Hematology service for patients with the following:
  - Presence of jaundice
  - Neurologic exam
  - Associated symptoms
  - Hemoglobin less than 8
  - Presumed acute splenic sequestration
  - Acute chest syndrome (refer to Sickle Cell Fever Guidelines). 
- Pain management

**ED Discharge Medication Management and Follow-up**
- For all degrees of pain (child moderate/severe) (i.e. medication and hospitalization for acute VOC or SCD fever;
- Pain management should be considered as first line of treatment for VMC.
- Initiate Sickle Cell Fever order set in Epic. Refer to Sickle Cell Fever Guidelines.
- Place child in the head of the bed with continuous oxygen saturation monitoring with continuous opioid infusion.
- Refer to Acute chest Syndrome Guidelines.
- Consider physical or psychological interventions such as heating pads, massage, warm baths, and other interventions.
- On-site consultation of the Child Life Specialist can recommend structured activity. Imaging and diagnostic tests should be performed as needed.
- Consult Hematology service for patients with the following:
  - Presence of jaundice
  - Neurologic exam
  - Associated symptoms
  - Hemoglobin less than 8
  - Presumed acute splenic sequestration
  - Acute chest syndrome (refer to Sickle Cell Fever Guidelines).
- Pain management

**3.0 References**


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4.0 Related documents
- Acute Chest Syndrome or Pneumonia: Guidelines for Management in Children with Sickle Cell Disease
- Pain Assessment Policy
- Pain Management Clinical Practice Guideline

Attachments:
- Discharge Criteria 2021 FINAL.pdf
- ED mediation management.pdf
- Inpatient Management 2021 FINAL.pdf
- Revision History.docx
- SC_Clinic Follow Up Revised 2021_FINAL.pdf
- SCD pain plan_july 2015.pdf
- VOC Care pathway 2021 FINAL.pdf