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 suspected SCD in a private room or spray booth or higher immediately into a team. Administer a code blue for SCD. Order Sickle Cell Panel. Order C-REACT, CRP, LFT, BUN, creatinine, and creatinine clearance. C-REACT on IV line if scheduled. Obtain a urine random and serum electrolytes.
- Refer to Acute chest Syndrome Guidelines.
- Place a child with sickle cell pain under a 4 to 6-point PCEA pump with pump settings per institution guidelines. Certain medications should be considered as first line of medication. T-IVSAR is recommended. Place child on continuous cardiological and respiratory monitoring if possible. Refer to ED medication management.
- Consider palivizumab prophylaxis for children with SCD. Risk factors include age less than 24 months, birth weight less than 1500 g, and presence of sickle cell trait.
- If child is admitted to hospital, refer to Pediatric Intake Physician.

**ED Discharge Medication Management and Follow-up:**
- For all degrees of pain, provide a prescribed dose of acetaminophen and ibuprofen. Provide a prescription of acetaminophen and ibuprofen. Consider acetaminophen and ibuprofen in children with SCD. Opiates should be used for patients with moderate to severe pain.
- Place child on a continuous cardiological and respiratory monitoring. Place child on continuous opioid monitoring. Consider palivizumab prophylaxis for children with SCD. Refer to Acute chest Syndrome Guidelines.
- Place child on an opioid management document. Consider palivizumab prophylaxis for children with SCD. Refer to Acute chest Syndrome Guidelines.
- Refer to Acute chest Syndrome Guidelines.
- Refer to Acute chest Syndrome Guidelines.
- Refer to Acute chest Syndrome Guidelines.

**Child discharged home with appropriate follow-up**
- Refer to Acute chest Syndrome Guidelines.
- Refer to Acute chest Syndrome Guidelines.
- Refer to Acute chest Syndrome Guidelines.

**PRINTABLE VERSION**

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 medication 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