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 episode (vaso-occlusive crisis):

**ED Initial Assessment and Management:**
- Place a child with documented SCD on “Hi bed”, or similar, and immediately into a team. Administrator medications frequently used, particularly in crisis, in Sickle Cell. Page Hematology/ICU.
- Initiate Sickle Cell Acute Pain Order Set in Epic.
- Place child after labor as soon as possible (based on CTAS score) and conduct a brief history and physical exam, including a family history and relevant medical problems. Include ABO and Rh typing.
- Collect initial labs: CBC, INR, PT, plasma volume, central line placement
- Perform sequential compression devices (SCD). Refer to Acute Vaso-occlusive Crisis Guidelines.
- Place IV antibiotics immediately (if chest or respiratory symptoms)
- Assess and document vital signs and baseline lactate
- Perform chest x-ray and monitor child (if chest or respiratory symptoms)
- Consider prophylactic antibiotics (such as heparin, vancomycin) and oral analgesics immediately (if chest or respiratory symptoms)
- Consider sedation or deep sedation (such as ketamine, midazolam) and oral analgesics immediately (if chest or respiratory symptoms)

**ED Discharge Medication Management and Follow-up:**
- For all degrees of pain (mild to severe), use acetaminophen and non-steroidal anti-inflammatory drugs (NSAIDs) as needed for pain management. Children with SCD may require additional medications for pain management.
- Place child with documented SCD on “Hi bed”, or similar, and immediately into a team. Administrator medications frequently used, particularly in crisis, in Sickle Cell. Page Hematology/ICU.
- Complete the following labs: CBC, INR, PT, glucose, creatinine.) should be measured prior to administration (refer to Fluid and Electrolyte Guidelines)
- Pain is managed by oral analgesics
- Place child after labor as soon as possible (based on CTAS score) and conduct a brief history and physical exam, including a family history and relevant medical problems. Include ABO and Rh typing.
- Obtain chest x-ray and monitor child (if chest or respiratory symptoms)
- Assess and document vital signs and baseline lactate
- Consider prophylactic antibiotics (such as heparin, vancomycin) and oral analgesics immediately (if chest or respiratory symptoms)

**Acute Painful Episodes Vaso-occlusive Crisis : Guidelines for Management in Children with Sickle Cell Disease**

**4.0 Related documents**

- [Acute Chest Syndrome or Pneumonia: Guidelines for Management in Children with Sickle Cell Disease](http://example.com)
- [Pain Assessment Policy](http://example.com)
- [Pain Management Clinical Practice Guideline](http://example.com)

**Attachments:**

- [Discharge Criteria 2021 FINAL.pdf](http://example.com)
- [ED medication management.pdf](http://example.com)
- [Inpatient Management 2021 FINAL.pdf](http://example.com)
- [Revision History.docx](http://example.com)
- [SC_Clinic Follow Up Revised 2021_FINAL.pdf](http://example.com)
- [SCD pain plan_july 2015.pdf](http://example.com)
- [VOC Care pathway 2021 FINAL.pdf](http://example.com)