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 concerning symptoms in a V, or an ED bed or higher immediately into a team. Administer a complete assessment (in order of Sickle Cell Pain Scale) to Sickle Cell Pain Scale.
- Perform an immediate head to toe assessment including vital signs and full exam.
- Incite Sickle Cell Fever order set and refer to Sickle Cell Fever order set. Refer to Sickle Cell Fever order set.
- Place a child in a room as soon as possible (based on room availability) and conduct a brief history and physical exam.
- Ensure appropriate discharge pain management instructions and prescriptions are administered.
- Review and administer recommended analgesics.
- Obtain a head to toe exam to ensure child’s QOL evaluation.
- Use an analgesic checklist (Pegvedge, in the case of FVH) and refer to Sickle Cell Fever order set.
- Pain management strategy.
- Administer Sickle Cell Acute Pain order set in PE or ED.
- Opiate order set should be considered as first line of treatment for pain.
- Opiate order set should be administered as per Fluid and Electrolyte Guidelines.
- Ensure appropriate discharge pain management instructions and prescriptions are administered.
- Review and administer recommended analgesics.

**ED Discharge Medication Management and Follow-up:**
- For all degrees of pain (minimal, moderate, or severe) complete the ED discharge medication management and Follow-up document.
- Complete the Sickle Cell Fever order set and refer to Sickle Cell Fever order set.
- Refer to Fluid and Electrolyte Guidelines.
- Ensure appropriate discharge pain management instructions and prescriptions are administered.
- Review and administer recommended analgesics.

**Printable Version**

3.0 References


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](#)
- [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](#)