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 symptoms consistent with VOC in the ED (OR) and call the Sickle Cell Team immediately. Order initial Sickle Cell Fever order set in Epic.
- Note that signs and symptoms of VOC are present at different ages. See Pediatric Vaso/occlusive Crisis (P-VOC) Assessment Chart.
- Refer to Acute chest Syndrome Guidelines.
- Check for potential modifiable risk factors.
- Refer to Acute Sickle Cell Fever and Apneic Crisis Guidelines.
- Initiate Sickle Cell Fever order set in Epic.
- Refer to Acute Splenic Sequestration Guidelines.
- Refer to Apheresis Guidelines.

ED Discharge Medication Management and Follow-Up:
- For all degrees of pain, child moderate to severe pain levels are treated in the initial evaluation and subsequent follow-up.
- If child has moderate to severe pain or dehydrated, refer to Corrective Pain Management Guidelines.
- Place child for immediate pain management interventions and prescription in order to prevent additional pain.
- Parent or caregiver must be present for child to prevent further pain.
- Refer to Acute chest Syndrome Guidelines.
- Refer to Acute Sickle Cell Fever and Apneic Crisis Guidelines.
- Child discharged home with appropriate follow up and refers to same instructions.

3.0 References


©The Hospital for Sick Children (“SickKids”). All Rights Reserved. This document was developed solely for use at SickKids. SickKids accepts no responsibility for use of this material by any person or organization not associated with SickKids. A printed copy of this document may not reflect the current electronic version on the SickKids Intranet. Use of this document in any setting must be subject to the professional judgment of the user. No part of the document should be used for publication without prior written consent of SickKids.

### 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](#)