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 documented SCD in triage, or if SCD is not known, triage on high probability for SCD (vascular occlusion crisis).
- If child is known to have SCD, refer to SCD acute pain order set in Epic.
- Assess for signs of sepsis.
- Assess for signs of stroke.
- Provide appropriate pain management before IV access.
- Consider administration of IV fluids, blood, and prescription pain medication.
- Place child in comfortable position.
- Ensure adequate oxygen saturation.
- Consider involvement of the Child Life Specialist.
- Initiate Sickle Cell Fever order set in Epic.
- Initiate ED Sickle Cell Acute Pain order set in Epic.

**ED Discharge Medication Management and Follow-up:**
- For all degrees of pain, child receives IV hydration and paracetamol and/or diclofenac and/or morphine.
- Continue oral analgesics as per Pain Management Guideline.
- Refer to Acute chest Syndrome Guidelines.
- Ensure appropriate discharge pain management instructions and prescriptions are documented.
- Ensure patient understanding of how to obtain SCD follow-up as per Sickle Cell Day Unit guidelines.
- Request blood cell counts (CBC), serum electrolytes (Na, K, glucose, creatinine) should be measured prior to administration (refer to Fluid and Electrolyte Guidelines).
- Test venous blood gas (VBG) levels.
- Verify and document child's O2 saturation.
- Take into consideration timing and dosing of previous medication.
- Monitor child carefully.
- Observe closely for signs of deterioration in clinical status through cardiac and respiratory work up.
- Continue cardiopulmonary monitoring.
- Select appropriate discharge pain medication instructions and prescriptions in consultation with ED provider.
- Take oral medication should be considered if child is tolerating fluids and is not vomiting.
- Refer to Fluid and Electrolyte Guidelines.
- Test for hypoglycemia and dehydration.
- Test for electrolyte disturbances.
- Contact the SCD provider.
- Refer to Acute chest Syndrome Guidelines.

**References**

---

**Printable Version**

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.

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](#)