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

ED Initial Assessment and Management:

- Place a child with suspected Sickle Cell Disease (SCD) in bed or higher immediately into a team. Administrator immediate medication (see Sickle Cell Pain Policy) in Sickle Cell Pain Policy (SCD) (see Sickle Cell Pain Policy).
- Place child in an upright position and maintain airway and chest ventilation.
- Place child in an upright position and maintain airway and chest ventilation.
- Continue until the child’s pain is controlled.

ED Discharge Medication Management and Follow-up:

- For all degrees of pain, child should be observed for at least 60 minutes in the ED and discharged home. Consider changing the pain management plan to meet the child’s needs.
- Doses of opioids are reviewed and administered, along with discharge pain management instructions and prescriptions.
- Continue acetaminophen and ibuprofen for use of this material by any person or organization not associated with SickKids. A printed copy of this document may not be distributed without the approval of SickKids.

Sickle Cell Pain Management & Follow-up

Are the child’s pain managed at a level analgesics? YES

- Continue with medication management
- To arrange outpatient follow-up
- Physical exam to include:
  - Temperature
  - Respiratory rate
  - Blood pressure
  - Heart rate
  - Respiratory rate
  - Blood pressure
  - Heart rate
- Laboratory tests:
  - CBC
  - Na
  - K
  - Glucose
  - Creatinine
  - Serology
  - Other
- Imaging to include:
  - CBC
  - Na
  - K
  - Glucose
  - Creatinine
  - Serology
  - Other
- Further investigation:
  - CBC
  - Na
  - K
  - Glucose
  - Creatinine
  - Serology
  - Other
- Follow-up confirmed

Child discharged home with appropriate follow-up

- Refer to Sickle Cell Pain Management & Follow-up
- Ensure appropriate discharge pain management instructions and prescriptions
- Follow-up confirmed

Child discharged home with appropriate follow-up and returns to same institution

- Refer to Sickle Cell Pain Management & Follow-up
- Ensure appropriate discharge pain management instructions and prescriptions
- Follow-up confirmed

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](#)