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**

- **For all degrees of pain:** mild, moderate and severe. Use medications and therapies aound the clock in addition to rectal suppositories and ibuprofen, as tolerated.
- **Pain assessment and management:**
  - Pain assessment should be performed at least every 4 hours.
  - Use a validated pain assessment tool.
- **Oral analgesics:** should be administered as per formulary for dosing.
- **Fever management:**
  - Temperature should be monitored every 4 hours.
  - If fever persists, consider oral acetaminophen or paracetamol.
- **Localizing signs of infection:**
  - Localizing signs of infection should be assessed and documented.
  - Fever should be monitored and documented.
- **Imagery and distraction:** are helpful.
  - Imagery and distraction are helpful.
- **Physical exam to include:**
  - All patients should be evaluated for signs of infection.
  - Physical exam to include.
- **Guideline has been developed by the Pain Management Committee, pediatric pain specialists and members of the Sickle Cell Care Team at SickKids.”

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3.0 References


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

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

Version: 5

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