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.
Clinical Practice Recommendations for Management of Vaso-occlusive Crisis

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

**Clinical Practice Recommendations for Management of Vaso-occlusive Crisis**

**ED Initial Assessment and Management**

1. **Pain** with documented fever at 38°C (100.4°F) or higher or at least 10°C (18°F) higher than core body temperature measured directly in the core. Administration of norepinephrine immediately (refer to Sick Kids Fever Guidelines).

2. **Initiate** SickKids Call Centre number at any time.

3. **Assess** child when possible based on CPEAS score, and establish a point-based pain scale with appropriate analgesics and consider other medical or respiratory symptoms or findings consistent with sickle cell crises.

4. **Assess** risk for bleeding, as per SickKids RBC Order Set in Epic.

5. **Assess** child’s vitals immediately:
   - **SBP** and **HR**
   - **Respiratory rate**
   - **Temperature**
   - **Pain score**
   - **Pulse oximetry**

6. **Interventions**
   - **Initiate** ED Sickle Cell Acute Pain Order Set in Epic
   - **Initiate** VOC Order Set in Epic
   - **Complete** the following:
     - **Epic** Sickle Cell Fever Order Set (police, vital signs, laboratory, radiology).
     - **Complete** a non-contrast head CT scan if symptoms of altered mental status.
   - **Monitor** for signs of deterioration in clinical status.

**ED Discharge Medication Management and Follow-up**

- **For all degrees of pain** with moderate or severe, use combination medication.
- **Eliminate** use of medication administered until home medications available.
- **Consult** physician or physician assistant for consultation within 24 hours of discharge.
- **Consult** child life specialist if possible.
- **Restore** to oral intake as tolerated.

**ED Discharge Home Care**

- **Notify** family on discharge of ED discharge in order to arrange outpatient follow-up and notify them of ED discharge in order to arrange outpatient follow-up.
- **Reassess** appropriate analgesics and pain medication.
- **Notify** any changes in condition or use of any medications.
- **Notify** the Child Life Specialist if possible.

**ED Discharge Planning Document**

- **Refer** to appropriate discharge planning document.
- **Notify** family on discharge of ED discharge in order to arrange outpatient follow-up.

**Child Discharged Home**

- **Notify** appropriate family member for follow-up.

**Initial Assessment and Management**

- **Rule out** infection:
  - **Temperature**
  - **Pain score**
- **Observe** closely for signs of deterioration in clinical status through cardiac and respiratory monitoring with continuous opioid infusion.
- **Consider** physical or psychological interventions such as heating pads, and other pain management strategies.
- **Consult** hematology/oncology specialists as needed.
- **Consult** palliative care experts as needed.

**Sickle Cell: Acute Painful Episodes (Vaso-occlusive Crisis)**

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

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References


Related documents

- [Acute Chest Syndrome or Pneumonia: Guidelines for Management in Children with Sickle Cell Disease](#)
- [Pain Assessment Policy](#)
- [Pain Management Clinical Practice Guideline](#)
- [Sickle Cell Pain Day Hospital](#)

Attachments:

- discharge criteria.pdf
- SCD pain plan july 2015.pdf
- care pathway_final.pdf
- Revision History.docx
- SC_clinic follow up.pdf

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ED medication management.pdf

inpatient management.pdf