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 suspected SCD and/or vaso-occlusive crisis (VOC) in the Red Zone as soon as possible to a team able to manage this correctly. 
  - Is the child a known patient at SickKids?
  - What is the CTAS score for the SCD patient (if known)?
  - Are there any contraindications for phlebotomy or drawing blood for laboratory tests?
- Is the child stable and able to safely receive medication?
- Are there other medical problems requiring attention?
- If ³ 14 years of age and at high risk for pneumococcal disease, consider starting pneumococcal conjugate vaccine (PCV).
- If ³ 2 years of age and at high risk for pneumococcal disease, consider starting pneumococcal polysaccharide vaccine (PPS).  
  - PPS should be given to all children with SCD and/or VOC as soon as possible. 
  - If the child is not ³ 2 years of age or if the child has already received PPS, then a vaccination program should be initiated in consultation with a pediatrician.
- Place child in a private and comfortable setting.
- Complete the following with prior to administration (refer to Fluid and Electrolyte Guidelines):
  - History and physical examination
  - Pain assessment
  - Initial treatment
- Place child in a private and comfortable setting.
- Complete the following with prior to administration (refer to Fluid and Electrolyte Guidelines):
  - History and physical examination
  - Pain assessment
  - Initial treatment
- If required, contact Pediatric Intake physician. 
  - Sickle Cell Day Hospital between 7:00 am and 5:00 pm.

**ED Discharge Medication Management and Follow-up:**
- All patients at high risk for VOC should be considered for home-based or inpatient management of VOC.
- If the child is discharged home with appropriate follow-up, refer to Sickle Cell Acute Pain order set in Epic.
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**INPATIENT MANAGEMENT:**
- If the child has VOC and requires inpatient management, initiate ED Sickle Cell Acute Pain order set in Epic.
- Place child in a private and comfortable setting.
- Complete the following with prior to administration (refer to Fluid and Electrolyte Guidelines):
  - History and physical examination
  - Pain assessment
  - Initial treatment
- Place child in a private and comfortable setting.
- Complete the following with prior to administration (refer to Fluid and Electrolyte Guidelines):
  - History and physical examination
  - Pain assessment
  - Initial treatment
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  - History and physical examination
  - Pain assessment
  - Initial treatment
- Place child in a private and comfortable setting.
- Complete the following with prior to administration (refer to Fluid and Electrolyte Guidelines):
  - History and physical examination
  - Pain assessment
  - Initial treatment

**PRIORITY CALL:**
- Reassess the child concurrently with other measures including pain assessment and initial treatment ideally within one hour.

**SICKKIDS INPATIENT MANAGEMENT GUIDELINES:**
- Place child in a private and comfortable setting.
- Complete the following with prior to administration (refer to Fluid and Electrolyte Guidelines):
  - History and physical examination
  - Pain assessment
  - Initial treatment
- Place child in a private and comfortable setting.
- Complete the following with prior to administration (refer to Fluid and Electrolyte Guidelines):
  - History and physical examination
  - Pain assessment
  - Initial treatment
- Place child in a private and comfortable setting.
- Complete the following with prior to administration (refer to Fluid and Electrolyte Guidelines):
  - History and physical examination
  - Pain assessment
  - Initial treatment
- Place child in a private and comfortable setting.
- Complete the following with prior to administration (refer to Fluid and Electrolyte Guidelines):
  - History and physical examination
  - Pain assessment
  - Initial treatment

**COMMUNICATION OF PATIENT INFORMATION DURING HOSPITAL TRANSFER:**
- Transfer all pertinent information to the receiving physician in a timely and reliable manner.

**ED DISCHARGE MEDICATION MANAGEMENT AND FOLLOW-UP:**
- For all degrees of acute pain (moderate, severe, critical), oral analgesics and parenteral analgesics should be provided.
- Oral analgesics are preferred unless contraindicated.
- If the child is discharged home with appropriate follow-up, refer to Sickle Cell Acute Pain order set in Epic.
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**RECOMMENDED PRACTICES:**
- Use acetaminophen and ibuprofen as first-line oral analgesics.
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**COMMENTS:**
- Place child in a private and comfortable setting.
- Complete the following with prior to administration (refer to Fluid and Electrolyte Guidelines):
  - History and physical examination
  - Pain assessment
  - Initial treatment
- Place child in a private and comfortable setting.
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  - History and physical examination
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  - History and physical examination
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- Place child in a private and comfortable setting.
- Complete the following with prior to administration (refer to Fluid and Electrolyte Guidelines):
  - History and physical examination
  - Pain assessment
  - Initial treatment

**PRINTABLE VERSION**

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](#)