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

Sickle Cell, Acute Painful Episodes (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 the child in a comfortable position (on bed, stretcher or chair) and immediately into a team. Administrate medications as per protocol;
- Place the child with acute pain either in a seated or standing position (if possible and based on CTAS score) and conduct a brief history and physical exam (Vital signs, Neurological examination, Oxygen saturation, and Pain management);
- Place the child with pain in the ED for 30 minutes from time before;
- Ensure appropriate discharge pain management instructions and prescriptions;
- If needed to arrange outpatient follow up and notify them of the ED discharge in order to continue follow up;

**ED Discharge Medication Management and Follow up:**
- For all degrees of pain, child moderate (i.e., mild, moderate, and severe pain) use acetaminophen and ibuprofen;
- Use acetaminophen and ibuprofen for dosage;
- Continue use of acetaminophen and ibuprofen as per protocol;
- Use acetaminophen and ibuprofen as per protocol;
- Refer to Sickle Cell Disease Guidelines;
- Ensure that the child is not vomiting;
- Ensure appropriate discharge pain management instructions and prescriptions;
- Place the child in the ED for 30 minutes from time before;

**To the child's pain managed at worst analgesics?**

**YES**
- Place the child in the ED for 30 minutes from time before;
- Use acetaminophen and ibuprofen for dosage;
- Continue use of acetaminophen and ibuprofen as per protocol;
- Refer to Sickle Cell Disease Guidelines;
- Ensure that the child is not vomiting;
- Ensure appropriate discharge pain management instructions and prescriptions;
- Place the child in the ED for 30 minutes from time before;

**NO**
- Place the child in the ED for 30 minutes from time before;
- Use acetaminophen and ibuprofen for dosage;
- Continue use of acetaminophen and ibuprofen as per protocol;
- Refer to Sickle Cell Disease Guidelines;
- Ensure that the child is not vomiting;
- Ensure appropriate discharge pain management instructions and prescriptions;
- Place the child in the ED for 30 minutes from time before;

**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](#)