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 symptoms consistent with VDC immediately into a team. Administrator the Timely Admission Document in Epic and Page Hematology for admission to Sickle Cell Day Hospital between 8am and 5pm.
- Place child in a safe area as soon as possible (based on CTCA data), and conduct a brief history and physical exam, including pain assessment and vital signs (with one hour of registration or 30 minutes from transfer).
- Administer O2 and monitor child’s O2 saturation.
- Perform initial blood work (e.g., CBC, electrolytes). Refer to the SickKids Sickle Cell Day Hospital admission at 1400.
- Ensure appropriate discharge planning instructions and prescriptions are given to the child and family.

**ED Discharge Medication Management and Follow-up:**
- For all degrees of pain: patient requires the use of acetaminophen and ibuprofen as directed (with caution to monitor bleeding risk).
- For pain > 3/10: dosing of opioids is increased.
- For pain > 4/10: continue with ketorolac.
- For pain > 5/10: continue with ketorolac and use acetaminophen and ibuprofen concurrently with other measures including pain assessment and initial treatment ideally within one hour.
- Place child on Cardiac Monitor and follow-up with the Sickle Cell Fever Order Set in Epic immediately.
- Use VDC Child’s Pain Scale.

**Child discharge form with appropriate follow-up: A**
- Make in (patient discharge plan summary document).
- Stable vitals (e.g., patient on oral analgesics).
- Tolerating fluids and is not vomiting.
- Pain is maintained by oral analgesics.
- Placement of chest drainage tube (if not contraindicated). 
- Pain is maintained by oral analgesics.
- Fever in a room as soon as possible.
- CBC, serum electrolytes, and AST/ALT (if changing medications).
- Pain is maintained by oral analgesics.
- Incentive spirometry to prevent acute pulmonary complications.
- Placement of chest drainage tube (if not contraindicated).
- Pain is maintained by oral analgesics.
- Physical exam to include:
  - Test venous blood gas and monitor child’s O2 saturation.
  - Observe closely for signs of deterioration in clinical status through cardiac and respiratory monitoring.

**Child discharged form with appropriate follow-up: B**
- Make in (patient discharge plan summary document).
- Stable vitals (e.g., patient on oral analgesics).
- Tolerating fluids and is not vomiting.
- Pain is maintained by oral analgesics.
- Placement of chest drainage tube (if not contraindicated).
- Pain is maintained by oral analgesics.
- Fever in a room as soon as possible.
- CBC, serum electrolytes, and AST/ALT (if changing medications).
- Pain is maintained by oral analgesics.
- Incentive spirometry to prevent acute pulmonary complications.
- Placement of chest drainage tube (if not contraindicated).
- Pain is maintained by oral analgesics.
- Physical exam to include:
  - Test venous blood gas and monitor child’s O2 saturation.
  - Observe closely for signs of deterioration in clinical status through cardiac and respiratory monitoring.

**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
- VOC Care pathway 2021 FINAL.pdf