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 child with symptoms of vaso-occlusive crisis (ACS) or SCD with fever or higher immediately into a team. Administer inpatient management document order to Sickle Cell, Page for document.
- Place child on bed rest as soon as possible (based on CT imaging), and conduct a brief history and physical exam. Include vital signs, pain assessment and initial treatment with hydroxyurea, and wing one hour of registration. If ED emerges from treatment.
- If severe or persistent pain, administer analgesics.
- Administer Sickle Cell Acute Pain order in the ED.
- Initiate Sickle Cell Fever order set in Epic.
- Order CCE (Child Life Specialist) to provide structured activity or play.
- Consider physical or psychological interventions such as heating pads, massage, warm baths, and other soothing methods.
- The CCE (Child Life Specialist) can recommend prompt activity or play.
- The CCE can assist in the patient's visit.
- Consider Hematology service for patients with the following:
  - Presence of jaundice
  - Neurologic exam
  - Presence of hemoglobin S <75%

ED Discharge Medication Management and Follow-up:
- Place child in inpatient management document.
- Administer hydroxyurea and other medications as per order.
- Refer child to Hematology service.
- Include Sickle Cell Fever order set in Epic.
- Perform physical exam to include:
  - Vital signs including O2 saturation
  - Pain score
  - Hemoglobin
  - Complete blood count
  - Electrolyte levels
  - Glucose

Does the child's pain management at risk analgesics?

YES
- Make in inpatient discharge planning documentation.
- Refer child to Hematology service.
- Order appropriate follow-up as per order.

NO
- If child has painful episode WITHOUT fever
- If child has moderate to severe pain or dehydration
- If child has high risk
- If child has severe pain, consult Pediatric Intake physician.
- If child has high risk for readmission
- If child has a concurrent fever

-child with Sickle Cell Disease (SCD) presents to ED with acute painful episodes (vaso-occlusive crisis).

Child discharged from ED with appropriate follow-up and returns to same instructions.

- Make in inpatient discharge planning document.
- Refer child to Hematology service.
- Order appropriate follow-up as per order.

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**