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

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 vaso-occlusive crisis on SC in ED; rest or higher intravenous into a team. Admission should be considered if not SC in ED. Place child on SC 24 hours.
- Initiate Sickle Cell Pain Score in ED to document.
- Place a child with suspected vaso-occlusive crisis on SC in ED; rest or higher intravenous into a team. Admission should be considered if not SC in ED. Place child on SC 24 hours.
- If severe or unstable, perform a rapid and reversal child’s Q2 evaluation.
- Initiate SC in ED for pain and reversal child’s Q2 evaluation.
- Pain management:
  - Administer Sickle Cell Acute Pain order set in Epic.
  - Monitoring vital signs (BP, HR, RR, O2 sat) and lab values (Hb, Hct, serum electrolytes, Na, K, glucose) should be measured prior to administration (refer to Fluid and Electrolyte Guidelines).
  - Administer SC in ED for pain and reversal child’s Q2 evaluation.
  - Ensure appropriate discharge pain management instructions and prescriptions are included in the ED discharge planning document.
  - Start cardiopulmonary monitoring and notify the Hematology service for patients with the following:
    - Presence of jaundice
    - Neurologic exam
    - Abdominal exam
    - Presence of fever
    - Severe vaso-occlusive crisis

ED Discharge Medication Management and Follow-up:
- For all degrees of pain ( mild, moderate, severe), ensure antiemetics and anti-inflammatory medications are ordered.
- Peds ED should never be asked to assess analgesia within an emergency.
- Severe pain should be managed within the first hour of admission, followed by oral medications and further analgesia if needed.
- Ensure appropriate discharge pain management instructions and prescriptions are included in the ED discharge planning document.
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- Place child with Sickle Cell Disease in hospital if:
  - Fever in a room as soon as possible
  - Stabilization of registration
  - Stable vital signs (HR, BP, RR, O2 sat)
  - Able to ambulate
  - Papilledema
  - No stroke
  - Acute splenic sequestration
  - Associated symptoms
  - Hemodynamic instability
  - Presence of jaundice
  - Neurologic exam
  - Presence of fever
  - Severe vaso-occlusive crisis

Physical exam to include:
- Complete physical exam to include:
  - BP
  - RR
  - O2 sat
  - Auscultation
  - Palpation
  - Neurologic exam
  - Urinalysis
  - Laboratory testing
- Start cardiopulmonary monitoring and notify the Hematology service for patients with the following:
  - Presence of jaundice
  - Neurologic exam
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Follow-up confirmed:
- Contact Pediatric Intake physician:
  - Refer to Sickle Cell Fever Guidelines
  - Refer to Acute chest Syndrome Guidelines
  - Refer to Acute splenic sequestration guidelines
  - Refer to Hematology service for patients with the following:
    - Presence of jaundice
    - Neurologic exam
    - Abdominal exam
    - Presence of fever
    - Severe vaso-occlusive crisis

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](http://example.com)
- [ED medication management.pdf](http://example.com)
- [Inpatient Management 2021 FINAL.pdf](http://example.com)
- [Revision History.docx](http://example.com)
- [SC_Clinic Follow Up Revised 2021_FINAL.pdf](http://example.com)
- [SCD pain plan_july 2015.pdf](http://example.com)
- [VOC Care pathway 2021 FINAL.pdf](http://example.com)