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 in a private room, on high-flow oxygen or higher immediately into a team. Administrator of care will be assigned. Be sure to ask if diabetic or renal issues.
- Indicate Sickle Cell Fever Order Set in Epic.
- Place child in a prone position if possible (based on CT scan results), and conduct a brief history and physical examination (including pulse rate, blood pressure, temperature), and review of systems within 4 hours of registration (or 30 minutes from transfer).
- Fervers in children and neonates should be taken to prevent constipation while on opiates.
- Turpentine baths are recommended to prevent or treat pruritus.
- Pain management should be considered as first line of medication if VOM is likely. Use of non-steroidal anti-inflammatory drugs and acetaminophen is contraindicated.
- Localizing signs of infection or meningitis or a fever above 38°C axilla should be considered in the differential. Refer to Acute chest Syndrome (ACS) Guidelines.
- Fever and antibiotics should be provided immediately with local chest x-ray and monitor child every 5 minutes from triage.
- Place child in the Sickle Cell Fever order set in Epic.
- Initiate VOC Order Set in Epic.
- Place child in the Sickle Cell Day Unit for Management of Vaso-occlusive Crisis.
- Discharge Medication Management and Follow-up
- Refer to Acute chest Syndrome Guidelines.
- Follow up confirmed.
- Determine risk for readmission.
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References

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4.0 Related documents

- [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](#)