Use of this guideline in any setting must be subject to the clinical judgment of those responsible for providing care.

IWK Health	Document Scope: Hospital-wide Patient Care	
	Document Type: Clinical Practice Guideline Approved on: January 2024 Next Review Date: January 2027	
	Acute Painful Episodes Vaso-occlusive Crisis: Guidelines for Management in Children with Sickle Cell Disease (adapted from the Hospital for Sick Children)	Version: 1

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.

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

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

ED Initial Assessment and Management:

1. Consult Hematology/Oncology
2. Place child with concurrent fever of 38.3ºC oral (or 37.8ºC axilla) or higher immediately into a room. Draw blood cultures and administer antibiotics immediately (refer to IWK or APPHON Sickle Cell Fever preprinted order)
3. Place child without fever in a room as soon as possible; and conduct a brief history and physical concurrently with other measures including pain assessment and initial treatment ideally within one hour of registration (or 30 minutes from triage).
4. If chest or respiratory symptoms:
   - Request a chest x-ray and monitor child s O_2 saturation to maintain O_2 sats > 93%.
   - Test venous blood gas (VBG) level. Refer to Acute chest Syndrome clinical care document.
6. Pain Management
   - Provide medication management. Oral medication should be considered as first line of treatment if child is tolerating fluids and is not vomiting. Take into consideration timing and dosing of previous medication. Refer to the pain management pre-printed orders, apphon-rohppa.com or IWK Access E-Forms.
   - If unable to obtain IV access after 2 attempts, consider ultrasound guided IV placement or page anesthesia/anesthesia assistant. Refer to IWK IV policy #1155.
   - Consider physical or psychological interventions such as heating pads, massage, warm baths, and other comfort measures.
   - The Child Life Specialist can recommend structured activity. Imagery and distraction are helpful.

History to Include:

- □ Nature, duration, and location of pain.
- □ Severity of pain.
- □ How pain compares to previous crises.
- □ What has worked in previous crises.
- □ Analgesics already used and doses.
- □ Associated symptoms.

Optimal labs:(only if clinically indicated or if obtaining IV access)

- □ Request blood cell counts (CBC, differential, and reticulocyte count)
- □ The plan includes IV fluids, serum electrolytes (Na, K, glucose, creatinine) should be measured prior to administration.

Physical exam to include:

- □ Vital signs including O2 saturation, pain score, cardiopulmonary and hydration status.
- □ Spleen size
- □ Neurologic exam
- □ Presence of jaundice
- □ Localizing signs of infection

Is child's pain managed by oral analgesics?

YES

ED Discharge Medication Management and Follow-up

- For all degrees of pain (mild-moderate-severe) use acetaminophen and ibuprofen around the clock in addition to morphine or hydromorphone unless contraindications exist. Continue for 1-2 days and then as needed. Refer to IWK drug dosing guidelines for dosing.
- PEG 3350 should be taken to prevent constipation while on opiates.
- Page Hematology/Oncology Consult Service and notify them of the ED discharge to arrange outpatient follow-up.
- Ensure appropriate discharge pain management instructions and prescriptions are reviewed and administered. Make sure then family can pick up prescriptions at the pharmacy.

Child discharged home with appropriate follow-up and return to care instructions.

NO

Inpatient Admission

- Refer to the IWK inpatient pain admission orders. If child has moderate to severe pain or dehydrated, consult Pediatric Hematology/Oncology for admission.
- Consider consult to acute pain service for hard to manage pain.
- Complete the following:
  - start cardiopulmonary monitoring; and
  - serum electrolytes (Na, K, glucose, creatinine) should be ordered prior to IV fluid administration.
  - Observe closely for signs of deterioration in clinical status through cardiac and O_2 saturation monitoring with continuous opioid infusion.
  - Monitor for acute chest syndrome.

Child discharged home with appropriate follow-up if:

- Stable vital signs i.e., afebrile unless clear viral source
- Tolerating fluids and medications by mouth
- Pain is maintained by oral analgesics
- Determine risk for re-admission (high risk: 3 admissions for VOC or 1 admission for ACS in the last 12 months)
- All patients at high risk for readmission should be seen in SCD clinic for follow-up within 14 days
- Does not have respiratory distress; and
- Follow up is confirmed.

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
- Hydromorphone continuous infusion high alert
- Morphine continuous infusion high alert