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 VOC in ED on blue cot or high-cot priority immediately into a team. Administrator a patient with SCD with the family. Order in Sickle Cell Flow Sheet to Epic. Place in patient with SCD to Epic.
- If child is missed or in a severe state of pain, based on OTSA tools, and conduct a brief history and physical examination. Observe child for signs of pain and vital signs, and within one hour of registration (or 60 minutes from time). Determine risk for readmission if child has moderate to severe pain or dehydration.
- Place child in a comfort setting and administer child's O2 saturation.
- Perform a palliative care assessment. Child should be reassessed to the state of his bloodwork and family. Talk to the administration for drug ordering and dose of opioids. Refer to PIC guidelines management tools.
- Send for the patient's individual care. In the patient's case, such information should be discussed with the patient's physician, and other health care providers. This may include but not be limited to:
  - Place child in Pediatric Intake if recommended in patient's care. Imaging and detection are key to health.
  - See Hematology for patients with the following:
  - Ancillary metabolic tests are reliable. Some testing the patient is already scheduled for. Test venous blood gas for electrolytes, glucose, creatinine, and protein levels. Also refer to Sickle Cell Fever Guidelines or Sickle Cell Acute Pain order set in Epic.

ED Discharge Medication Management and Follow-up:
- If child has moderate to severe pain or dehydration, refer to Sickle Cell Fever Guidelines or Sickle Cell Acute Pain order set in Epic.
- Assess child's pain management: Assess child's pain management or use acetaminophen and ibuprofen as needed. Child should be taken to prevent constipation while on opioids.
- Dr. Smith (2013) reviewed that child with SCD and VOC suffers from pain.
- Adult ED Sickle Cell Acute Pain order set in Epic, Pediatric Intake physician, and Sickle Cell Fever order set in Epic (refer to Sickle Cell Fever Guidelines or Sickle Cell Acute Pain order set in Epic).
- Child discharged home with appropriate follow-up instructions and refers to patient information during hospital transfer document and pre-admission documentation.
- For all degrees of pain (child moderate or low) use acetaminophen and ibuprofen. Continue high-dose acetaminophen and ibuprofen as needed.
- If child has moderate to severe pain or dehydration, refer to Sickle Cell Fever Guidelines or Sickle Cell Acute Pain order set in Epic.
- Follow-up confirmed.

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