Sickle Cell with Pain Crisis

May 24, 2019

LEGAL DISCLAIMER: The information provided by Dell Children’s Medical Center of Texas (DCMCT), including but not limited to Clinical Pathways and Guidelines, protocols and outcome data, (collectively the "Information") is presented for the purpose of educating patients and providers on various medical treatment and management. The Information should not be relied upon as complete or accurate; nor should it be relied on to suggest a course of treatment for a particular patient. The Clinical Pathways and Guidelines are intended to assist physicians and other health care providers in clinical decision-making by describing a range of generally acceptable approaches for the diagnosis, management, or prevention of specific diseases or conditions. These guidelines should not be considered inclusive of all proper methods of care or exclusive of other methods of care reasonably directed at obtaining the same results. The ultimate judgment regarding care of a particular patient must be made by the physician in light of the individual circumstances presented by the patient. DCMCT shall not be liable for direct, indirect, special, incidental or consequential damages related to the user's decision to use this information contained herein.
**Definition**

Children with sickle cell disease presenting with acute pain event

**Epidemiology**

Sickle cell pain crisis is very common in both pediatric and adult patients; it is the most common reason for patients to seek medical attention. In one 2010 study, there were approximately 200,000 emergency department visits by children and adults with sickle cell disease, with 67 percent for pain alone. In comparison, visits for chest symptoms (pain, shortness of breath, cough) and fever accounted for only 20 and 6 percent of visits, respectively. In one adult study [Pain in Sickle Cell Epidemiology Study (PiSCES)], patients reported pain on 54.5% of the 31,017 days surveyed. Almost 30% of respondents had pain on more than 95% of the days surveyed. In the pediatric population, Dampier et al. studied children and adolescents (ages 6–21 years) with sickle cell disease for 18,377 days. Children commonly reported pain, with 514 distinct pain episodes occurring over 2592 days and 2326 nights. Acute pain is a known hallmark of sickle cell disease, with chronic pain often occurring frequently as well.

References- 1,2,3, &4

**Etiology**

Sickle cell disease is due to a single amino acid substitution in the gene encoding the β-globin subunit. Polymerization of deoxygenated sickle hemoglobin leads to decreased deformability of red blood cells. Through adhesive events among blood cells, these erythrocytes can obstruct the vasculature, producing pain, hemolytic anemia, organ injury, and early mortality. Although the molecular basis of SCD is well characterized, the complex mechanisms underlying vaso-occlusion have not been fully established. Preferential adhesion of low-density SS-RBCs and reticulocytes in immediate postcapillary venules leads to trapping of the older, more dense, and misshapen SS-RBCs. Precapillary obstruction by a small number of dense SS-RBCs also contributes to VOC. Recent data indicates other blood cell elements that are not directly affected by the sickle cell mutation play a direct role in VOC. Theories have been proposed in which the process is viewed as multistep and multicellular cascade driven by inflammatory stimuli and the adherence of leukocytes.

References- 5

**Guideline Eligibility Criteria**

All children presenting to Dell Children's Medical Center with a history of sickle cell disease and with acute pain episode.

**Guideline Exclusion Criteria**

Sickle cell patients presenting with symptoms of acute chest, fever.
Differential Diagnosis

Vaso-occlusive Crisis (VOC), Pneumonia, Pulmonary Embolism, Acute Chest Syndrome, Reactive Airway Disease, Asthma, Cardiomyopathy, Myocardial Infarction, Gastroesophageal Reflux, Cholelithiasis, Mesenteric Ischemia, Hemolysis, Splenic Sequestration, Aplastic Crisis, Priapism, Avascular Necrosis, Osteomyelitis, Septic Arthritis, Stroke

Methods

<table>
<thead>
<tr>
<th>Existing External Guideline/Clinical Pathway</th>
<th>Organization and Author</th>
<th>Last Update</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sickle Cell Patients in Acute Pain Crisis Guideline</td>
<td>Zora Rogers</td>
<td>2012</td>
</tr>
<tr>
<td>Pediatric Emergency Department Clinical Guideline: Sickle Cell Disease (SCD) Patients With Pain</td>
<td>University of Chicago</td>
<td>9/29/2015</td>
</tr>
<tr>
<td>Sickle Cell Disease in Vaso-Occlusive Crisis Evidence-Based Guideline</td>
<td>TEXAS CHILDREN’S HOSPITAL</td>
<td>July 2017</td>
</tr>
<tr>
<td>Vanderbilt Pain Algorithm</td>
<td>Vanderbilt</td>
<td>No Date</td>
</tr>
<tr>
<td>ED SCD Pathway</td>
<td>OU Children’s</td>
<td>No Date</td>
</tr>
<tr>
<td>ED Pathway for Evaluation/Treatment of Children with Sickle Cell Disease and Pain</td>
<td>Children’s Hospital of Philadelphia</td>
<td>May 2017</td>
</tr>
</tbody>
</table>

Any published clinical guidelines have been evaluated for this review using the AGREE II criteria. The comparisons of these guidelines are found at the end of this document. AGREE II criteria include evaluation of: Guideline Scope and Purpose, Stakeholder Involvement, Rigor of Development, Clarity of Presentation, Applicability, and Editorial Independence.

Evaluating the Quality of the Evidence

The GRADE criteria were used to evaluate the quality of evidence presented in research articles reviewed during the development of this guideline. The table below defines how the quality of evidence is rated and how a strong versus a weak recommendation is established.
Diagnostic Evaluation

**Labs:** CBC w/diff, Abs Retic, Type & Screen, CMP, UA, Blood Culture (if febrile), other lab studies at provider discretion.

**Radiology studies:** dependent on physical presentation / exam results, provider discretion.

Clinical Management

- **Initial Management:** Appropriate Triage, Establish Venous Access, Initiate Pain Management
- **Secondary Management:** Admission to hospital may be necessary as well if pain is uncontrolled, or patient is febrile / ill appearing. Consult Pedi Hematology if pain uncontrolled after initial management, patient is febrile, or admission deemed necessary.
**Inclusion Criteria:**
Presenting with sickle cell disease and with acute pain episode.

**Triage Level 2**

**Obtain order for intranasal fentanyl 2mcg/kg (max dose 100mcg) & administer**

- IV/Port Access
- CBC, Retic count, BMP
- Blood Culture if febrile

**NS bolus 10cc/kg x1 (max 1 liter) then start 1-1.5x maintenance hydration**

**Acute Chest Syndrome Present**

- Yes → **Manage off Protocol**
- No → Present with fever

**Assess pain 20 minutes after administration of IN Fentanyl**

- Pain Improved
  - Yes → Reassess pain after 30 min
  - No → Did patient take home oral opioids within 2 hrs or NSAID within 6 hrs of arrival
    - Yes → Reassess pain 20 min after 1st opioid administered
    - No → Pain Improved
      - Yes → Reassess pain 20 min after 2nd opioid administered
      - No → Reassess pain 20 min after 3rd opioid administered

**Discharge Criteria:**
- Observe for 1 hr post opioid
- Discuss with patient/family desire to go home and home pain management
- Assess medications needed x 48hrs
- Discharge on oral ibuprofen 10mg/kg (max 600mg) & oxycodone 0.1mg/kg (max 10mg)
- Encourage PO intake
- Complete discharge pain management sheet
- Inform hematology team regarding decision to d/c
- Follow up in clinic

**ED to Inpatient Handoff:**
- There must be a nurse practitioner (NP) verbal handoff with the resident or attending before patient is admitted

**Important Triage Questions**

- History of acute chest syndrome
- Last pain crisis

**Acute Chest Symptoms**

- Chest pain
- Cough
- Fever
- Hypoxia (Low Oxygen Level)
- Lung infiltrates
Child Admitted from ED for Sickle Cell Pain

**Mild Pain** (Score 1-3)

- Administer oral Ibuprofen or IV Ketorolac 1mg/kg if 1st dose, then 0.5mg/kg every 6hr not to exceed 3 days (max 30mg/dose); Alternating with PO Tylenol. Consider PRN Opioid
- Assess pain 30 min after administration of Ketorolac
- Pain well managed?
  - Yes: Continue pain assessment per policy
  - No: Consider alternative pain methods (Table 2)

**Moderate to Severe Pain** (Score 4-10)

- Initiate PCA basal + q15min demand. Schedule Ketorolac/po NSAIDs alternate with Tylenol
- Assess pain 1hr after initiation of PCA
- Pain well managed?
  - Yes: Consider alternative pain methods (Refer to Table 1)
  - No: Consider alternative pain methods (Table 2)

**Pain remains well managed?**

- Yes: Consider alternative pain methods (Table 2)
- No: Begin DC Planning (Table 3)

**Pain well controlled?**

- Yes: Notify Attending for pain management
- No: Determine whether demand or continuous needs to be adjusted. Reassess every 30 min x2

**Oral or IV?**

- Oral: Review Home Dosing of Opioid
- IV: IV Morphine (0.2mg-0.5mg/kg) or Dilaudid 0.015 mg/kg q 2hrs pm

**Continue pain assessment every hour**

- Pain well managed?
  - Yes: Consider alternative pain methods (Table 2)
  - No: Consider extra demand dose and reassess every 30 min x2
Sickle Cell Pain Management (Inpatient) Pathway
Evidence-Based Outcomes Center

Table 1
Pain – Questions to ask to Evaluate PCA Effectiveness:

- **Is the demand dose helping?**
  - Yes, but it makes me fall asleep every time, but doesn’t last the full 15/20 minutes → Consider decreasing the bolus dose and interval
  - Yes, but it doesn’t last the full 10/15/20 minutes → Consider decreasing the dosing interval
  - No, I don’t feel it at all → Consider increasing the demand dose
- **Do you feel your pain has improved since starting the PCA?**
  - Yes, a little bit, but I’m still hitting my button a lot → Consider increasing the continuous +/- demand (depending on demand answers above)
  - Yes, it’s helping a lot → Continue as is!
  - No, I’m still a 10/10 → Consider increasing both the continuous and demand (as demand questions above as well)

Table 2
Alternative/Adjuvant Pain Management to consider:

- TENS unit (PT consult)
- Lidocaine patch
- Virtual Reality (Social Work)
- Psychology/Psychiatry consult?
- Simple/Exchange Transfusion
- Prolonged NSAID use
- For those NOT on a PCA, consider PCA with demand only to give some control of their pain management

Table 3
Discharge Planning

- Start Methadone/wean PCA
- Consult Case management 48 hours prior to discharge in case prior authorization/home health needed
- Provider should send prescriptions to DCOP/home pharmacy 48 hours prior to discharge
- Social work- school plan
References


Executive Summary

Approved by the Pediatric Evidence-Based Outcomes Center Team

Revision History
Original Date Approved: May 2019
Revision Dates:
Next Review Date: May 2023

Sickle Cell with Pain EBOC Team:
Dory Collette, RN, CCRN
Robert Mignacca, MD
Mark Tabarrok, MD
Molly McNaull, PharmD
Daryl Mozygemba, RN, MSN, CPNP-PC
Amber Bills, MSN, RN, CPN, CPON
Debra Rodriguez, MSN, RN
Denita Lyons, BSN, RN, CPEN
Anne Raines, MSN, RN, CPON
Frank James, MBA
Carmen Garudo, PM

EBOC Leadership Committee:
Lynn Thoreson, DO
Sarmistha Hauger, MD
Terry Stanley, DNP
Sujit Iyer, MD
Tory Meyer, MD
Meena Iyer, MD
Nilda Garcia, MD

Recommendations
Practice recommendations were directed by the existing evidence and consensus amongst the content experts. Patient and family preferences were included when possible.

Approval Process
EBOC guidelines are reviewed by DCMC content experts, the EBOC committee, and are subject to a hospital wide review prior to implementation. Recommendations are reviewed and adjusted based on local expertise.

LEGAL DISCLAIMER: The information provided by Dell Children’s Medical Center (DCMC), including but not limited to Clinical Pathways and Guidelines, protocols and outcome data, (collectively the "Information") is presented for the purpose of educating patients and providers on various medical treatment and management. The Information should not be relied upon as complete or accurate; nor should it be relied on to suggest a course of treatment for a particular patient. The Clinical Pathways and Guidelines are intended to assist physicians and other health care providers in clinical decision-making by describing a range of generally acceptable approaches for the diagnosis, management, or prevention of specific diseases or conditions. These guidelines should not be considered inclusive of all proper methods of care or exclusive of other methods of care reasonably directed at obtaining the same results. The ultimate judgment regarding care of a particular patient must be made by the physician in light of the individual circumstances presented by the patient. DCMC shall not be liable for direct, indirect, special, incidental or consequential damages related to the user's decision to use this information contained herein.