Sickle Cell with Pain Crisis

May 24, 2019

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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.

<table>
<thead>
<tr>
<th>Recommendation</th>
<th>Type of Evidence</th>
</tr>
</thead>
<tbody>
<tr>
<td>Strong</td>
<td>Desirable effects clearly outweigh undesirable effects or vice versa</td>
</tr>
<tr>
<td>Weak</td>
<td>Desirable effects closely balanced with undesirable effects</td>
</tr>
<tr>
<td><strong>Type of Evidence</strong></td>
<td></td>
</tr>
<tr>
<td>High</td>
<td>Consistent evidence from well-performed RCTs or exceptionally strong evidence from unbiased observational studies</td>
</tr>
<tr>
<td>Moderate</td>
<td>Evidence from RCTs with important limitations (e.g., inconsistent results, methodological flaws, indirect evidence, or imprecise results) or unusually strong evidence from unbiased observational studies</td>
</tr>
<tr>
<td>Low</td>
<td>Evidence for at least 1 critical outcome from observational studies, from RCTs with serious flaws or indirect evidence</td>
</tr>
<tr>
<td>Very Low</td>
<td>Evidence for at least 1 critical outcome from unsystematic clinical observations or very indirect evidence</td>
</tr>
</tbody>
</table>
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

**Presents with fever**
- Yes: Start Sepsis Protocol
- No

Assess pain 20 minutes after administration of IN Fentanyl

**Pain Improved**
- Yes
- No

Did patient take home oral opioids within 2 hrs or NSAID within 6hrs of arrival

**IV opioid (up to 3 doses), Give one of 3 listed:**
- Morphine IV 0.1mg/kg (max dose 8mg)
- Fentanyl IV 2mcg/kg (max dose 100mcg)
- Dilaudid IV 0.015 to 0.02mg/kg (max dose 1 mg)

**Pain Improved**
- Yes
- No

Administer appropriate oral opioid, then NSAID per home routine or Consider Toradol 0.5mg/kg (<16yo max 15mg; >16yo max 30)

*Use with caution if known renal dysfunction*

**Reassess pain after 30 min**

**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

**Last Updated May 24, 2019**
Executive Summary

Approved by the Pediatric Evidence-Based Outcomes Center Team

Revision History
Original Date Approved: May 2019
Revision Dates:
Next Review Date: May 2021

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

EBOC Committee:  
Lynn Thoreson, DO  
Sarmistha Hauger, MD  
Terry Stanley, DNP  
Deb Brown, RN  
Sujit Iyer, MD  
Tory Meyer, MD  
Nilda Garcia, MD  
Meena Iyer, MD  
Michael Auth, DO  
Jorge Ganem, 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.

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References


