Population: Patients with any sickle hemoglobinopathy (most commonly HbSS, HbSC, HbS-beta\(^0\) thalassemia, HbS-beta\(^+\) thalassemia) presenting with pain.

History and Physical Examination

• Characteristic of vaso-occlusive pain episode (VOE)
  o Location, character, severity, and duration of pain; comparison to usual VOE
• Pain assessment
  o Use the Wong-Baker 0-10 Faces Scale to assess children (as young as 3 y/o) and cognitively impaired patients
  o Use the 0-10 Numeric Pain Scale to assess cognitively developed children (usually \(\geq 8\) y/o)
• Assess pain management prior to presentation
  o Duration of home management, medication name(s), frequency, dose, time of last dose
• History of SCD complications: acute chest syndrome (ACS), sepsis, cerebral vascular accident (CVA), splenic sequestration, priapism, aplastic crisis
• Medications and allergies
• Transfusion history (simple and exchange)
• Assess vital signs
  o If febrile, refer to Clinical Guideline for Sickle Cell Patients with Fever and continue pain management concurrent with management of fever
• Assess circulatory, respiratory, and neurologic status
• Evaluation of liver and spleen size (compare to baseline exam in medical records)
• General assessment for evidence of focal infection
• Consider other etiologies for pain (e.g. cholecystitis, appendicitis, trauma)

*After initial assessment, contact Pediatric Hematology/Oncology fellow at pager 4363 to discuss planned interventions and to notify the Sickle Cell Disease Research Group of patient’s visit

Diagnostic Evaluation *(If IV placement required)*

• CBC with differential
• Reticulocyte count
• Baseline metabolic panel (ER Panel I) and hepatic function panel (ER Panel II)
• Consider Type and Screen if patient is in shock, ill-appearing, unusually pale, persistently tachycardic, or if there is concern for splenic sequestration
  o If transfusion is required, notify Blood Bank as patients may be difficult to cross-match due to higher antigen-antibody cross-reactivities
• Obtain CXR if patient presents with respiratory symptoms or new chest pain
  o If evidence of ACS, begin management for ACS
    ▪ Patient will likely require admission; pain management should continue concurrent with ACS management

Therapeutic Interventions

• If minimal pain management at home (< 24-48 hours or PO analgesia given on PRN basis) or mild-to-moderate pain (Pain Score < 5), refer to Clinical Pathway 1.
• If aggressive pain management at home (> 48 hours or PO analgesia given on scheduled basis) or moderate-to-severe pain (Pain Score \(\geq 5\)), refer to Clinical Pathway 2.

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Updated 9/29/2015
Pediatric Emergency Department Clinical Guideline:
Sickle Cell Disease (SCD) Patients with Pain

ED Management
• Refer to the included Dosing Guideline and Clinical Pathways
• Administer oral antihistamines for patients who experience pruritus secondary to opioid administration, and then every 4 to 6 hours as needed
• Admit if pain remains inadequately controlled after receiving 3-4 doses of IV opioid within 4 hours in ED

Dosing Guideline

<table>
<thead>
<tr>
<th>Intranasal (IN) Analgesia</th>
<th>fentaNYL</th>
</tr>
</thead>
<tbody>
<tr>
<td>≥ 3 years</td>
<td>1-2 mcg/kg</td>
</tr>
<tr>
<td></td>
<td>(max 2 doses, 5 minutes apart)</td>
</tr>
</tbody>
</table>

Intravenous (IV) Analgesia

<table>
<thead>
<tr>
<th>Ketorolac</th>
<th>0.5 mg/kg</th>
<th>(up to max dose 30 mg if &gt; 16 years and &gt; 50 kg)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Morphine</td>
<td>Loading dose: 0.1-0.15 mg/kg</td>
<td>*Repeat opioid doses at 25-50% of loading IV dose</td>
</tr>
<tr>
<td>Hydromorphone</td>
<td>Loading dose: 0.015-0.02 mg/kg</td>
<td></td>
</tr>
</tbody>
</table>

Oral (PO) Analgesia

<table>
<thead>
<tr>
<th>Ibuprofen</th>
<th>10 mg/kg</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hydrocodone and acetaminophen (<strong>Preferred oral opioid)</strong></td>
<td></td>
</tr>
<tr>
<td>2-13 years or &lt; 50 kg</td>
<td>Hydrocodone 0.1-0.2 mg/kg/dose</td>
</tr>
<tr>
<td>≥ 13 years or ≥ 50 kg</td>
<td>Hydrocodone 2.5-10 mg/dose</td>
</tr>
</tbody>
</table>

Acetaminophen and codeine

| 1-3 years | Codeine 0.5-1 mg/kg/dose |
| 3-6 years | Codeine 12 mg |
| 7-12 years | Codeine 24 mg |
| > 12 years | Codeine 30-60 mg |

Disposition
• Discharge criteria
  o Absence of other complications of SCD
  o Pain adequately controlled for a minimum of 30-60 minutes following last PO analgesia dose
  o **Patient should be discharged home with PO opioid prescription for one week’s duration; additional prescriptions will be provided by the patient’s hematologist at the follow-up appointment as needed.**

• La Rabida Admission Criteria
  o Uncomplicated VOE requiring multiple IV doses of analgesics
  o No concern for ACS or other complications of SCD

• Comer Inpatient Admission Criteria
  o Complicated VOE
  o Concern for ACS, CVA, splenic sequestration, aplastic crisis, or other complications of SCD

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Updated 9/29/2015
**Clinical Pathway 1: Minimal Pain Management at Home or Pain Score < 5**

1. **IN fentanyl x 2 doses**
   - Reassess pain after 30 minutes

2. **Patient declines IN fentanyl**
   - **PO Opioid**
   - **Ibuprofen**
   - Reassess pain after 30-60 minutes

   - **Pain Score < 5**
     - Patient comfortable and pain well controlled
     - Discharge home
   - **Pain Score ≥ 5**
     - Proceed to Clinical Pathway 2B

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**Pain Assessment**
- Use the Wong-Baker 0-10 Faces Scale to assess children (as young as 3 y/o) and cognitively impaired patients
- Use the 0-10 Numeric Pain Scale to assess cognitively developed children (usually ≥ 8 y/o)

*After initial assessment, contact Pediatric Hematology/Oncology fellow at pager 4363 to discuss planned interventions and to notify the Sickle Cell Disease Research Group of patient’s visit*

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Updated 9/29/2015
Clinical Pathway 2: Aggressive Pain Management at Home or Pain Score ≥ 5

Pain Assessment
- Use the Wong-Baker 0-10 Faces Scale to assess children (as young as 3 y/o) and cognitively impaired patients
- Use the 0-10 Numeric Pain Scale to assess cognitively developed children (usually > 8 y/o)

*After initial assessment, contact Pediatric Hematology/Oncology fellow at pager 4363 to discuss planned interventions and to notify the Sickle Cell Disease Research Group of patient’s visit

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Updated 9/29/2015