Caring for Children with Sickle Cell Disease in the ED

What is Sickle Cell Disease?
• An inherited blood disorder, most common among people of African, Mediterranean, or Hispanic descent
• Causes breakdown of red blood cells (sickle cell anemia)
• Distorts cells into a sickle shape that blocks blood and oxygen flow
• Triggers severe pain (vaso-occlusive episodes) and multisystem complications

Acute Complications of SCD
If not recognized or managed timely, complications may be life-threatening or result in lifelong disabilities.

ASSESSMENT
Signs and symptoms vary based on system affected, and may start to appear as early as 5 months of age.

- Fever
- Anemia
- Pain
- Dactylitis (infants)
- Priapism
- Abdominal tenderness or rigidity
- Extremity weakness
- Shock

PAIN (vaso-occlusive episodes)
• Sickled cells block blood flow in small vessels to chest, abdomen, and bones
• Sudden onset; excruciating and debilitating

STROKE (!)
• Sickled cells block vessels impairing blood flow to brain
• 10% children with SCD are affected
• Blood transfusion may be indicated*

ACUTE CHEST SYNDROME (!)
• Impaired oxygen flow to lungs
• Shortness of breath, chest pain, cough, fever
• Blood transfusion may be indicated*

FEVER/INFECTION
• High risk for infection and sepsis due to splenic damage
• Pneumonia leading cause of death in children with SCD

ANEMIA
• Red blood cells breakdown early
• Not enough RBCs to carry oxygen through body
• Blood transfusion may be indicated*

SPLENIC SEQUESTRATION (!)
• Typically occurs in children under 5 years
• Large amount sickled cells trapped in spleen
• Causes sudden splenic enlargement and left-sided abdominal pain

*Only with hematology consult

(!) = Life-threatening; requires immediate assessment and treatment
**Acute Pain**

Painful vaso-occlusive episodes can be induced by dehydration, stressors, and exposure to cold or infection. Opioids may be required for pain relief.

**Fever**

Children with SCD have compromised immunity and are at high risk for infection and sepsis.

Fever ≥ 101.3°F (38.5°C)

a. Physical exam
b. Evaluate for source of fever
c. Labs
   i. Blood cultures; additional cultures as needed
   ii. CBC with differential, reticulocyte count
d. Administer empiric parenteral antibiotics (within 1 hour)
e. Chest X-ray for chest pain, shortness of breath, tachypnea, cough or rales
f. Admission vs. Discharge
   i. Discharge following empiric antibiotics only if:
      1. Child non-ill appearing
      2. High likelihood for good follow up (next day)
      3. No other reasons for admission

Fever ≥ 103.1°F (39.5°C)

a. Admit
   i. Close observation
   ii. Parenteral antibiotics

**Patient and Family Education**

Provide patients and caregivers with education on healthy, self-management behaviors.

- Maintain hydration, nutrition, and rest
- Avoid environmental triggers (e.g., extreme hot and cold temperatures)
- Take hydroxyurea (improves quality of life; decreases hospitalizations and chances for ACS; associated with improved survival)
- Take all medications as directed
- Maintain Influenza and Pneumococcal vaccinations
- Use tools for pain management
  - Pediatric Self-Management Model - bit.ly/3kDsa0W
  - Adolescent Pediatric Pain Tool - bit.ly/3pszmke

**Individualized Prescribing and Monitoring Care Protocols**

Use of an individualized VOE protocol significantly decreases hospital admissions, and allows for consistent care across the healthcare continuum.

- Collaboratively develop individualized care protocol with child/family, primary care provider, case management, and hematologist
- Include specific pain medications, current doses, non-pharmacological pain therapies, and current pain scale used by the child or adolescent

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