Foundations Frameworks

Approach to the Sickle Cell Crisis

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Evaluate the following in every sickle cell patient:

- Cardiopulmonary signs/symptoms
- Hemoglobin and reticulocyte count
- Signs/symptoms concerning for vaso-occlusive or anemic crisis

Vaso-occlusive Crisis

- Typical SCC, consistent with previous exacerbations of pain
- Caused by anemia, dehydration, viral processes
- These patients typically have higher resting hemoglobin levels and won't have a drop in hemoglobin with vaso-occlusive crisis
- Management: pain control and rehydration
    - Fluids:
        - PO vs IV depending on level of dehydration/ability to take PO
        - IV fluid of choice: D5 ½ NS at maintenance rate
        - Severe dehydration/hypotension: NS bolus followed by D5 ½ NS
    - Opioids for pain control
        - If requiring ≥ 3 doses of pain medications, consider admission
    - Acetaminophen
    - Avoid NSAIDs as sickle cell patients typically have underlying renal dysfunction
- Evaluate for Acute Chest Syndrome
    - Leading cause of death in SC patients
    - 80% of patients have respiratory symptoms or fever
    - Diagnosis: infiltrate on XR chest plus hypoxia, fever, CP, or respiratory signs/symptoms
    - Treatment: antibiotics, fluid resuscitation as needed, exchange transfusion (or RBC transfusion to temporize), hematology consult, consider admission to ICU

Anemic Crisis

- > 2 g/dL drop from baseline hemoglobin
- Obtain hemoglobin, reticulocyte count
- Rule-out other sources of bleeding (GI)
- 3 general categories:
    - Transient Red Cell Aplasia
        - Typically, from viral infection (classically Parvovirus B19)
        - Well appearing
        - Low reticulocyte counts
        - Keep away from pregnant females
    - Splenic Sequestration

        - Typically occurs in pediatric patients (because they haven't auto-infarcted their spleen yet)
        - Large spleen on exam, LUQ pain
        - Rapid drop in hemoglobin, may be in hemodynamic shock/extremis
        - Need emergent resuscitative transfusion
        - High reticulocyte count
        - Rare in adults but possible
    - Hemolytic Crisis
        - Typical hemolytic crisis in SC patient
        - Lab abnormalities associated with hemolysis include:
            - Elevated LDH, elevated reticulocyte count, elevated LFTs/bilirubin
        - Transfuse when indicated

Special Circumstances

- Always consider/evaluate the following diagnoses in sickle cell patients:
    - Hyphemia, priapism, CVA, gallstones, osteomyelitis, sepsis, and fever (no functional spleen)

References:

- Adams, J et al. Emergency Medicine: Clinical Essentials, Second Edition. Emergency Management of Red Blood Cell Disorders, pp. 1702-1713. 2013.
- Glassberg, J, Weingart, S. Sickle Cell Disease Update. August and September 2012. Emrap.org