Sickle Cell Disease: Acute Complications

Complications of sickle cell disease arise because Hemoglobin S polymerizes and becomes rigid upon deoxygenation, causing vaso-occlusion and hemolysis.

Triggers for hypoxic states include:

infection

dehydration

stress

extreme temps

Vaso-Occlusive Crisis (Acute Pain Episode)

Vaso-occlusion → hypoxic-ischemic and reperfusion injury

Presentation: Acute pain (commonly long bones, chest, abdomen).

Investigations: Usually a clinical diagnosis, but rule out other causes.

Initial Management:

- Analgesia: acetaminophen, NSAIDs, opioids
- IV fluids

Stroke

Ischemic stroke most common in children, hemorrhagic in adults. High risk of stroke recurrence (60-90%) without secondary prevention.

Presentation: Headache, nausea & vomiting, focal neurologic deficits, seizures, altered level of consciousness.

Investigations: CT/MRI

Initial Management:

- Stabilize vitals
- Red blood cell exchange transfusion

Sepsis

Primary cause of mortality in children. ↑ risk of sepsis from encapsulated organisms as functionally asplenic.

Presentation: Fever and unwell-appearing. Highest risk if <5 years old.

Investigations: CBC+diff, blood culture, other infectious workup as appropriate.

Initial Management:

- 3rd gen cephalosporin +/- vancomycin

Splenic Sequestration

Vaso-occlusion leads to trapping of erythrocytes with rapid painful enlargement of the spleen and acute drop in hemoglobin.

Presentation: Hypotension, tachycardia, LUQ pain, splenomegaly.

Investigations: CBC+diff with retic count

Initial Management:

- Transfusion of pRBCs in small aliquots (beware auto-transfusion from spleen).
- Reassess vitals, hemoglobin, and spleen size regularly.

**If recurrent or life-threatening, consider splenectomy after acute event has resolved.*

Acute Chest Syndrome

Clinically defined as new infiltrate on CXR with respiratory symptoms or fever. Often precipitated by infection.

⚠ May progress rapidly to respiratory failure

Presentation: Chest pain, cough, SOB, tachypnea, fever, hypoxia.

Investigations: CXR, CBC+diff with retic count

Initial Management:

- Optimize ventilation. Incentive spirometry.
- Hydration and analgesia.
- Antibiotics: 3rd generation cephalosporin and macrolide (cover *Mycoplasma, S. pneumoniae*).
- Simple or exchange red blood cell transfusion.

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