SICKLE CELL DISEASE

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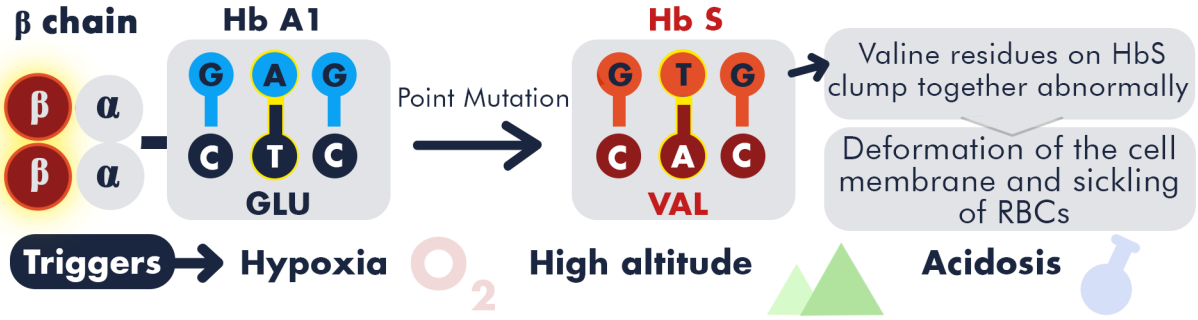


TREATMENT

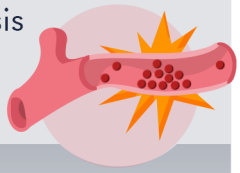
Hydroxyurea

- ↑ Fetal hemoglobin (HbF)
- Interrupts HbS polymerization

Vaso-Occlusion

- Sickled cells clog the vasculature

- **Pain**
  - Due to ischemic tissue injury
- **Acute Chest Syndrome**
  - Occlusion of pulmonary vasculature
- **Organ dysfunction**
  - Renal necrosis
  - Avascular necrosis
  - Stroke



Treatments

- Adakveo - monoclonal Ab
  - P-selectin inhibitor
  - decreases sickle cell adhesion
  - reduces vaso-occlusive crises
- L-glutamine - an amino acid
  - decreases oxidative agents
  - reduces triggers for sickling and vaso-occlusion / pain

COMPLICATIONS

Hemolysis

- Sickled blood cells are recognized as abnormal by complement system and the spleen and marked for destruction

RBC lifespan

90-120 days	10 - 20 days
normal	sickle cell

Anemia

RBC destruction causes a drop in Hb

Hemolysis

Leads to increased unconjugated bilirubin
- jaundice
- gall stones

Treatment

- Voxelotor
  - Increases hemoglobin's affinity for oxygen

Splenic Dysfunction

- Spleen filters all sickled RBCs in circulation, which clogs the organ
- Can lead to splenic infarction, dysfunction, and asplenia

Increased infection rate

-especially encapsulated bacteria

- Pneumonia
  - typical + atypical
- Osteomyelitis
  - S. aureus
  - salmonella, etc
- Bacteremia

Treatments

- Age appropriate vaccines
  - Pneumococcus
  - Meningococcus
  - Influenza
  - etc...

UpToDate vaccine recommendations

Clinical Pearl: Beware of iron overload from transfusion therapy as another complication

Sickle Cell Pain Episode Management

- **Prompt analgesia** and reassessment - the initial symptom is often pain
  - Starting dose of opioid is chosen based on the intensity of the pain compared with previous pain and effective doses
  - If previous dose is unknown: Start with **IV dilaudid** or **IV morphine** with rapid reassessment of pain
  - If pain relief is inadequate, pain medication should be re-dosed
  - If pain control continues to be inadequate, may need to use PCA

- **Hydration** is also important, but be thoughtful if the patient has cardiac and/or renal impairment
  - Fluid resuscitation and maintenance: generally **hypotonic fluids (D5W or ½ NS)**

Acute Chest Syndrome

- Etiology: Vaso-occlusion within the pulmonary vasculature
- Diagnosis: New radiodensity on CXR accompanied by fever and/or respiratory symptoms

Differential Diagnosis

- Parenchymal process: (infection, asthma, hypoventilation due to splinting, etc.)
- Necrosed marrow products (bone marrow and fat emboli released from necrosed/infarcted marrow)

Management

1 - RBC exchange
2 - Pain management
3 - IV Fluids as needed
4 - Empiric antibiotics
  - including atypicals
5 - Supplemental oxygen