Types of sickle cell crisis: Aplastic, Vaso-occlusive, Hemolytic and Sequestration

Sickle Cell Crisis refers to acute clinical episodes triggered by sickled red blood cells (RBCs) occluding microvasculature, leading to **ischemia, pain, and organ dysfunction**. Crises can be life-threatening and are a hallmark of **Sickle Cell Disease (SCD)**—a hereditary hemoglobinopathy caused by a mutation in the **HBB gene**, leading to the production of abnormal hemoglobin S (HbS).

Pathophysiology

- Under **hypoxic**, **acidotic**, or **dehydrated** conditions, RBCs containing HbS polymerize, forming a rigid, sickle shape.
- These misshapen cells obstruct capillaries ? ? blood flow ? **ischemia**, **infarction**, and **pain**.
- Sickled cells are fragile ? **hemolysis** ? chronic **anemia**.
- Chronic hypoxia ? organ damage across multiple systems.

Types of Sickle Cell Crisis

1. Vaso-Occlusive Crisis (VOC)

- **Most common type**.
- Caused by obstruction of microcirculation by sickled cells ? tissue ischemia and inflammation.
- **Clinical features**:
  - Severe localized pain (often bones, joints, chest, abdomen)
  - Fever, swelling, tenderness
- **Pediatric features**:
  - **Dactylitis (hand-foot syndrome)**: Painful, swollen hands/feet; often first sign in infants.
  - **Stroke (CVA)**: ~11% of children have overt strokes; up to 20% silent infarcts before age 18.
- **Complications**:
  - **Acute Chest Syndrome (ACS)**
  - **Priapism**
  - **Retinopathy**
  - **Bone infarction**

2. Sequestration Crisis

- **Pooling of blood** in spleen or liver ? **hypovolemia** and **shock**.
- **Common in children 3 months–5 years**.
- **Spleen** can hold ~20% of blood volume.
- **Clinical signs**:

    - Rapidly enlarging spleen
    - Pallor, tachycardia, hypotension
    - Sudden drop in hemoglobin
- **Management**:
    - Immediate transfusion
    - Splenectomy in recurrent cases (post-age 5)

3. Aplastic Crisis

- **Transient suppression of erythropoiesis**, often due to **Parvovirus B19**.
- Leads to sudden, severe anemia.
- **Signs**:
    - Pallor, fatigue, tachycardia
    - Very low reticulocyte count
- **Highly contagious**; isolation required.

4. Hemolytic Crisis

- **Acute accelerated RBC destruction** exceeds compensatory erythropoiesis.
- Can occur spontaneously or with triggers (infection, stress).
- **Findings**:
    - Jaundice
    - Dark urine
    - Elevated LDH and indirect bilirubin
- **Liver involvement** is prominent.

Acute Chest Syndrome (ACS)

Definition:

A life-threatening complication of SCD marked by new pulmonary infiltrate on imaging plus ?1 of the following:

- Fever
- Chest pain
- Cough
- Hypoxia
- Wheezing

Pathophysiology:

- Triggered by **infection**, **fat emboli**, or **VOC hypoventilation**.
- Sickled cells adhere to pulmonary endothelium ? ? inflammation ? ? O2 exchange.

Diagnosis:

- Chest X-ray: new infiltrate
- ABGs: hypoxia

- CBC: leukocytosis, anemia

Management:

- Oxygen therapy
- **Empiric antibiotics** (cephalosporins + macrolide)
- Pain control (opioids)
- Incentive spirometry
- Blood transfusion (simple or exchange)
- **Monitor for respiratory failure**

Management & Treatment

Pain Management

- **IV opioids**: Morphine 0.1 mg/kg IV q20min then q2–4h
- **PCA pump** for persistent pain
- **NSAIDs**, acetaminophen adjuncts
- Avoid meperidine (seizure risk)
- Reassess pain regularly

Hydration

- Oral or IV fluids to reduce sickling

Oxygen Therapy

- If hypoxic (SpO? < 94%)

Transfusions

- For severe anemia, ACS, CVA, or persistent VOC

Infection Prophylaxis & Treatment

- **Penicillin** prophylaxis (children)
- **Vaccination**: Pneumococcus, H. influenzae, Meningococcus
- Empiric antibiotics during febrile episodes

Disease-Modifying Therapies

- **Hydroxyurea**:
  - ? HbF ? ? sickling
  - ? frequency of VOC and ACS
- **L-glutamine**: Reduces oxidative stress
- **Crizanlizumab (P-selectin inhibitor)**: Decreases VOC frequency
- **Hydroxyurea**:
  - ? HbF ? ? sickling

    - ? frequency of VOC and ACS
- **Voxelotor**: Prevents Hb polymerization
- **Bone marrow transplant**: Potential cure

Complications of Sickle Cell Disease

System	Complication
Neurological	Stroke, silent cerebral infarct
Respiratory	Acute chest syndrome, pulmonary HTN
Hematologic	Anemia, aplastic crisis, hemolysis
Spleen	Splenic sequestration, functional asplenia
Renal	Hematuria, enuresis, hyposthenuria
Eyes	Retinopathy, blindness
Skeletal	Osteonecrosis, bone infarcts

High-Yield NCLEX & USMLE Notes

? **Hand-Foot Syndrome** is often the **first manifestation in infants**
? **Hydroxyurea** increases HbF ? ? sickling episodes
?? **ACS** is a leading cause of death; treat as emergency
? Children are at risk of **overwhelming sepsis** due to **functional asplenia**
? **Transfusions** are vital in stroke, ACS, and aplastic crisis
? **Parvovirus B19** ? aplastic crisis ? ? reticulocytes
? Recurrent **splenic sequestration** ? splenectomy after age 5
? Monitor for **reticulocytopenia**, **LDH**, **bilirubin**, **hemoglobin trends**