Hematology Alterations:
Altered Erythrocyte Function
Normocytic Anemias and Polycythemia

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Normocytic Anemia

- **Characteristics**
  - **Erythrocytes:** Normal size
  - Normal Hgb
  - Decreased number

- **Types:**
  - Aplastic
  - Post-hemorrhagic
  - Hemolytic
  - Sickle cell
  - Anemia of Chronic Diseases
Aplastic Anemia

- Pathophysiology
  - Bone marrow suppression
    - Hypocellular bone marrow
    - ↑ fat content of bone marrow
  - Pancytopenia - absence or severe depletion of all three blood cells:
    - RBC
    - WBC
    - Platelets
Aplastic Anemia

Types

- Primary acquired – unknown etiology
  - Autoimmune – cytotoxic T-cells
  - Bone marrow fails to produce blood cells

- Secondary
  - Radiation
  - Chemotherapy
  - Drugs

Refer to Table 28-4

Normocytic

Resolves with removal of offending agent
Clinical Manifestations

- Sx reflect the cell type most affected

- Usually rapidly progressing
  - Hypoxemia
  - Pallor
  - Weakness

- Disease progresses rapidly → death
  - Infection - fever
  - Bleeding
Posthemorrhagic Anemia

■ Cause
  ○ Acute blood loss

■ Effects
  ○ Internal blood loss
    ■ Iron is recovered from destroyed RBC
  ○ External blood loss
    ■ Severe iron deficiency $\rightarrow$ ↓ erythropoesis

■ Unique Clinical Manifestations
  ○ .5 – 1L: CV compensation
  ○ > 2L: poor CV compensation $\rightarrow$ shock
Hemolytic Anemia

Cause
- Premature, accelerated erythrocyte destruction

- Compensation
  - ↑ erythropoietin
  - ↑ erythropoiesis

- Hemolysis

- Anemia
  - Hemolysis > Compensation

Refer to Table 28-6
Hemolytic Anemia

Types

- Congenital defects
  - Erythrocyte cell membrane
  - Hemoglobin synthesis
    - Sickle cell anemia

- Acquired
  - Autoimmune
  - Allogenic: Transfusion reactions (mismatched RBC)
  - Allergic: Drug-induced
  - Mechanical: Prosthetic cardiac valve – RBC destruction
Hemolytic Anemia

- Unique Clinical Manifestations
  - Jaundice
    - Heme destruction → exceeds liver’s ability to conjugate and excrete bilirubin
  - Aplastic Crisis
    - No bone marrow production of RBC

Normocytic
Sickle Cell Disease

- **Description**
  - Abnormal form of Hgb S within erythrocytes
  - **Genetic mutation (autosomal recessive)**
    - Most prevalent in Blacks and African Americans
  - **Conditions cause the Hgb S changes RBC to fragile, sickle-like shape**
    - Hypoxemia
    - Acidosis
    - Dehydration
    - Hypovolemia
    - Hypothermia
  - **Sickling is not permanent – will return to normal with improved PO$_2$**
    - Pneumococcal vaccine improved mortality rates

**Normocytic**
Sickle Cell Disease

Pathophysiology

- **Sickle-shaped RBCs:**
  - Deliver less oxygen to tissues
  - Cannot pass through small blood vessels
    - Vascular occlusion
    - Pain
    - Organ ischemia \(\rightarrow\) infarction

- Spleen – Hemolysis of sickled cells
  - Vascular stasis \(\rightarrow\) infarction
  - Atrophy and ↓ function
Sickle Cell Disease

- Unique Clinical Manifestations
  - Usually appears at age 6-12 months
    - (fetal Hgb replaced by Hgb S)
  - Isolated local manifestations
    - Any body area with hypoxia
  - Chronic disease with acute exacerbations
    - Pain
    - Pallor
    - Fatigue
    - Jaundice
    - Irritability
Sickle Cell Crisis

- Vasoocclusive (thrombotic)
  - Precipitated by physiologic alterations that produce sickling
  - Vascular occlusion and vasospasm

- Aplastic
  - Transient cessation of RBC production

- Sequestration
  - Pooling of blood in liver and spleen

- Hyperhemolytic Anemia
  - Accelerated hemolysis
Anemia of Chronic Diseases

- Definition
  - Mild – moderate anemia
  - Normocytic, normochromic
  - Chronic systemic disease
    - Cancer
    - Infections
    - Autoimmune disorders
  - Asymptomatic → Progressive symptoms
    - Microcytic and hypochromic
Anemia of Chronic Diseases

- **Pathophysiology**
  - **Chronic inflammation**
    - ↑ release of cytokines by lymphocytes and macrophages
    - ↓ erythrocyte life span
    - ↓ erythropoietin production
    - Ineffective bone marrow response to erythropoietin
    - Altered iron metabolism or iron sequestration in macrophages → iron deficiency
Polycythemia

Definition
- Overproduction of RBC

Causes
- Exogenous
  - Radiation/drugs
- Endogenous
  - Physiologic compensation
  - Immune disorders
Polycythemia

Types

- **Relative polycythemia**
  - Hemoconcentration of blood – dehydration

- **Absolute**
  - Primary - Polycythemia vera
    - ↑ RBC
    - ↑ WBC
    - ↑ platelets
  - Secondary
    - ↑ erythropoietin 2° hypoxemia
Polycythemia vera

Clinical Manifestations

- **Splenomegaly**
  - Pain

- **↑ viscosity and thrombocythemia**
  - Thrombosis (peripheral and GI organs)
  - Plethora (red color of face, hands, feet, ears, mucous membranes)

- **Vascular**
  - Compensation by CV system
  - ↓ peripheral circulation – Raynaud syndrome
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