Chapter 33
Hematologic Disorders

Learning Objectives

- Identify the anatomy of the hematopoietic system
- Describe the components of blood and volume, volume control in relation to the hematopoietic system
- Identify and describe blood-forming organs and how/where blood is formed

Learning Objectives (Cont'd)

- Describe normal red blood cell production, function, lifespan, and destruction
- Explain the significance of hematocrit in regard to red blood cell size and number
- Explain correlations of red blood cell count, hematocrit, and hemoglobin values
Learning Objectives (Cont’d)

- Describe normal white blood cell production, function, and destruction
- Identify characteristics of the inflammatory process
- Identify alterations in immunological response

Learning Objectives (Cont’d)

- Describe the number, normal function, types, and lifespan of leukocytes
- Identify differences between cellular and humoral immunity
- Describe platelets in regard to normal function, lifespan, and numbers

Learning Objectives (Cont’d)

- Describe components of the hemostatic mechanism
- Describe the function of coagulation factors, platelets, and blood vessels necessary for normal coagulation
- Describe intrinsic and extrinsic clotting systems and identification of factor deficiencies in each stage
Learning Objectives (Cont'd)

- Define fibrinolysis
- Describe disseminated intravascular coagulation and its precipitating factors
- Identify blood groups
- Define anemia

Learning Objectives (Cont'd)

- Describe the pathology, clinical manifestations, and prognosis associated with:
  - Aplastic anemia
  - Hemoglobinopathy (including sickle cell disease)
  - Hemolytic anemia
  - Iron-deficiency anemia
  - Methemoglobinemia

Learning Objectives (Cont'd)

- Describe the pathology and clinical manifestations associated with disorders of hemostasis: platelet dysfunction, thrombocytopenia, decreased production, platelet destruction, sequestration, and hemophilia
- Describe the pathology and clinical manifestations associated with leukocyte disorders: leukemia, lymphoma, and multiple myeloma
Learning Objectives (Cont’d)

- Identify the components of physical assessment in relation to the hematological system; integrate pathophysiological principles into the assessment of hematological disease

Introduction

- Hematology
  - Study of blood, parts, and functions
- Blood
  - Carries nourishment, oxygen, waste products
  - Regulates temperature, balances fluid and electrolytes, regulates pH, prevents fluid loss, prevents disease
  - Maintains homeostasis

Anatomy and Physiology of the Hematopoietic System

- Functions and characteristics of blood
  - Connective tissue
  - Cells, cell fragments suspended in plasma
  - Supply oxygen, nutrients to cells
  - Transport carbon dioxide, nitrogenous wastes from tissues to lungs, kidneys
  - Carry hormones from endocrine glands to target tissues
Anatomy and Physiology of the Hematopoietic System (Cont’d)

- Functions and characteristics of blood
  - Regulate body temperature
  - Regulate pH through buffering components
  - Keep fluid, electrolytes balanced through salt, plasma proteins
  - Regulate the immune system through white blood cells, antibodies
  - Form clots through platelets

- Blood composition
  - 55% plasma
  - 45% formed cellular fragments
  - Hematocrit
  - Plasma
  - Formed elements

- Hematopoietic stem and progenitor cells
  - Formation of blood/blood cells in body
  - Starts with stem cell, matures into RBCs, WBCs, platelets
    - Originates in bone marrow
    - Have the ability to renew
    - As more differentiated, lose ability to self-renew
Anatomy and Physiology of the Hematopoietic System (Cont’d)

- Red blood cells
  - Normal erythropoiesis
    - tissues need oxygen for aerobic metabolism
    - Production
    - Major components, including RBC structure, function, turnover
    - Bone marrow capacity to produce new RBCs
    - Growth factor regulation

Anatomy and Physiology of the Hematopoietic System (Cont’d)

Development of Cells of the Hematological System

- Red blood cell structure
  - Allows maximal flexibility as it travels through microvasculature
  - Mature cell, biconcave disc
  - Lacks nucleus, mitochondria
  - Hemoglobin
  - 100-200 day lifespan
Anatomy and Physiology of the Hematopoietic System (Cont’d)

- Clinical and laboratory measures
  - Hematocrit
    - Volume of packed red cells/packed cell volume
    - Measures volume of whole blood composed of RBCs
    - Spun in centrifuge, measure red cell column height, compare to total height of whole blood column
    - Portion of total blood volume occupied by red cell mass
    - Depends mostly on RNCs number
    - 42-52% males, 36-48% females
    - Three times hemoglobin value
    - May be affected by altitude, patient position, heavy smoking

- RBC count
  - Number of RBCs/microliter
  - Estimate of hemoglobin content of blood

- Hemoglobin
  - Molecule of RBC carries oxygen
  - Blood capacity
  - Total blood hemoglobin depends on RBC number
White blood cells

- Normal myelopoiesis
  - Produce differentiated cells, provide body's host defense
  - Myeloid cell production, share common precursor cell
  - With acute infection, WBCs release colony-stimulating factor, prompts marrow to increase WBC production
  - Leukocytosis
  - Leukopenia

Types
- Neutrophils
- Eosinophils
- Basophils
- Lymphocytes
Anatomy and Physiology of the Hematopoietic System (Cont’d)

● Platelets
  ➢ Formation of clots, coagulation
  ➢ Platelet, vessel wall, von Willebrand factor, fibrinogen must work together for adhering, aggregation, clot formation
  ➢ Function
  ➢ Normal count: 150-400 million/mL of blood
  ➢ Factors influencing platelet count include exercise, racial origin
  ➢ 10 day lifespan

Anatomy and Physiology of the Hematopoietic System (Cont’d)

● Platelets
  ➢ Normal hemostasis
    • Bleeding cessation after endothelial cell injury
    • Blood vessels lined with vascular endothelium
    • Vascular endothelium disruption activates coagulation cascade
    • Achieved through vascular constriction, platelet plug formation, coagulation activation, clot formation
    • von Willebrand factor needed for adhesion

Anatomy and Physiology of the Hematopoietic System (Cont’d)

● Platelets
  ➢ Normal hemostasis
    • Coagulation cascade
    • Fibrin clot formation
    • Fibrinolysis
Blood typing and transfusions: ABO and Rh blood groups

Agglutinogens

• Specific blood type antigens found on RBC surface
• Different blood types, antigens on RBCs
• ABO blood types
• Rh blood groups
Anatomy and Physiology of the Hematopoietic System (Cont'd)

Agglutination reactions

Blood typing and transfusions: ABO and Rh blood groups
- Transfusions
  - Both ABO, Rh must match
  - AB-positive, universal recipient
  - O-negative, universal donor

Red Blood Cell Disorders
- Aplastic anemia
  - Decreased production of 1+ major hematopoietic lineages within bone marrow
  - Marrow failure, differentiation of pluripotent stem cells
  - Classic marrow failure disorder
Red Blood Cell Disorders (Cont'd)

- Aplastic anemia
  - Inherited in autosomal recessive pattern
  - Growth retardation, congenital skeleton defects
  - Heterogeneous phenotypic presentation
  - Physical deformities, hematological abnormalities
  - Acute manifestations

Red Blood Cell Disorders (Cont'd)

- Hemoglobinopathy
  - Single amino acid substitution in one globin chain
  - Sickle cell
  - Physical findings
    - Recurrent pain, chronic complications
      - Anemic
      - Vasoocclusion
      - Growth retardation, psychosocial problems, susceptibility to infection
Red Blood Cell Disorders (Cont'd)

- Hemolytic anemia
  - Premature RBC destruction
  - Hereditary, acquired glucose-6-phosphate dehydrogenase (G6PD) deficiency
  - Increased fatigue, decreased exercise tolerance
  - Congestive heart failure
  - Iron-deficiency anemia

Red Blood Cell Disorders (Cont'd)

- Methemoglobinemia
  - Oxidation of iron in hemoglobin from ferrous to ferric state
  - Causes
  - Suspect with unexplained cyanosis, with normal PaO₂

Disorders of Hemostasis (Cont'd)

- Thrombocytopenia
  - Platelet count <150,000/mm³
  - Decreased platelet production, increased destruction, sequestration
  - Causes
Disorders of Hemostasis (Cont’d)

- Hemophilia
  - Delayed clotting, difficulty controlling hemorrhage
  - Factor VIII/IX reduced, genetic mutations of portion of the X chromosome
  - One in 1000 births
  - Hemophilia A
  - Hemophilia B

Disorders of Hemostasis (Cont’d)

- Disseminated intravascular coagulation
  - Coagulation cascade triggered abnormally
  - Systemic thrombohemorrhagic disorder
  - Intravascular fibrin production, procoagulants and platelet consumption
  - Tissue damage leads to blood clots
  - Excessive clotting

White Blood Cell Disorders

- Acute leukemia
  - Immature hematopoietic progenitor cells, rapidly multiply, displace normal elements within marrow, peripheral blood
  - Fatal if untreated
  - Spontaneous cell turnover
  - Life-threatening elevations of uric acid, potassium, phosphate in tumor lysis syndrome
White Blood Cell Disorders (Cont'd)

- **Hodgkin’s lymphoma**
  - Reed-Sternberg cell
  - Peaks in second and third decades of life, again in sixth and seventh decades
  - Fever, night sweats, weight loss, pruritis
  - Lymph node histology
  - Bone marrow transplantation

White Blood Cell Disorders (Cont'd)

- **Non-Hodgkin’s lymphoma**
  - Derived from B cells
  - Adenopathy, abdominal mass
  - Classified through morphology, immunophenotype, genetic features, clinical characteristics
  - Indolent lymphomas
  - Chemotherapy can cure
  - Lymphoblastic lymphoma, B cell-derived
  - Burkitt’s lymphoma

White Blood Cell Disorders (Cont'd)

- **Multiple myeloma**
  - Plasma-cell dyscrasia
  - Lose ability to respond to signals from immune cells
  - Divide, form abnormal proteins, damage to bone, bone marrow, other organs
  - Rapid, repeated plasma cell production interferes with normal blood cell production
  - Plasma cells cause lytic lesions in skeleton/soft tissue masses
Prehospital Management

- History and physical examination
  - ABCs
  - Assess primary complaint
  - Inquire about pertinent systems review
  - Past medical history
  - Medication
  - Illicit drug use
  - Allergies
  - Family history

Prehospital Management (Cont’d)

- Management
  - Stabilize hemodynamics
  - Secure airway, breathing
  - Bag-mask
  - Supplemental high-flow O₂
  - Endotracheal intubation
  - Shock, suspected blood loss, large-bore IV lines
  - Administer pain medication
  - Rapid transport

Prehospital Management (Cont’d)

- Epidemiology
  - Hypoxia, infection, anemia
Prehospital Management (Cont’d)

- Therapeutic interventions
  - ABCs
  - IV
  - Basic, immediately reversible causes of altered mental status
  - Bleeding, standard measures, direct pressure, elevation
  - Sickle cell, analgesics, isotonic IV

Prehospital Management (Cont’d)

- Patient and family education
  - Reassurance, comfort
  - Avoid head trauma, intracranial hemorrhage

Chapter Summary

- Blood is connective tissue; it consists of cells and cell fragments and comprises approximately 8% of total body weight (5 to 6 L)
- Blood composed of 55% plasma and 45% formed cellular fragments
- RBCs developed through a carefully regulated process known as erythropoiesis
Chapter Summary (Cont’d)

- WBCs are the body’s normal host defense; they include neutrophils, eosinophils, basophils, monocytes, and macrophages
- Platelet function requires cohesion among platelet, vessel wall, von Willebrand factor, and fibrinogen

Chapter Summary (Cont’d)

- Agglutinogens are specific blood type antigens on the surface of the RBC membrane
  - Antibodies are made of agglutinogens (i.e., antigens).
  - A, B, O, Rh agglutinogens must be matched before transfusion

Chapter Summary (Cont’d)

- Diagnosis of the specific cause of a patient’s hematological symptoms in the field is impossible; most patients with hematological disorders show symptoms of hypoxia, infection, and anemia
- Each clinical disorder can present with acute, life-threatening manifestations; must manage the ABCs and be prepared for aggressive resuscitation
Questions?