Chapter 32
Hematology

Learning Objectives
• Describe the physiology of blood and its components
• Discuss the pathophysiology and signs and symptoms of specific hematological disorders.
• Outline the general assessment and management of patients with hematological disorders.
Blood and Blood Components

- Blood composed of cell and formed elements surrounded by plasma
  - 95 percent volume of formed elements consists of red blood cells (RBCs; erythrocytes)
  - 5 percent consists of white blood cells (WBCs; leukocytes) and cell fragments (platelets)

Blood and Blood Components

- Continuous blood movement keeps formed elements dispersed throughout plasma
  - Where available to carry out chief functions
    - Delivery of substances needed for cellular metabolism in tissues
    - Defense against invading microorganisms and injury
    - Acid-base balance
Blood and Blood Components

- Blood cells formed within red bone marrow
  - Present in all tissues at birth
- Adult red bone marrow primarily found in membranous bone
  - Vertebrae, pelvis, sternum, ribs
- Yellow marrow produces some white cells
  - Composed mainly of connective tissue and fat

Blood and Blood Components

- Other blood-forming organs
  - Lymph nodes
    - Produce lymphocytes and antibodies
  - Spleen
    - Stores large quantities of blood
    - Produces lymphocytes, plasma cells, antibodies
  - Liver
    - Blood-forming organ only during intrauterine life
    - Plays important role in coagulation process

Plasma

- Clear portion of blood, is about 92 percent water
- Contains three important proteins
  - Albumin
    - Most plentiful protein
    - Similar to egg white
    - Gives blood gummy texture
    - Keeps water concentration low so water diffuses readily from tissues into blood
Plasma

- Contains three important proteins
  - Globulins (alpha, beta, and gamma)
    - Transport other proteins
    - Provide immunity to disease
  - Fibrinogen
    - Responsible for blood clotting
    - Maintaining blood pH (acting as either acid or base)
    - Transporting fat-soluble vitamins, hormones, carbohydrates
    - Allowing body to digest them temporarily for food

Red Blood Cells

- Most abundant cells in body
  - Primarily responsible for tissue oxygenation
  - Appear as small rounded disks with nearly hollowed-out centers
  - Comprised mainly of water and red protein hemoglobin

Red Blood Cells

- Production continues throughout life
  - Replace blood cells that grow old and die, killed by disease or lost through bleeding
  - After production occurs in marrow, new cell divides until there are 16 RBCs
  - Cells produce hemoglobin protein until concentration of protein becomes 95 percent of dry weight of cell
  - Cell expels nucleus, giving cell its characteristic pinched look
  - New shape increases surface area of cell and oxygen-carrying potential
Red Blood Cells

- Life span of about 120 days
  - As aging occurs, internal chemical machinery weakens
  - Lose elasticity
  - Become trapped in small blood vessels in bone marrow, liver, spleen
  - Destroyed by specialized WBCs (macrophages)
  - Most components of destroyed hemoglobin molecules used again
    - Some broken down to waste product bilirubin

Red Blood Cells

- Each RBC contains about 270 million hemoglobin molecules
  - Each molecule carries 4 oxygen molecules
- Normal amount of hemoglobin about 15 g/100 mL
  - Normally a little higher in males than in females
- Number of RBCs is about 4.2 to 6.2 million cells/mm²
White Blood Cells

• Arise from bone marrow
  – Released into bloodstream
  – Destroy foreign substances (e.g., bacteria and viruses)
  – Clear bloodstream of debris

• Leukocyte production increases in response to infection
  – Causes elevated WBC count in blood
  – Bone marrow and lymph glands continually produce and maintain reserve
  – Not many WBCs in healthy bloodstream

• Normal WBC count is about 5,000 to 10,000 cells/mm²
  – Monocytes make up about 5 percent of total WBC count
  – Increase with chronic infections
  – Lymphocytes account for about 27.5 percent
  – Neutrophils about 65 percent
  – Eosinophils and basophils together about 2.5 percent
White Blood Cells

- Increased WBC count is specific for various illnesses
  - Bacterial infection
  - Inflammation
  - Leukemia
  - Trauma
  - Stress

White Blood Cells

- Differential count (also called diff)
  - Identifies different types of leukocytes present in blood
  - Test performed by
    - Spreading drop of blood on microscope slide
    - Staining slide
    - Examining under microscope

White Blood Cells

- Differential count (also called diff)
  - Identified by
    - Shape and appearance of nucleus
    - Color of cytoplasm
    - Presence and color of granules
  - Percentage of each cell type is reported
  - Red cells and platelets are examined for abnormalities in appearance
What body functions are impaired if the WBC number or function is diminished?

Platelets

• Platelets (thrombocytes) are small, sticky cell fragments
  – Important role in blood clotting
  – When blood vessel is cut
    • Travel to site and swell into odd, irregular shapes
    • Adhere to damaged wall
    • Plug the leak
    • Allow other cells to stick and form clot

• Platelets (thrombocytes) are small, sticky cell fragments
  – If damage is too great, platelets chemically signal complex clotting process or clotting cascade
    • Repair millions of ruptured capillaries each day
    • Often make rest of clotting cascade unnecessary
Hemostasis

- Initial physiological response to wounding, causes bleeding to cease
- Initiated when break in integrity of vascular endothelium

Hemostasis

- Vascular reaction or physiology of hemostasis involves
  - Vasoconstriction
    - Resulting from injury is rapid but temporary
    - In response to injury, severed blood vessels constrict and retract with aid of surrounding subcutaneous tissues
    - Vessel spasm slows blood loss immediately
    - Usually sustained as long as 10 minutes
    - Blood coagulation mechanisms activated to produce clot

Hemostasis

- Vascular reaction or physiology of hemostasis involves
  - Formation of platelet plug
    - Adhere to injured vessels and collagen in connective tissue that surrounds injured vessel
    - Contact collagen, they swell, become sticky, and secrete chemicals that activate other surrounding platelets
    - Process causes platelets to adhere to one another
    - If opening in wall is small, plug may be sufficient to stop blood loss completely
    - If opening is large, a blood clot is necessary to arrest blood flow
### Hemostasis

- Vascular reaction or physiology of hemostasis involves
  - Coagulation
  - Growth of fibrous tissue into clot that permanently closes and seals injured vessel

### Hemostasis

- Coagulation occurs as result of chemical process that begins within seconds of severe vessel injury
  - Progresses rapidly; within 3 to 6 minutes after vessel rupture, entire end of vessel filled clot
  - Within 30 minutes, clot retracts and vessel is sealed further

### Hemostasis

- Coagulation occurs as result of chemical process that begins within seconds of severe vessel injury
  - Clotting mechanism is complex process and includes three mechanisms
    - Prothrombin activator is formed in response to rupture or damage of blood vessel
    - Prothrombin activator stimulates conversion of prothrombin to thrombin
    - Thrombin in presence of calcium ions act as enzyme to convert fibrinogen into fibrin threads
    - Threads entrap platelets, blood cells, and plasma to form clot
Hemostasis

- Process of hemostasis usually is protective and required for survival
  - Can result in responses that threaten life and function
    - Myocardial infarction or stroke

Specific Hematological Disorders

- Disorders include
  - Anemia
  - Leukemia
  - Leukopenia
  - Lymphomas
  - Polycythemia
  - Disseminated intravascular coagulopathy
  - Hemophilia
  - Sickle cell disease
  - Multiple myeloma
Specific Hematological Disorders

• Anemia
  – Condition in which concentration of hemoglobin or erythrocytes is below normal
  – Precipitating causes
    • Chronic or acute blood loss
    • Decreased production of erythrocytes
    • Increased destruction of erythrocytes
  – Symptom of disease

Specific Hematological Disorders

• Anemia
  – Persons at greatest risk are those with
    • Chronic kidney disease
    • Diabetes
    • Heart disease
    • Cancer
    • Chronic inflammatory conditions
    • Persistent infections
  – Conditions interfere with production of oxygen-carrying RBCs

Iron Deficiency Anemia

• Iron is critical part of a hemoglobin molecule
  – Gives ability to bind oxygen
  – Lack of iron prevents bone marrow from making enough hemoglobin for RBCs
    • RBCs produced are small and have pale center
    • Reduced oxygen-carrying capacity
Iron Deficiency Anemia

• Most common causes
  – Blood loss from menstrual bleeding or intestinal bleeding
  – Diet low in iron usually is cause in children
  – Vitamin deficiencies can produce anemia
    • Lack of folic acid (B vitamins) is most common form of vitamin-deficiency

Can you predict the signs and symptoms of anemia?
Hemolytic Anemia

• Cause
  – Premature destruction of RBCs in blood (hemolysis) causes hemolytic anemia
    • Can result from inherited disorder inside RBC
    • Can result from disorder outside cell
    • Condition usually acquired later in life

Inherited Disorders

• Hemolysis
  – Can occur as result of abnormal rigidity of cell membrane
    • Causes cell to become trapped at an early stage of its life span in smaller blood vessels (usually of spleen)
    • In these smaller blood vessels, RBC is destroyed by macrophages

• Hemolysis
  – Can occur from genetic defect in hemoglobin within cell (e.g., sickle cell anemia and thalassemia)
  – Can occur from defect in one of the enzymes in cell that helps protect cell from chemical damage during infectious illness
    • Deficiency glucose-6-phosphate dehydrogenase is common in African-Americans
Acquired Disorders

• Acquired hemolytic anemia results from
  – Disorders in which normal RBCs are disrupted as result of mechanical forces
    • Abnormal blood vessel linings
    • Blood clots
  – Autoimmune disorders
    • Can destroy RBCs with antibodies that are produced by immune system
    • Drug-induced hemolytic anemia
    • Incompatible blood transfusion
  – Conditions that can cause hemolytic anemia when RBCs are destroyed by microorganisms in blood (e.g., malaria)

Signs and Symptoms of Anemia

• All forms of anemia share signs and symptoms
  – Fatigue and headaches
  – Sore mouth or tongue
  – Brittle nails
  – Breathlessness and chest pain

Signs and Symptoms of Anemia

• Other patient complaints are related to abnormal decrease in number of WBCs (leukopenia) or reduction in platelets (thrombocytopenia) and may include
  – Bleeding from mucous membranes
  – Cutaneous bleeding
  – Fatigue
  – Fever
  – Lethargy
Diagnosis and Treatment

• Diagnostic tools
  – Signs and symptoms
  – Patient history
  – Examination of patient's blood through blood tests and bone marrow biopsy
  – Example
    • Iron deficiency anemia usually reveals RBCs that are smaller than normal
    • Hemolytic anemia shows RBCs that are immature and abnormally shaped

• Treatment
  – Indicated to correct, modify, or diminish mechanism or process leading to defective RBC production or reduced RBC survival

Leukemia

• Refers to any of several types of cancer in which abnormal proliferation of WBCs usually occurs in bone marrow
  – Excess production of leukemic cells crowds and impairs normal production of RBCs, WBCs, and platelets
  – More common in males than females
  – More common in Caucasians than African-Americans
  – In 2008, about 46,000 American were diagnosed (2,500 of them children)
Leukemia

- Exact cause is not known
  - Genetics may play role
  - Abnormal chromosomes associated with congenital disorders (e.g., Down’s syndrome) and HIV-type viruses are associated with rare form of disease
- Other factors
  - Exposure to radiation
  - Viral infections
  - Immune defects
  - Various chemicals in home and work environments

Leukemia Classifications

- Classified as acute or chronic
  - Acute
    - Cancer cells begin proliferating at early stage of their development (arrested as immature cells)
  - Chronic
    - Implies abnormal proliferation of more mature but not fully differentiated cells
  - Classified further according to type of WBC involved
Leukemia Classifications

• Two common forms of leukemia
  – Acute lymphocytic leukemia
    • Affects mostly children under age 15
    • Sometimes called childhood leukemia
  – Acute myelogenous leukemia
    • Affects mostly middle-aged adults

Leukemia Classifications

• Two common forms of leukemia
  – In both types, abnormal WBCs are produced in such large amounts that they eventually accumulate in vital organs (liver, spleen, lymph, brain)
    • Impedes function of these organs and leads to death
  – Chronic forms of leukemia can develop slowly, often over many years
    • Often are discovered by chance during routine blood analysis

Leukemia Signs and Symptoms

• Proliferation of leukemic cells or resulting inadequate production of other normal blood cells makes patient highly susceptible to
  – Serious infections
  – Anemia
  – Bleeding episodes
Leukemia Signs and Symptoms

- Signs and symptoms
  - Abdominal fullness
  - Bleeding
  - Bone pain
  - Elevated body temperature and diaphoresis
  - Enlargement of lymph nodes

- Enlargement of the liver, spleen, and testes
- Fatigue
- Frequent bruising
- Headache
- Heat intolerance
- Night sweats
- Weight loss

If a child presents with a lot of unusual bruises, what would you suspect if a diagnosis of leukemia is not known?

Leukemia Diagnosis and Treatment

- Diagnosis
  - Confirmed by bone marrow biopsy
- Severity assessed by
  - Degree of liver and spleen enlargement
  - Anemia
  - Lack of platelets in blood
Leukemia Diagnosis and Treatment

- Treatment: acute
  - Transfusion of blood and platelets
  - Antibiotic therapy to manage anemia and infection
  - Anticancer drugs
  - Radiation
  - Bone marrow transplant
- Treatment: chronic leukemia
  - Managed effectively with medication
  - Many patients require no treatment in its early stages

Lyphomas

- General term applied to any neoplastic disorder of lymphoid tissue
  - Hodgkin’s disease is one type
    - All others are called non-Hodgkin’s lymphomas
    - All lymphomas are malignant (cancerous tumors that tend to metastasize)

Hodgkin’s Disease

- Characterized by painless, progressive enlargement of lymphoid tissue found mainly in lymph nodes and spleen
  - Left unchecked, cancer cells multiply and eventually displace healthy lymphocytes
  - Suppresses immune system
Hodgkin’s Disease

• Signs and symptoms
  – Swollen lymph nodes in neck, armpits, groin
  – Fatigue
  – Chills
  – Night sweats
  – Sometimes itching, persistent cough, weight loss, shortness of breath, chest discomfort

Hodgkin’s Disease

• Rare cancer of unknown cause that may have heritable component
  – More common in males than females
  – Peak incidence in persons in their 20s and in persons between 55 and 70 years of age
  – Confirmed by identification of Reed-Sternberg cells in lymph nodes or organs affected by cancer
Hodgkin’s Disease

- Treatment
  - Depends on level of lymph node and organ system involvement (stage of disease)
  - Can consist of radiation and chemotherapy with anticancer drugs
  - Hodgkin’s disease is one of most curable cancers

Non-Hodgkin’s Lymphomas

- Vary in their malignancy according to nature and activity of abnormal cells
  - At least 10 types of non-Hodgkin’s lymphoma identified
- Ranked as low, intermediate, high grade
  - Ranking based on how aggressively disease behaves
    - Low-grade
      - Progress slowly
      - Tend not to spread beyond lymphatic system
    - High-grade
      - Can spread to distant organs within few months

Non-Hodgkin’s Lymphomas

- Signs and symptoms
  - Painless swelling of one or more groups of lymph nodes
  - Enlargement of liver and spleen
  - Fever
  - In rare cases, abdominal pain and GI bleeding
Non-Hodgkin’s Lymphomas

• Cause largely unknown
  – Burkitt’s lymphoma
    • Childhood cancer
    • In Africa, strongly associated with infection by Epstein-Barr virus
  – Other types worldwide have been linked to infection by HIV-type viruses and other conditions that affect immune system

Non-Hodgkin’s Lymphomas

• Treatment
  – Radiation therapy
  – Anticancer drugs
  – Sometimes bone marrow transplantation

Polycythemia

• Increase in total RBC mass of blood
  – May be natural response to chronic hypoxia (secondary polycythemia)
  – May occur for unknown reasons (primary polycythemia)
  – Can result from dehydration (apparent polycythemia)
    • RBC production does not exceed upper limits of normal
Secondary Polycythemia

- Can be naturally present in persons who live in or visit areas of high altitude
  - Due to reduced air pressure and low O₂
  - When O₂ supply to blood is reduced, kidneys produce hormone erythropoietin
    - Stimulates RBC production in bone marrow to make up for reduced O₂ supply
    - Result is increase in oxygen-carrying efficiency of blood

Secondary Polycythemia

- RBC numbers return to normal when person returns to sea level
  - Can be present in heavy smokers
  - Can be caused by chronic bronchitis and conditions that increase erythropoietin production (e.g., liver cancer and some kidney disorders)

Primary Polycythemia

- Also known as polycythemia vera
  - Rare disorder of bone marrow
  - Increased production of RBCs causes blood to thicken
  - Primarily develops in persons 50 or older
Primary Polycythemia

- Can lead to several physiological problems:
  - Blurred vision
  - Dizziness
  - Generalized itching
  - Headache
  - Hypertension
  - Red hands and feet; red-purple complexion
  - Splenomegaly

Other complications
- Platelet disorders, which cause bleeding or clot formation
- Stroke
- Development of other bone marrow diseases (e.g., leukemias)

Treatment
- Phlebotomy
  - Slow removal of blood through vein
- Anticancer drug therapy
  - Controls overproduction of RBCs in marrow
Disseminated Intravascular Coagulopathy

• Complication of severe injury, trauma, or disease
• Common abnormal clotting disorder
  – Most often seen in critical care setting
  – Disrupts balance among
    • Procoagulants
    • Inhibitors
    • Thrombus formation
    • Lysis

Disseminated Intravascular Coagulopathy

• Signs and symptoms
  – Dyspnea
  – Bleeding
  – Those associated with hypotension and hypoperfusion

Disseminated Intravascular Coagulopathy

• Occurs in two phases
  – First phase characterized by
    • Free thrombin in blood
    • Fibrin deposits
    • Aggregation of platelets
  – Second phase characterized by
    • Hemorrhage caused by depletion of clotting factors
Disseminated Intravascular Coagulopathy

- Clinical consequences predispose patient to multiple-system organ failure from bleeding and coagulation disorders caused by
  - Loss of platelets and clotting factors
  - Fibrinolysis
  - Fibrin degradation interference
  - Small vessel obstruction, tissue ischemia, RBC injury, and anemia from fibrin deposits

Disseminated Intravascular Coagulopathy

- Confirmed through laboratory tests

- Treatment
  - Aimed at reversing underlying illness or injury that triggered event
  - In effort to control depletion of clotting factors, in-hospital care includes
    - Replacement of platelets
    - Coagulation factors
    - Blood
  - At same time, attempts are made to manage primary process
Hemophilia

• Refers to a medical condition that causes uncontrolled bleeding and loss of bleeding control mechanisms
  – Group of inherited bleeding disorders
  – Hemophilia A is due to a deficiency in factor VIII
    • Factor essential to process of blood clotting
  – Hemophilia B is caused by a deficiency of factor IX
    • Also is known as Christmas disease
  – All types present with similar problems
    • Specific factor involved determines severity of bleeding
  – About 18,000 people in U.S. have hemophilia
  • About 400 are born with disorder each year

Hemophilia

• Bleeding from hemophilia can occur spontaneously, even after minor injury
  – Can occur during some medical procedures (e.g., tooth extraction)
  – Hemorrhage can occur anywhere in body

Hemophilia

• Bleeding from hemophilia can occur spontaneously, even after minor injury
  – Most common sites
    • Joints
    • Deep muscles
    • Urinary tract
    • Intracranial sites
  – Head trauma is potentially life threatening
  – CNS bleeding is major cause of death for patients in all age groups
Hemophilia

- Controlled by infusions of concentrates of factor VIII
  - Can be administered by patient
  - Serious or unusual bleeding often requires hospitalization
  - Patients are advised to avoid activities that may increase risk of injury
  - Most patients are knowledgeable about their disease
  - Most seek emergency care only when complicated problems and trauma-related issues arise

Imagine that you are caring for a patient with hemophilia who has fallen 15 feet from a ladder. This patient refuses care and transportation. What should you do?

Thrombocytopenia

- Low platelet count
  - In healthy people, blood normally contains 150,000 to 450,000 platelets/microliter of blood
  - At levels of 20,000 to 30,000 platelets/microliter of blood, bleeding can occur with relatively minor trauma
  - At levels less than 20,000 platelets/microliter of blood, spontaneous bleeding can occur, increasing risk for shock and death
    - Especially true if bleeding occurs in brain
    - Bleeding on skin is usually first sign of low platelet count
Thrombocytopenia

- Bleeding of skin may appear as
  - Small red or purple spots on skin (petechiae), often on lower legs
  - Purple, brown, red bruises (purpura) that happen easily and often
  - Prolonged bleeding, even from minor cuts
  - Bleeding or oozing from mouth or nose, especially nosebleeds or bleeding from brushing teeth
  - Unusually heavy menstrual flow

Thrombocytopenia

- Can occur when body either doesn’t produce enough platelets
  - If too many platelets are destroyed
  - If spleen holds on to too many platelets

Thrombocytopenia

- Often associated with
  - Leukemia or lymphoma
  - Aplastic anemia
  - Vitamin B12 or folic acid deficiency anemias
  - Enlarged spleen
  - Infectious diseases such as HIV/AIDS
  - Massive blood transfusions
Thrombocytopenia

- Two diseases that occur because of increased destruction of platelets
  - Idiopathic thrombocytopenic purpura (ITP)
    - Occurs when antibodies attack and destroy body's platelets for unknown reasons
    - In children, can be acute condition that occurs after infection
    - Acute ITP is rare in adults
    - Chronic ITP most frequently affects women ages 20 to 40 years

- Thrombotic thrombocytopenic purpura (TTP)
  - Life-threatening disease that occurs when small blood clots form suddenly throughout body
  - Can result in cardiac hemorrhage and death
  - Occurs more often in women and is associated with pregnancy, metastatic cancer, chemotherapy, HIV/AIDS, some prescription drugs
  - Patients experience kidney failure or decreased kidney function, fever, neurological complications

Thrombocytopenia

- Treatment depends on cause and severity
  - Some only require careful monitoring of platelet counts
  - More serious cases
    - Corticosteroids (prednisone)
    - Transfusion of platelets
    - Rarely, surgical removal of spleen
Sickle Cell Disease

- Inherited blood disorder that affects red blood cells
- Several types, most common is sickle cell anemia
- Debilitating and unpredictable genetic illness

Sickle Cell Disease

- Affects persons of African descent, and less commonly, persons of Mediterranean origin
  - 1 in 12 African-Americans
  - More than 70,000 Americans of different ethnic origins have disease
  - In U.S., about 1,000 are born with disease each year
  - 12.5 million Americans have sickle cell trait

Sickle Cell Disease

- Signs and symptoms
  - Delayed growth, development, and sexual maturation in children
  - Jaundice
  - Priapism in adolescent and adult males
  - Splenomegaly
  - Stroke
Sickle Cell Pathophysiology

- Produces abnormal type of hemoglobin called hemoglobin S
  - Abnormal type has inferior oxygen-carrying capacity
  - When hemoglobin S is exposed to low O₂ states, it crystallizes
    - Distorts RBCs into sickle shape
    - Sickle-shaped cells are fragile and easily destroyed
    - Unable to pass easily through tiny blood vessels and block flow to various organs and tissues
    - This causes vasoocclusive sickle cell crisis that can be life threatening

Sickle Cell Pathophysiology

- As fewer RBCs pass through congested vessels, tissues and joints become starved for O₂ and other nutrients
  - Causes excruciating pain
Sickle Cell Pathophysiology

- Signs and symptoms
  - Increased weakness
  - Aching
  - Chest pain with shortness of breath
  - Sudden and severe abdominal pain
  - Bony deformities
  - Icteric (jaundice) sclera
  - Fever
  - Arthralgia (joint pain)
How do you think a client with such chronic pain must feel at the beginning of a sickle cell crisis?

Sickle Cell Pathophysiology

- Sickle cell crisis
  - Can occur in any part of body
  - Can vary in intensity from one person to next and from one crisis to next
  - Over time can destroy spleen, kidneys, gallbladder, other organs
  - May occur for no apparent reason

Sickle Cell Pathophysiology

- It also may be triggered by
  - Dehydration
  - Exposure to extremes in temperature
  - Infection
  - Lack of O₂
  - Strenuous physical activity
  - Stress
  - Trauma
Sickle Cell Pathophysiology

• 3 less common types of sickle cell crisis
  – Aplastic
    • Bone marrow temporarily stops producing RBCs
  – Hemolytic
    • RBCs break down too rapidly to be replaced adequately
  – Splenic sequestration
    • Childhood difficulty that occurs when blood becomes trapped in spleen
    • Causes organ to enlarge and may lead to death

Sickle Cell Management

• No cure exists
• Because of eventual damage to spleen
  – Patients are at increased risk for septicemia if infected by certain types of bacteria
  – Children with disease should be current with all immunizations

Sickle Cell Management

• When in crisis, require prompt treatment with
  – O₂ if hypoxic
  – IV therapy to manage dehydration
  – Antibiotics to manage infection
  – Analgesics (e.g., morphine) to manage pain
**Sickle Cell Management**

- In severe cases, blood transfusion may be indicated
  - Effect temporary replacement of hemoglobin S
  - Can be advised during pregnancy to reduce risk of a crisis that can be fatal to mother and fetus
  - May be advised before surgery because anesthesia can be hazardous to those with disease

**Multiple Myeloma**

- Malignant neoplasm of bone marrow
  - Tumor, composed of plasma cells, destroys bone tissue (especially in flat bones)
  - Causes
    - Pain
    - Fractures
    - Hypercalcemia
    - Skeletal deformities

- Neoplastic cells produce large amounts of protein (M protein) that affect viscosity of blood
- Masses of coagulated protein can accumulate within tissues and impair function
Multiple Myeloma

• Some patients die of kidney failure
  – Kidneys fail because of buildup of proteins that infiltrate kidneys and block renal tubules
  – In many ways, resembles leukemia
  – Plasma cell proliferation generally is confined to bone marrow

Multiple Myeloma

• Other associated disorders
  – Proteinuria
  – Anemia
  – Weight loss
  – Pulmonary complications from rib fracture
  – Recurrent infections from suppression of immune system

Multiple Myeloma

• Patient complaints
  – Weakness
  – Skeletal pain
  – Hemorrhage
  – Hematuria
  – Lethargy
  – Weight loss
  – Frequent fractures
Multiple Myeloma

- Occurs rarely before 40 years of age, then occurs increasingly with age
  - Disease is more common in males than females and may have heritable component
- Diagnosed through
  - X-ray films
  - Blood studies
  - Tumor biopsy

Multiple Myeloma

- Treatment
  - Chemotherapy with anticancer drugs
  - Radiation
  - Plasma exchange
  - Bone marrow transplantation

General Assessment and Management

- Most patients are knowledgeable about their disease
  - Often call EMS to help manage “change” in their condition
  - May call to arrange for transportation to an emergency department for physician evaluation
  - Situations that invoke call for emergency care vary by patient and disease
  - Common chief complaints can be classified by body system
Prehospital Care

- Mainly supportive, must perform
  - General assessment
  - Focused history
  - Focused physical examination
    - Will guide patient care
    - Will help determine appropriateness of emergency transport
- Some patients will have complex medical histories
  - When possible, should be transported to primary hospital where they usually receive medical care

Prehospital Care

- Patient may have variety of complaints and physical findings
  - Some may be vague
    - Can further complicate assessment

Prehospital Care

- After ensuring adequate airway, ventilatory, circulatory status
  - Assess vital signs
  - Perform physical examination
  - Assess skin for color and turgor, noting any cyanosis or jaundice, warmth or coolness, bruising, edema, or ulcerations
Prehospital Care

- Ascertain any new onset of
  - Fever
  - Weakness
  - Cough
  - Rash
  - Spontaneous bleeding (e.g., bleeding gums, epistaxis)
  - Vomiting
  - Diarrhea

- Some hematological disorders can involve ability of blood to deliver enough oxygen to tissues
  - Question all patients with hematological disorders specifically about
    - Recent dizziness
    - Syncope
    - Difficulty breathing
    - Heartbeat irregularities

- Other key elements
  - Identify existing hematological disease
    - Including any family history of hematological disease
  - Significant medical history or recent injury
  - Medication use
  - Allergies
  - Alcohol or illicit drug use
Prehospital Care

- Based on patient’s condition, prehospital care:
  - O₂ administration
  - IV fluid replacement
  - Antidysrhythmics
  - Analgesics for pain management
  - Some of these patients will be gravely ill
    - Calming and comfort measures for patient and family

Summary

- Blood is composed of cells and formed elements surrounded by plasma
  - About 95 percent of volume of formed elements consists of RBCs (erythrocytes)
    - Remaining 5 percent consists of WBCs (leukocytes) and cell fragments (platelets)

Summary

- Anemia is condition in which amount of hemoglobin or erythrocytes in blood is below normal
  - Two common forms of anemia are iron deficiency anemia and hemolytic anemia
  - All forms of anemia share signs and symptoms
    - Include fatigue and headaches, sometimes a sore mouth or tongue, brittle nails, and, in severe cases, breathlessness and chest pain
    - Diagnosis is made by history and from blood tests and bone marrow biopsy
Summary

• Leukemia refers to any of several types of cancer in which abnormal proliferation of WBCs usually occurs in bone marrow
  – Proliferation of leukemic cells crowds and impairs normal production of RBCs, WBCs, and platelets
  – Leukemia is classified as acute or chronic
  – Proliferation of leukemic cells makes the patient highly susceptible to serious infections, anemia, and bleeding episodes
  – Diagnosis is confirmed by bone marrow biopsy

Summary

• Lymphoma refers to a group of diseases that range from slowly growing chronic disorders to rapidly evolving acute conditions
  – Hodgkin’s disease is one type; all others are called non-Hodgkin’s lymphomas

Summary

• Polycythemia is characterized by an unusually large number of RBCs in blood as a result of their increased production by bone marrow
  – Polycythemia may be natural response to hypoxia
    • Known as secondary polycythemia
  – Polycythemia also may occur for unknown reasons
    • Known as primary polycythemia
Summary

• Disseminated intravascular coagulopathy is a complication of severe injury, trauma, or disease
  – Disrupts balance among procoagulants, thrombin formation, inhibitors, and lysis
  – Signs and symptoms of disseminated intravascular coagulation include dyspnea, bleeding, and those associated with hypotension and hypoperfusion
  – Treatment aimed at reversing underlying illness or injury that triggered event

Summary

• Hemophilia A is caused by deficiency of blood protein called factor VIII
  – Hemophilia B is caused by deficiency of factor IX
  – Bleeding from hemophilia can occur spontaneously, after even minor injury, or during some medical procedures

Summary

• Thrombocytopenia is a low platelet count
  – Can occur when body either doesn’t produce enough platelets; if too many platelets are destroyed; or if spleen holds on to too many platelets
  – Bleeding is chief complication of thrombocytopenia
Summary

- Sickle cell disease is a debilitating and unpredictable recessive genetic illness
  - Affects persons of African descent
  - Less often, affects persons of Mediterranean origin
  - Sickle cell anemia produces abnormal type of hemoglobin
    - Called hemoglobin S
    - Has inferior oxygen-carrying capacity
  - Complications include episodes of severe pain, fatigue, pallor, jaundice, stroke, delayed growth, hematuria, priapism, and splenomegaly

Summary

- Multiple myeloma is a malignant neoplasm of the bone marrow
  - Tumor destroys bone tissue (especially flat bones) and causes pain, fractures, hypercalcemia, and skeletal deformities
- In many cases of hematological disorders, prehospital treatment is supportive
  - Treatment includes ensuring adequate airway, ventilatory, and circulatory support

Questions?