HEMATOLOGIC SYSTEM

- The hematologic system consists of the blood and the sites where blood is produced, including the bone marrow and the **reticuloendothelial system** (RES).
- Blood is a specialized organ that differs from other organs in that it exists in a fluid state. Blood is composed of plasma and various types of cells.
- **Plasma** is the fluid portion of blood; it contains various proteins, such as albumin, globulin, **fibrinogen**, and other factors necessary for clotting, as well as electrolytes, waste products, and nutrients. About 55% of blood volume is plasma.
- **Serum** is plasma minus the clotting factors.

BLOOD CELLS

Cell Type	Major Function
WBC (Leukocyte) Normal: 4500-11,000/mm3	Fights infection
Granulocytes	
Neutrophil	Essential in Preventing or limiting bacterial infection via phagocytosis
Eosinophil	Involved in allergic reactions (neutralizes histamine), digests foreign proteins
Agranulocytes	
Monocyte	Enters tissue as macrophage; highly phagocytic, especially against fungus; immune surveillance
T lymphocyte	Responsible for cell- mediated immunity
Plasma Cell	Secretes immunoglobulin (Ig/ antibody) Most mature form of B lymphocytes
RBC (Erythrocyte) Hemoglobin Male: 13-18 g/dL Female: 12-16 g/dL Hematocrit Male: 42-52% Female: 35-47%	Carries hemoglobin to provide oxygen to tissues; average lifespan is 120 days
Platelet (Thrombocyte) Normal: 150,000 450,000/ mm3	Fragment of megakaryocyte; provides basis for coagulation to occur; maintains hemostasis; average lifespan is 10 days.

IRON DEFICIENCY ANEMIA (IDA)

- Results when the intake of dietary iron is inadequate for hemoglobin synthesis
- **Predisposing Factors**
  - Chronic blood loss due to trauma, menorrhagia, GI bleeding (hematemesis, melena, hematochezia)
  - Inadequate Fe+2 in the diet
  - Impaired Fe+2 absorption due to
    - ✓ Chronic diarrhea
    - ✓ Malabsorption syndrome
    - ✓ Gastrectomy
    - ✓ Celiac disease

- **Clinical Manifestations**
  - Fatigue/Easy fatigability – hallmark sign
  - Brittle hair, spoon-shaped nails **(koilonychia)** due to atrophy of epidermal cells
  - Palpitations, cold sensitivity
  - Pallor, fatigue
  - Smooth, sore tongue
  - **Plummer Vinson's Syndrome** - atrophic glossitis, stomatitis, dysphagia due to atrophy of papilla of the tongue, mouth and pharyngeal cells
  - **Pica-** due to neuronal degeneration that affects cognitive functions
  - **Angular cheilosis-** ulceration of the corner of the mouth
  - **Cerebral hypoxia**- dizziness, dyspnea

❖ **Diagnostic Procedures**
- RBC, Hemoglobin, Hematocrit, Reticulocyte count, Serum Fe+2, Ferritin

❖ **Management**
- Correction of chronic blood loss.
- Oral or parenteral iron therapy

❖ **Nursing Management**
- Monitor all signs of bleeding
- CBR
- Increase intake of Fe+2 rich foods such as:
  - ✓ Green, leafy, vegetables,
  - ✓ Organ liver meat
  - ✓ Egg yolk
  - ✓ Legumes
  - ✓ California raisins
  - ✓ Red Meats
  - ✓ Molasses

- Administer Iron preparation as ordered.
  - ✓ FeSO4, Fe+2 gluconate, Fe+2 Fumarate
  - ✓ Taken with Vit C to increase absorption
  - ✓ Instruct the patient to avoid taking antacids and dairy products (it decreases iron absorption)
  - ✓ Best absorbed on an empty stomach, in between meals to prevent GI upset
  - ✓ Monitor S/E: anorexia, N/V, abdominal pain, diarrhea/constipation, black stools

MEGALOBLASTIC ANEMIA: PERNICIOUS ANEMIA

❖ Most dangerous of all chronic anemia due to deficiency of intrinsic factor leading to Vit. B12 malabsorption

❖ Predisposing factors
- Unknown cause
- Subtotal gastrectomy
- Hereditary
- Inflammation disorders of the ileum
  - ✓ Crohn's Disease
- Absence of intrinsic factor
- Strict vegetarian diet

❖ **Clinical Manifestations**
- Headache, dizziness, dyspnea, palpitations, cold sensitivity, general body malaise, extreme pallor
  - ✓ Sore mouth, anorexia, nausea, vomiting, loss of weight, indigestion, epigastric discomfort, recurring diarrhea or constipation.
  - ✓ **Red-beefy tongue/ Glossitis** - pathognomonic sign
  - ✓ **Paresthesia** in the extremities, difficulty maintaining their balance, lose position sense (proprioception)

❖ **Diagnostic procedures**
- ✓ CBC and blood smear decreased hemoglobin and hematocrit
- ✓ **Schilling's test** for absorption of vitamin B12 —patient receives small amount of radioactive B12 orally and 24-hour urine collection is obtained
  - **Positive:** Vitamin B12 absent in urine
  - **Negative:** Vitamin B12 present in urine

❖ **Nursing Management**
- Enforce CBR and ensure safety
- Administer Vit. B12 injections at monthly intervals for lifetime as ordered.
- Diet
  - ✓ Small frequent bland soft food
  - ✓ Increase CHO, CHON, iron & Vit C
- Avoid irritating mouthwashes. Use of soft bristled toothbrush is encouraged
- Avoid excessive heat and cold temperature
- Administer parenteral Iron preparation as ordered
  - ✓ Administer using Z-tract method to prevent discoloration and leakage to tissues
  - ✓ Do not massage the injection site, encourage ambulation instead
  - ✓ Monitor S/E such as:
    - ➢ Pain at injection site
    - ➢ Localized abscess
    - ➢ Lymphadenopathy
    - ➢ Fever and chills
    - ➢ Skin rashes
  - ✓ Watch out for anaphylaxis due to parenteral Iron supplement

MEGALOBLASTIC ANEMIA: FOLIC ACID DEFICIENCY

- Occurs if there is folic acid deficiency within 4 months
- **Risk Factors**
  - People who rarely eat uncooked vegetables
  - Alcoholism
  - Chronic hemolytic anemia
  - Pregnancy
  - Malabsorptive diseases of the small bowel such as sprue
- **Clinical Manifestations**
  - Fatigue, weakness
  - Pallor, dizziness, headache
  - Tachycardia.
  - Sore tongue, cracked lips
- **Diagnostic Procedure**
  - CBC will show decreased RBC, hemoglobin, and hematocrit with increased mean corpuscular volume and mean corpuscular hemoglobin concentration
- **Management**
  - Administer 1mg of folic acid daily
  - Folic acid intramuscular for patients with malabsorption problem
  - Small frequent meals of bland, soft food
    if sore mouth and tongue are present
  - Diet: food rich in folic acid such as beef liver, peanut butter, red beans, oatmeal, broccoli, asparagus

APLASTIC ANEMIA

- ✓ A rare disease caused by a decrease in or damage to marrow stem cells, damage to the microenvironment within the marrow, and replacement of the marrow with fat resulting in pancytopenia (decreased RBCs, WBCs' and platelets)

Predisposing Factors

- Chemicals (Benzene & its derivatives, pesticides)
- Radiation
- Immunologic injury
- Drugs causing bone marrow depression
- Broad spectrum antibiotic
  - ✓ Chloramphenicol
  - ✓ Sulfonamides — Bactrim
- Chemo therapeutic agents
  - ✓ Methotrexate
  - ✓ Nitrogen mustard
  - ✓ Vincristine
- Attack of T-cells against bone marrow
- Infections and pregnancy

Clinical Manifestations

- Signs of such as pallor, weakness, fatigue, exertional dyspnea, palpitations, fatigue
- **Infections** associated with Leukopenia: fever, headache, malaise, abdominal pain, diarrhea, erythema, pain, exudate at wounds or sites of invasive procedures, Lymphadenopathies and Splenomegaly
- **Thrombocytopenia:** bleeding from gums, nose, GI or GU tracts; purpura, petechiae, ecchymoses, retinal hemorrhage, oozing of blood from venipuncture site

Diagnostic Procedures

- Bone Marrow Aspiration shows an extremely hypoplastic or even aplastic (very few to no cells) marrow replaced with fat.
- CBC and peripheral blood smear shows decreased RBC, WBC and platelets (pancytopenia)

Management

- Removal of causative agent or toxin.
- Bone Marrow Transplantation (BMT) or Peripheral Blood Stem Cell Transplant (PBSCT)
- Immunosuppressive therapy
- Supportive treatment includes platelet and RBC transfusions, antibiotics, and antifungal administration

Nursing Management

- Administration of immunosuppressants as ordered
- Blood transfusion as ordered
- Complete bed rest
- O2dministration
- Teach patient how to minimize risk of infection

- Reverse isolation due leukopenia
- Monitor signs of infection
- Avoid SQ, IM injections Use only soft toothbrush for mouth care and electric razor for shaving

SICKLE CELL ANEMIA

❖ Is a severe hemolytic anemia that results from inheritance of the sickle hemoglobin gene. This gene causes the hemoglobin molecule to be defective. The abnormal sickle hemoglobin (HbS) acquires a crystal-like formation when exposed to low oxygen tension.

❖ **Clinical Manifestations**
- Severe pain in various parts of the body
- Tachycardia, murmurs & cardiomegaly
- Chest pain, dyspnea
- Jaundice
- Enlarged skull & facial bones due to bone marrow expansion

❖ **Complications**
- Hypoxia, ischemia, infection, poor wood healing
- Impotence
- Cerebrovascular accident
- Renal failure
- Heart failure
- Pulmonary hypertension

❖ **Sickle Cell Crisis**
- **Sickle crisis -** most common and very painful
  - ✓ Results from tissue hypoxia and necrosis due to inadequate blood flow to a specific region of tissue or organ
- **Aplastic crisis** - results from infection with the human parvovirus
- **Sequestration crisis** - results when other organs pool the sickled cells

❖ **Treatment**
- Bone marrow transplant
- **Hydroxyurea** = Increases production
- Long term RBC transfusion
- **Splenectomy**
- Peripheral **Blood Stem Cell Transplant**

❖ **Nursing Management**
- **MANAGING PAIN**
  - ✓ Treat the **triggering factors**.
    - Hypoxia: Provide oxygen support.
    - Dehydration: Infuse intravenous fluids. Encourage increased oral fluid intake.
    - Infection: Administer antibiotic medications as prescribed.
  - ✓ Support & elevate acutely inflamed joint
  - ✓ Relaxation techniques

- **PREVENT AND MANAGE INFECTION**
  - ✓ Monitor patient for signs and symptoms of infection
  - ✓ Initiate prompt antibiotic therapy

- **MONITOR AND PREVENT POTENTIAL COMPLICATIONS**
  - ✓ Always provide adequate hydration
  - ✓ Avoid cold temperature that may cause vasoconstriction
  - ✓ Protect leg from trauma and contamination to prevent leg ulcer
  - ✓ Aseptic technique

THALASSEMIA

❖ Group of hereditary anemias characterized by hypochromia (an abnormal decrease in the hemoglobin content of erythrocytes), extreme microcytosis (smaller-than-normal erythrocytes), destruction of blood elements (hemolysis), and variables degrees of anemia

❖ Associated with defective synthesis of hemoglobin; the production of one or more globulin chains within the hemoglobin molecule is reduced

- 2 classifications:
  - **Alpha-thalassemia** occur mainly in people from Asia and the Middle East
    - ✓ Milder than the beta forms and often occurs without symptoms; the erythrocytes are extremely microcytic, but the anemia, if present, is mild
  - **Beta-thalassemia** are most prevalent in people from Mediterranean regions
    - ✓ Patients with mild forms have microcytosis and mild anemia
    - ✓ Severe beta-thalassemia (i.e., thalassemia major or Cooley's anemia) can be fatal within the first few years of life if untreated
- **Management**
  - Bone Marrow Transplant
  - Blood transfusion of Packed RBSs
- **Thalassemia Major (Cooley's Anemia)**
  - Characterized by severe anemia, marked hemolysis, and ineffective erythropoiesis
  - With early regular transfusion therapy, growth and development through childhood are facilitated
  - Management: PBSCT before liver damage occurs
  - Watch out for iron overload which results from excessive iron in multiple packed RBC
    - ✓ Management: regular chelation therapy

POLYCYTHEMIA VERA (PRIMARY POLYCYTHEMIA)

- Proliferative disorder of the myeloid stem cells
- The bone marrow is hypercellular.
- Elevated levels of blood cells (erythrocyte, leukocyte, platelets)

- **Clinical Manifestations**
  - Ruddy complexion
  - Splenomegaly
  - Headache and dizziness
  - Tinnitus, fatigue and paresthesia
  - Blurred vison
  - Increased blood viscosity: angina, claudication, dyspnea and thrombophlebitis
  - Elevated blood pressure
  - Uric acid maybe elevated resulting in gout and renal stone formation
  - Generalized pruritus
  - Erythromyalgia (burning sensation in fingers and toes)

- **Diagnostic Procedures**
  - CBC
  - Bone Marrow Aspiration

- **Complications**
  - Cerebrovascular Accident
  - Myocardial Infarction
  - Bleeding due to dysfunctional large amount of platelet

- **Management**
  - Phlebotomy – removing enough blood (initially 500 mL once or twice weekly) to reduce blood viscosity and to deplete the patient's iron stores
  - Chemotherapeutic agents (eg, hydroxyurea) can be used to suppress marrow function
  - Anagrelide (Agrylin) – inhibits platelet aggregation
  - Interferon alfa-2b (Intron-A) – for management of pruritus (WOF: flulike syndrome and depression)
  - Antihistamine
  - Allopurinol

- **Nursing Management**
  - Instruct the patient to avoid sedentary behaviours, crossing of legs, wearing tight or restrictive clothing
  - Avoid aspirin and aspirin-containing medications
  - Minimize alcohol intake
  - Instruct the patient to avoid iron supplements
  - For pruritus:
  - Bathing in tepid or cool water
  - Avoiding vigorous toweling off after bathing
  - Use of cocoa butter or oat meal-based lotions and bath products
  - Dissolved baking soda in bath water

HEMOPHILIA

- Inherited bleeding disorder
- Hemophilia A – caused by genetic disease that results in deficient or defective factor VIII
- Hemophilia B (Christmas Disease) – genetic defect that causes deficient or defective factor IX
- Both types of hemophilia are inherited as X-linked traits, so almost all affected people are males; females can be carriers
- Recognized in early childhood, usually in the toddler age group

- **Clinical Manifestations**
  - Hemorrhages into various parts of the body
  - Hemarthroses and hematomas
  - 75% of all bleeding occurs into joints
  - Chronic pain or ankylosis (fixation) of the joint occurs
  - Spontaneous hematuria and GI bleeding
  - Intracranial or extracranial bleeding – most dangerous

- **Management**
  - Administration of factor VIII and factor IX concentrates
  - Infusion of fresh frozen plasma
  - Plasmapheresis or concurrent immunosuppressive therapy
  - Aminocaproic acid inhibits fibrinolysis and therefore stabilizes the clot
  - Desmopressin (DDAVP) – induces a significant but transient rise in factor VII levels

- **Nursing Management**
  - Assist the child in coping with the condition
  - Encouraged to be self-sufficient and to maintain independence by preventing unnecessary trauma that can cause acute bleeding episodes
  - Instruct the patient to avoid OTC medications such as aspirin, NSAIDs, herbs, nutritional supplements and alcohol
  - Nasal packing should be avoided, because bleeding frequently resumes when the packing is removed
  - All injections should be avoided
  - Splints and other orthopedic devices may be useful in patients with joint or muscle hemorrhages
  - Warm baths promote relaxation, improve mobility, episodes
  - Provide genetic testing and counselling to female carriers