University of THi-Qar
College of Nursing

Hematologic System, Oncologic Disorders & Anemias

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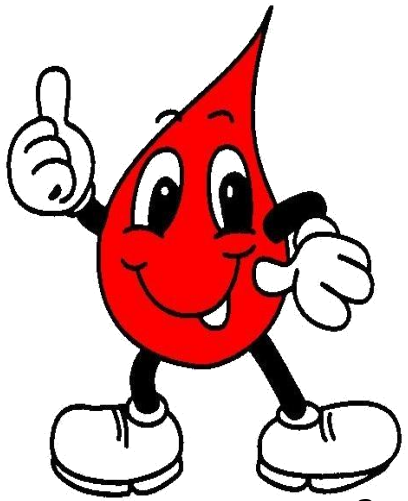
Hematology

- Study of blood and blood forming tissues
- Key components of hematologic system are:
  - Blood
  - Blood forming tissues
    - Bone marrow
    - Spleen
    - Lymph system



What Does Blood Do?

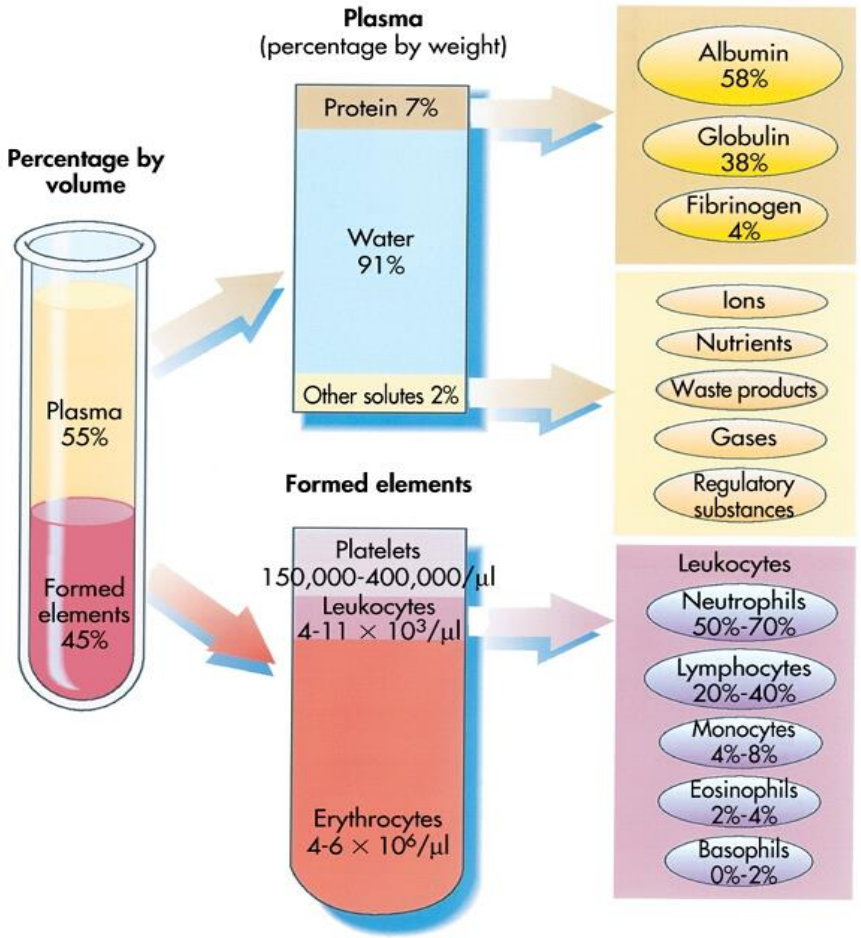
- **Transportation**
  - Oxygen
  - Nutrients
  - Hormones
  - Waste Products
- **Regulation**
  - Fluid, electrolyte
  - Acid-Base balance

What Does Blood Do? (cont'd)

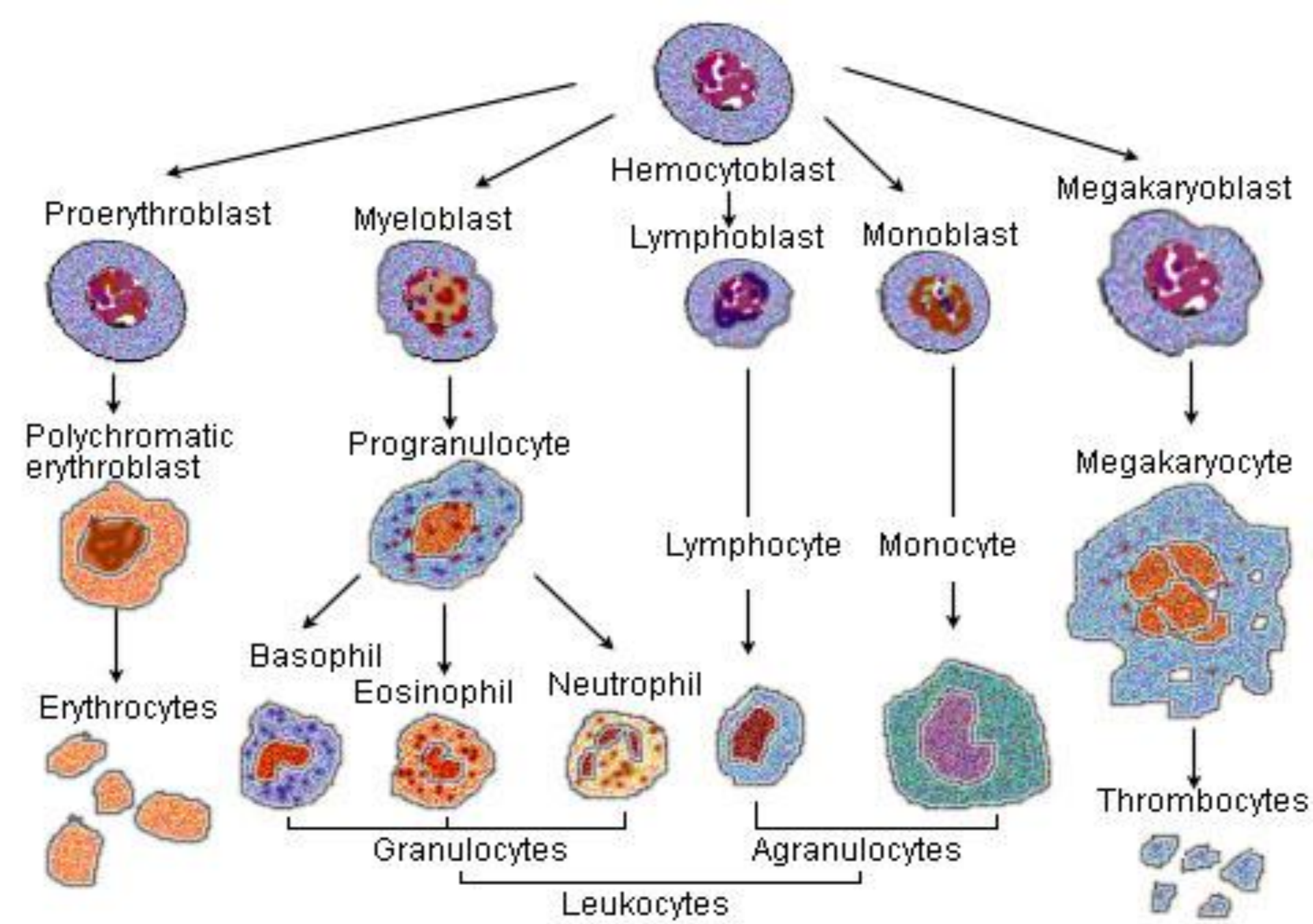
- **Protection**
  - Coagulation
  - Fight Infections

Components of Blood

- **Plasma**
  - 55%
- **Blood Cells**
  - 45%
  - Three types
    - **Erythrocytes/RBCs**
    - **Leukocytes/WBCs**
    - **Thrombocytes/Plat**



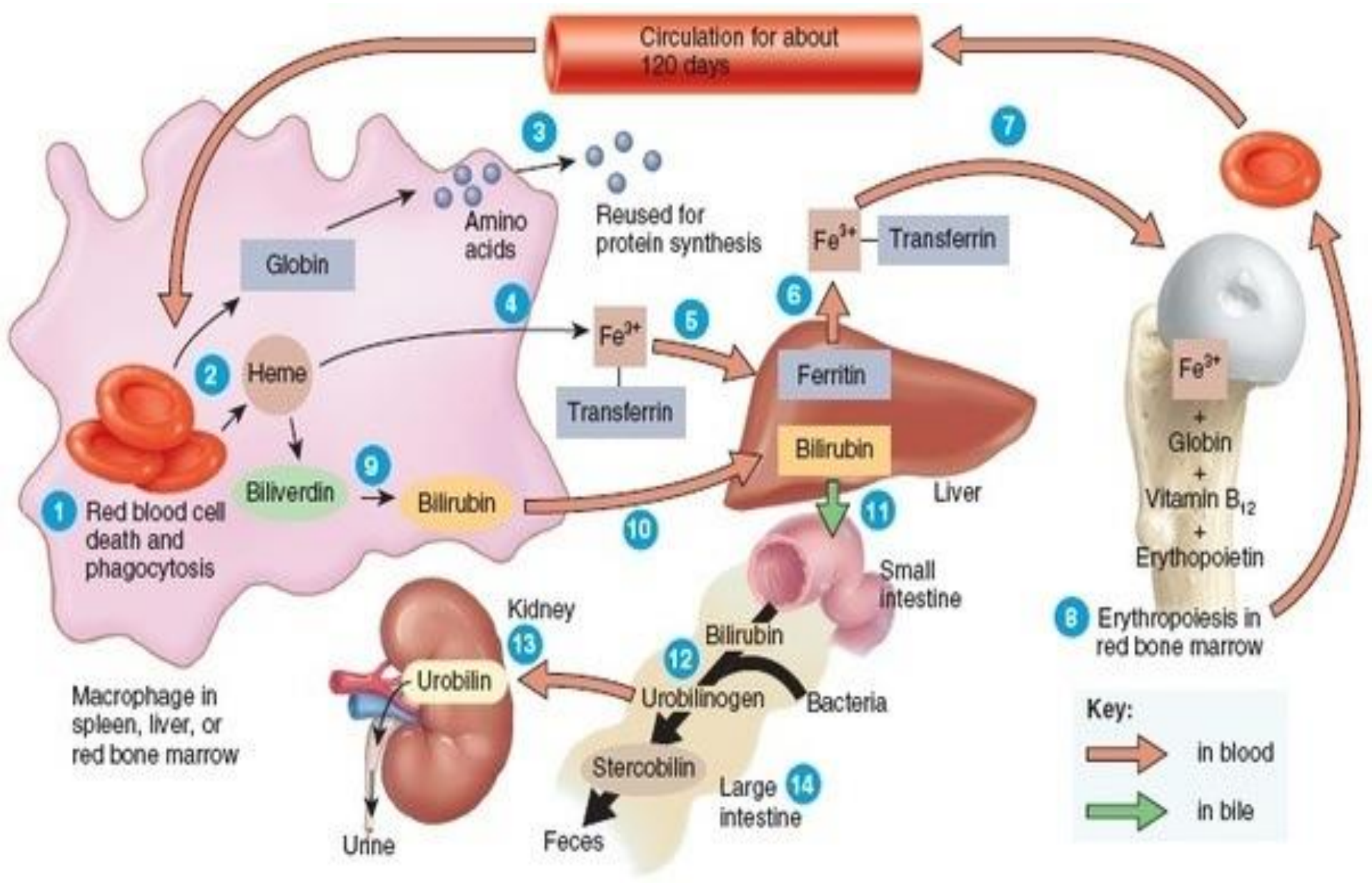
From Thibodeau GA, Patton KT: *The human body in health and disease*, ed 3, St. Louis, 2002, Mosby.


Hemocytoblast
Proerythroblast
Myeloblast
Lymphoblast
Monoblast
Megakaryoblast
Polychromatic erythroblast
Progranulocyte
Megakaryocyte
Lymphocyte
Monocyte
Basophil
Eosinophil
Neutrophil
Erythrocytes
Thrombocytes
Granulocytes
Agranulocytes
Leukocytes

Erythrocytes/Red Blood Cells

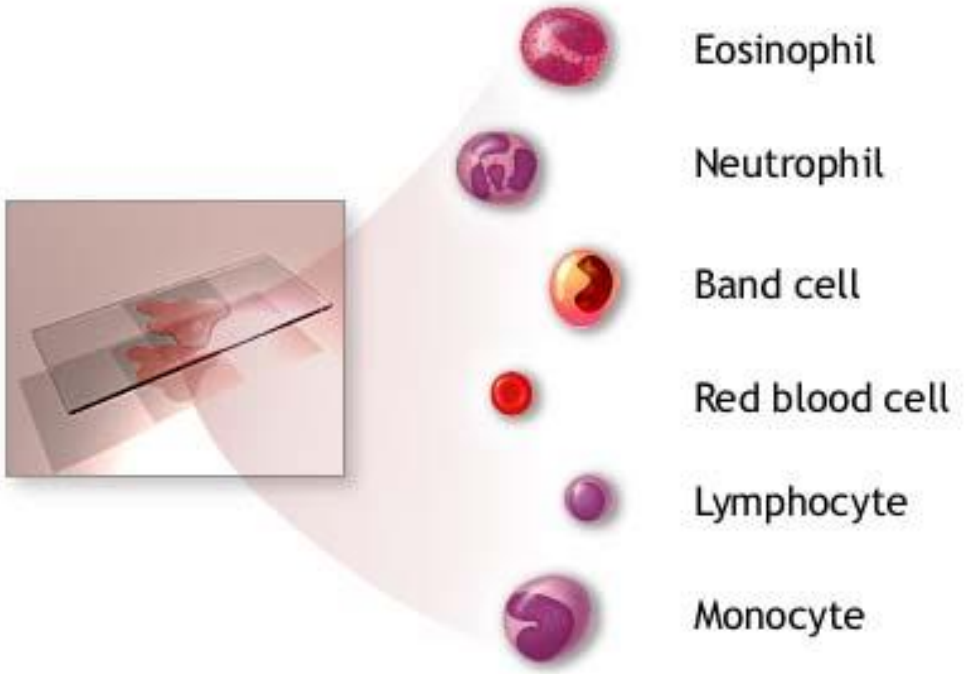
- Composed of hemoglobin
- Erythropoiesis
  - = RBC production
    - **Stimulated by hypoxia**
    - **Controlled by erythropoietin**
      - Hormone synthesized in kidney
- Hemolysis
  - **= destruction of RBCs**
  - **Releases bilirubin into blood stream**
  - **Normal lifespan of RBC = 120 days**

Normal lifespan of RBC = 120 days



Leukocytes/White Blood Cells

- 5 types
  - Basophils
  - Eosinophils
  - Neutrophils
  - Monocytes
  - Lymphocytes



Types and Functions of Leukocytes

TYPE	CELL FUNCTION
<u>Granulocytes</u>	
Neutrophil	Phagocytosis, early phase of inflammation
	Phagocytosis, parasitic infections
Eosinophil	Inflammatory response, allergic response
Basophil	
<u>Agranulocytes</u>	
Lymphocyte	Cellular, humoral immune response
Monocyte	Phagocytosis; cellular immune response

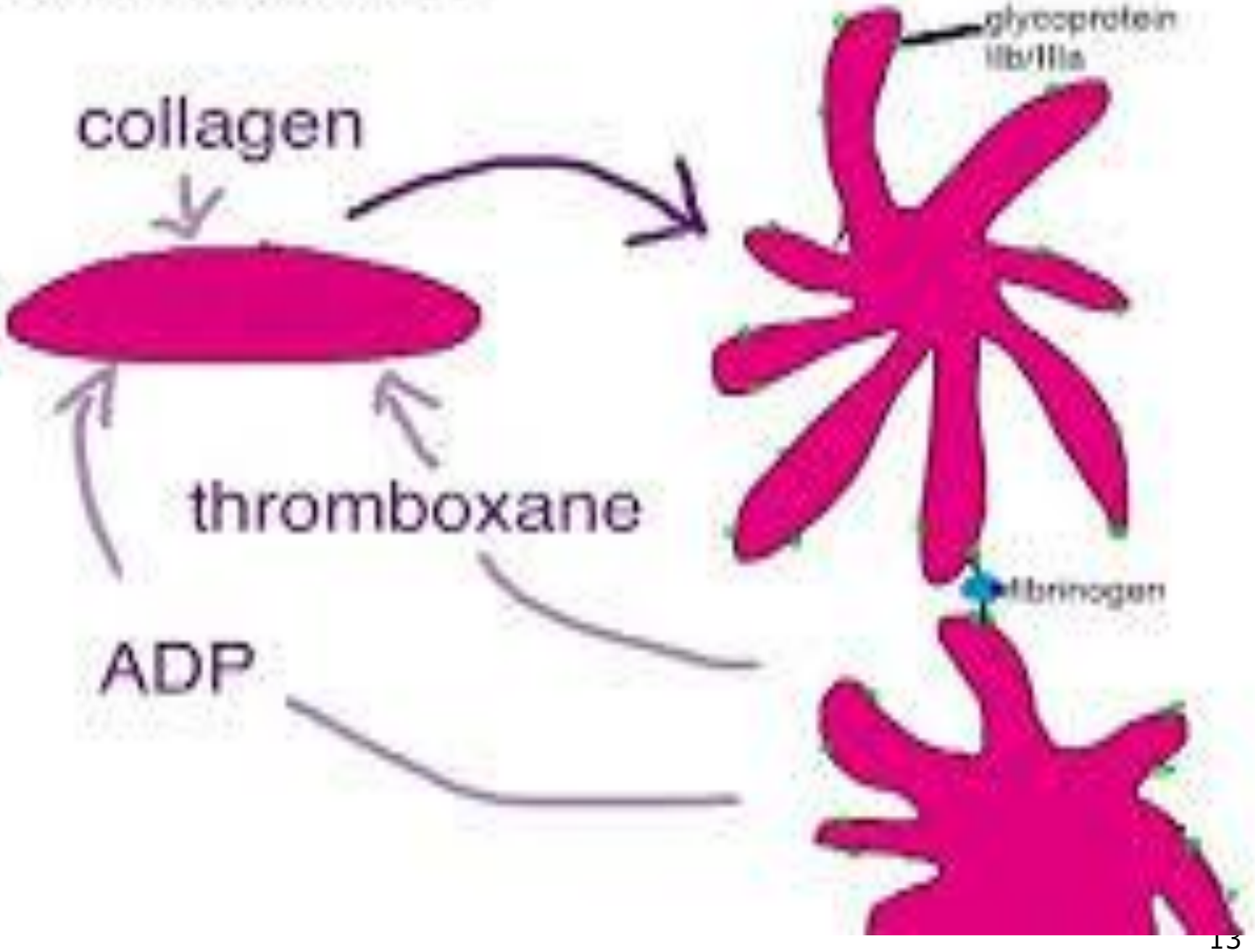
Thrombocytes/Platelets

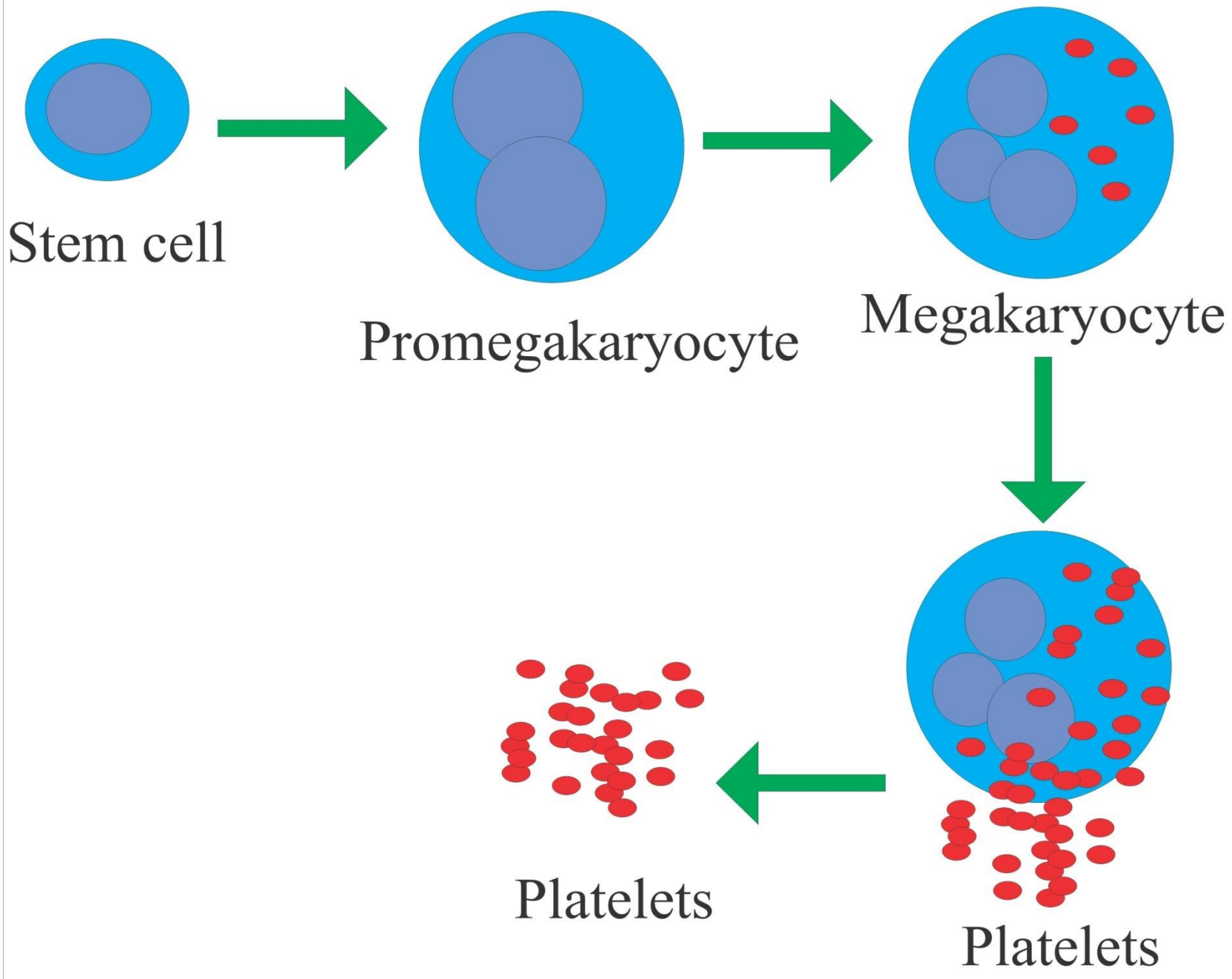
- Must be present for clotting to occur
- Involved in hemostasis

Normal Clotting Mechanisms

- **Hemostasis**
  - Goal: Minimizing blood loss when injured
    1. **Vascular Response**
       - vasoconstriction
    2. **Platelet response**
       - Activated during injury
       - Form clumps (agglutination)
    3. **Plasma Clotting Factors**
       - **Factors I – XIII**
       - **Intrinsic pathway**
       - **Extrinsic pathway**

Platelet activation




Stem cell
Promegakaryocyte
Megakaryocyte
Platelets
Platelets

Anticoagulation

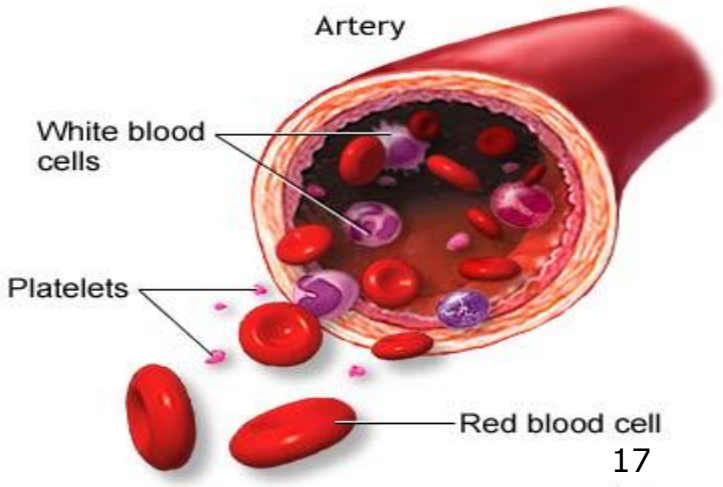
- Elements that interfere with blood clotting
- Counter mechanism to blood clotting—keeps blood liquid and able to flow.

Structures of the Hematologic System

- Bone Marrow
- Liver
- Lymph System

Bone Marrow

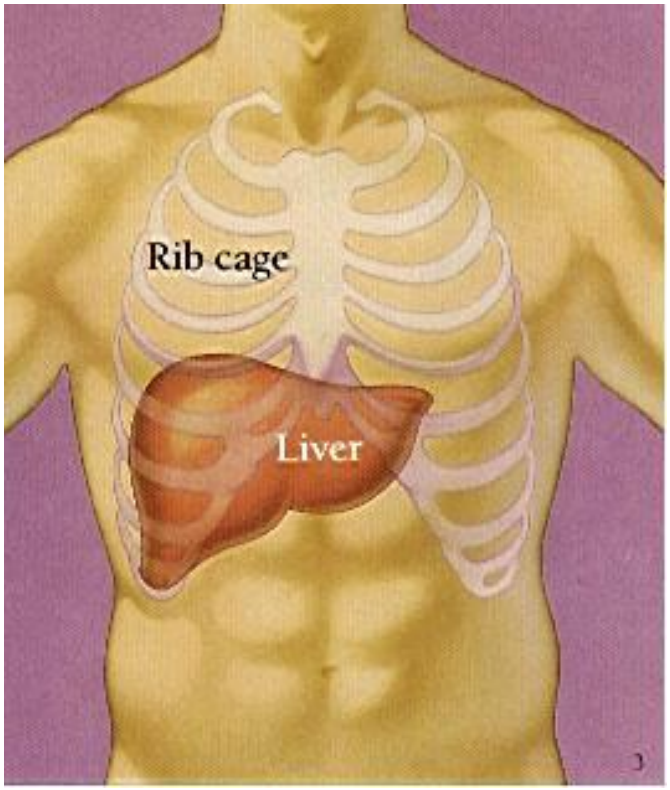
- Bone Marrow
  - Soft substance in core of bones
  - Blood cell production (Hematopoiesis):The production of all types of blood cells generated by a remarkable self-regulated system that is responsive to the demands put upon it.
    - RBCs
    - WBCs
    - Platelets



Liver

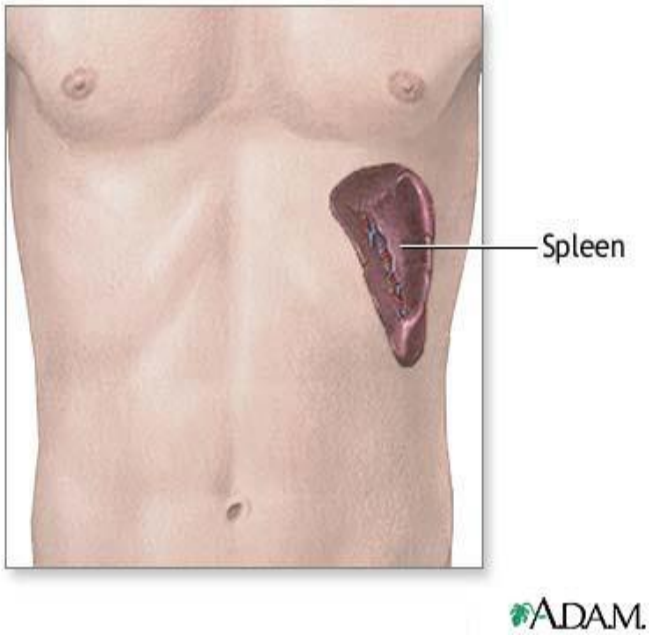
Receives 24% of the cardiac output (1500 ml of blood each minute)

- Liver has many functions
- Hematologic functions:
  - Liver synthesis plasma proteins including **clotting factors** and **albumin**
  - Liver clears damaged and non-functioning RBCs/erythrocytes from circulation



Spleen

- Located in upper L quadrant of abdomen
- Functions
  - Hematopoietic function
    - Produces fetal RBCs
  - Filter function
    - Filter and reuse certain cells
  - Immune function
    - Lymphocytes, monocytes
  - Storage function
    - 30% platelets stored in spleen



Effects of Aging on the Hematologic System

- **CBC Studies**
  - ↓ Hemoglobin (Hb or Hgb)
  - ↓ response to infection (WBC)
  - Platelets=no change
- **Clotting Studies**
  - ↓ PTT

Assessment of the Hematologic System

- Subjective Data
  - Important Health Information
    - Past health history
    - Medications
    - Surgery or other treatments

Assessment of the Hematologic System(cont.)

- Functional Health Patterns
  - Health perception – health management
  - Nutritional – metabolic
  - Elimination
  - Activity – exercise
  - Sleep – rest
  - Cognitive – perceptual
  - Self-perception – self-concept

Assessment of the Hematologic System(cont.)

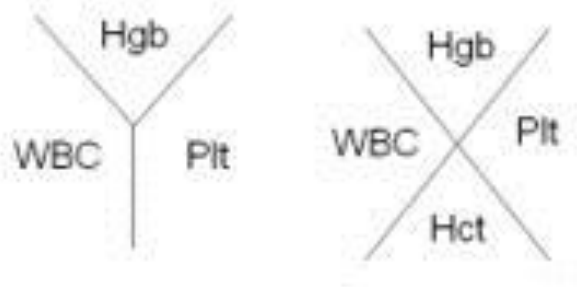
- Functional Health Patterns(cont.)
  - Role – relationship
  - Sexuality – reproductive
  - Coping – stress tolerance
  - Value – belief

Assessment of the Hematologic System(cont.)

- Objective Data
  - Physical Examination
    - Skin
    - Eyes
    - Mouth
    - Lymph Nodes
    - Heart and Chest
    - Abdomen
    - Nervous System
    - Musculoskeletal System

Diagnostic Studies of the Hematologic System: Complete Blood Count (CBC)

- **WBCs**
  - Normal 4,000 -11,000 μ/ℓ
  - Associated with infection, inflamn death
  - *Leukopenia*-- ↓ WBC
  - *Neutropenia* -- ↓ neutrophil count



- **RBC**
  - ♂ 4.5 – 5.5 x $10^6/\ell$
  - ♀ 4.0 – 5.0 x $10^6/\ell$

- **Hematocrit (Hct)**
  - The hematocrit is the percent of whole blood that is composed of red blood cells. The hematocrit is a measure of both the number of red blood cells and the size of red blood cells.

Diagnostic Studies of the Hematologic System: Complete Blood Count (CBC)

- Platelet count
  - Normal 150,000- 400,000
  - Thrombocytopenia-↓ platelet count
  - Spontaneous hemorrhage likely when count is below 20,000

- *Pancytopenia*
  - Decrease in number of RBCs, WBCs, and platelets

Diagnostic Studies of the Hematologic System: Complete Blood Count (CBC)

- Radiologic Studies
  - CT/MRI of lymph tissues
- Biopsies
  - Bone Marrow examination
  - Lymph node biopsies

TABLE 19-1 Common Laboratory Tests for Hematologic and Lymphatic Disorders

TEST	NORMAL ADULT VALUES	EXPLANATION	NURSING IMPLICATIONS
Complete Blood Count (CBC)			
Red Blood Cell (RBC) count ■ Men ■ Women	 4.6–6.0 million/μL (mm^3) 4.0–5.0 million/μL(mm^3)	Number of circulating RBCs in one microliter (cubic millimeter, or mm^3) of blood Reduced in hemorrhage, anemia, and chronic kidney disease Increased (polycythemia) in high altitude, cardiopulmonary disease	No fasting or special client preparation is necessary. Explain the test and the reason it is being done. Results may be affected by deficient or excess fluid volume.
Reticulocyte count	0.5%–1.5% of total RBC	Percentage of immature RBCs Used to help diagnose anemias and their underlying cause	No fasting or special client preparation is necessary. Explain the test and the reason it is being done.
Hemoglobin (Hgb) ■ Men ■ Women	 13.5–18 g/dL 12–15 g/dL	Amount of hemoglobin in 100 mL (1 dL) of blood Used to help diagnose anemias	No fasting or special client preparation is necessary. Do not draw a sample from an arm in which an IV is infusing. Explain why the test is being done. Results may be affected by deficient or excess fluid volume. No special preparation is required. Explain that these tests are used to help identify the underlying cause or type of anemias.
Hematocrit (Hct) ■ Men ■ Women	 40%–54% 36%–46%	Packed volume of RBCs in 100 mL of blood; reported as a percentage Used to help diagnose acute blood loss, anemias, and to monitor chronic diseases	
Mean corpuscular volume (MCV)	80–98 cuμ (fL)	Average volume of individual RBCs	
Mean corpuscular hemoglobin (MCH)	27–31 pg	Weight of the hemoglobin in an average RBC	
Mean corpuscular hemoglobin concentration (MCHC)	32%–36%	Average concentration (percent) of hemoglobin within RBC	

TABLE 19-1 Common Laboratory Tests for Hematologic and Lymphatic Disorders (continued)

TEST	NORMAL ADULT VALUES	EXPLANATION	NURSING IMPLICATIONS
WBC count	4,500–10,000/μL (mm^3)	Measures the number of WBCs in circulating blood	No food or fluid restriction is required. Inquire about manifestations of acute infection or known chronic conditions that may affect WBC count. Decreased WBCs are seen in disorders affecting blood cell production and some infections. Increased WBCs are present in acute infection, leukemias, stress responses, and some acute and chronic diseases.
Differential WBC count		Provides more specific information about infections and disease processes	
Neutrophils	50%–70% (2,500–7,000/μL)	Rapid responders to infection and tissue damage Increase in acute infection and inflammation	
Eosinophils	1%–3% (100-300/μL)	Increase during allergic and parasitic conditions	
Basophils	0.4%–1.0% (40–100/μL)	Increase during healing; decrease in stress and allergic reactions	
Lymphocytes	25%–35% (1,700–3,500/μL)	Play a major role in immune response with B lymphocytes and T lymphocytes	
Monocytes	4%–6% (200–600/μL)	Second line of defense against bacterial infection and foreign substances	
Platelets	150,000–400,000/μL (mm^3)	The number of circulating platelets in the blood Low platelet count associated with bleeding; increased count may increase risk for abnormal clotting	No client preparation is required. Observe for manifestations of bleeding. Monitor count in clients undergoing chemotherapy.
Bleeding time	3–7 minutes	Used to screen for disorders caused by platelet dysfunction	Bleeding time is prolonged by ingestion of aspirin and anti-inflammatory drugs.

Common Laboratory Tests for Hematologic and Lymphatic Disorders

Coagulation Studies			
Prothrombin time (PT or protime)	10–13 seconds (varies by laboratory)	Evaluates the extrinsic clotting pathway; prolonged in warfarin (Coumadin) therapy	No food or fluid restrictions are necessary.
INR (International Normalized Ratio)	2–3.0	Used to evaluate Coumadin therapy (see Chapter 18 for therapeutic values)	The INR provides a more standardized measure of Coumadin therapy.
Partial thromboplastin time (PTT) Activated partial thromboplastin time (APTT, PTT)	60–70 20–35 seconds	Used to evaluate clotting pathways and monitor heparin therapy More sensitive than PTT; evaluates the intrinsic clotting pathway; prolonged in heparin therapy	No food or fluid restriction is required. Values are increased in clotting factor deficiencies, heparin therapy, and aspirin ingestion.
Coombs' test	Negative	Performed to diagnose hemolytic anemias and evaluate transfusion reactions. The expected results are no detected antibodies to RBCs (indirect Coombs') or no detected RBC antigen–antibody complexes (direct Coombs').	No food or fluid restriction is required. Ask about previous transfusions or transfusion reactions. Report manifestations of transfusion reactions.
Hemoglobin electrophoresis	■ Hb A_1 95%–98% ■ Hb A_2 1.5%–4% ■ Hb F less than 2% ■ Hb C 0% ■ Hb D 0% ■ Hb S 0%	Performed to detect abnormal forms of hemoglobin associated with genetic hemolytic anemias (e.g., sickle cell anemia, thalassemia)	No food or fluid restrictions are required. Assess for and report manifestations of hemolytic anemias. Encourage the client to obtain genetic counseling.

TABLE 19-1 Common Laboratory Tests for Hematologic and Lymphatic Disorders (continued)

TEST	NORMAL ADULT VALUES	EXPLANATION	NURSING IMPLICATIONS
		Serum Iron Studies	
Iron	50–150 mcg/dL (10–27 mol/L)	Serum iron and body iron stores are measured to evaluate iron deficiency anemia.	Antibiotics, estrogen and testosterone, oral contraceptives, aspirin, and ethanol affect results.
Total iron-binding capacity	250–450 µg/dL	Measures the maximum amount of iron that can bind to transferrin, the protein that transports it	
Ferritin	Men: 15–445 ng/mL (15–445 µg/L) Women: 10–310 ng/mL (10–310 µg/L)	A measure of the amount of iron stored in body tissues	No food or fluid restrictions are required. Results in women are affected by age and use of oral contraceptives.
Transferrin	200–430 mg/dL (2.0–4.3 g/L)	Measures the protein that transports iron to the bone marrow for use in synthesizing hemoglobin	Avoid iron supplements for 12 hours before testing. Results are affected by pregnancy and use of oral contraceptives.
D-dimer	Negative	D-dimer is a fragment produced when fibrinolysis occurs. It is used primarily to diagnose disseminated intravascular coagulation.	No food or fluid restriction is required. Report manifestations such as unexplained bleeding. Monitor vital signs.

Common Laboratory Tests for Hematologic and Lymphatic Disorders

Schilling test	10%–40% of vitamin B_{12} excretion in 24 hr	Primarily used to diagnose pernicious anemia. This timed test evaluates the body's ability to absorb vitamin B_{12} from the GI tract. An oral dose of radioactively tagged vitamin B_{12} and an intramuscular vitamin B_{12} injection are administered, followed by collection of a 24-hour urine specimen.	Verify that client has given informed consent. Instruct the client to: ■ Withhold food and fluids for 8–12 hours before the test. ■ Avoid taking vitamin B supplements for 3 days before the test. May eat and drink after vitamin B_{12} injection is given. Observe for manifestations of anaphylaxis for at least 1 hour after administration of radioactive vitamin B_{12}. Collect a 24-hour urine sample (see Box 28-3), using rubber gloves to handle urine.

Anemia

- Anemia is a reduction in the number of RBCs, the quantity of hemoglobin, or the volume of RBCs
- Because the main function of RBCs is oxygenation, anemia results in varying degrees of hypoxia

Anemia

- **Prevalent conditions**
  - Blood loss
  - Decreased production of erythrocytes
  - Increased destruction of erythrocytes

Anemia(cont'd)

Symptoms of Anemia

- **Decreased oxygenation**
  - Exertional dyspnea
  - Dyspnea at rest
  - Fatigue
  - Bounding pulses
  - Lethargy, confusion

Anemia(cont'd)

Symptoms of Anemia(cont'd)

- **Decreased volume**
  - Fatigue
  - Muscle cramps
  - Postural dizziness
  - syncope

Anemia(cont'd)

Nursing Management:

- Direct general management toward addressing the cause of anemia and replacing blood loss as needed to sustain adequate oxygenation.
- Promote optimal activity and protect from injury.
- Reduce activities and stimuli that cause tachycardia and increase cardiac output.
- Provide nutritional needs.
- Administer any prescribed nutritional supplements.
- Patient and family education.

Nursing Actions for a Patient who is Anemic or Suffered Blood Loss

- Administer oxygen as prescribed.
- Administer blood products as prescribed.
- Administer erythropoietin as prescribed.
- Allow for rest between periods of activity.
- Elevate the pt's head on pillows during episodes of shortness of breath.
- Provide extra blankets if the pt feels cool.
- Teach the pt/family about underlying. pathophysiology and how to manage the symptoms of anemia.

Anemia caused by Decreased Erythrocyte Production

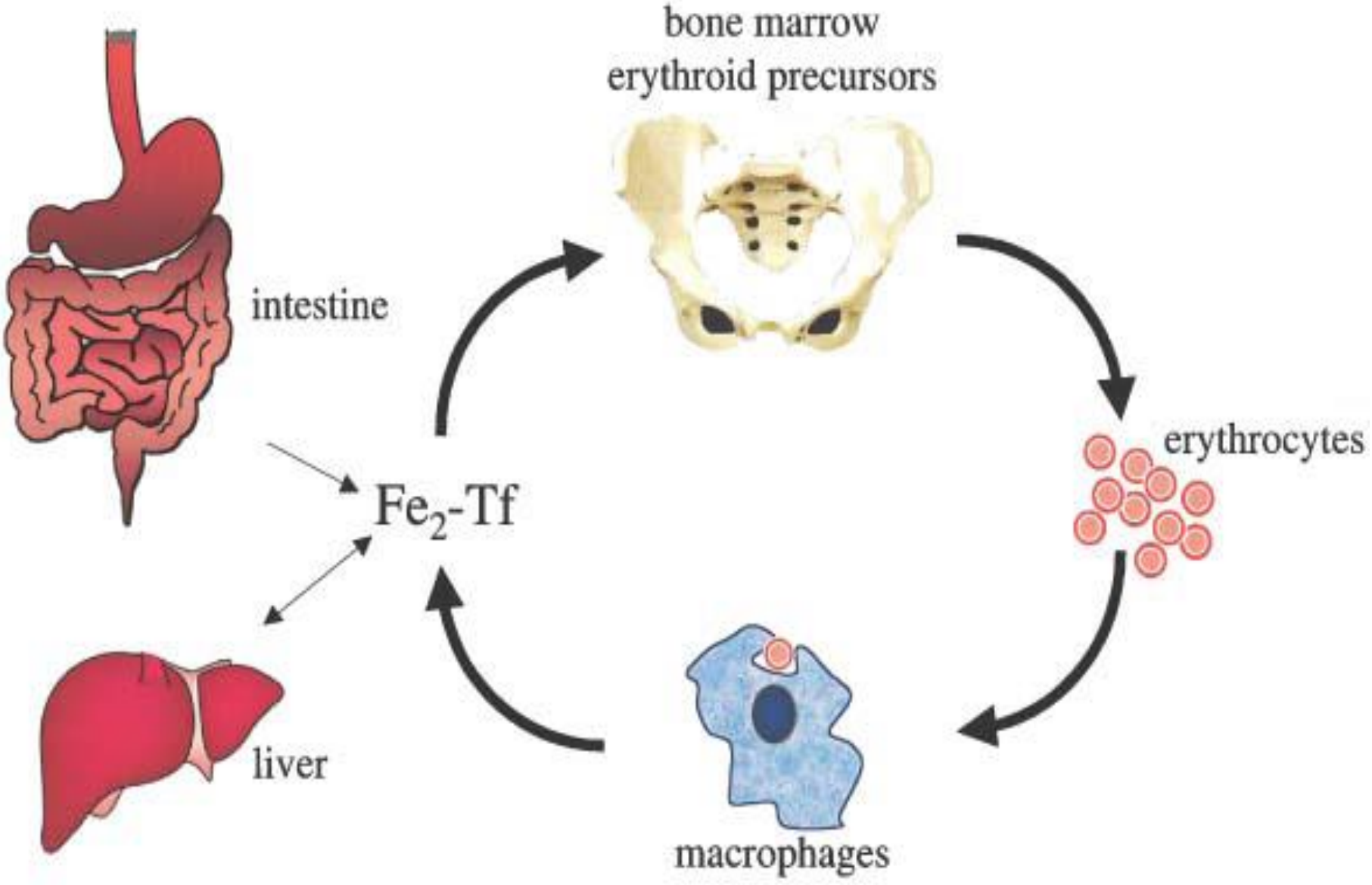
- Iron Deficiency Anemia
- Thalassemia
- Megablastic Anemia

Iron-Deficiency Anemia

Etiology

1. **Inadequate dietary intake**
   - Found in 30% of the world's population
2. **Malabsorption**
   - Absorbed in duodenum
   - GI surgery
3. **Blood loss**
   - 2 mls blood contain 1mg iron
   - GI, GU losses
4. **Hemolysis**

IRON METABOLISM



Iron-Deficiency Anemia

Clinical Manifestations

- Most common: pallor
- Second most common: inflammation of the tongue (glossistis)
- Cheilitis=inflammation/fissures of lips
- Sensitivity to cold
- Weakness and fatigue
- Nail changes
  - Brittle/fragility
  - Koilonchia/spooning

Iron-Deficiency Anemia

Clinical Manifestations(cont'd)

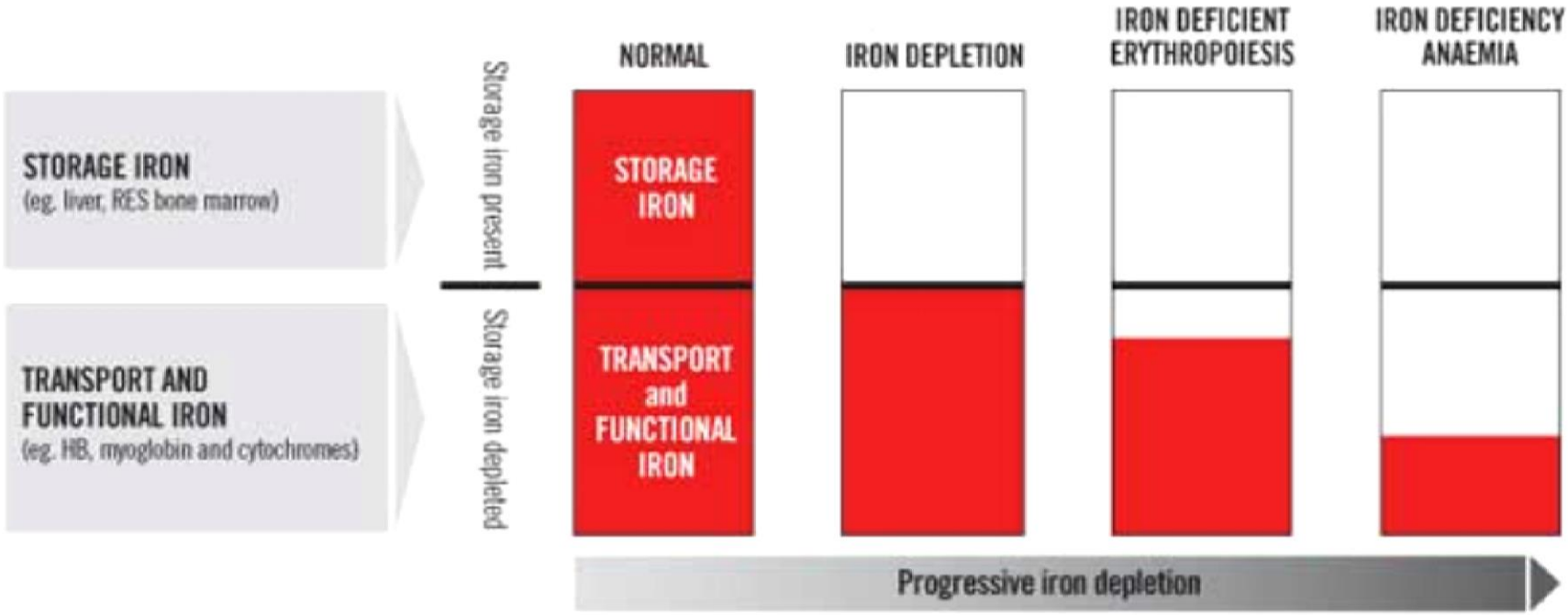
- Angular stomatitis
- Pica (appetite for bizarre substances e.g. ice, paint, clay)
- Brittle hair
- Dry skin
- Dysphagia (esophageal web, Plummer-Vinson ring

Iron-Deficiency Anemia

Diagnostic Studies

- CBC
- Iron studies Diagnostics:
- Iron levels: Total iron-binding capacity (TIBC), Serum Ferritin.
- Endoscopy/Colonscopy

SPECTRUM OF IRON DEFICIENCY

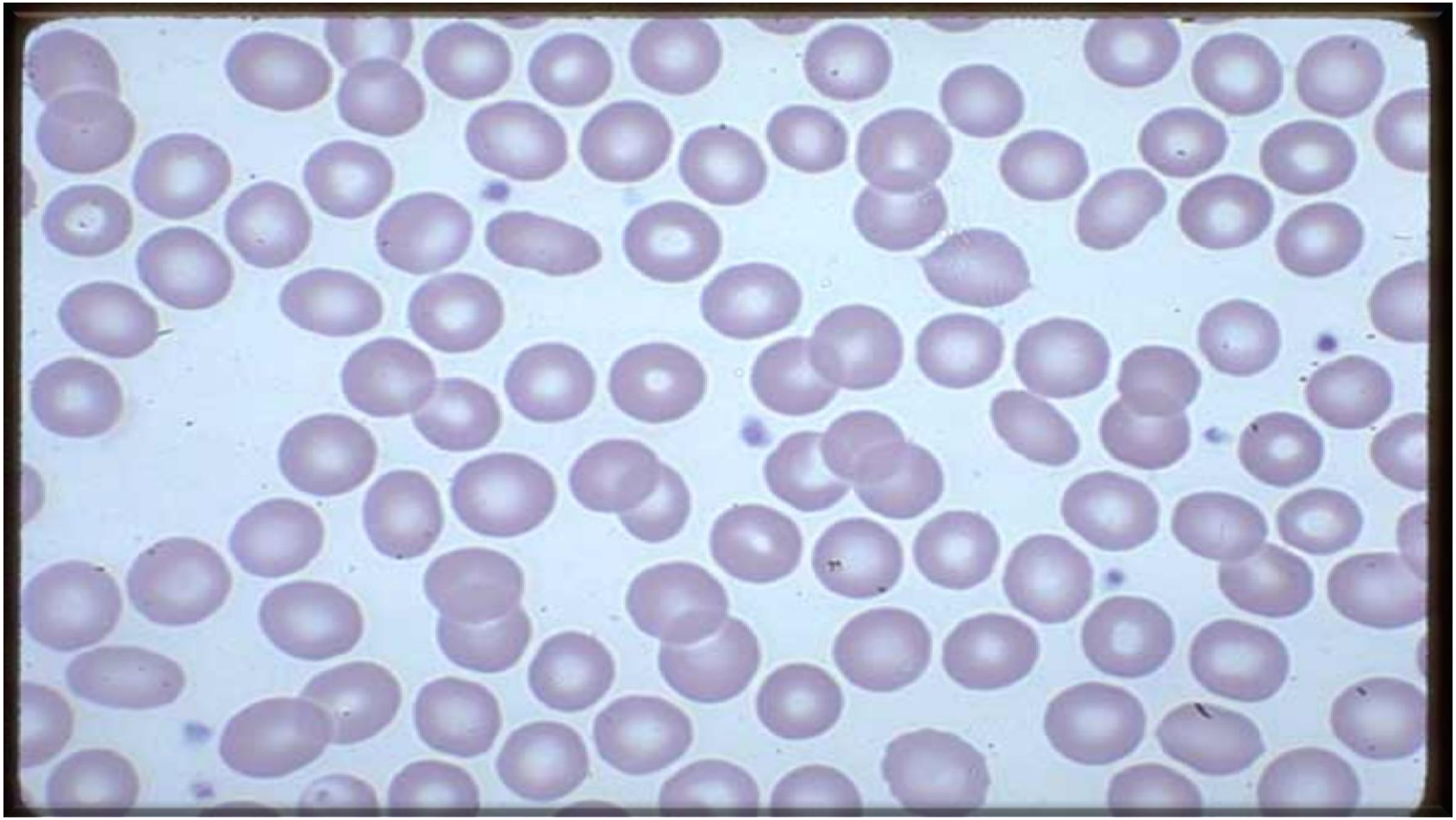


Example of laboratory profile

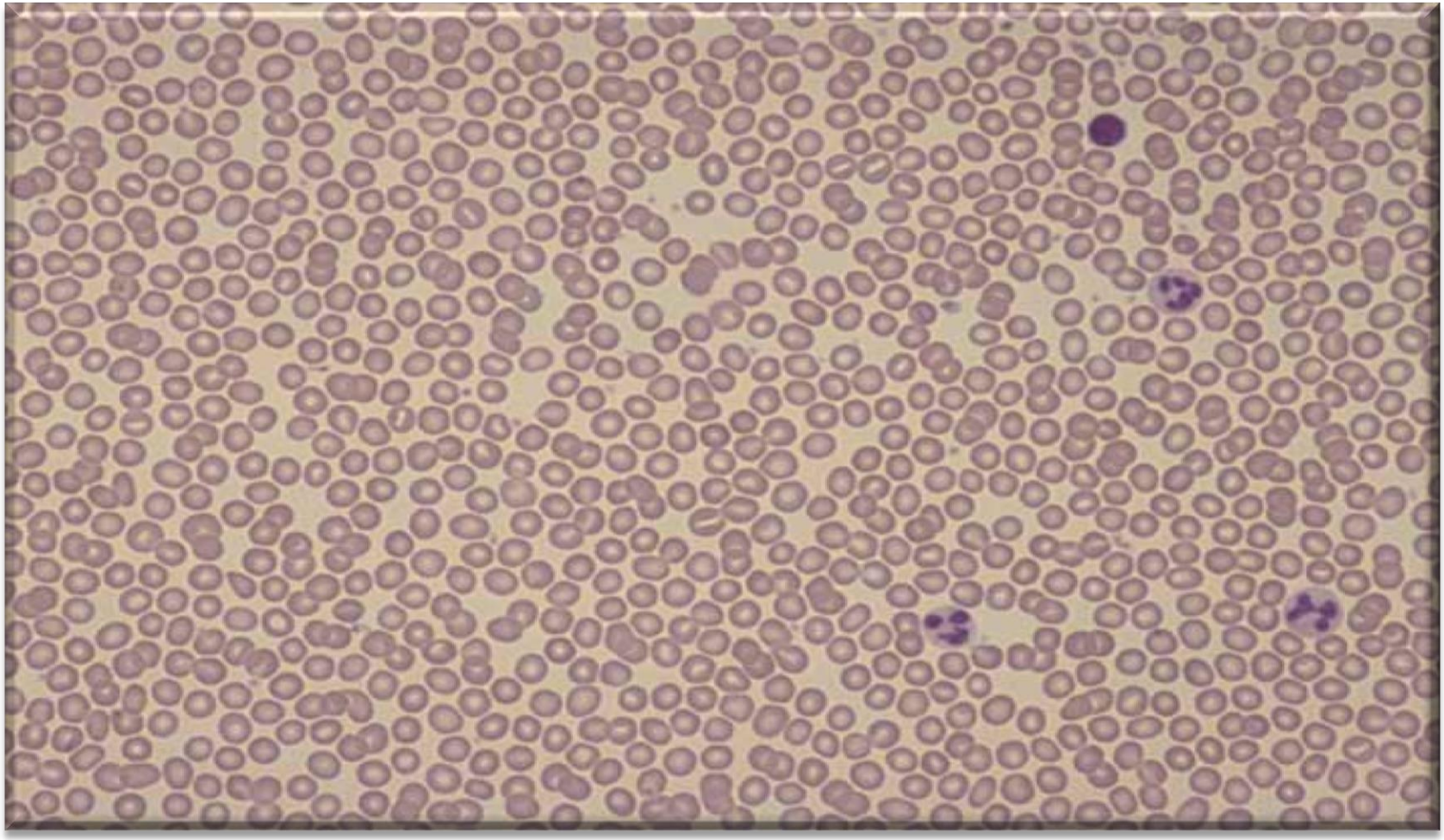
Serum ferritin (µg/L)	60	<15	<15	<15
Transferrin saturation (%)	35	35	<15	<15
Haemoglobin (g/L) – female	>120	>120	>120	<120
Haemoglobin (g/L) – male	>130	>130	>130	<130

Image Source: http://www.transfusion.com.au

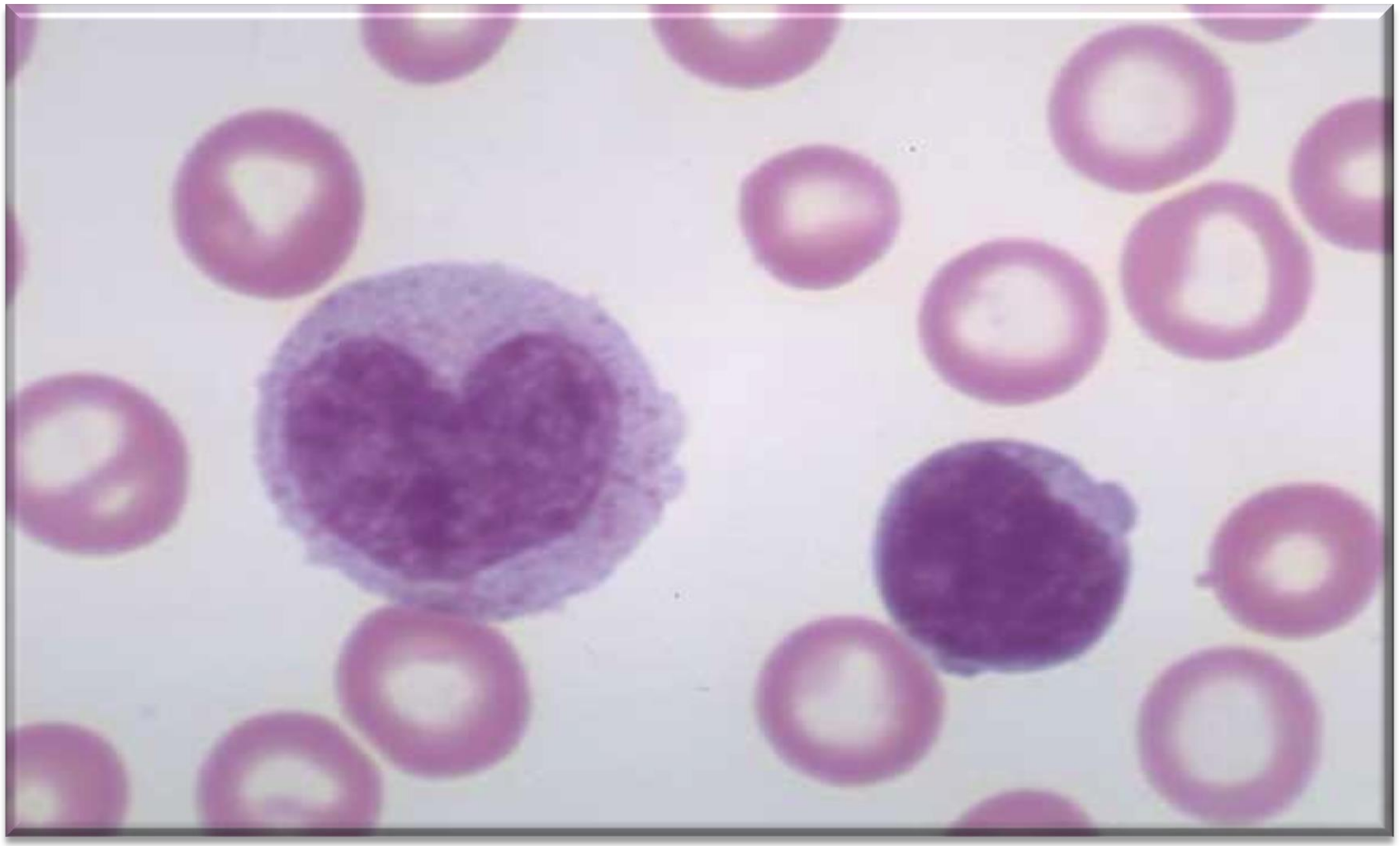
Normal Blood Film



MICROCYTES



HYPOCHROMIA



Findings	Normal	Prelatent period	Latent period	Iron def. anemia Early	Late
Hb g/dl	N	N	N	8-14	<8
MCV fl	N	N	N	N, ↓	↓
S. Ferr.	N	↓	<12	<12	<12
T. Sat.	N	N	<16	<16	<16
BM iron	N	↓	-	-	-
FEP	N	N	↑	↑↑	↑↑
Symptom	-	-	-	+	+
Ept. change	-	-	-	-	+

Iron-Deficiency Anemia

Collaborative Care

- Treatment of underlying disease/problem
- Replacing iron
- Diet
- Drug Therapy
  - Iron replacement
    - Oral iron
      - Feosol, DexFerrum, etc
        - Absorbed best in acidic environemtn
        - GI effects

Iron replacement

Iron-Deficiency Anemia

- **Parenteral iron**
  - IM or IV
  - Less desirable than PO

Iron-Deficiency Anemia

Nursing Management

- Assess cardiovascular & respiratory status
- Monitor vital signs
- Recognizing s/s bleeding
- Monitor stool, urine and emesis for occult blood
- Diet teaching—foods rich in iron
- Provide periods of rest

Iron-Deficiency Anemia

Nursing Management(cont'd)

- Supplemental iron
- Discuss diagnostic studies
- Emphasize compliance
- Iron therapy for 2-3 months after the hemoglobin levels return to normal

The Thalassemias

Introduction

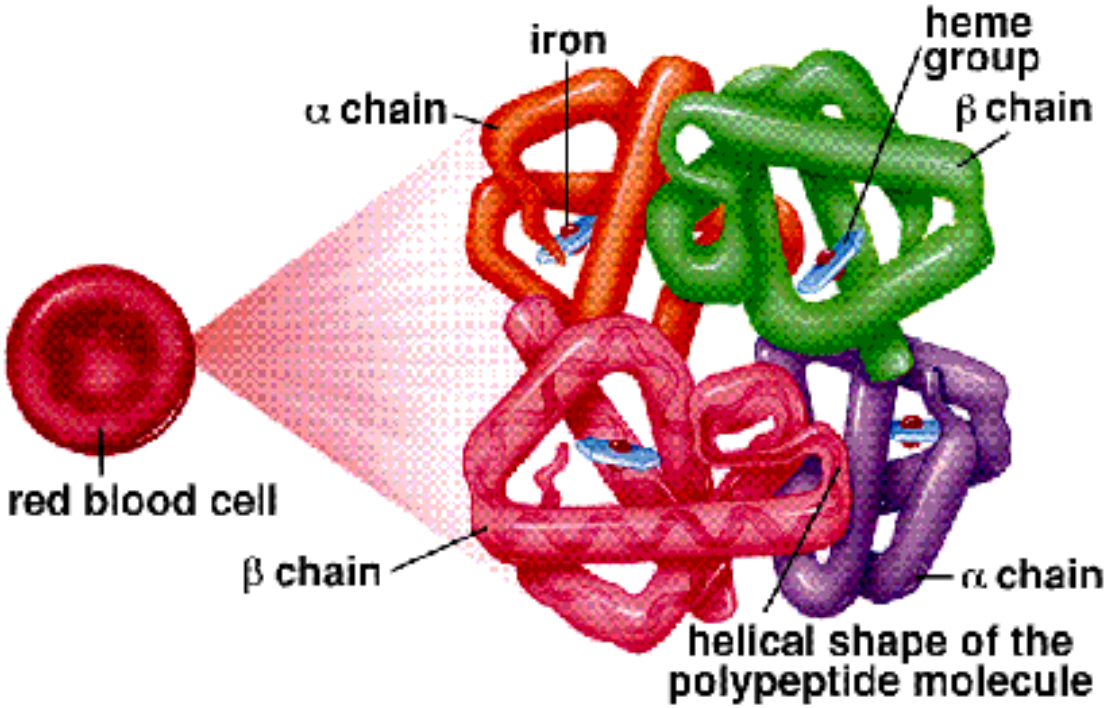
- Heritable, hypochromic anemias-varying degrees of severity.
- Genetic defects result in decreased or absent production of mRNA and globin chain synthesis.
- At least 100 distinct mutations.
- High incidence in Asia, Africa, Mideast, and Mediterrenean countries.

Hemoglobin Review

- **Each complex consists of :**
  - Four polypeptide chains, non-covalently bound
  - Four heme complexes with iron bound
  - Four O2 binding sites

Hemoglobin Structure

- Four subunits
  - two α
  - two β
- Iron
- Heme
- Binds 4 O_2



Globin Chains

- **Alpha Globin**
  - 141 amino acids
  - Coded for on Chromosome 16
  - Found in normal adult hemoglobin, A1 and A2
- **Beta Globin**
  - 146 amino acids
  - Coded for on Chromosome 11, found in Hgb A1
- **Delta Globin**
  - Found in Hemoglobin A2--small amounts in all adults
- **Gamma Globin**
  - Found in Fetal Hemoglobin
- **Zeta Globin**
  - Found in embryonic hemoglobin

Hemoglobin Types

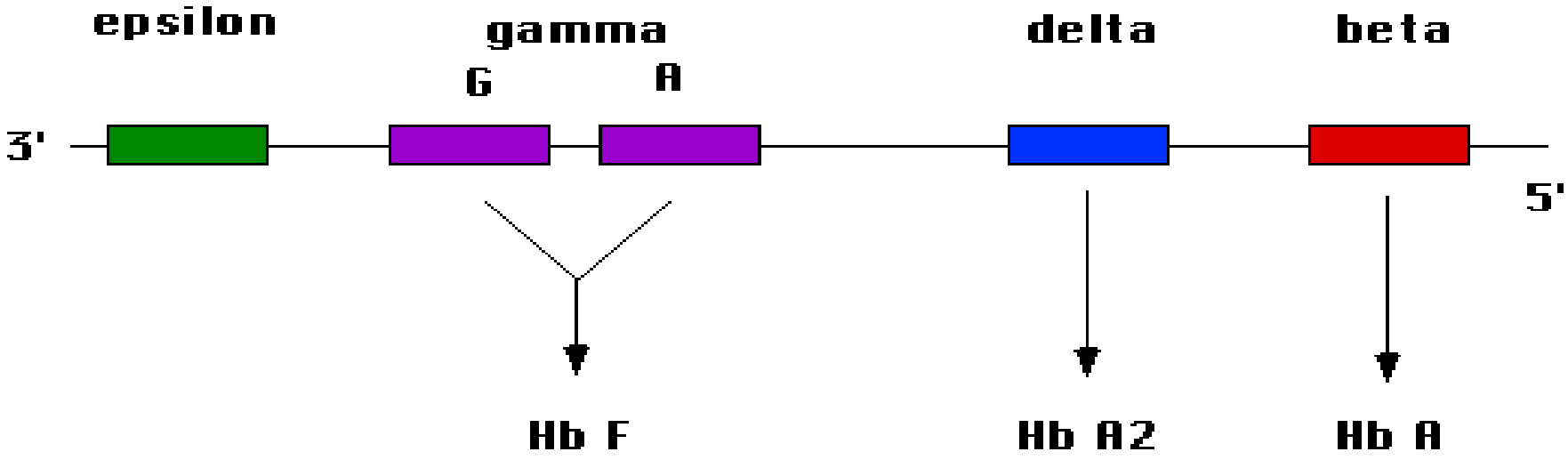
Hemoglobin Type Globin Chains

- Hgb A1—92%-------- $\alpha_2\beta_2$
- Hgb A2—2.5%------- $\alpha_2\delta_2$
- Hgb F — <1%------- $\alpha_2\gamma_2$
- Hgb H ---------- β_4
- Bart's Hgb---------- γ_4
- Hgb S---------- $\alpha_2\beta_{26}^{glu\rightarrow val}$
- Hgb C---------- $\alpha_2\beta_{26}^{glu\rightarrow lys}$

Genetics

- Alpha globins are coded on chromosome 16
  - Two genes on each chromosome
  - Four genes in each diploid cell
  - Gene deletions result in Alpha-Thalassemias
- Also on chromosome 16 are Zeta globin genes—Gower's hemoglobin (embryonic)
- Beta globins are coded on chromosome 11
  - One gene on each chromosome
  - Two genes in each diploid cell
  - Point mutations result in Beta-Thalassemias
- Also on chromosome 11 are Delta (Hgb A2) and Gamma (Hgb F) and Epsilon (Embryonic)

Beta Globin Gene Cluster
Chromosome 11



Alpha Globin Gene Cluster
Chromosome 16

Alpha Thalassemias

- Result from gene deletions
- One deletion—Silent carrier; no clinical significance
- Two deletions—α Thal trait; mild hypochromic microcytic anemia
- Three deletions—Hgb H; variable severity, but less severe than Beta Thal Major
- Four deletions—Bart's Hgb; Hydrops Fetalis; In Utero or early neonatal death

Alpha Thalassemias

- Usually no treatment indicated
- 4 deletions incompatible with life
- 3 or fewer deletions have only mild anemia

Beta Thalassemias

- Result from Point Mutations on genes
- Severity depends on where the hit(s) lie
  - β^0-no β-globin synthesis;
  - β^+ reduced synthesis
- Disease results in an overproduction of a-globin chains, which precipitate in the cells and cause splenic sequestration of RBCs
- Erythropoiesis increases, sometimes becomes extramedullary

β-Thal--Clinical

- **β-Thalassemia Minor**
  - **Minor point mutation**
  - Minimal anemia; no treatment indicated
- **β-Thalassemia Intermedia**
  - Homozygous minor point mutation or more severe heterozygote
  - Can be a spectrum; most often do not require chronic transfusions
- **β-Thalassemia Major-Cooley's Anemia**
  - Severe gene mutations
  - Need careful observation and intensive treatment

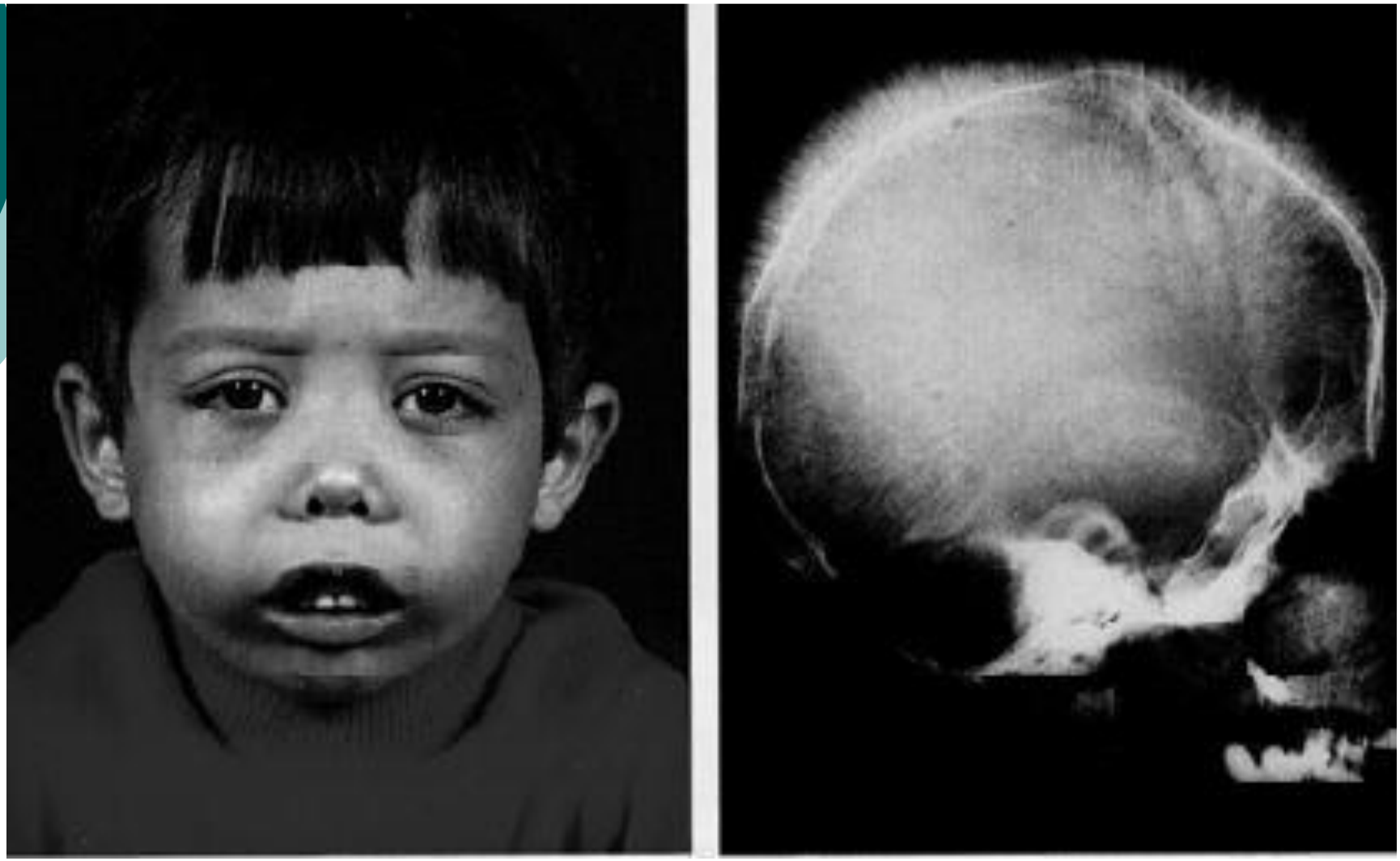
Beta Thalassemia Major

- **Reduced or nonexistent production of β-globin**
  - Poor oxygen-carrying capacity of RBCs
    - Failure to thrive, poor brain development
  - Increased alpha globin production and precipitation
    - RBC precursors are destroyed within the marrow
- **Increased splenic destruction of dysfunctional RBCs**
  - Anemia, jaundice, splenomegaly

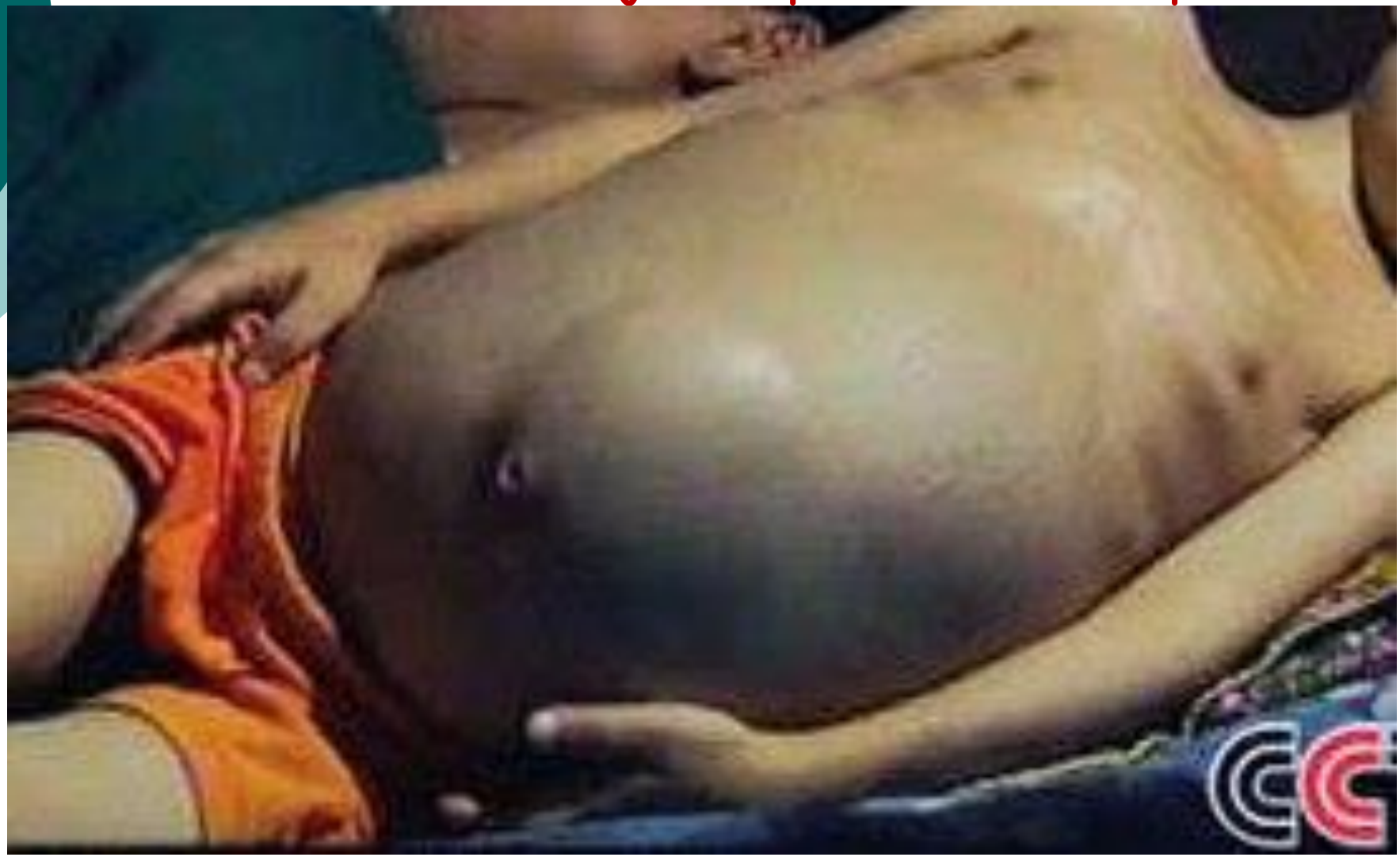
Beta Thalassemia Major

- **Hyperplastic Bone Marrow**
  - Ineffective erythropoiesis—RBC precursors destroyed
    - Poor bone growth, frontal bossing, bone pain
  - Increase in extramedullary erythropoiesis
- **Iron overload—increased absorption and transfusions**
  - Endocrine disorders, Cardiomyopathy, Liver failure

Thalassemia Major; Bone Changes



Thalassemia Major; a protuberant spleen

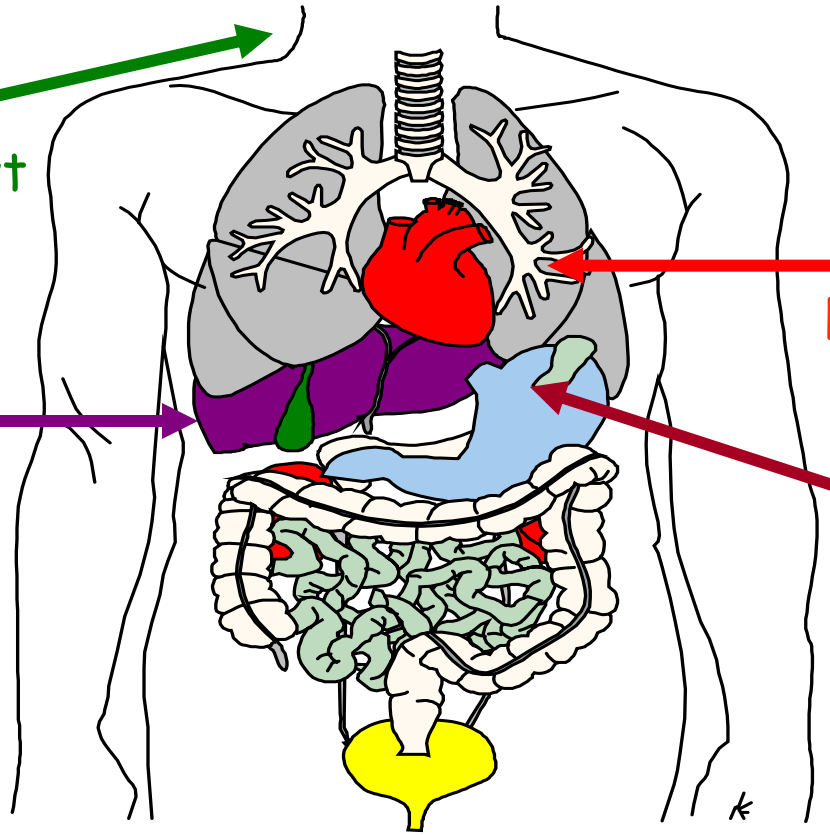


Thalassemia Major; the beneficial effect of transfusions

β-Thalassemia Major—Lab findings

- **Hypochromic, microcytic anemia**
  - Target Cells, nucleated RBCs, anisocytosis
- **Reticulocytosis**
- **Hemoglobin electrophoresis shows**
  - Increased Hgb A2—delta globin production
  - Increased Hgb F—gamma globin production
- **Hyperbilirubinemia**
- **LFT abnormalities (late finding)**
- **TFT abnormalities, hyperglycemia (late endocrine findings)**

Tissue Iron Concentrations in Transfusion-dependent Thalassemia Patients



Thyroid
Fe = 1.6 – 6.8% d wt

Liver
Fe = 3.7-6.8% d wt

Heart
Fe = 0.6 –1.3% d wt

Pancreas
Fe = 1.4-3.9% d wt

Adapted from ***Modell & Berdoukas, 1984***

β-Thalassemia Major--Treatment

- **Chronic Transfusion Therapy**
  - Maximizes growth and development
  - Suppresses the patient's own ineffective erythropoiesis and excessive dietary iron absorption
  - PRBC transfusions often monthly to maintain Hgb 10-12
- **Chelation Therapy**
  - Binds free iron and reduces hemosiderin deposits
  - 8-hour subcutaneous infusion of deferoxamine, 5 nights/week
  - Start after 1year of chronic transfusions or ferritin>1000 ng/dl

β-Thalassemia Major--Treatment

- **Splenectomy--indications**
  - Trasfusion requirements increase 50% in 6mo
  - PRBCs per year >250cc/kg
  - Severe leukopenia or thrombocytopenia

β-Thalassemia Major Complications and Emergencies

- **Sepsis—Encapsulated organisms**
  - Strep Pneumonia
- **Cardiomyopathy—presentation in CHF**
  - Use diuretics, digoxin, and deferoxamine
- **Endocrinopathies—presentation in DKA**
  - Take care during hydration so as not to precipitate CHF from fluid overload

Anticipatory Guidance and Follow Up

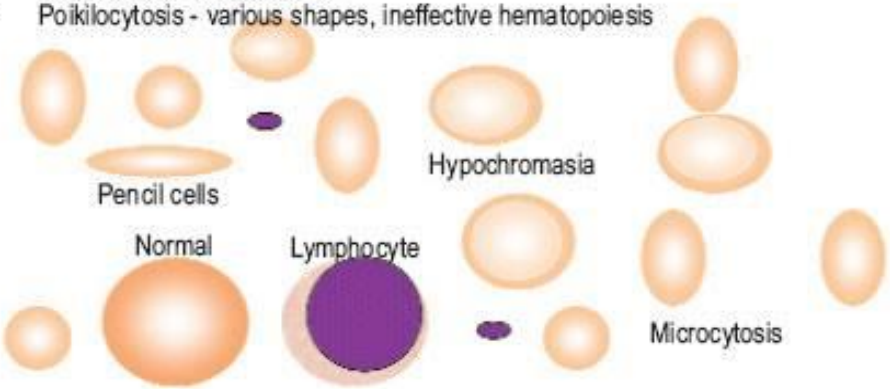
- Immunizations—Hepatitis B, Pneumovax
- Follow for signs of diabetes, hypothyroid, gonadotropin deficiency
- Follow for signs of cardiomyopathy or CHF
- Follow for signs of hepatic dysfunction
- Osteoporosis prevention
  - **Diet, exercise**
  - **Hormone supplementation**
  - **Osteoclast-inhibiting medications**
- Follow ferritin levels

On The Horizon

- **Oral Chelation Agents**
- **Pharmacologically upregulating gamma globin synthesis, increasing Hgb F**
  - Carries O2 better than Hgb A2
  - Will help bind α globins and decrease precipitate
- **Bone Marrow transplant**
- **Gene Therapy**
  - Inserting healthy β genes into stem cells and transplanting

Microcytic Anemia - Iron Deficiency anemia

- Anisocytosis - varying sizes
- Poikilocytosis - various shapes, ineffective hematopoiesis

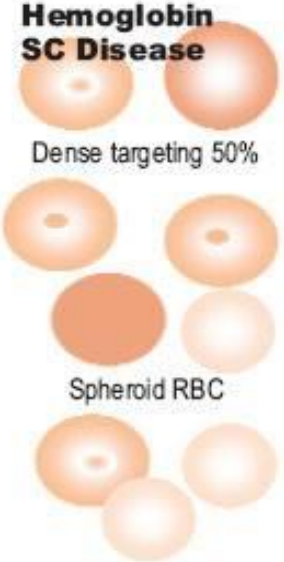


Anemia & Thalassemia: Differential Diagnosis

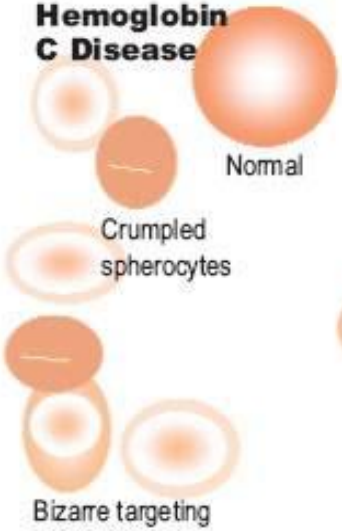
Differential Features	Iron Deficiency Anemia	Thalassemia
Mentzer's index = MCV/RBC*	> 13	< 13
RDW (red cell distribution width)*	High (wide)	Normal
Iron, ferritin, transferrin saturation**	Low	Normal
TIBC (transferring)	High	Normal
Other diagnostic tools	Work up the cause of iron deficiency	α-thal (Asian): molecular test; β-thal: electrophoresis

*High sensitivity, low specificity; index values defined in Mentzer WC, Lancet 1973;1:882.
**Higher specificity, MCV, mean corpuscular volume

Hemoglobin SC Disease



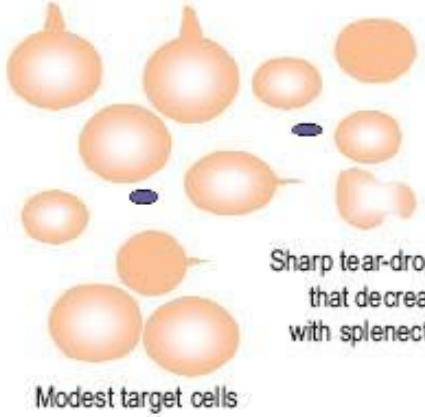
Hemoglobin C Disease

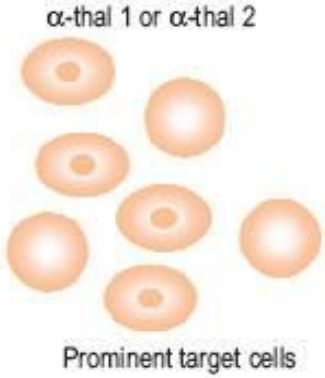


Hemoglobin E Disease

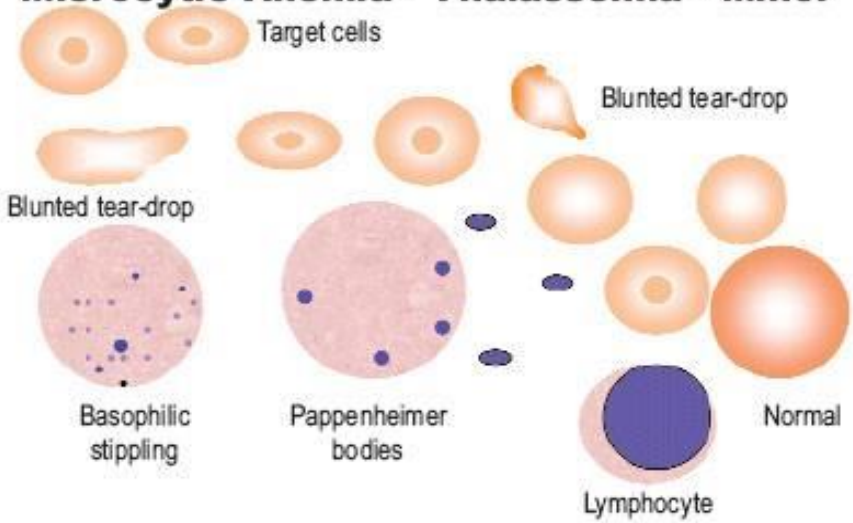
Microcytic Anemia - Thalassemia - Major

- Anisocytosis - varying sizes
- Poikilocytosis - various shapes



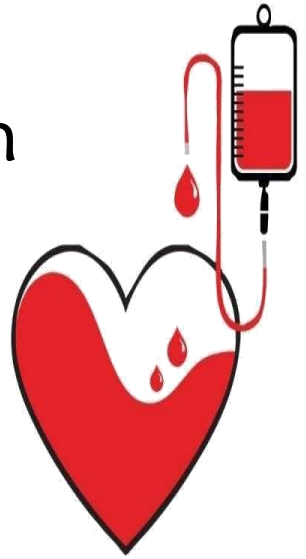


Microcytic Anemia - Thalassemia - Minor



Thalassemia

Collaborative Care

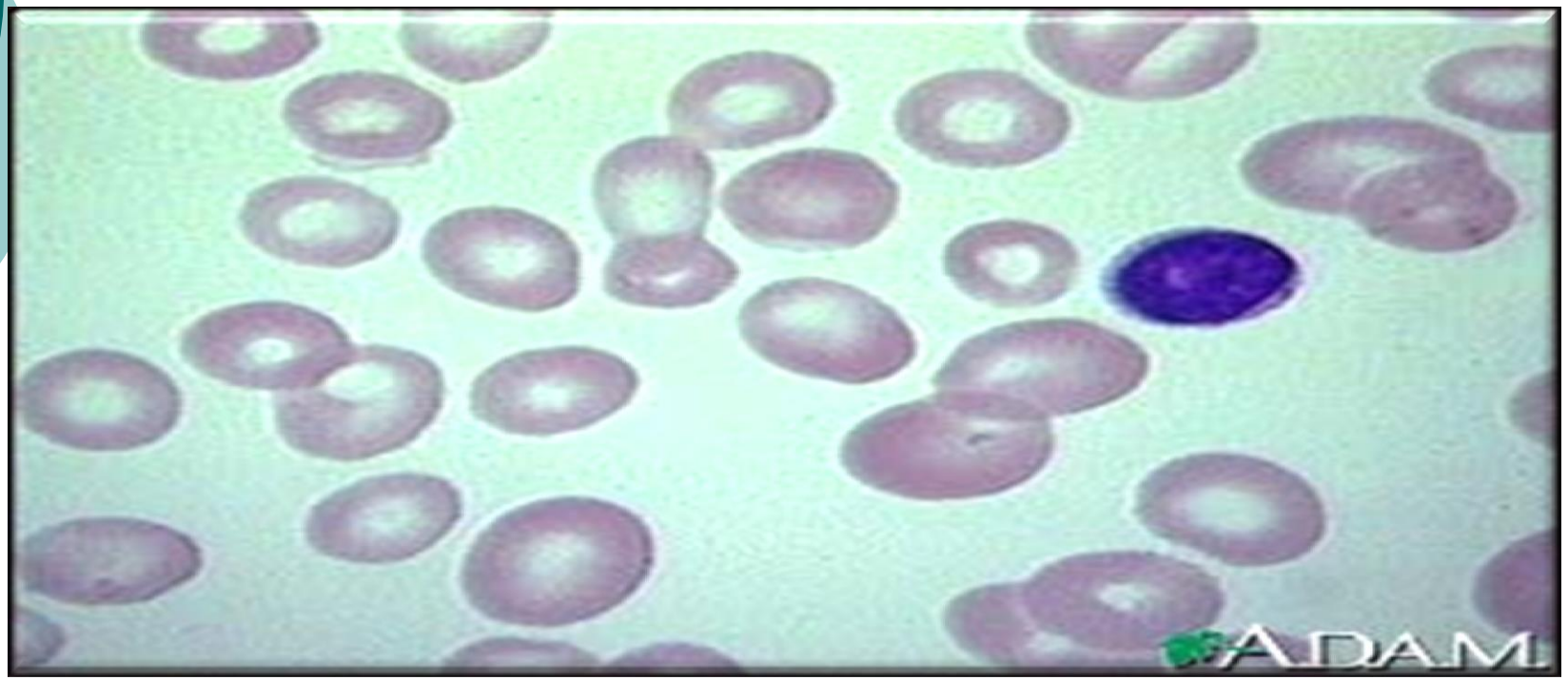


- No specific drug or diet are effective in treating thalassemia
- Thalassemia minor
  - Body adapts to ↓ Hgb
- Thalassemia major
  - Blood transfusions with IV deferoxamine (used to remove excess iron from the body)

Megaloblastic Anemias

- Characterized by large RBCs which are fragile and easily destroyed
- Common forms of megaloblastic anemia
  1. Cobalamin deficiency
  2. Folic acid deficiency

Megaloblastic Anemias



This picture shows large, dense, oversized, red blood cells (RBCs) that are seen in megaloblastic anemia.

Cobalamin (Vitamin B_{12}) Deficiency

Cobalamin Deficiency: formerly known as pernicious anemia.

- **Vitamin B_{12} :**(cobalamin) is an important water-soluble vitamin.
- **Intrinsic factor (IF) :**is required for cobalamin absorption.

Cobalamin (Vitamin B_{12}) Deficiency

Causes of cobalamin deficiency

- Gastric mucosa not secreting IF
- GI surgery →loss of IF-secreting gastric mucosal cells
- Long-term use of H_2-histamine receptor blockers cause atrophy or loss of gastric mucosa.
- Nutritional deficiency
- Hereditary defects of cobalamine utilization

Transportation path of Vit B12

Diet

Stomach

B12

B12 + IF

IF

Ileum B12+IF TCII

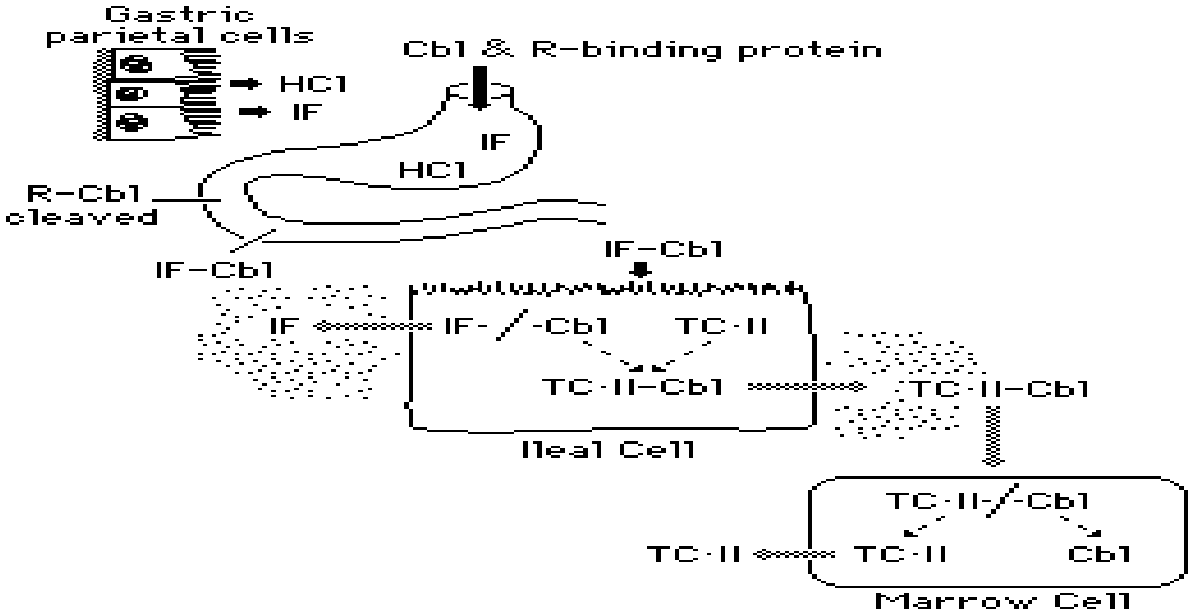
B12 +TCII

Circulation

Bone marrow (DNA synthesis)

Liver (storage)

Other tissues (DNA synthesis)



Cobalamin (Vitamin B_{12}) Deficiency

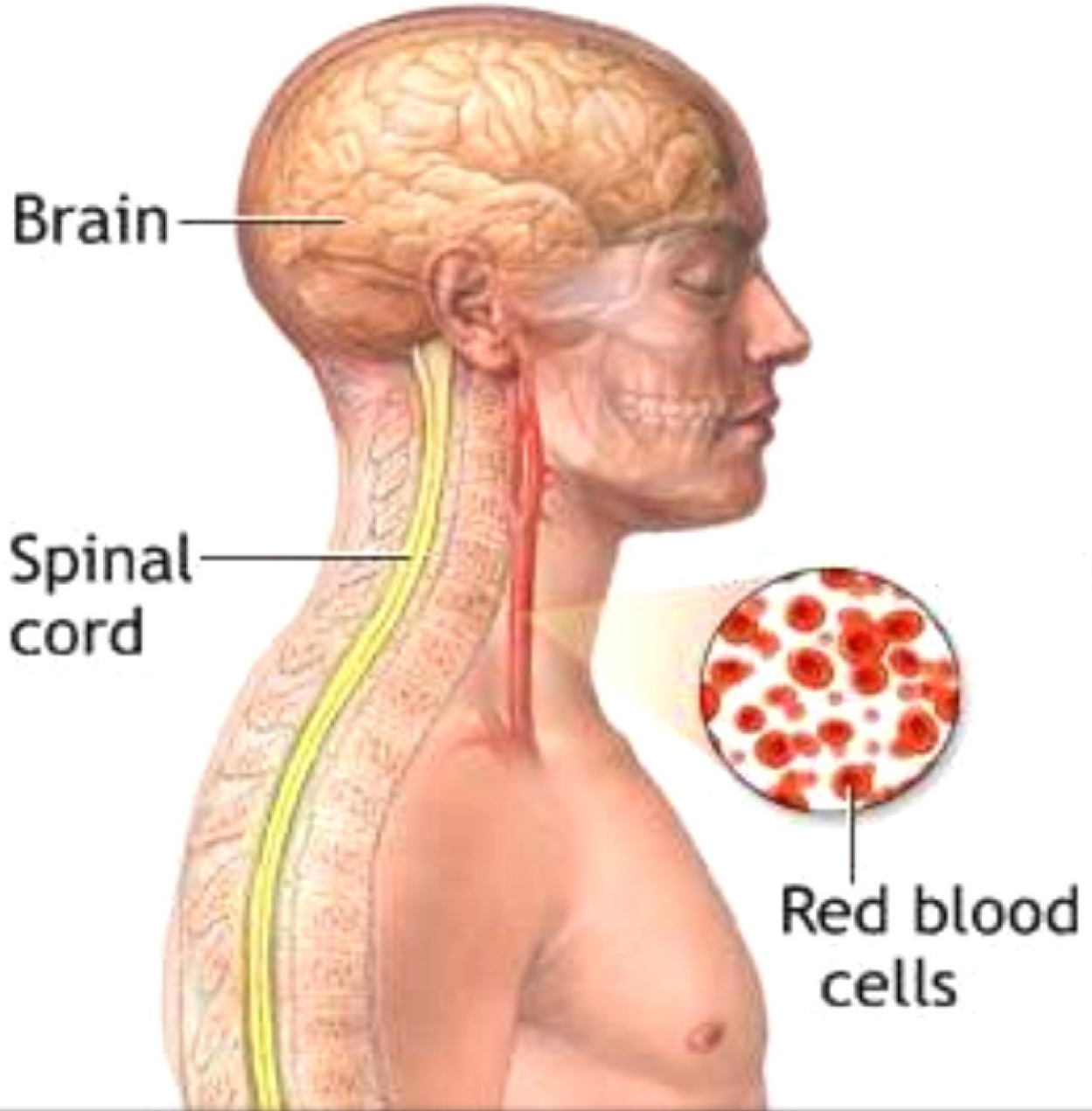
- Clinical manifestations
  - General symptoms of anemia
  - Sore tongue
  - Anorexia
  - Weakness
  - Parathesias of the feet and hands
  - Altered thought processes
    - Confusion → dementia

Cobalamin (Vitamin B_{12}) Deficiency

Neurological effects

- Deficiency results in damage to dorsal columns (sensory) and lateral columns (motor) of spinal cord
- Decreased vibration sense and position sense of joints detectable, and may affect gait, etc.
- May have positive Romberg's test
- Severe effects may include ataxia and dementia

Vitamin B12



Vitamin B12 is important for metabolism, the formation of red blood cells, and the maintenance of the central nervous system, which includes the brain and spinal cord

Symptoms of Vitamin B12 Deficiency

1 Mental Problems

Problems such as brain fog, Alzheimer's and dementia are symptoms of low B12

2 Fatigue

The most classic B12 Deficiency Symptom

3 Hair Problems

Thinning hair and grey hair can be symptoms too

6 Chronic Pain

Fibromyalgia, back pain, , neuropathy and more

5 Infertility

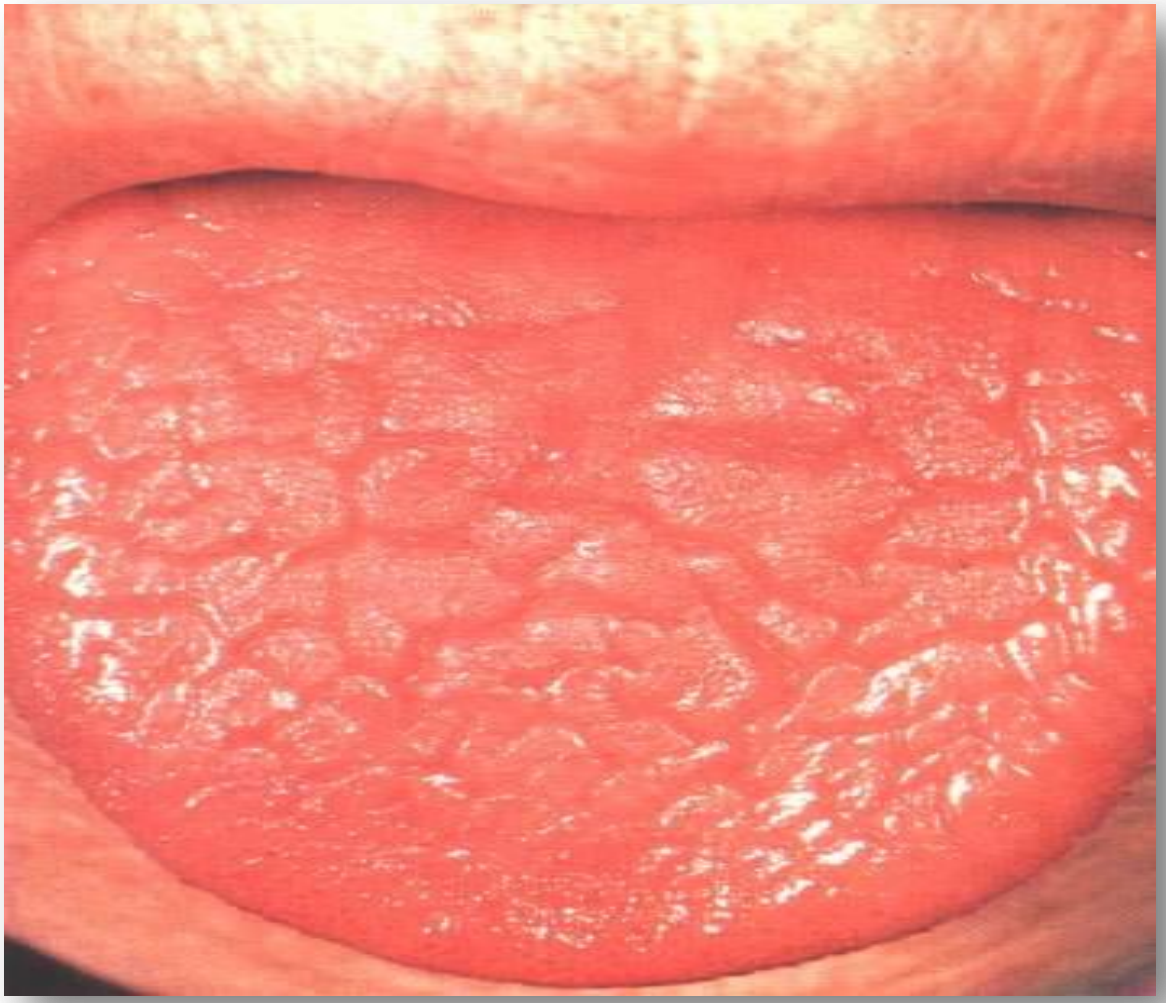
In both women and men

4 Blood Disorders

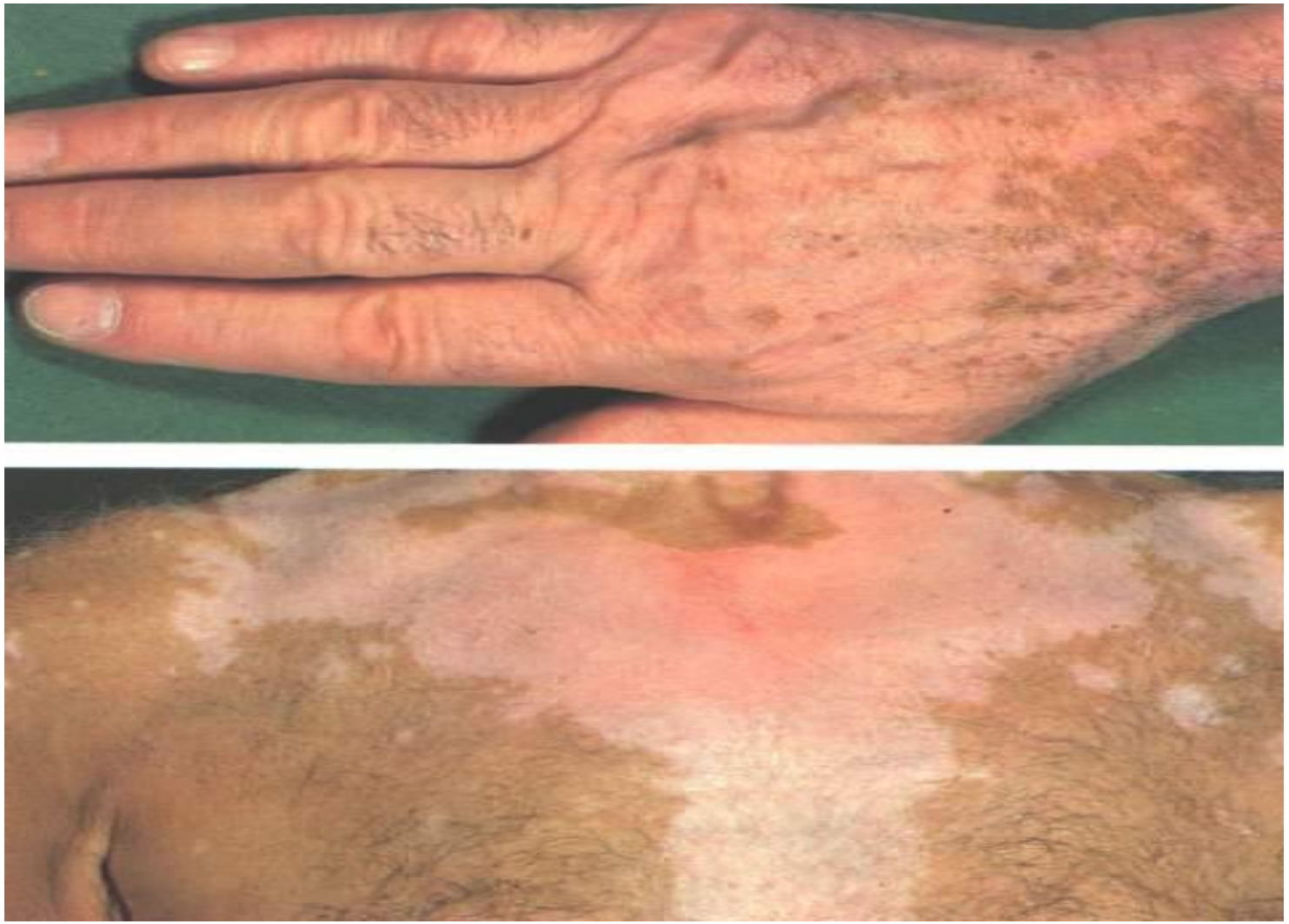
Blood disorders such as elevated MCV are symptoms

Easy-immune-health.com

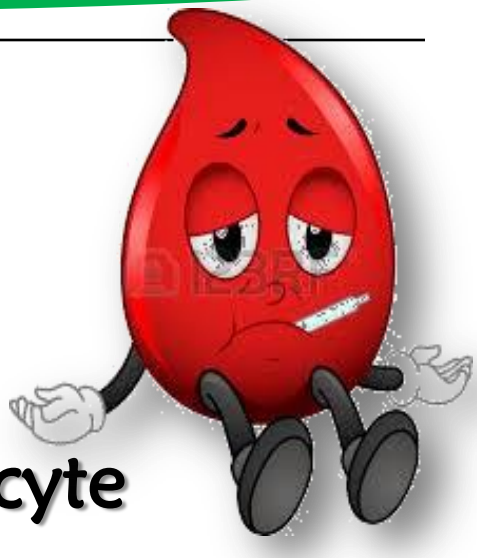
Red beefy tongue



Vitiligo

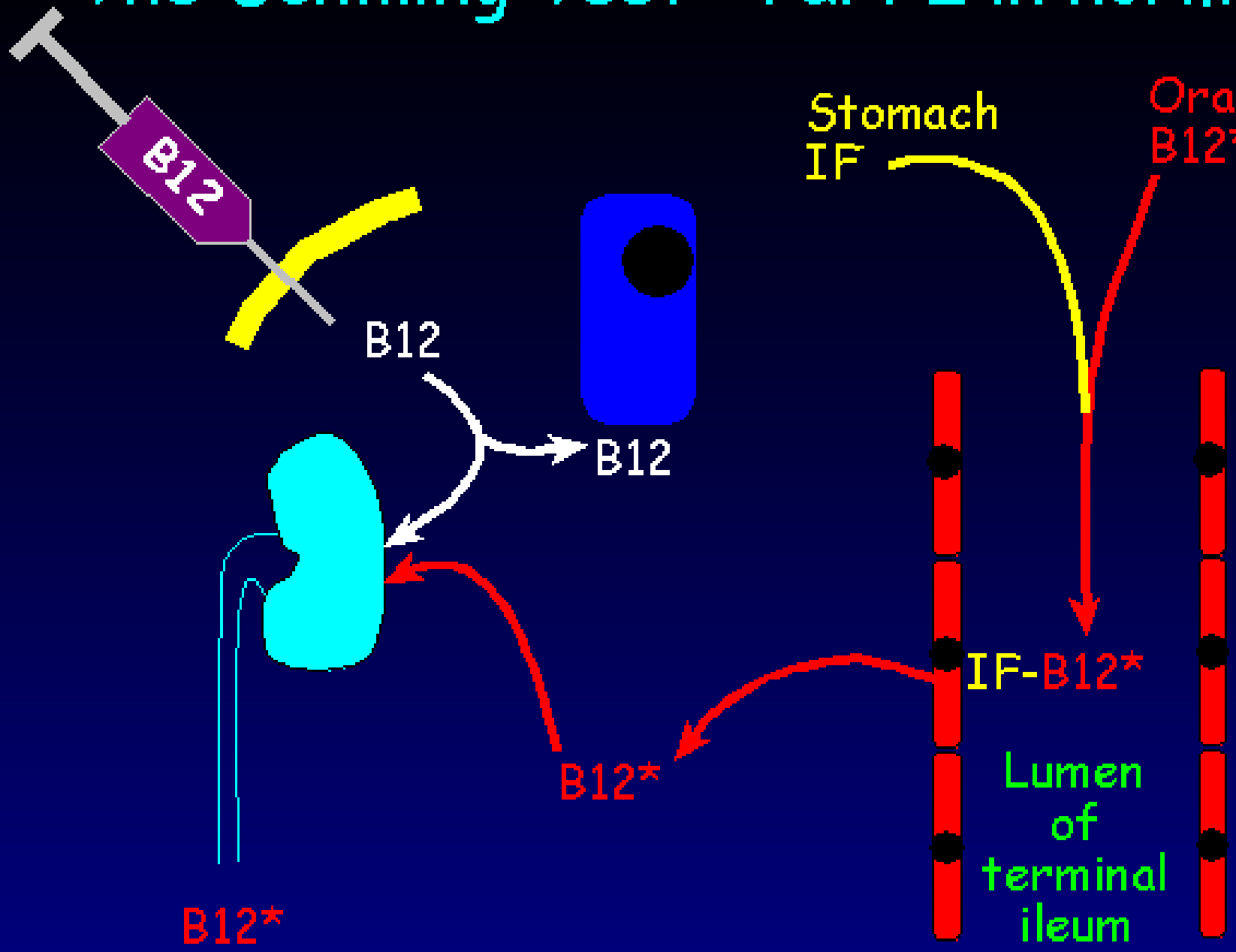


Cobalamin (Vitamin B_{12}) Deficiency



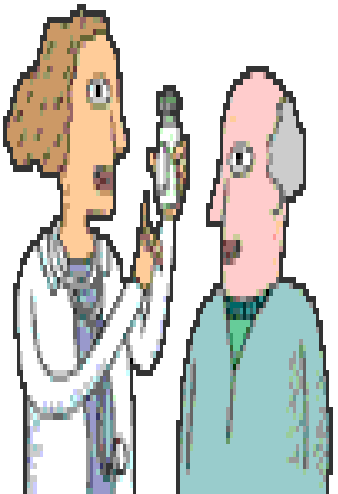
- Diagnostic Studies
  - RBCs appear large
  - Abnormal shapes
  - Structure contributes to erythrocyte destruction
  - Schilling Test: a medical investigation used for patients with vitamin B12 deficiency. The purpose of the test is to determine if the patient has pernicious anemia.

The Schilling Test - Part I in normal



Cobalamin (Vitamin B_{12}) Deficiency

- Collaborative Care
  - Parenteral administration of cobalamin
  - ↑ Dietary cobalamin does not correct the anemia
    - Still important to emphasize adequate dietary intake
  - Intranasal form of (Nascobal) is availablecyanocobalamin
  - High dose oral cobalamin and SL cobalamin can use be used



Cobalamin (Vitamin B_{12}) Deficiency

- Nursing Management
  - Familial disposition
    - Early detection and treatment can lead to reversal of symptoms
  - Potential for Injury r/t patient's diminished sensations to heat and pain
  - Compliance with medication regime
  - Ongoing evaluation of GI and neuro status
    - Evaluate patient for gastric carcinoma frequently

Vitamin B12

Food sources of vitamin B12:

Eggs, meat, poultry, shellfish, milk and milk products

A.D.A.M.

Folic Acid Deficiency

- Folic Acid Deficiency also causes megablastic anemia (RBCs that are large and fewer in number)
- Folic Acid required for RBC formation and maturation
- **Causes**
  - **Poor dietary intake**
  - **Malabsorption syndromes**
  - **Drugs that inhibit absorption**
  - **Alcohol abuse**
  - **Hemodialysis**

Folic Acid Deficiency

- Clinical manifestations are similar to those of cobalamin deficiency
- Insidious onset: progress slowly
- Absence of neurologic problems
- Encourage patient to eat foods with large amounts of folic acid
  - Leafy green vegetables
  - Liver
  - Mushrooms
  - Oatmeal (الشوفان المجروش)
  - Peanut butter
  - Red beans

Folic Acid Deficiency

- Treated by folate replacement therapy

❑ never give folate alone to individual with megaloblastic anemia because it

will mask B12 deficiency and neurological degeneration will continue

❑ folic acid 15 mg PO/day x 3 months; then 5 mg PO/day maintenance if cause not reversible

❑ folic acid supplementation 1 mg PO/day will protect against elevated homocysteine levels (risk factor for CAD)

Sources of Folic Acid

- Liver
- Yeast
- Nuts
- Dried beans
- Whole grains
- Spinach and other leafy greens
- Oranges
- Avacados

Source: The Nutrition Bible

Anemia of Chronic Disease

- Underproduction of RBCs, shortening of RBC survival
- 2nd most common cause of anemia (after iron deficiency anemia
- Generally develops after 1-2 months of sustained disease
- Causes
  - Impaired renal function
  - Chronic, inflammatory, infectious or malignant disease
  - Chronic liver disease
  - Folic acid deficiencies
  - Splenomegaly
  - Hepatitis

Anemia of Chronic Disease

Management

- resolves if underlying disease is treated
- erythropoietin may normalize the hemoglobin value
- dose of erythropoietin required higher than for patients with renal disease
- only treat patients who can benefit from a higher hemoglobin level

Case Study

- Pt is a 43 year old woman who was in her usual state of health until 03/16/11 when she presented to outside ER with left flank pain and dark urine for two days.
- Found to have hct of 26 with platelet count of 26k.
- No hemolysis on labs.
- Peripheral smear reveals no atypical cells with few large platelets. Initially thought to have ITP- given IVIG x 1. Did not response to IVIG.
- Required daily platelet transfusions.

Aplastic Anemia

- **Characterized by Pancytopenia**
  - ↓ of all blood cell types
    - RBCs
    - White blood cells (WBCs)
    - Platelets
  - Hypocellular bone marrow

↓WBC→infection.
↓Hb →anemia.
↓platelets →bleeding

- **Etiology**
  - **Congenital**
    - Chromosomal alterations
  - **Acquired**
    - Results from exposure to ionizing radiation, chemical agents, viral and bacterial infections

Aplastic Anemia

- Etiology
  - Low incidence
    - Affecting 4 of every 1 million persons
  - Manageable with erythropoietin or blood transfusion
  - Can be a critical condition
    - Hemorrhage
    - Sepsis

Aplastic Anemia

Pathophysiology:

The primary defect is a reduction in or depletion of hematopoietic precursor stem cells

- This may be due to quantitative or qualitative damage to the pluripotential stem cell.
- In rare instances it is the result of abnormal hormonal stimulation of stem cell proliferation or the result of a defective bone marrow microenvironment or from cellular or humoral immunosuppression of hematopoiesis.

Aplastic Anemia

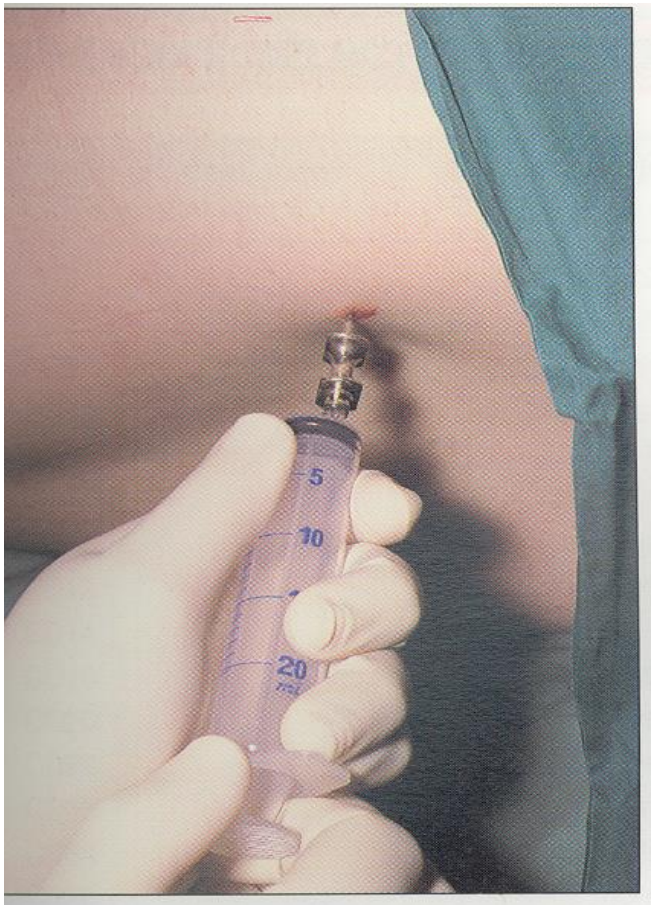
Clinical Manifestations

- Gradual development
- Symptoms caused by suppression of any or all bone marrow elements
- General manifestations of anemia
  - Fatigue
  - Dyspnea
  - Pale skin
  - Frequent or prolonged infections
  - Unexplained or easy bruising
  - Nosebleed and bleeding gums
  - Prolonged bleeding from cuts
  - Dizziness
  - headache

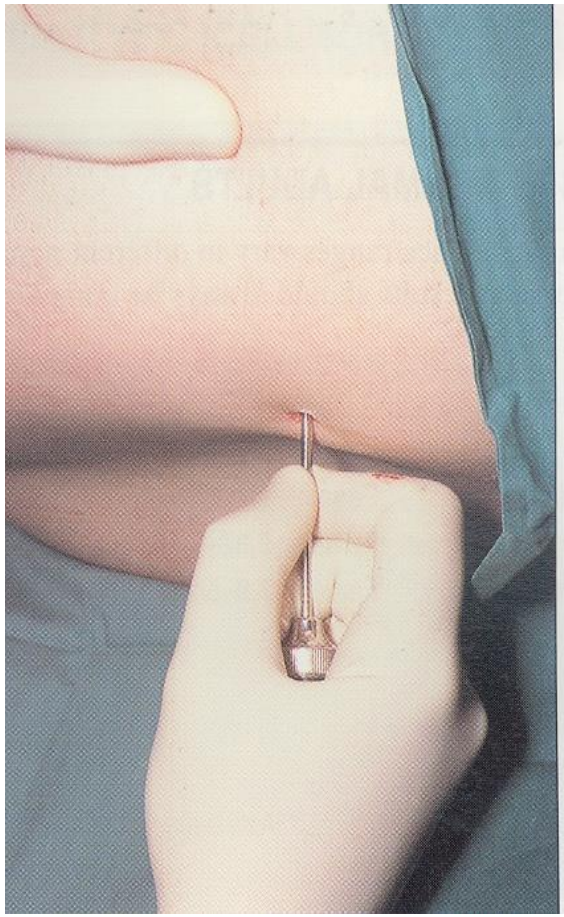
Aplastic Anemia

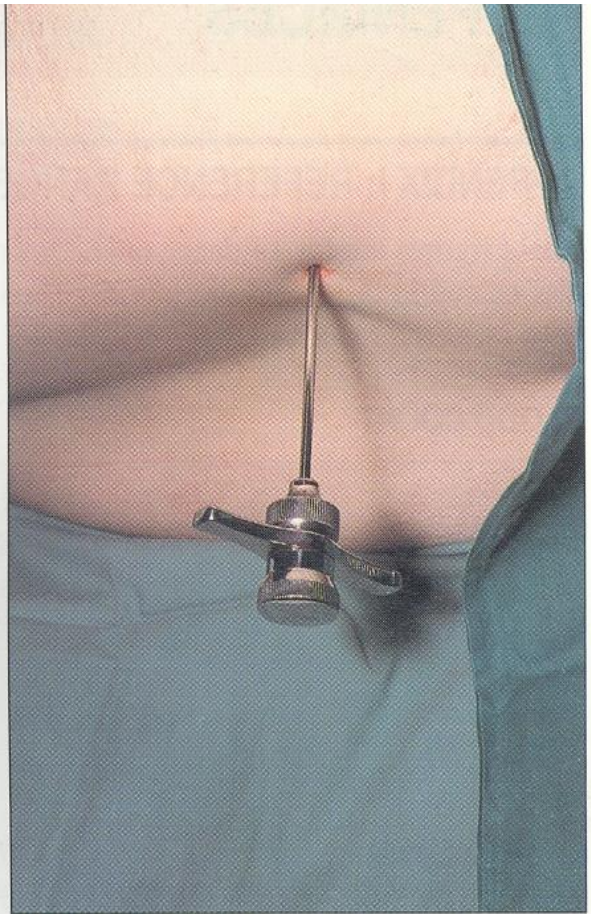
DIAGNOSIS??

- Blood tests
  - CBC
- Bone marrow biopsy

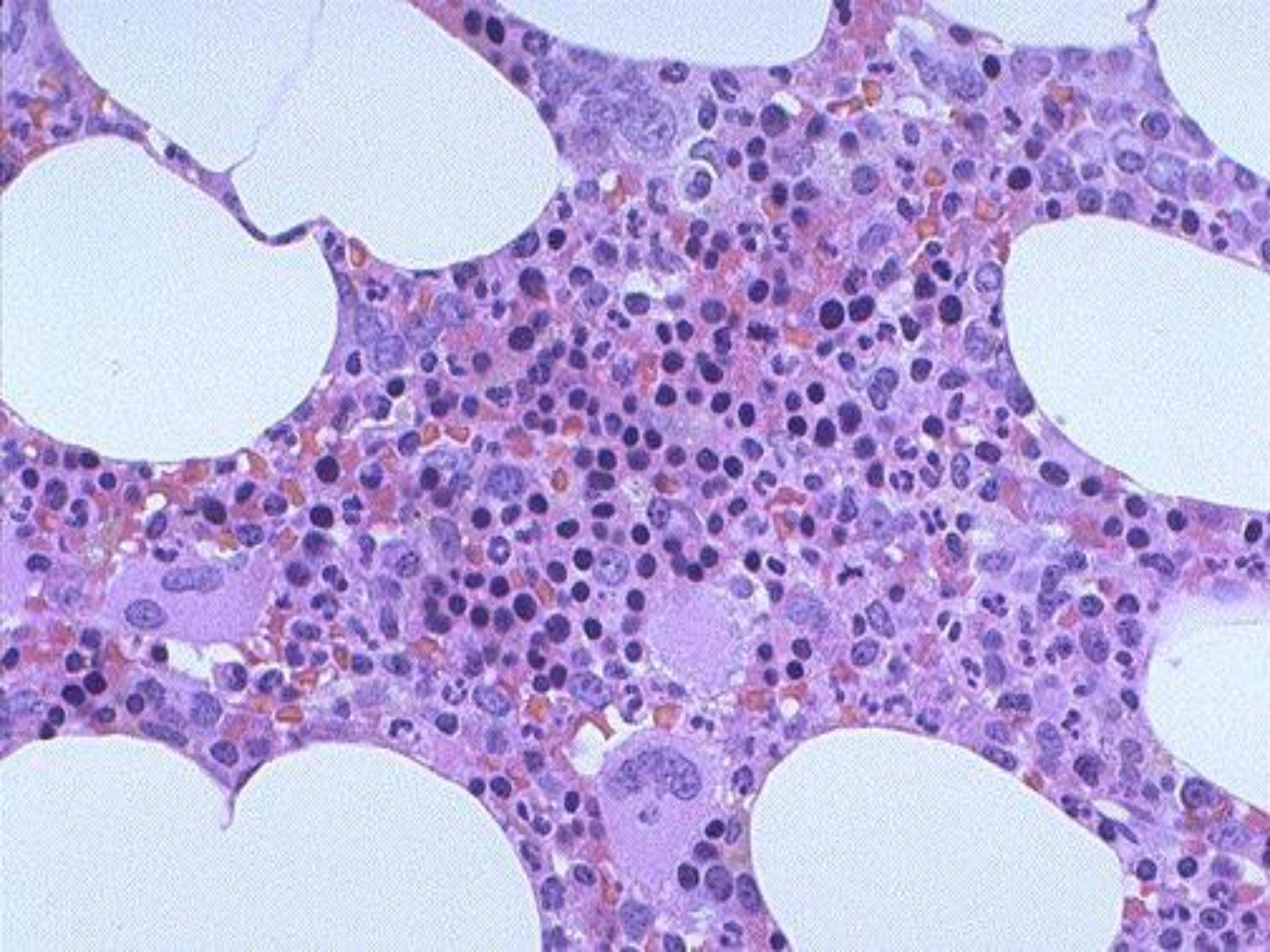


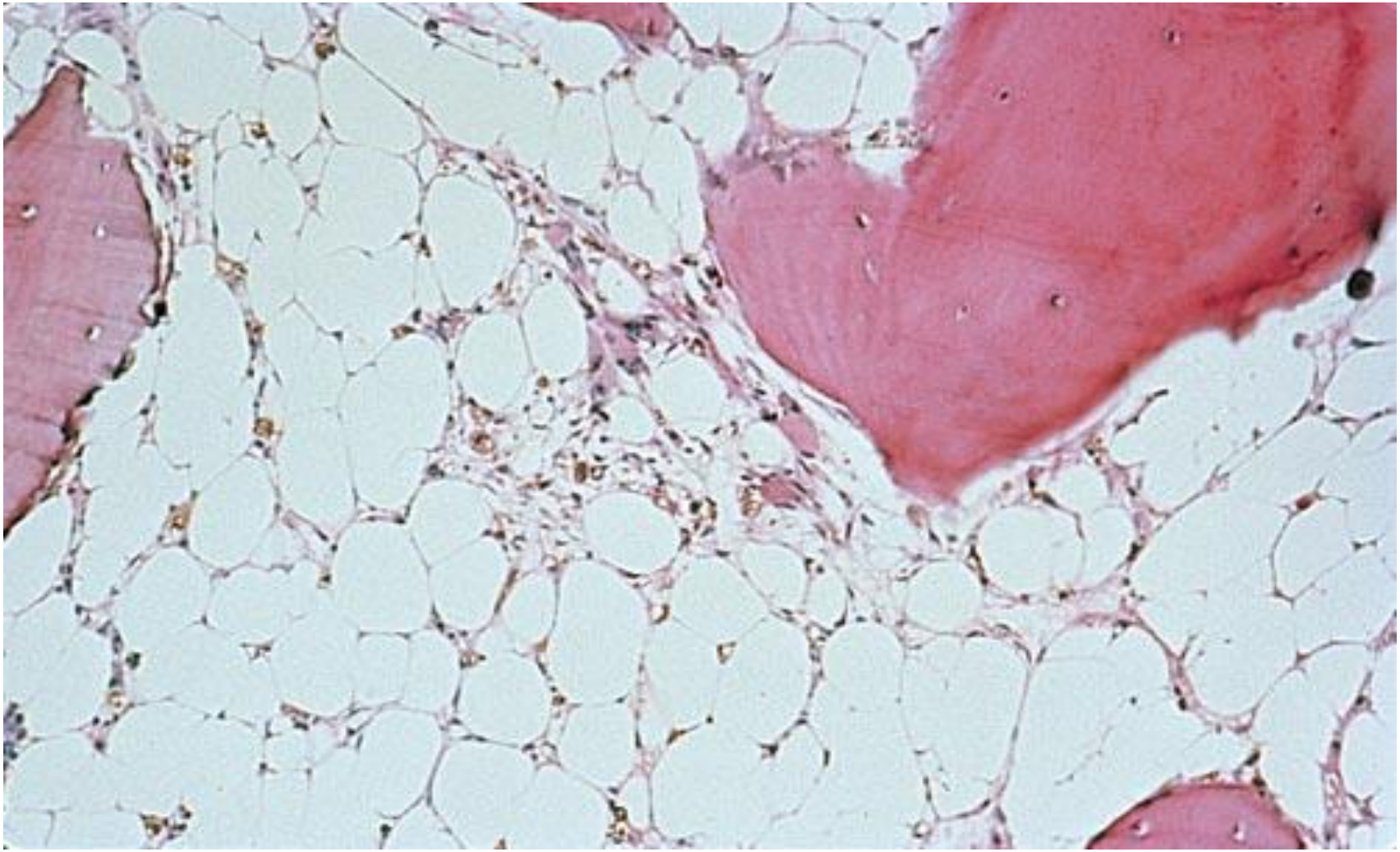
BM Aspiration





BM Biopsy





BM biopsy

hypocellular ,increased fat spaces

Aplastic Anemia

- **Treatment**
  - Identifying cause
  - Blood transfusions
  - Antibiotics
  - Immunosuppressants (neoral, sandimmune)
    - Corticosteroids (Medrol, solu-medrol)
  - Bone marrow stimulants
    - Filgrastim (Neupogen)
    - Epoetin alfa (Epogen, Procrit)
  - Bone marrow transplantation

Aplastic Anemia

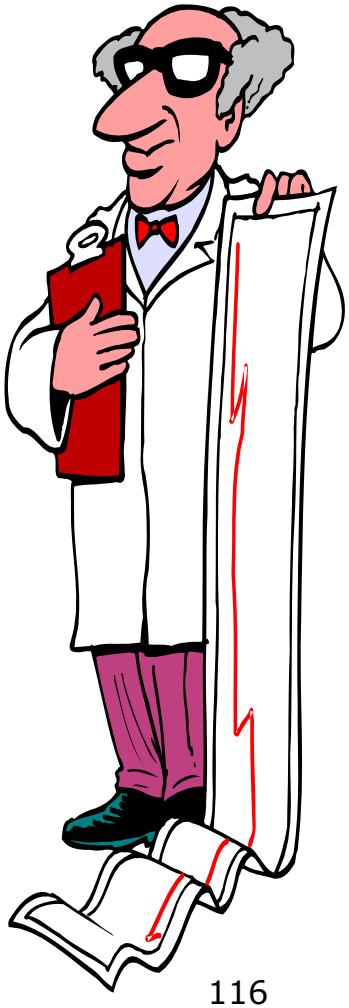
- **Nursing Management**
  - Preventing complications from infection and hemorrhage
  - Prognosis is poor if untreated
    - 75% fatal

Anemia Caused By Blood Loss

- Acute Blood Loss
- Chronic Blood Loss

Acute Blood Loss

- **Result of sudden hemorrhage**
  - Trauma, surgery, vascular disruption
- **Collaborative Care**
  1. Replacing blood volume
  2. Identifying source of hemorrhage
  3. Stopping blood loss

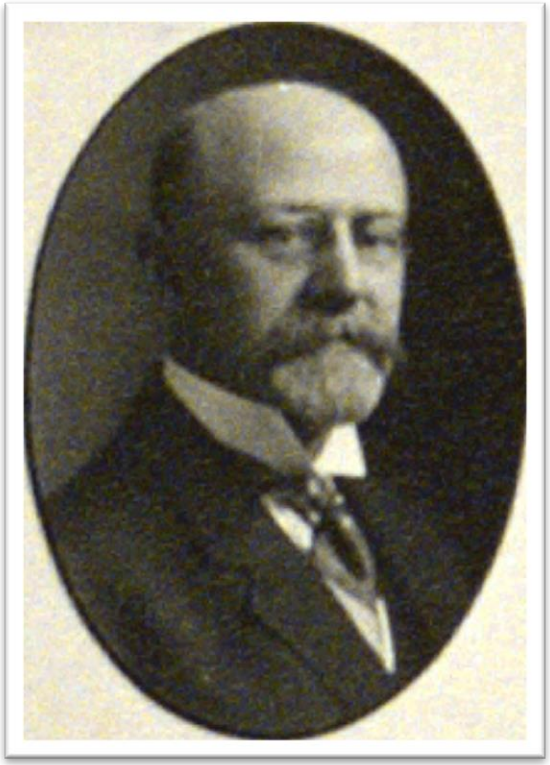


Chronic Blood Loss

- **Sources/Symptoms**
  - Similar to iron deficiency anemia
  - GI bleeding, hemorrhoids, menstrual blood loss
- **Diagnostic Studies**
  - Identifying source
  - Stopping bleeding
- **Collaborative Care**
  - Supplemental iron administration

Anemia caused by Increased Erythrocyte Destruction

- Hemolytic Anemia
  - Sickle Cell disease (peds)
  - Acquired Hemolytic Anemia
  - Hemochromatosis
  - Polycythemia



Hemolytic Anemia

- Destruction or hemolysis of RBCs at a rate that exceeds production
- Third major cause of anemia
- Intrinsic hemolytic anemia
  - Abnormal hemoglobin
  - Enzyme deficiencies
  - RBC membrane abnormalities
- Extrinsic hemolytic anemia
  - Normal RBCs
  - Damaged by external factors
    - Liver
    - Spleen
    - Toxins
    - Mechanical injury (heart valves)

Sequence of Events in Hemolysis

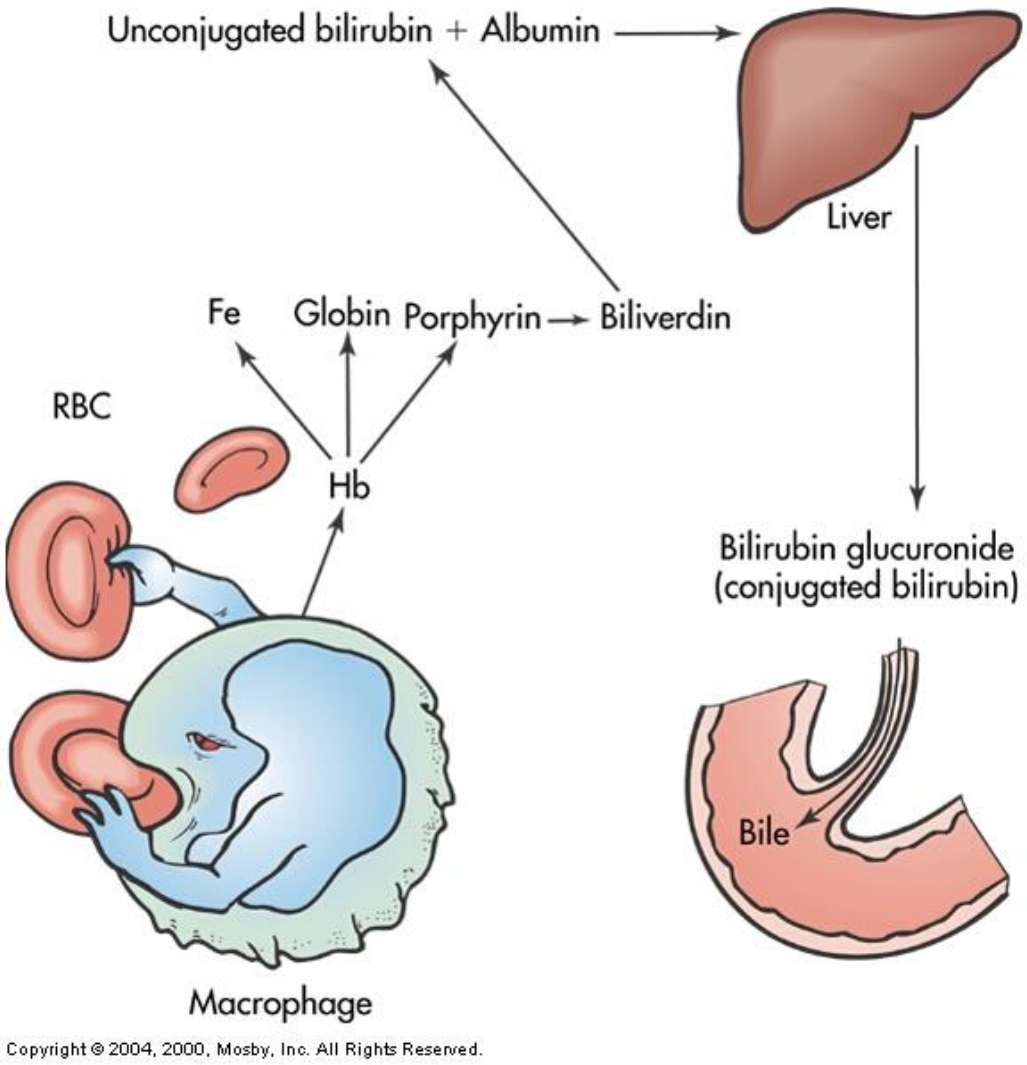


Fig. 30-1

Acquired Hemolytic Anemia

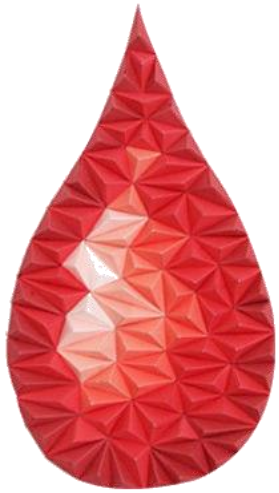
- Causes
  - Medications
  - Infections
- Manifestations
  - S/S of anemia
- Complications
  - Accumulation of hemoglobin molecules can obstruct renal tubules → Tubular necrosis
- Treatment
  - Eliminating the causative agent

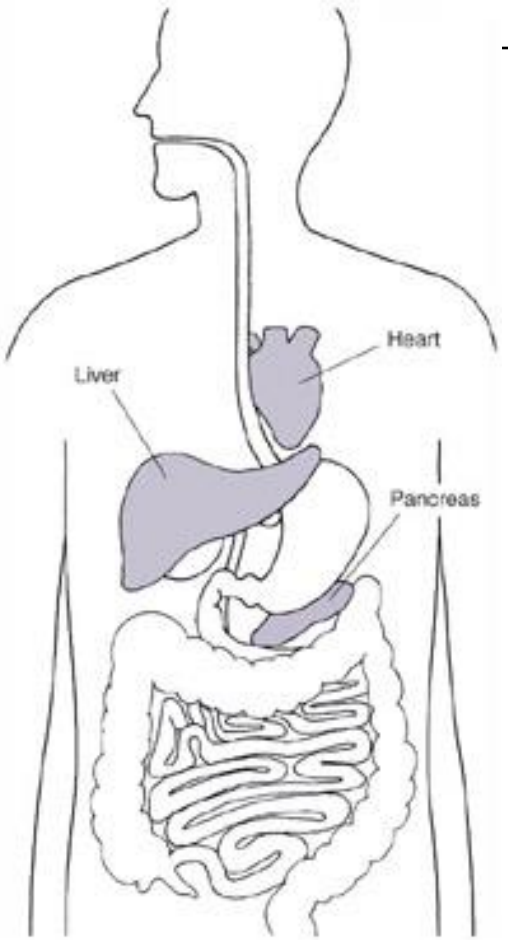
Potential Nursing Dx for Patients with Anemia

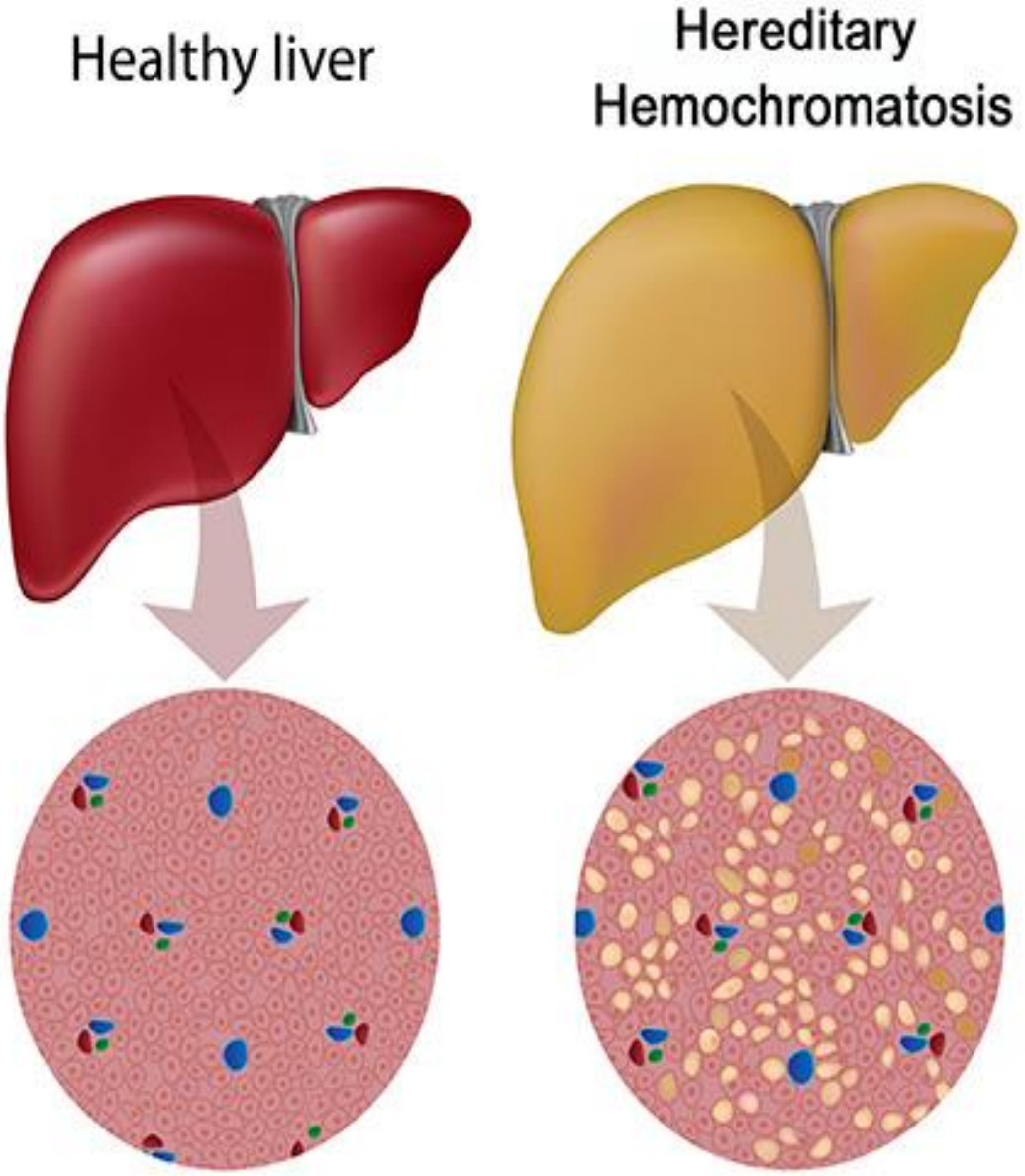
- **Activity Intolerance** r/t weakness, malaise m/b difficulty tolerating ↑'d activity
- **Imbalance nutrition: less than body requirements** r/t poor intake, anorexia, etc. m/b wt loss, ↓ serum albumin, ↓ iron levels, vitamin deficiencies, below ideal body wt.
- **Ineffective therapeutic regimen management r/t** lack of knowledge about nutrition/medications etc. m/b ineffective lifestyle/diet/medication adjustments
- Collaborative Problem: **Hypoxemia** r/t ↓hemoglobin

Hemochromatosis

- Iron overload disease
- Over absorption and storage of iron causing damage especially to liver heart and pancreas






Healthy liver
Hereditary Hemochromatosis

Polycythemia

- **Polycythemia** is a condition in which there is a net increase in the total number of red blood cells
- Overproduction of red blood cells may be due to
  - a primary process in the bone marrow (a so-called myeloproliferative syndrome)
  - or it may be a reaction to chronically low oxygen levels or
  - malignancy

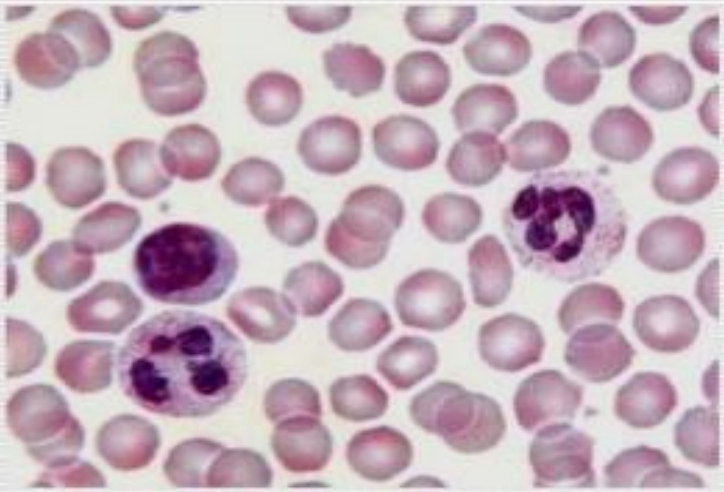
Polycythemia

Result from increased blood volume and viscosity.

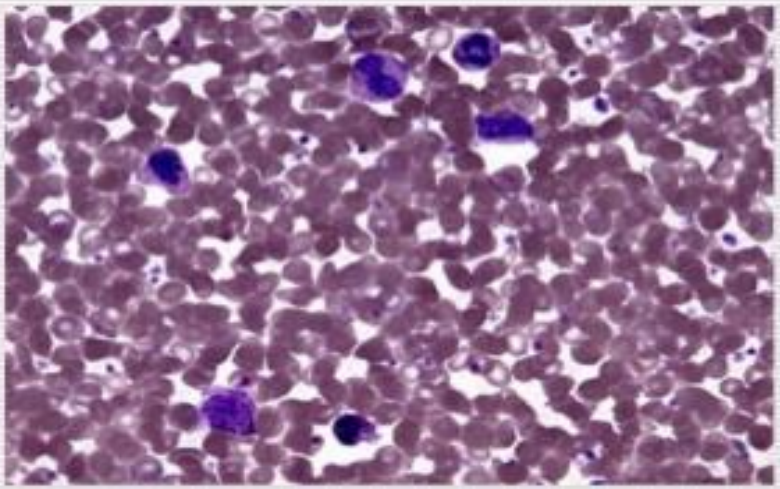
1. Reddish purple skin and mucosa, pruritus (especially after bathing).
2. Splenomegaly, hepatomegaly.
3. Epigastric discomfort, abdominal discomfort.
4. Painful fingers and toes from arterial and venous insufficiency, paresthesia ,dyspnea and orthopnea .

Vaquez' disease (Polycythemia vera)

Tumor induced hyperplasia of bone marrow



Normal blood smear



Polycytemia vera

Polycythemia

- Complications
  - ↑d viscosity of blood
  - → hemorrhage and thrombosis
- Treatment
  - Phlebotomy
    - if symptoms are due to erythrocytosis alone and platelet count normal or only slightly increased
  - Myelosupressive agents: A number of new therapeutic agents such as, interferon alfa-2b (Intron A) therapy, agents that target platelet number (e.g., anagrelide [Agrylin]), and platelet function (e.g., aspirin).

Thrombocytopenia

- Disorder of decreased platelets
- platelet count below 150,000
- Causes
  - Low production of platelets
  - Increased breakdown of platelets
- Symptoms
  - Bruising
  - Nosebleeds
  - Petechiae (pinpoint microhemorrhages)

Thrombocytopenia

- Types of Thrombocytopenia
  - Immune Thrombocytopenic Purpura
    - Abnormal destruction of circulating platelets
    - Autoimmune disorder
    - Destroyed in hosts' spleen by macrophages
  - Thrombotic Thrombocytopenic Purpura
    - ↑d agglutination of platelets that from microthrombi

Heparin-Induced Thrombocytopenia (HIT)

- **HIT**
  - Associated with administration of heparin
  - Develops when the body develops an antibody, or allergy to heparin
  - Heparin (paradoxically) causes thrombosis
  - Immune mediated response that casues intense platelet activation and relaese of procoaggulation particles.
- **Clinical features**
  - Thrombocytopenia
  - Possible thrombosis after heparin therapy
    - Can be triggered by any type, route or amount of heparin

Thrombocytopenia

Diagnostic Studies

- Platelet count
- Prothrombin Time (PT)
- Activated Partial Thromboplastin Time (aPTT)
- Hgb/Hct

Treatment

- Based on cause
- Corticosteroids
- Plasmaphoresis
- Splenectomy
- Platelet transfusion

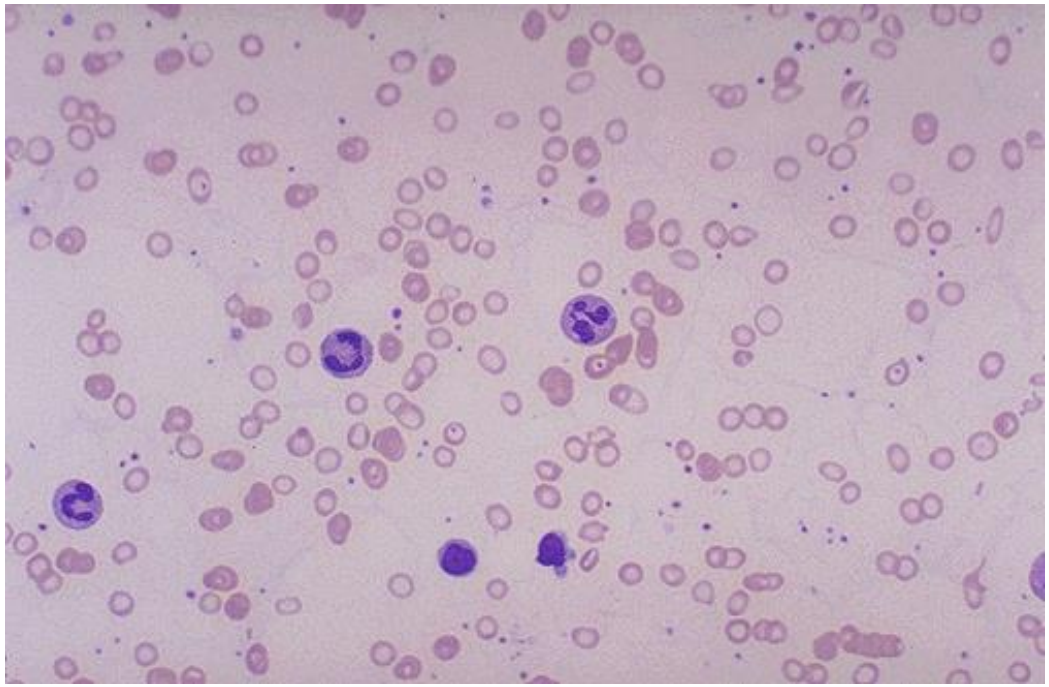
Case Study

Anemia Case Study 1

A 72 year old male has the CBC findings shown. Peripheral RBCs are hypochromic & microcytic.

Anemia Case Study 1

- **What test would you order for this patient?**
  - A-Hemoglobin Electrophoresis
  - B-Retic count
  - C-Stool for occult blood
  - D-B12 Assay
  - E-Bone marrow biopsy



Anemia Case Study 1

- Two questions:
  - What is your diagnosis?
  - What is the next step for this patient?

Anemia Case Study 2

- A 67 year old woman with a history of IDDM and treated hypothyroidism is referred to you for evaluation of anemia. Her complaints leading to this diagnosis included weakness, fatigue, weight loss, and mild numbness in her feet bilaterally.
- Physical exam was essentially normal except for mild loss of proprioception in her feet bilaterally.

Anemia Case Study 2

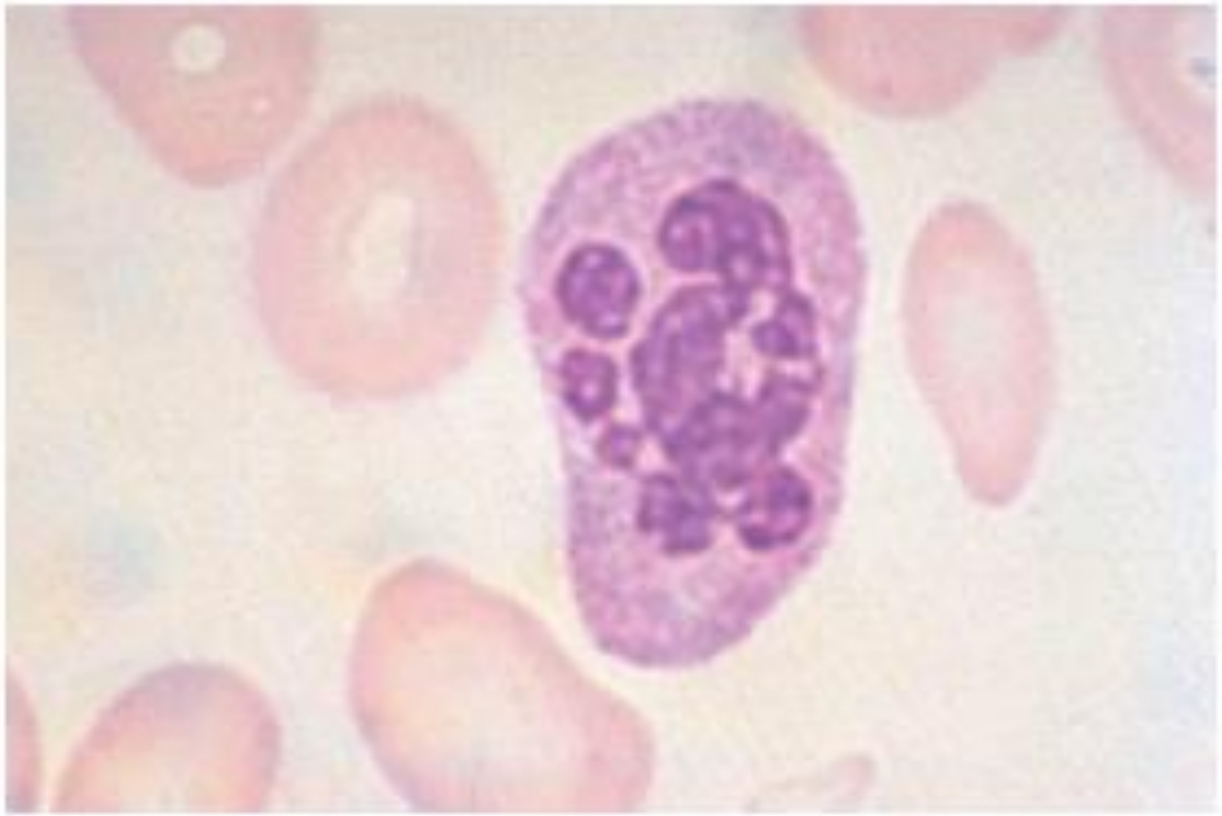
Current Labs:

WBC	3,700/mm^3 (4-10 thou/mm^3)
Hemoglobin	8.3 g/dL (12-16 g/dL)
Hematocrit	25% (35-45%)
Platelet	152,000/mm^3 (150-450 thou/mm^3)
MCV	123 μ^3 (80-100)
Retic count	.3% (.3-1.7%)

2 years ago:

WBC	4800/mm^3 (4-10 thou/mm^3)
Hemoglobin	11.2 g/dL (12-16 g/dL)
Hematocrit	33% (35-45%)
Platelet	187,000/mm^3 (150-450 thou/mm^3)
MCV	103 μ^3 (80-100)
Retic count	1.1% (.3-1.7%)

How do you interpret these values?



*Hypersegmented neutrophil, macroovalocytes

- What tests or procedures do you want to perform to further evaluate this patient?

Folate	8.3 ng/mL (>3)
Vitamin B12	73 pg/mL (180-914)
TSH	1.5 mIU/mL (.4-4.0)

You diagnose B12 deficiency and prescribe B12 injections 1000ug weekly x4 then 1000ug a month indefinitely.

- In 1 month the patient feels remarkably better and her blood counts have all improve