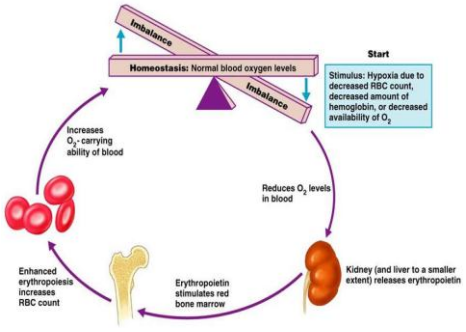
Blood Disorders

PART TWO
Roslyn G. Nott MSN, RN
Semester 2

REVIEW YOUR OBJECTIVES

1

Concept Review



2

Hemolytic Anemia

3

Hemolytic Anemia: 3rd Major Cause

- Destruction of RBC's
- Occurs in the bone marrow, liver and spleen
- Byproduct of hemolysis is bilirubin
- Under normal circumstances
  - Liver should be able to conjugate and excrete all bilirubin released

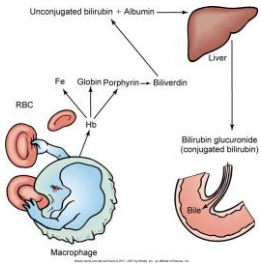
4

Hemolytic Anemia

- Destruction or hemolysis of RBC's
  - Rate which exceeds production
- Intrinsic hemolytic anemia: Usually hereditary
  - *Talk about this with Sickle Cell Anemia
  - Abnormal hemoglobin (hgb)
  - Enzyme deficiencies
  - RBC membrane abnormalities

5

Sequences of Events in Extravascular Hemolysis



6

Acquired Hemolytic Anemia: Most Common

- Four extrinsic categories
  - Physical factors
    - Destruction of RBC's for multiple reasons
      - Example: Hemodialysis, traumatic events
  - Microangiopathic destruction
    - RBC's are sheared or fragmented causing destruction
      - TTP or DIC

7

Acquired Hemolytic Anemia

- Four extrinsic categories
  - Immune reactions
    - Antigen-antibody reactions/ autoimmune reactions
    - Isoimmune reactions: antibodies develop after blood transfusions
  - Infectious agents & toxins
    - ↑ Hemolysis
      - Invades RBC's and destroys them, release of hemolytic substances

8

Hemolytic Anemia: Clinical Manifestations

- Jaundice
  - Destroyed RBC's cause ↑ bilirubin
- Enlarged spleen & liver
  - Enlarged d/t phagocytosis of defective RBC's
- **Renal Tubular Necrosis**
  - Accumulation of hgb molecules can obstruct renal tubules
    - Treatment focus: Maintain renal function

9

Hemolytic Anemia: Collaborative Care

- Remove causative agent
  - Malaria/ bacteria / toxins
- Aggressive hydration & electrolyte therapy
- If necessary:
  - Administer corticosteroids
  - Blood products
  - Splenectomy

10

Thalassemia

11

Thalassemia

- Autosomal recessive genetic disorder
- Inadequate production of normal hemoglobin
- Common ethic groups
  - Mediterranean Sea
  - Equatorial regions
    - Asia
    - Middle East
    - Africa

12

Thalassemia: Etiology

- Abnormal **Hgb** synthesis
  - Problem with globulin protein
  - Hemolysis occurs
- Thalassemia Minor
  - One gene
- Thalassemia Major
  - Two gene's

13

Thalassemia: Clinical Manifestations

- Thalassemia Minor
  - Frequently asymptomatic
  - Moderate anemia
    - Microcytosis (small size)
    - Hypochromia (pale color)

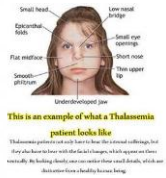
14

Thalassemia: Clinical Manifestations

- Thalassemia Major
  - Life-threatening
  - Physical & Mental Growth deficits
  - Symptoms develop in childhood

15

Thalassemia Major



16

Thalassemia: Physical Exam

- Thalassemia Major
  - Pallor
  - Splenomegaly
  - Hepatomegaly
  - Jaundice
  - Chronic bone marrow hyperplasia
    - Expansion of bone marrow space

17

Thalassemia: Severe Complications

- Hypertension
- Cardiac complications
- Pulmonary disease
- Liver issues (May have Hep C) >Cirrhosis >HCC
- Thombosis

18

Thalassemia: Collaborative Care

- No specific drugs or diet is effective in treatment
- Thalassemia Minor
  - Body adapts to ↓ in Hgb
- Thalassemia Major
  - Splenectomy
  - Enlarged spleen due to increased RBC lysis

19

Thalassemia: Collaborative Care

- Thalassemia Major
  - **Iron Chelation Therapy**
  - Blood transfusions with oral deferasirox (Exjade) or IV deferoxamine (Desferal)
    - Binds with excess iron and excretes it (reducing iron overload)
    - Patients are living longer
    - Goal: Hgb: 10g/ dL

20

Thalassemia Major: Collaborative Care

- Hematopoietic Stem Cell Transplantation (HSCT)
  - Only cure, but risky procedure
  - Risks may outweigh benefits

21

Megaloblastic Anemia

Group of disorders

22

Megaloblastic Anemia

- Characterized by **LARGE** RBC's: megaloblasts
- Cause: Impaired DNA synthesis

23

Megaloblastic Anemia

- Megaloblasts
  - Easily destroyed d/t fragile cell membrane
- **Majority result from nutritional deficiency of:**
  - Cobalamin (vit B12)
  - Folic Acid

24

Megaloblastic Anemia: Classification

- Cobalamin deficiency (Vit B12)
- Folic Acid deficiency
- Suppression of DNA synthesis – drug induced
- Erythroleukemia
  - Blood disorder ↑ erythropoietic cells in bone marrow

25

Cobalamin (B12) Deficiency

- Intrinsic Factor (IF)
  - Protein secreted by the parietal cells of the gastric mucosa
- IF is required for cobalamin absorption in the small intestine
- If intrinsic factor is not secreted cobalamin will not be absorbed

26

Pernicious Anemia

- **#1 Cause of cobalamin deficiency**
  - Caused by the absence of IF
    - IF is not secreted d/t gastric mucosal atrophy or
    - Autoimmune destruction of parietal cells
- FYI: ↓ HCL in stomach in pernicious anemia
  - Parietal cells secrete hydrochloric acid

27

Pernicious Anemia

- Begins usually after 40
  - Most commonly diagnosed at 60
- Occurs frequently
  - Northern European ancestry
  - African Americans
    - Usually onset earlier
    - Higher frequency in women
    - Often severe

28 Ethnic Link

Cobalamin Deficiency: Other Causes

- GI Surgery
  - Gastrectomy/ Gastric bypass/ Crohn's disease
    - Page 640 table 31-8
- Nutritional deficit
- Chronic alcoholism

29

Cobalamin Deficiency: Other Causes

- Long-term users of H2 histamine receptor blockers
- Acid environment is required for secretion of IF

Generic Name	Brand Name
cimetidine	Tagamet
famotidine	Pepcid
ranitidine	Zantac

30 Need acid environment to secrete IF

Cobalamin Deficiency: Clinical Manifestations (Table: 31-3)

- **Impaired thought process**
  - Range: confusion → dementia
- Neuromuscular weakness
- Ataxia & muscle weakness
- Paresthesia of feet & hands
- GI symptoms: N/V, abdominal pain, anorexia

31

All develop due to tissue hypoxia

Cobalamin Deficiency: Diagnostic Studies:

- RBC's appear large and abnormally shaped
- ↓ Serum cobalamin levels
- Normal serum folate levels

32

Cobalamin Deficiency: Diagnostic Studies:

- Serum test for anti-IF antibodies
  - Specific for pernicious anemia
- ↑ Serum methymalonic acid (MMA)
  - Elevated mainly in cobalamin deficiency
- ↑ Serum homocysteine
  - Elevated in both cobalamin & folic acid

33

Cobalamin Deficiency: Nursing Management

- Familial disposition
  - Early detection & treatment can reverse the symptoms (Pernicious Anemia)
- Injury prevention
  - Due to diminished sensation to heat & pain
- Encourage compliance with treatment
- Frequently evaluate for gastric carcinoma

34

Cobalamin Deficiency:Collaborative Care

- Treatment of choice: Parenteral or intranasal cobalamin
  - **Lifelong treatment necessary**
- Emphasize adequate diet
  - However ↑ dietary cobalamin does not correct the anemia
    - Lack IF
- Longstanding neuromuscular complications may not be reversible

35

Megaloblastic Anemia: Folic Acid Deficiency

- Folic acid is required:
  - DNA synthesis
  - RBC formation & maturation

36

Folic Acid Deficiency: Common Causes

- Dietary/nutritional deficiency
  - Malabsorption syndrome
  - Alcohol abuse
  - Anorexia
- Loss during hemodialysis

37

Folic Acid Deficiency: Clinical Manifestations

- Similar to cobalamin deficiency
- *__Absence of neurological problems__
- Treated by replacement therapy
  - Folic Acid 1mg daily po
    - In malabsoption states (chronic ETOH) up to 5mg may be necessary

38

Folic Acid Deficiency: Clinical Manifestations

- Encourage patient to eat foods with ↑ amount of folic acid
  - Green leafy veggies, liver, meat, fish, legumes, whole grains, orange juice, peanuts

39 Page 637 Table: 31-5

Aplastic Anemia

40

Aplastic Anemia

- Pancytopenia
  - ↓ in all blood cell types
    - RBC's
    - WBC's
    - Platelets
- Hypocellular bone marrow
  - With increased yellow marrow (fat content)

41

Aplastic Anemia

- Low incidence
  - Statistically: 2 of every 1 million persons
- Types
  - Congenital
    - Chromosomal alterations
  - Acquired
    - Autoimmune: Approx 75%
    - Exposure:
      - Ionizing radiation, chemical agents, viral & bacterial infections

42

Aplastic Anemia: Clinical Manifestations

- Fatigue/ Dyspnea
- Cardiovascular: ↑ HR, palpitations
- Cerebral: Confusion, HA, vertigo
- Neutropenia
  - Low grade fever should be considered an emergency

43

Aplastic Anemia: Diagnostics

- Lab studies
  - Normocytic/ Normochromic Anemia
  - ↓ Hgb/ Hct
  - ↓ Retic count, prolonged bleeding time
  - ↑ Serum iron & TIBC
    - Initial signs of erythropoiesis suppression

44

Page 638 table 31-6

Aplastic Anemia: Nursing Management

- Identify & remove cause (if possible)
- Preventing complications from infection & hemorrhage
- Untreated prognosis is poor: 70% fatal

45

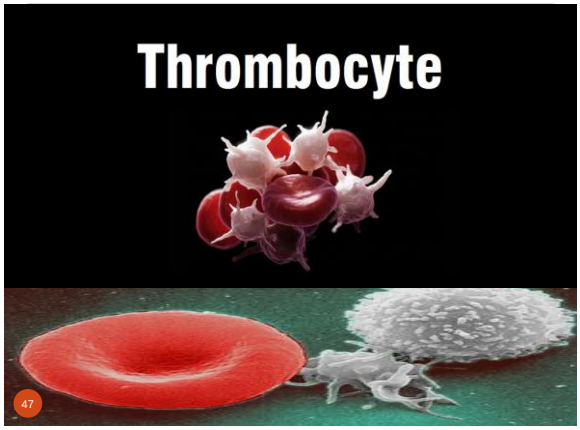
Aplastic Anemia: Nursing Management

Treatment options

- Immunosuppressive therapy
  - ATG, Cyclosporine or high-dose Cytoxan

    *Significantly improving outcomes
- Stem cell transplantation
  - Best results in younger patients who have not had previous blood transfusions
- High dose corticosteroids may be used

46



47

Thrombocytopenia

- Reduction of platelets
  - Below 150,000
- Clinical Manifestation
  - Prolonged or spontaneous bleeding
    - May be without injury

48

Thrombocytopenia: Etiology

- Primary
  - Inherited
- Acquired
  - Majority are acquired
  - Common cause: ingestion of certain herbs or drugs
    - Usual mechanism is **accelerated platelet destruction**
    - Antibodies attack the platelets

49 Chemo drugs or ganciclovir (antiviral)

Acquired Thrombocytopenia

- **Immune** thrombocytopenia purpura (ITP)
- **Thrombotic** thrombocytopenia purpura (TTP)
- **Heparin-induced** thrombocytopenia (HIT)

50

Immune Thrombocytopenia Purpura (ITP)

- Most common
- Primarily an autoimmune disease
- Syndrome of abnormal destruction of circulating platelets
  - Platelets are coated with antibodies
  - Spleen recognizes these platelets as foreign and they are destroyed by macrophages

51

Immune Thrombocytopenia Purpura (ITP)

- Acute condition when affecting children
- Chronic condition in adults
  - Gradual onset with transient remissions
    - Women between 15-40 years of age
    - Older adults
- Treated with multiple therapies

52

Collaborative Care: Immune Thrombocytopenia Purpura (ITP)

- Therapy initiated when platelet count <30,000
- Corticosteroids
  - Prednisone (example)
    - ↓ phagocytic response of the splenic macrophages
- High doses of IV Immunoglobin (IVIG)

53

Collaborative Care: Immune Thrombocytopenia Purpura (ITP)

- Splenectomy
  - 60-70% of patients have a complete or partial remission
- Platelet Transfusion
  - Life-threatening hemorrhage
    - * Concern: Antibody formation
    - Usually platelet count: <10,000 or if there is anticipated bleeding from a procedure

54 Renal biopsy

Thrombotic Thrombocytopenia Purpura (TTP)

- Uncommon Syndrome
- Called TTP-HUS
  - Most always associated with hemolytic-uremic syndrome
- Associated with ↑ aggregation of platelets that form into **microthrombi**

55

Thrombotic Thrombocytopenia Purpura (TTP)

- Causes:
  - Plasma enzyme deficiency (most cases)
    - ADAMTS13 >>> von Willebrand clotting factor (vWF)
      - vWF attach to platelets → ↑aggregation
  - Secondary cause: unclear
    - Autoimmune or drug toxicities
- Medical Emergency
  - Bleeding & clotting occur simultaneously

56

Ms. Najera will talk about von Willebrand

Collaborative Care: Thrombotic Thrombocytopenia Purpura (TTP)

- Remove the causative agent
  - Treat infection
- If untreated usually results
  - Irreversible renal failure & death
- **Plasma**pheresis (blood purification procedure)
  - Removal of enzymes & molecules that bind with platelets

57

Plasma contain antibodies → Plasma exchange

Collaborative Care: Thrombotic Thrombocytopenia Purpura (TTP)

- Corticosteroids
- Immunosuppressants
  - Ex: Cyclosporine
- If difficult to treat (unresponsive to tx)
  - Splenectomy

58

Heparin-induced Thrombocytopenia (HIT)

- Associated with heparin use
- Two major responses to an immune-mediated response to heparin:
  - Platelet destruction
  - Vascular endothelial injury
- Life threatening

59

Heparin-induced Thrombocytopenia (HIT)

- Develops 5-10 days after heparin therapy is started
- Platelet count drops 50% from baseline
- Major clinical problem
  - #1 Venous Thrombosis
  - Arterial thrombosis may occur
  - DVT & PE often result

60

Thrombocytopenia: Clinical Manifestations

- Asymptomatic (often)
- Most common symptom
  - Mucosal or cutaneous bleeding
    - Petechiae- microhemorrhages
    - Purpura- bruise from numerous petechiae
    - Ecchymosis- larger lesions from hemorrhage

61

Heparin-induced Thrombocytopenia (HIT)

From Forbes CD, Jackson WF: Colour atlas and text of clinical medicine, ed 3, London, 2003, Mosby.

62

Thrombocytopenia: Clinical Manifestations

- Prolonged bleeding after routine procedures
  - Internal bleeding may manifest as:
    - Tachycardia, hypotension, Weak/ dizzy, abdominal pain
- Major complication
  - Hemorrhage
    - May be obvious: Cerebral hemorrhage or
    - Subtle initially: hemorrhage of the joints

63

Thrombocytopenia: Lab Findings

- ↓ Platelet count
  - Prolonged bleeding < 50,000/uL
  - Hemorrhage <20,000 u/L
- Peripheral blood smear
- *Trend your lab values

64

Thrombocytopenia

- Medical History & clinical examination is key
  - Subtle confusion
  - HA
  - Seizures
  - Coma
- S/S may be subtle astute & thorough patient assessment is essential

65

Thrombocytopenia

- Diagnostics
  - Lab tests for coagulation may be normal
    - PT/ PTT
- Bone Marrow aspiration/ biopsy
  - Rule out: leukemia, aplastic anemia or other causes of thrombocytopenia
- Specific assays
  - C-serotonin release assay, ELISA

66

Nursing Diagnosis: Thrombocytopenia

- Examples:
  - Risk for bleeding r/t ↓ platelets
  - Deficient knowledge r/t lack of information
  - Impaired oral mucous membrane r/t ↓ platelets
- Potential Complications: PE, Stroke, Intracranial Hemorrhage

67

Thrombocytopenia: Collaborative Care

- Based on the **etiology** of the disorder
- Removal of tx or cause of the disorder
  - **Stop heparin**
- **Avoid aspirin**
  - And other medications that affect platelet function or production

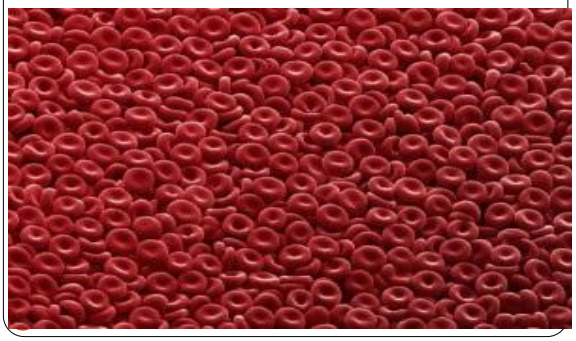
68

Educational Needs

- Prevention of bleeding
  - Minor nosebleeds or new petechiae
- Injury prevention strategies
- Discourage OTC medications
  - Many contain Asa
  - NSAIDS

69

Polycythemia



Polycythemia

- Production & presence of ↑ RBC's
- Results in ↑ blood viscosity & blood volume
  - Hyperviscosity & Hypervolemia

71

Polycythemia Vera (Primary)

- Chronic Problem
  - ↑ RBC's, WBC's & platelets
- Problem: ↑ blood in organs & tissues
- Common S/s
  - Splenomegly: 90% of patients
  - Hepatomegly
  - Hypercoagulopathies
    - Prone to clotting issues

72

Polycythemia Vera

- Genetic Link-small percentage
  - Genetic link: JAK2 gene
    - Begins with 1 or more DNA mutations of a hematopoietic stem cell
- Most polycythemia vera are not inherited
  - Occur with genetic changes in certain cells during lifetime

73

Secondary Polycythemia

- Hypoxia driven
  - Stimulates erythropoietin (EPO) production
- Hypoxia independent: need for 02
  - High altitude
  - Pulmonary/Cardiovascular disease
  - Tissue hypoxia
- Initially ↑ Serum EPO levels
- Splenomegaly does not occur

74

Polycythemia: Signs & Symptoms

- Circulatory
  - HTN, hypervolemia, hyperviscosity
    - C/O HA, dizziness, visual disturbances
    - Generalized pruritus
    - Painful burning & redness to hands and feet
- Most common serious complication
  - Stoke secondary thrombosis

75

Disease process ↑ RBC, WBC and platelets

Polycythemia: Signs & Symptoms

- Hemorrhagic Phenomena
  - Petechiae/ Ecchymosis
  - GI bleed/ Epistaxis
- Ruddy complexion (plethora)
- Hyperuricemia
  - RBC destruction

76

Polycythemia: Diagnostics

- Labs
  - ↑ Hgb & RBC
  - ↑ WBC/ platelets
  - ↑ Histamine, uric acid & cobalamin
  - **Normal or ↓ EPO in polycythemia vera**
  - **EPO ↑ (in secondary polycythemia only)**
- Bone marrow exam: hypercellularity of RBC's WBC's & platelets

77

↑ stem cells in bone marrow

Polycythemia: Treatment Goals

- Goal: Reduce blood volume/ viscosity & bone marrow activity
- Phlebotomy
  - 300-500mL removed every other day
  - Goal: ↓ hct <45-48%
    - Complication: iron deficiency (seldom symptomatic)

78

Polycythemia: Treatment Goals

- Hydration therapy
- Myelosuppressive agents
  - Hydroxyurea, Myleran & Chlorambucil
- Low-dose Asa
- Agrylin
  - Used to ↓ platelet count and aggregation

79 ↓ rate of blood cell production

Polycythemia: Collaborative Care

- Hydration Therapy
  - Evaluate I & O's
  - Monitor for fluid volume overload
- Assess nutritional status
  - Risk for inadequate nutrition
    - GI symptoms: fullness/ pain/ dyspepsia
- Teach medication side effects

80 Hepatomegaly, organ engorgement

Blood Disorders

The end of part 2

Roslyn G. Nott MSN, RN
Semester 2

81