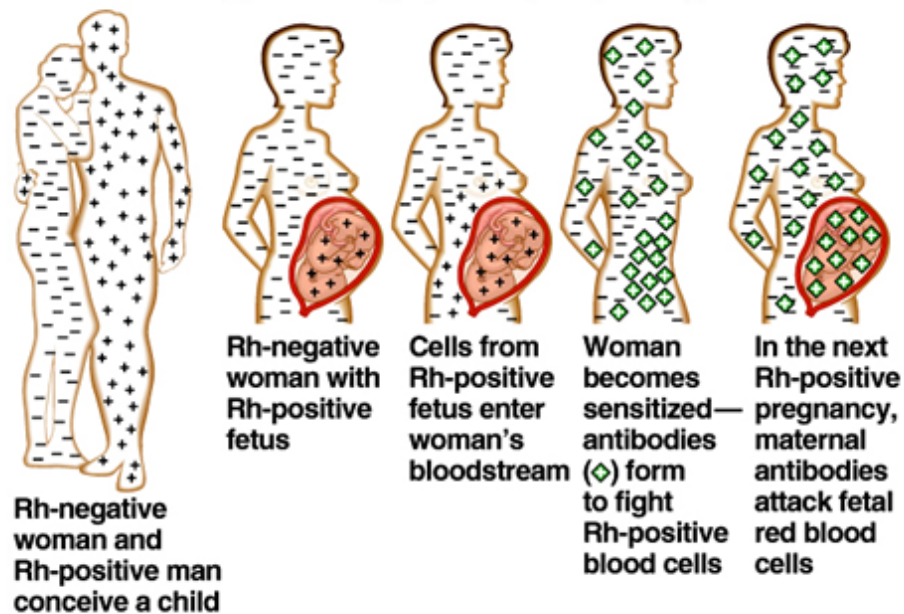


Pediatric Hematology Pathology

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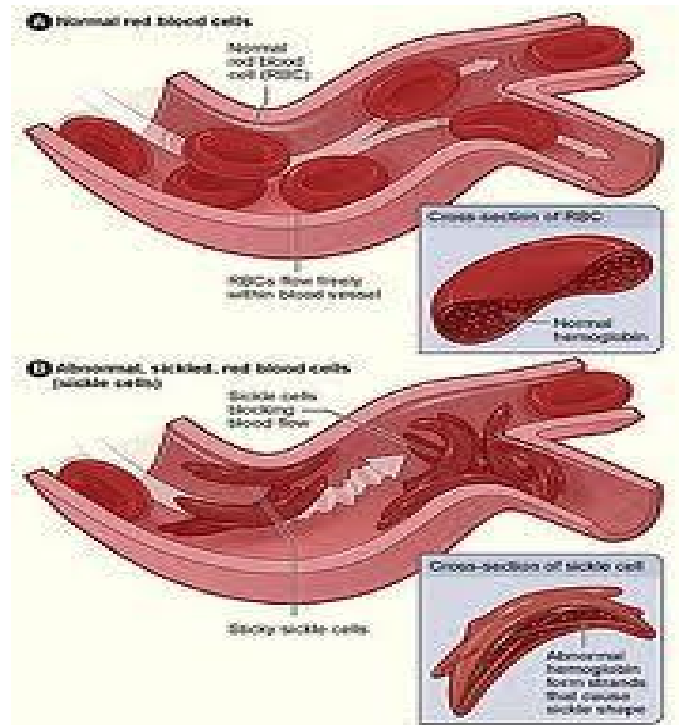
Acquired Disorders of Erythrocytes

- Iron deficiency anemia
 - Most common blood disorder of infancy and childhood
 - Lack of iron intake or blood loss
 - Manifestations:
 - Irritability
 - Decreased activity tolerance
 - Weakness
 - Lack of interest in play
- Acquired congenital hemolytic anemia
 - Hemolytic disease of the newborn (HDN)
 - Maternal antibody directed against fetal antigens
 - ABO incompatibility occurs in 20% to 25% of cases
 - Rh incompatibility occurs in less than 10%
 - Also termed erythroblastosis fetalis
- Hemolytic disease of the newborn (HDN)
 - Manifestations:
 - Anemia
 - Hyperbilirubinemia
 - Icterus neonatorum
 - Kernicterus
 - Glucose-6-phosphate dehydrogenase deficiency (G6PD)



Inherited Disorders of Erythrocytes

- Sickle cell disease
 - Disorders characterized by the presence of an abnormal hemoglobin (HbS)
 - Deoxygenation and dehydration cause the red cells to solidify and stretch into an elongated sickle shape
 - Sickle cell trait
 - Child inherits HbS from one parent and HbA from another
 - Can result in:
 - Vaso-occlusive crisis (thrombotic crisis)
 - Aplastic crisis
 - Sequestration crisis
 - Hyperhemolytic crisis
 - Other forms:
 - Sickle cell-thalassemia disease
 - Sickle cell-HbC disease
- Sickle-cell anemia – results from a defective gene coding for an abnormal hemoglobin called hemoglobin S (HbS)
- This defect causes RBCs to become sickle-shaped in low oxygen situations



Inherited Coagulation and Platelet Disorders

- Hemophilias
 - Serious bleeding disorders
 - Involve gene deletions or point mutations
 - First signs by age 3 to 4 years include episodes of persistent bleeding from minor injuries
 - Hemophilia A (factor VIII deficiency)
 - von Willebrand disease
 - Hemophilia B (factor IX deficiency)
 - Hemophilia C (factor XI deficiency)

Antibody-Mediated Hemorrhagic Disease

- Idiopathic thrombocytopenic purpura
 - Autoimmune or primary thrombocytopenic purpura
 - Platelet destruction rate that exceeds production
 - 70% with antecedent viral disease
- Idiopathic thrombocytopenic purpura
 - Manifestations of bruising and petechial rash
 - Prognosis excellent with 80% regaining normal platelet counts within 6 months after onset

Leukemia

- Most common malignancy of childhood
- 80% to 85% are acute lymphoblastic leukemias (ALL); remainder acute myelogenous leukemia (AML)
- Manifestations of pallor, fatigue, purpura, and fever
- Blast cell is hallmark of acute leukemia
- 5-year survival rate is 80%

Lymphoma

- Non-Hodgkin lymphoma (NHL)
 - Nodular or diffuse (most in childhood are diffuse)
 - Childhood NHL subdivided
 - Large cell (histiocytic)
 - Lymphoblastic
 - Small noncleaved cell (Burkitt or non-Burkitt)
 - Etiology viral, chronic immunostimulation, congenital immunodeficiency syndromes, and genetics
 - Manifestations specific to site involved
 - Mediastinal mass
 - Treatment of chemotherapy and radiation
 - 60% to 80% cure rate
- Hodgkin lymphoma
 - Rare in childhood
 - Infectious mode of transmission
 - Many children with Hodgkin lymphoma demonstrate a high antibody titer to Epstein-Barr virus (EBV)
 - Manifestations :
 - Most common: painless adenopathy with/without fever
 - Others: anorexia, malaise, and weight loss