Alterations of Hematologic Function in Children

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Acquired Disorders of Erythrocytes

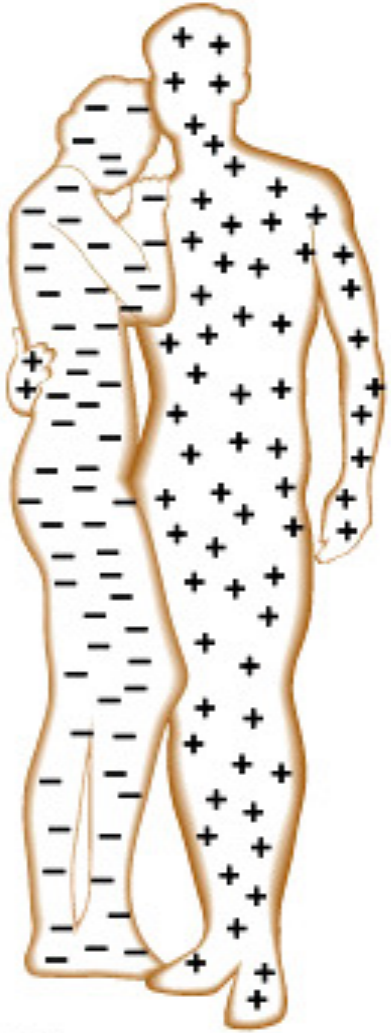
- Iron deficiency anemia
  - Most common blood disorder of infancy and childhood
  - Lack of iron intake or blood loss
  - Manifestations:
    - Irritability
    - Decreased activity tolerance
    - Weakness
    - Lack of interest in play

Acquired Disorders of Erythrocytes

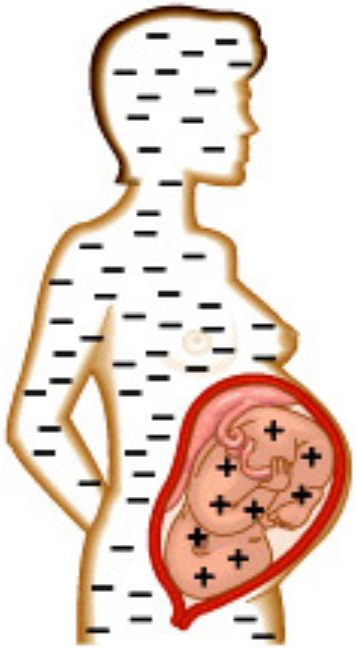
- Acquired congenital hemolytic anemia
  - Hemolytic disease of the newborn (HDN)
    - Alloimmune disease
    - Maternal antibody directed against fetal antigens
    - ABO incompatibility occurs in 20% to 25% of cases
    - Rh incompatibility occurs in less than 10%
    - Also termed erythroblastosis fetalis

Acquired Disorders of Erythrocytes (cont'd)

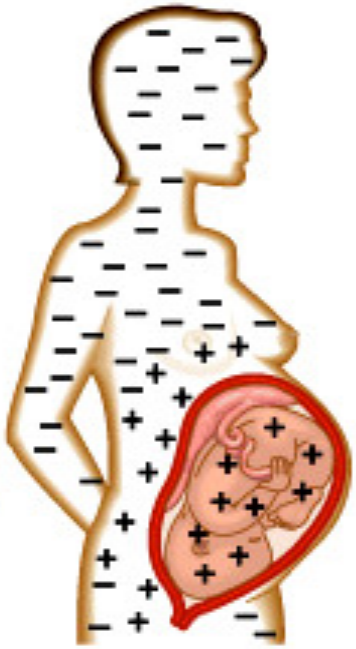
- Hemolytic disease of the newborn (HDN)
  - Manifestations:
    - Anemia
    - Hyperbilirubinemia
    - Icterus neonatorum
    - Kernicterus
    - Glucose-6-phosphate dehydrogenase deficiency (G6PD)



Rh-negative woman and Rh-positive man conceive a child

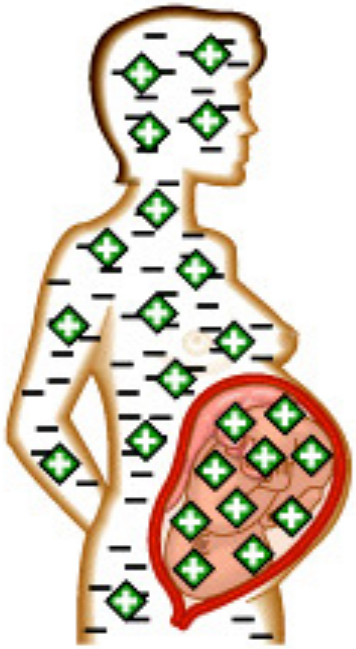


Rh-negative woman with Rh-positive fetus



Cells from Rh-positive fetus enter woman's bloodstream

Woman becomes sensitized—antibodies (◇) form to fight Rh-positive blood cells



In the next Rh-positive pregnancy, maternal antibodies attack fetal red blood cells

Inherited Disorders of Erythrocytes

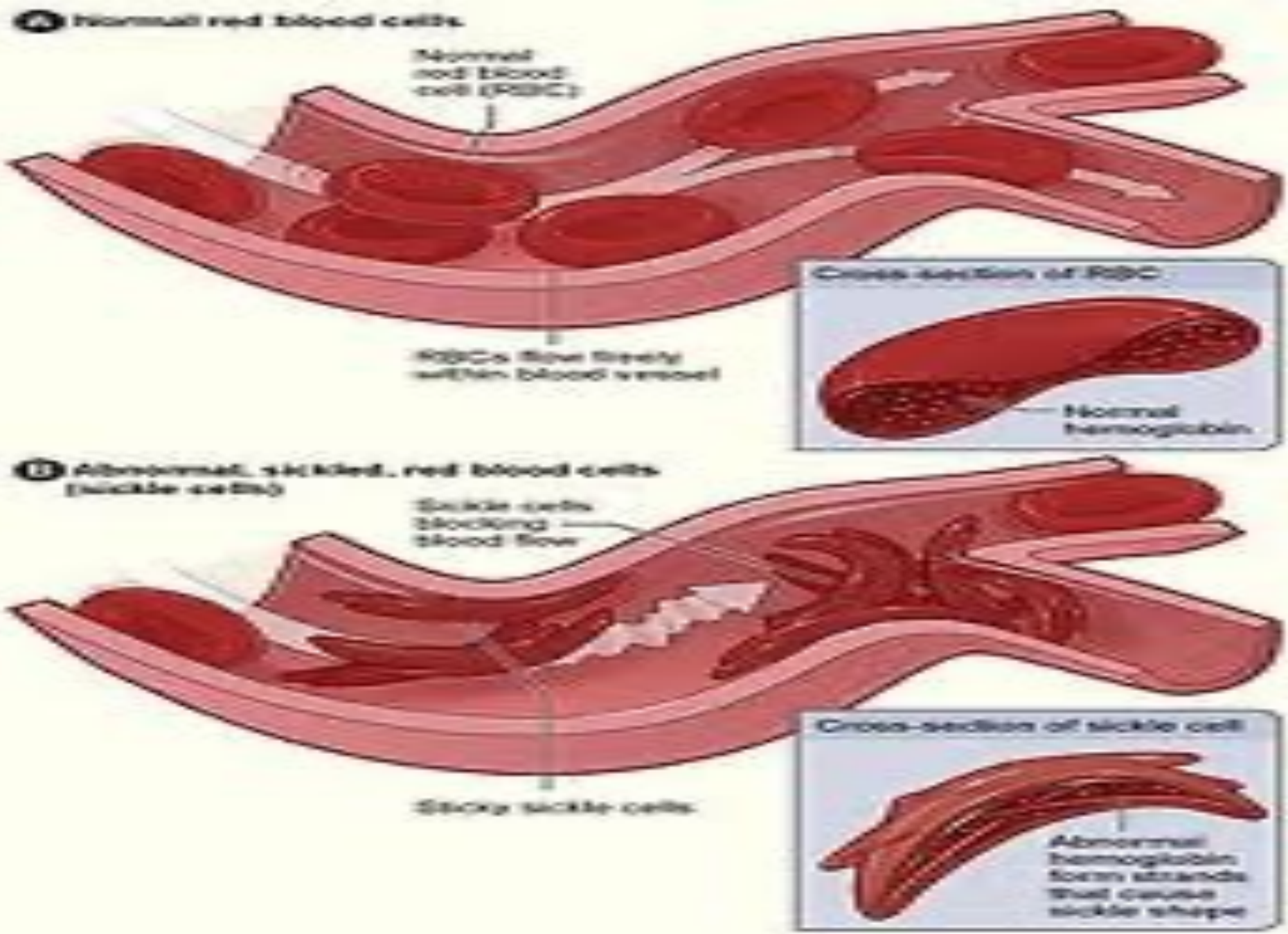
- Sickle cell disease
  - Disorders characterized by the presence of an abnormal hemoglobin (HbS)
  - Deoxygenation and dehydration cause the red cells to solidify and stretch into an elongated sickle shape

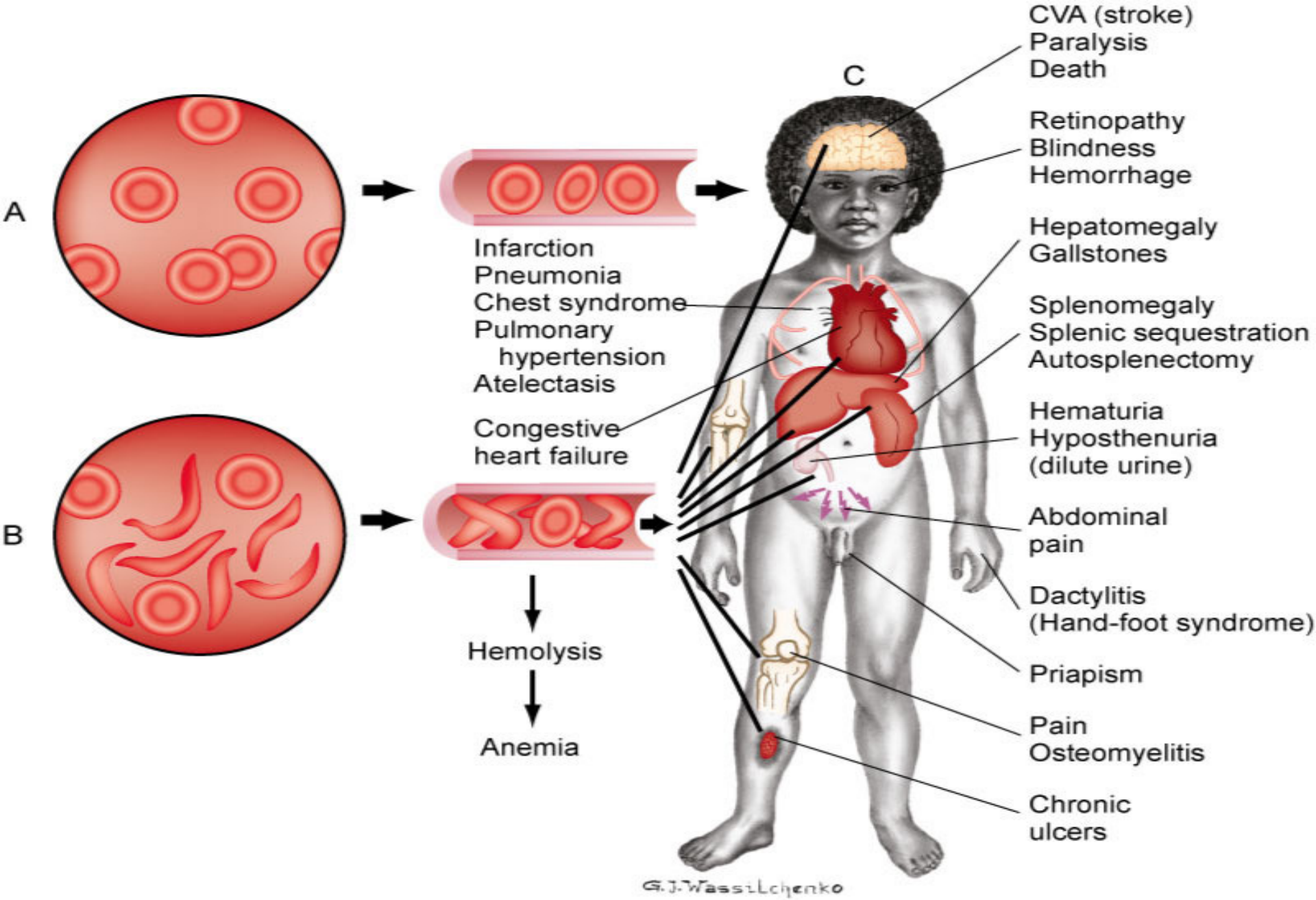
Inherited Disorders of Erythrocytes (cont'd)

- Sickle cell disease (cont'd)
  - 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

Anemia: Abnormal Hemoglobin

- 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


A Normal red blood cells
Normal red blood cell (RBC)
RBCs flow freely within blood vessel
Cross-section of RBC
Normal haemoglobin
B Abnormal, sickled, red blood cells (sickle cells)
Sickle cells blocking blood flow
Sticky sickle cells
Cross-section of sickle cell
Abnormal haemoglobin form strands that cause sickle shape



(**A** and **B** adapted from Hockenberry MJ et al, editors: *Wong's nursing care of infants and children*, ed 8, St Louis, 2007, Mosby.)

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

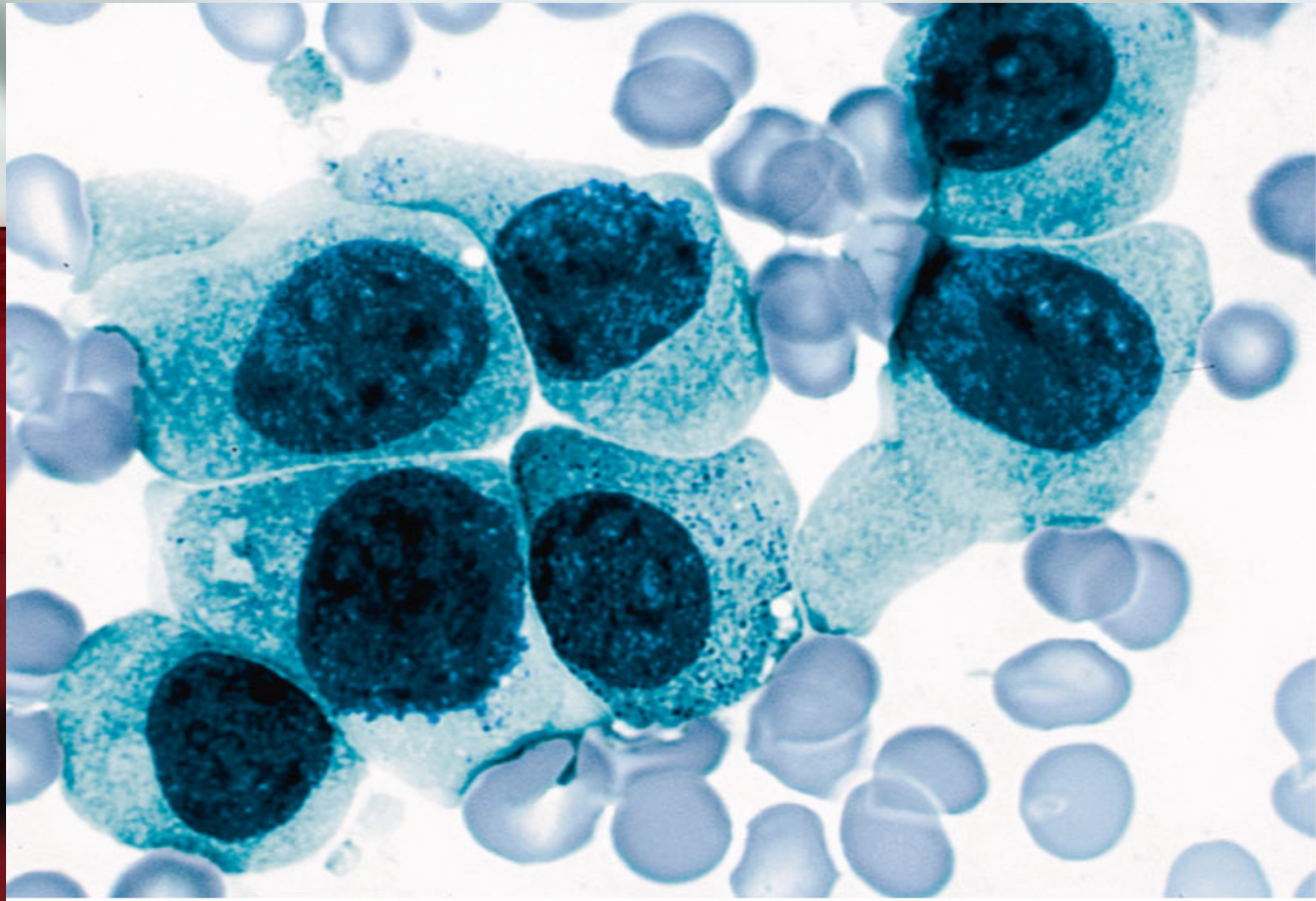
- Idiopathic thrombocytopenic purpura
  - Autoimmune or primary thrombocytopenic purpura
  - Platelet destruction rate that exceeds production
  - 70% with antecedent viral disease

Antibody-Mediated Hemorrhagic Disease (cont'd)

- 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%



(From Damjanov I, Linder J, editors: *Anderson's pathology*, ed 10, St Louis, 1996, Mosby.)

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)

Lymphoma (cont'd)

- Non-Hodgkin lymphoma (NHL) (cont'd)
  - 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

Lymphoma (cont'd)

- 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