Aplastic Anemia

Aplastic Anemia

- Reduction of erythroid, granulocytic/monocytic, and megakaryocytic cell lines in the bone marrow and their progeny in the peripheral blood.

Table

Table 4-1 Types of Constitutional and Acquired Aplastic Anemia and Red Cell Aplasia

Constitutional aplastic anemia	Constitutional red cell aplasia
Fanconi's anemia	Diamond-Blackfan anemia
Dyskeratosis congenita	
Shwachman-Diamond syndrome	
Acquired aplastic anemia	Acquired red cell aplasia
Idiopathic	Transient erythroblastopenia of childhood
Secondary to drugs, toxins, infections, and miscellaneous disorders/conditions	Parvovirus infection* (usually transient)
	Idiopathic pure red cell aplasia
	Sustained pure red cell aplasia secondary to neoplasms, immune disorders, infections, and drug treatment
Paroxysmal nocturnal hemoglobinuria (clonal)	

*Parvovirus infection may be sustained in an immunocompromised host.

Clinical Findings

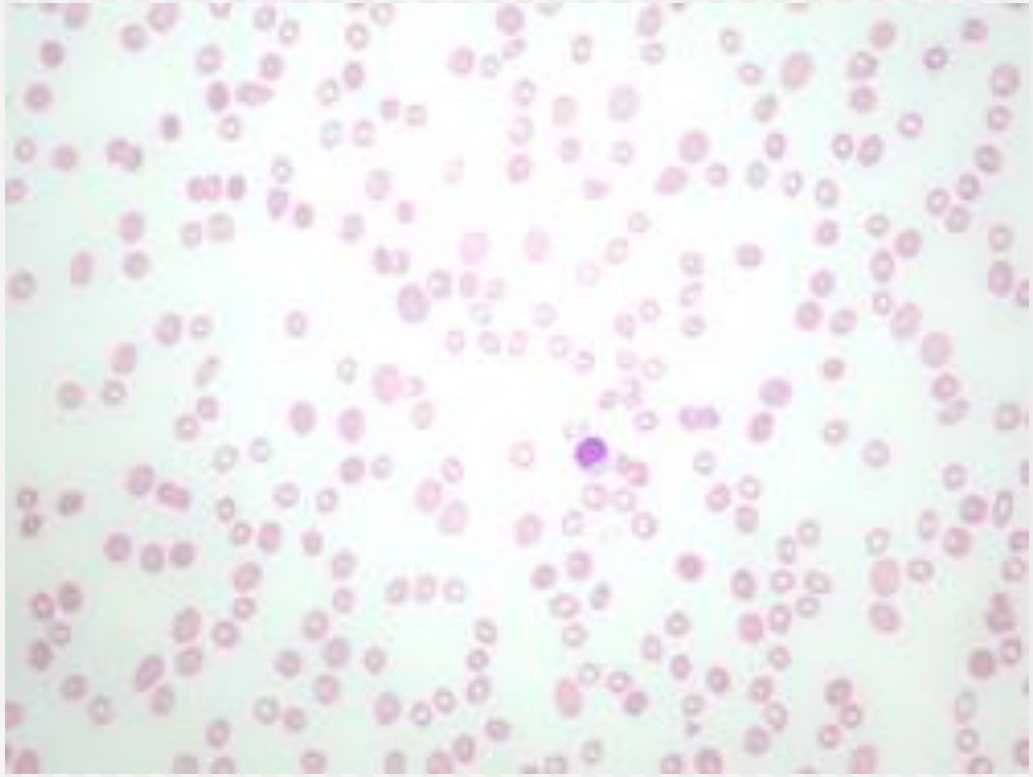
- Anemia- weakness, fatigue, pallor
- Granulocytopenia- fever, infection
- Thrombocytopenia- petechiae, ecchymosis, mucosal bleeding
- No hepatosplenomegaly or lymphadenopathy

- Phenotypic abnormalities- bony defects, mental retardation, skin/nail abnormalities

Blood Findings

- Pancytopenia
  - Normocytic/normochromic anemia
  - RBC, platelets, granulocytes have normal morphology
- Elevated erythropoietin
- Decreased reticulocytes

Pancytopenia



Bone Marrow Findings

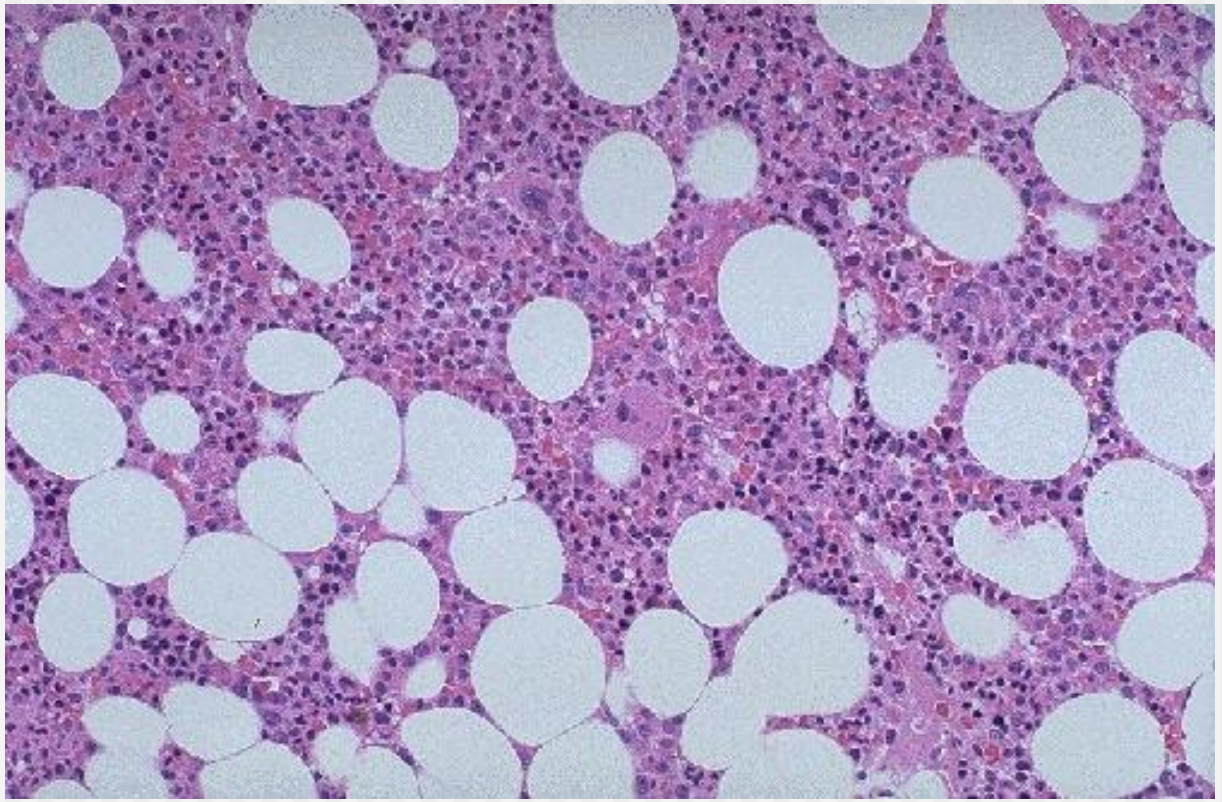
- Hypocellular bone marrow
  - Rare residual hematopoietic elements
  - Replaced by fat
- What is hypocelluar?

Bone Marrow Cellularity

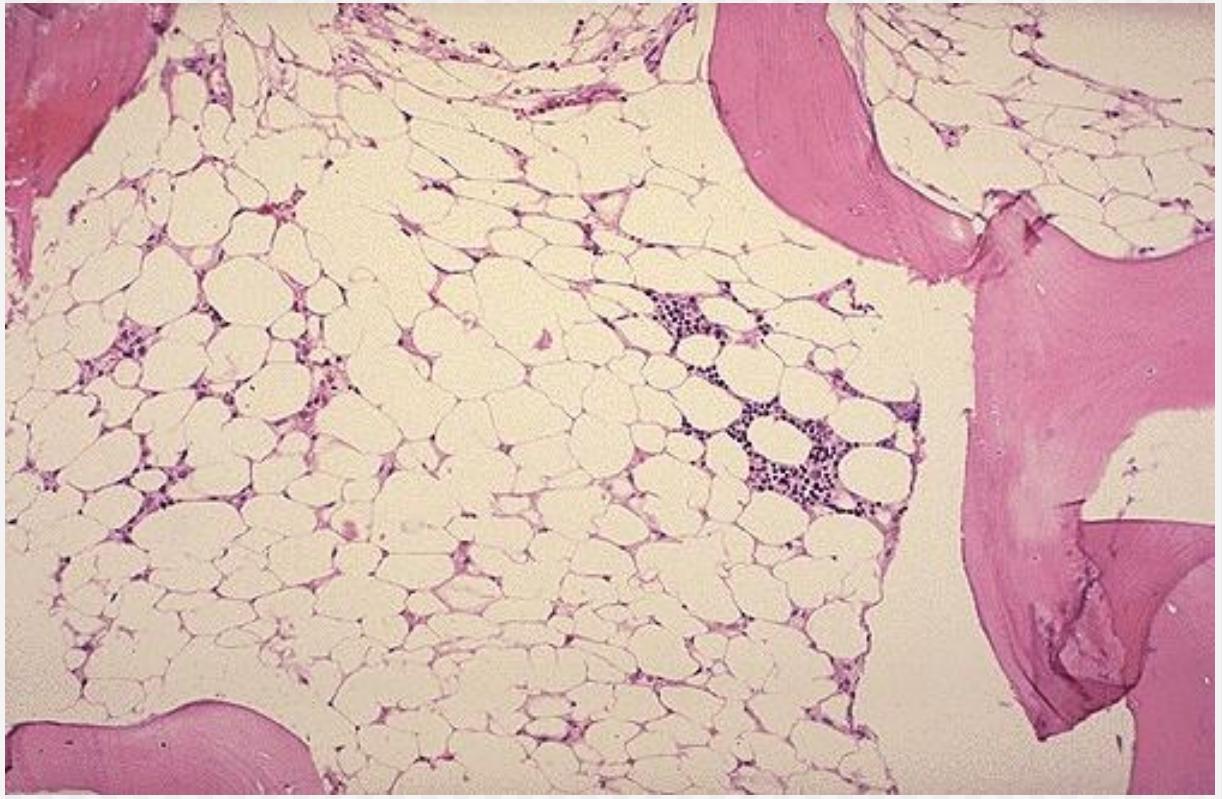
- Newborn 75-100%
- Adolescent 50-90%
- Adult 30-80%
- >65 years old 20-50%

- Hypocellular: <20%

Normocellular Bone Marrow



Hypocellular Bone Marrow



Aplastic Anemia- Causes

- Constitutional- including Fanconi's
- Idiopathic
- Secondary
  - Chemical/drug
  - Radiation
  - Infection
  - Others

Fanconi's Anemia

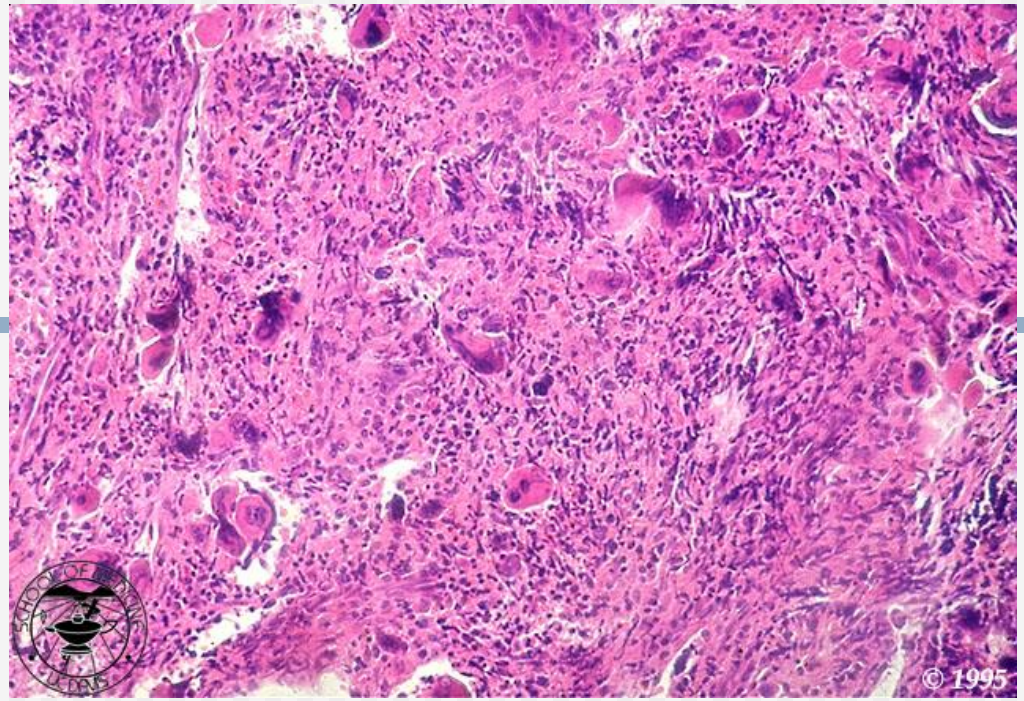
- 2/3 of constitutional aplastic anemia
- Autosomal recessive
  - DNA repair defect
- Phenotypic abnormalities- bone, skin, mental retardation
- Aplasia develops by mid childhood
- Progression to AML

Chemical/Drug

- Dose-dependant
  - Chemotherapy
  - Benzene
- Idiosyncratic
  - CHLORAMPHENICOL, anticonvulsants, sulfonamides, gold, NSAIDs

Ionizing Radiation

- Aplastic anemia- acute
- Long-term effects are myelofibrosis and leukemia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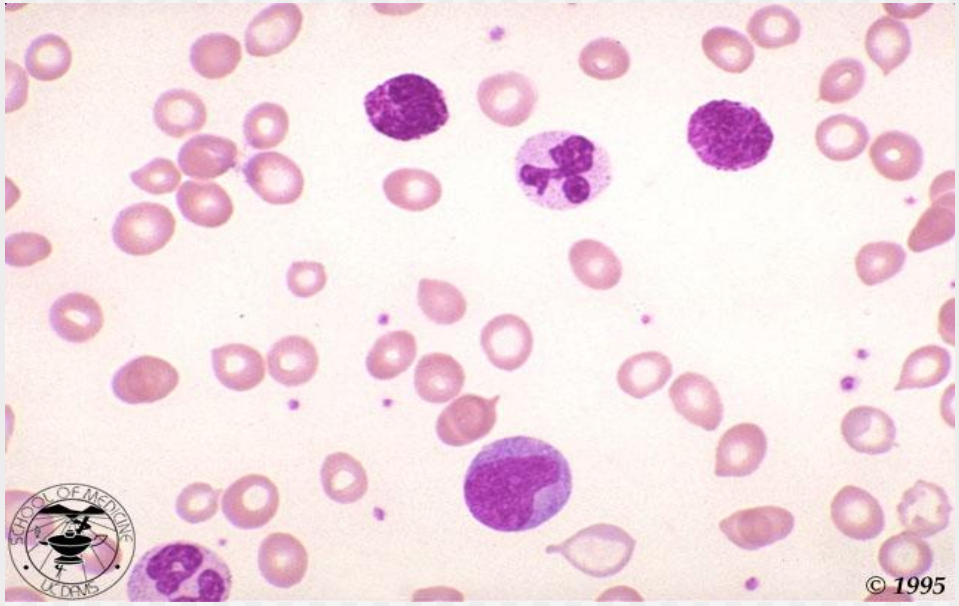
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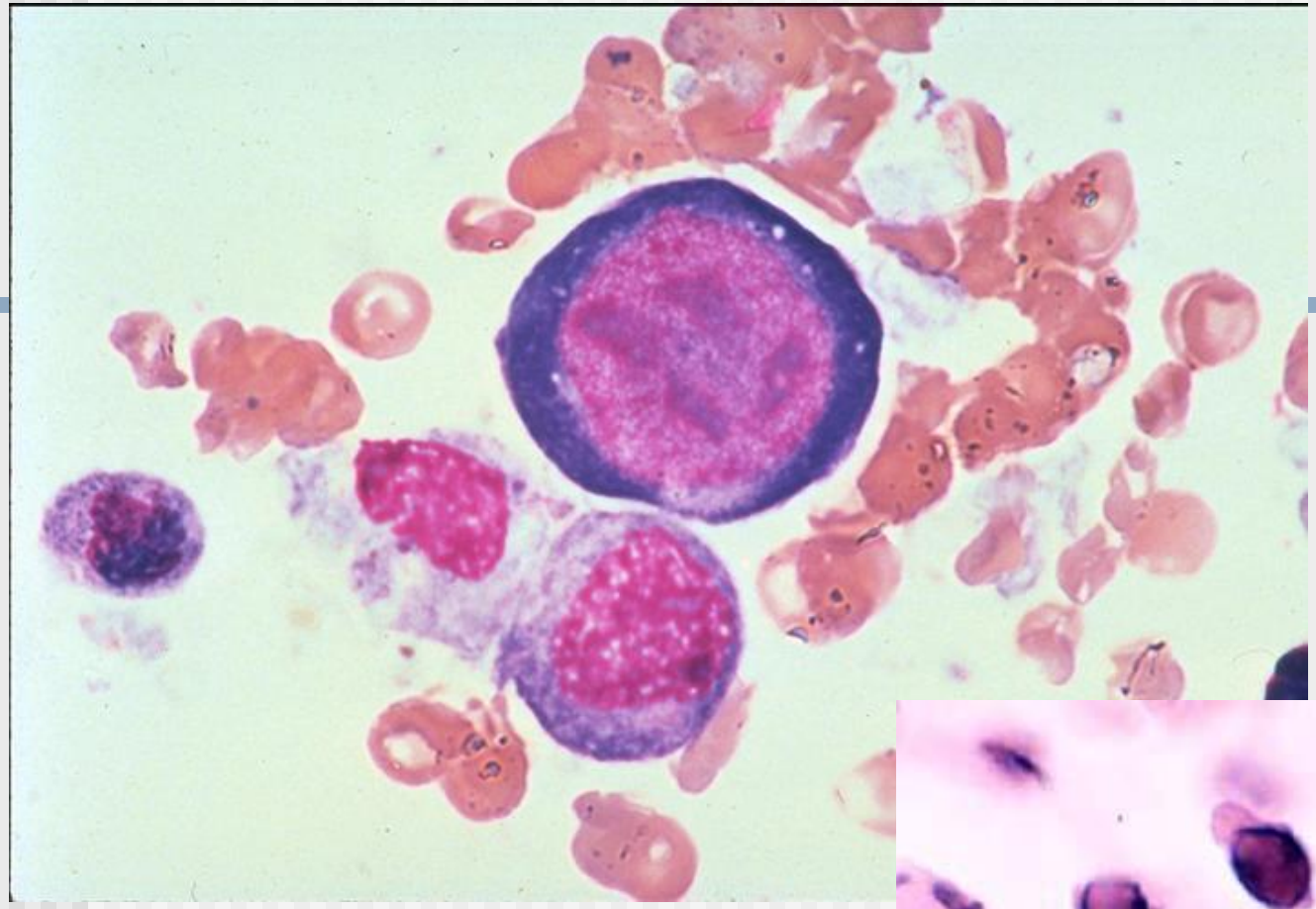
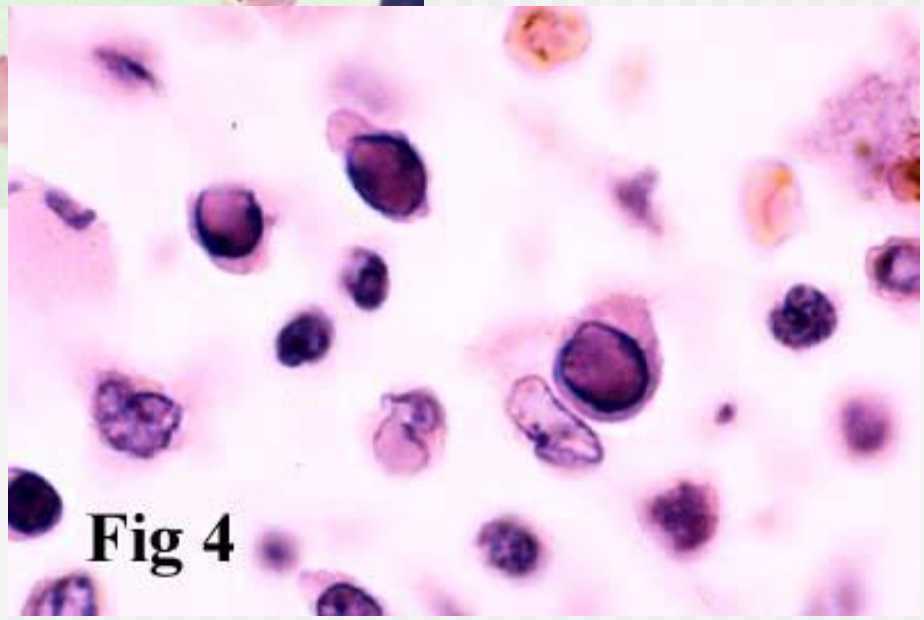
Infections

- Hepatitis
  - Rare, often fatal
  - Non-A, non-B, non-C, non-G
  - Usually 6 wks after clinical symptoms
  - No relation to severity of illness

Parvovirus B19

- Usually red cell aplasia
  - Selective cytotoxic invasion of erthryoblasts
- Mild reduction in granulocyte and megakaryocyte production
- Chronic hemolytic anemia pts. at risk
  - Sensitive to cessation in RBC production
  - Transient aplastic crisis
- Self limited





Parvovirus B19

Other Causes

- Transfusion-associated Graft-vs.-Host disease
- Eosinophilic Fasciitis

Treatment

- Bone marrow transplant
- Androgens, immune modulation, cytokine therapy
- Transfusions

…Finally

- Pancytopenia without reticulocytosis
- Bone marrow cellularity <20%
- Hypoplasia not dysplasia
- Causes
  - Idiopathic most common
  - Constitutional, drugs, infections