Anemia (3).ms4.25.Oct.15

Hemolytic Anemia

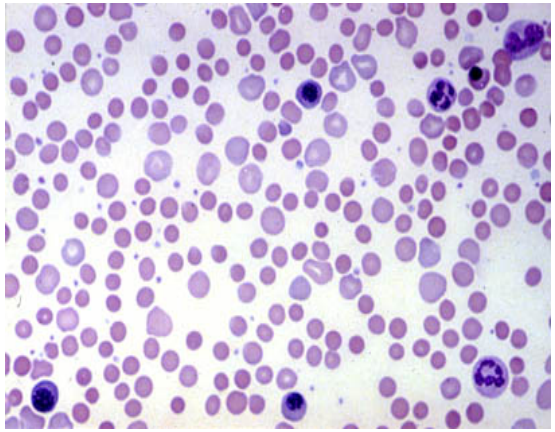
Abdallah Abbadi

Case 3

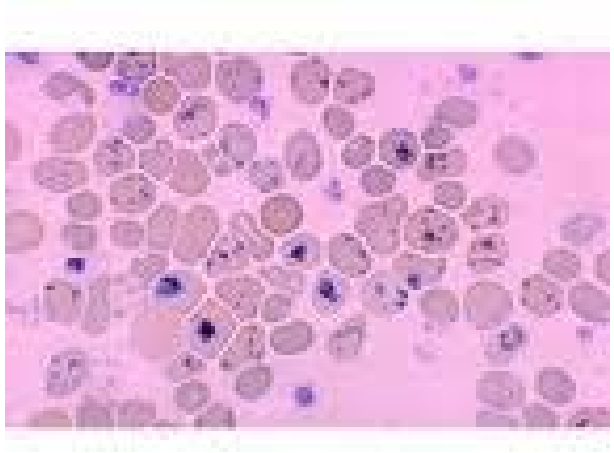
24 yr old female presented with “anemia syndrome” and jaundice. She was found to have splenomegaly.

Hb 8, wbc 12k, Plt 212k, retics© 12%, LDH 1400, bilirubin 7mg/dl,d 2.5mg/dl, DAT +3.Bld film spherocytosis, polychromasia.

Bld film

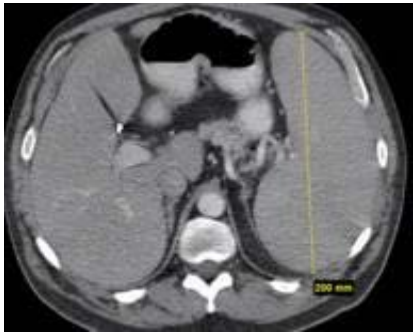


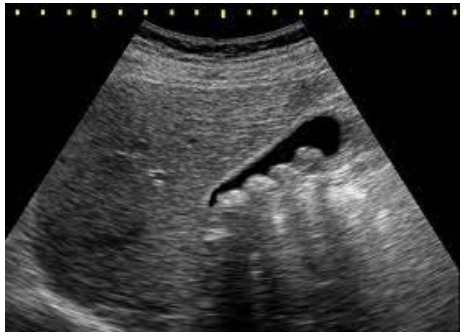
Supravital stain(retics)

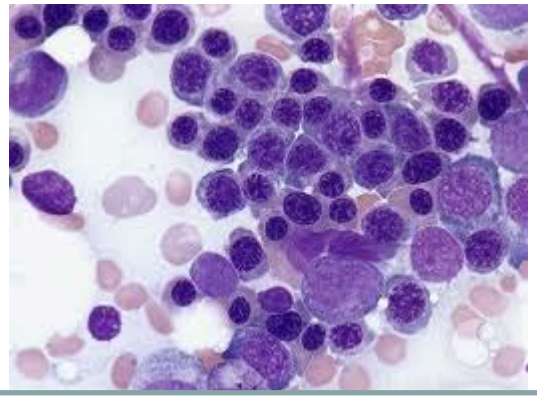


Case 3

CT Abdomen AbdominalUS BM aspirate







BM:erythroid hyperplasia with megaloblastoid changes

Diagnosis: AIHA. Treated with steroids + folic acid, complete response, but 9 months later had NHL.

Hemolysis= RBC destruction= Shortend RBC Survival with or without anemia

Hemolytic Anemias –Classification

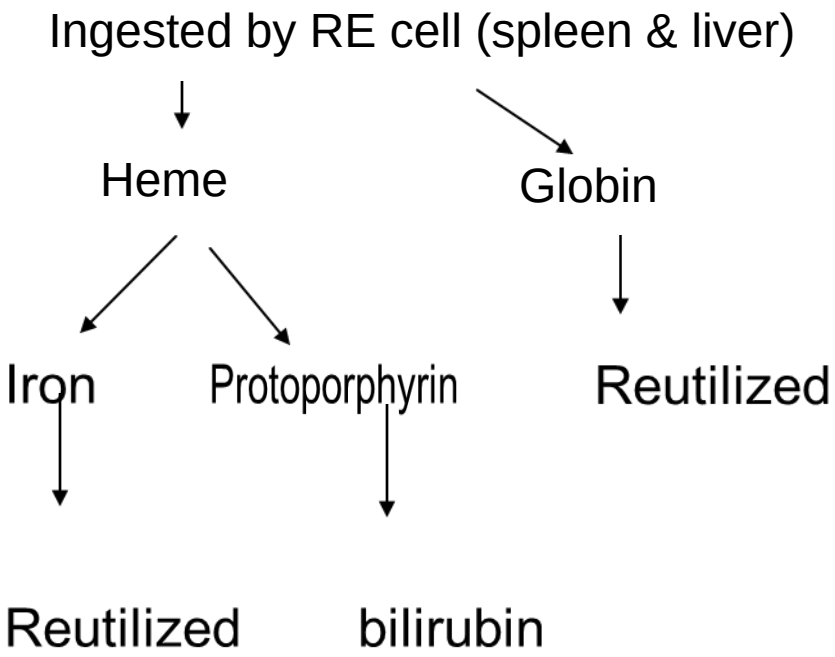
- **By sites of red cell destruction: intra *v* extravascular**

- **Acquired (immune, Non-immune).**

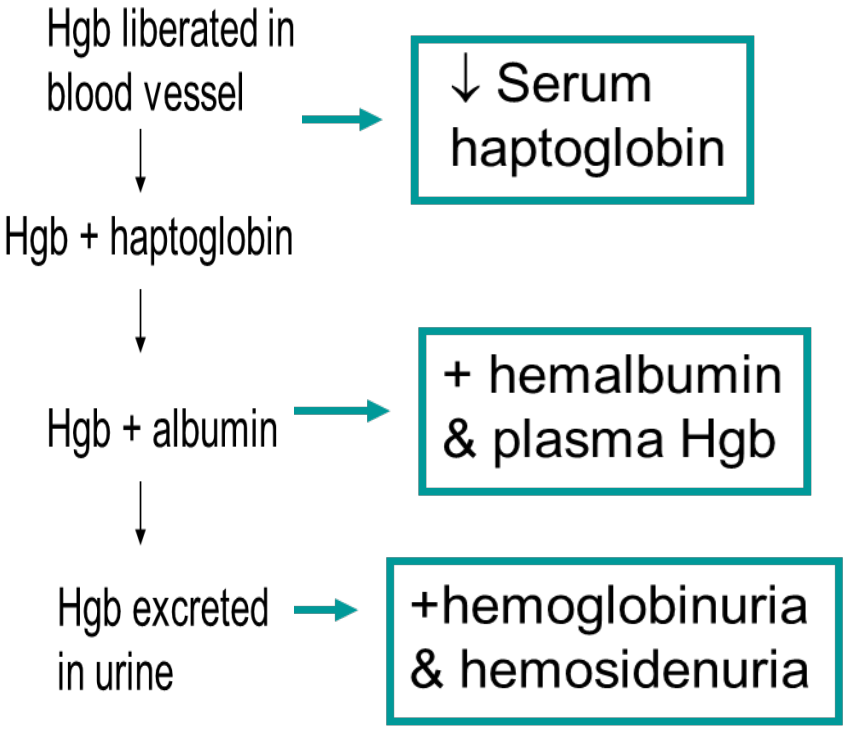
v **congenital (membrane: HS, Enzymopathies: G6PD def/PK, Hb-pathies: Thal, ss)**

- **By mechanism of red cell damage:**

Extravascular Hemolysis



Intravascular Hemolysis



Hemolysis

Evidence for increased red cell production

- **In the blood:**

– Elevated reticulocyte count (corrected/RPI)
– Circulating NRBCs may be present

- **In the bone marrow:**

– erythroid hyperplasia
– reduced M/E (myeloid/ erythroid erythroid ratio)

- **In the bone:**

– Deforming changes in the skull and long bones (“ frontal bossing ”)

General Clinical Features

1- Anemia syndrome
2- Splenomegaly
3-gallstones.
4- Dark urine (tea-colored or red)
5- Patients may have chronic ankle ulcers.
6- Aplastic crises associated with Parvovirus B19, may occur
7- Increased requirement for folate

Gallbladder stones/ biliary/ pigment stones

Parvovirus B19

- **Non-encapsulated DNA virus.**
- **Infects and lyses RBC precursors in marrow, causing 7-10d cessation of erythropoiesis.**
- **Normal individuals have no significant hematologic effect, since RBCs have normal life span.**
- **In pts with hemolytic anemias , loss of red cell production causes Aplastic Crisis**

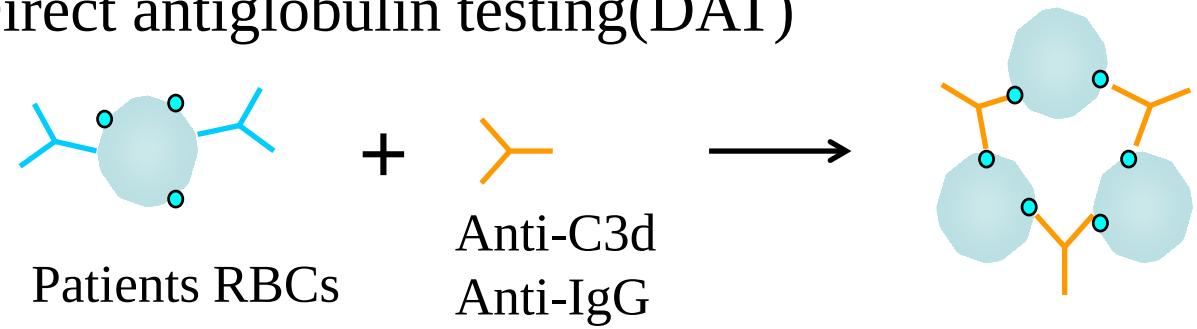
Autoimmune Hemolytic Anemia

- Warm antibodies (IgG-mediated)
  - Primary 45%
  - Secondary 40%
    - Lymphoproliferative disease
    - Connective tissue disease
    - Infectious disease
  - Drug-induced 15%

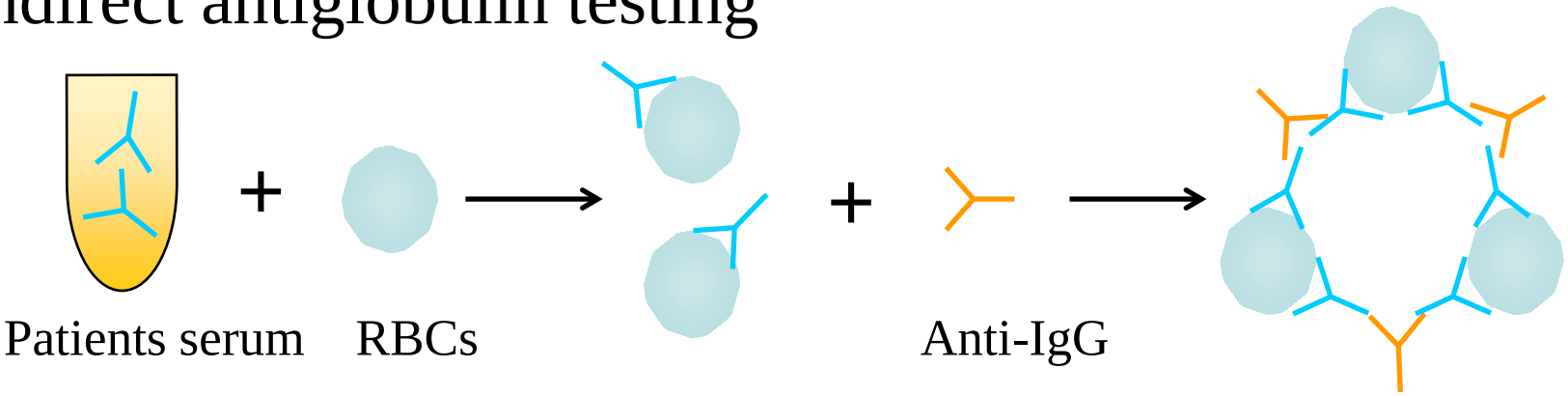
- Laboratory testing
  - Normocytic/macrocytic anemia
  - Peripheral smear - spherocytosis

Anti-Globulin (Coombs) Testing

Direct antiglobulin testing(DAT)



Indirect antiglobulin testing



Treatment of Autoimmune Hemolytic Anemia (Warm Antibody type)

- Treat underlying disease if indicated

- Prednisone (1 mg/kg/day for two weeks, then taper)

- Splenectomy ??

- Other
  - Immunosuppressive agents
  - IVIG

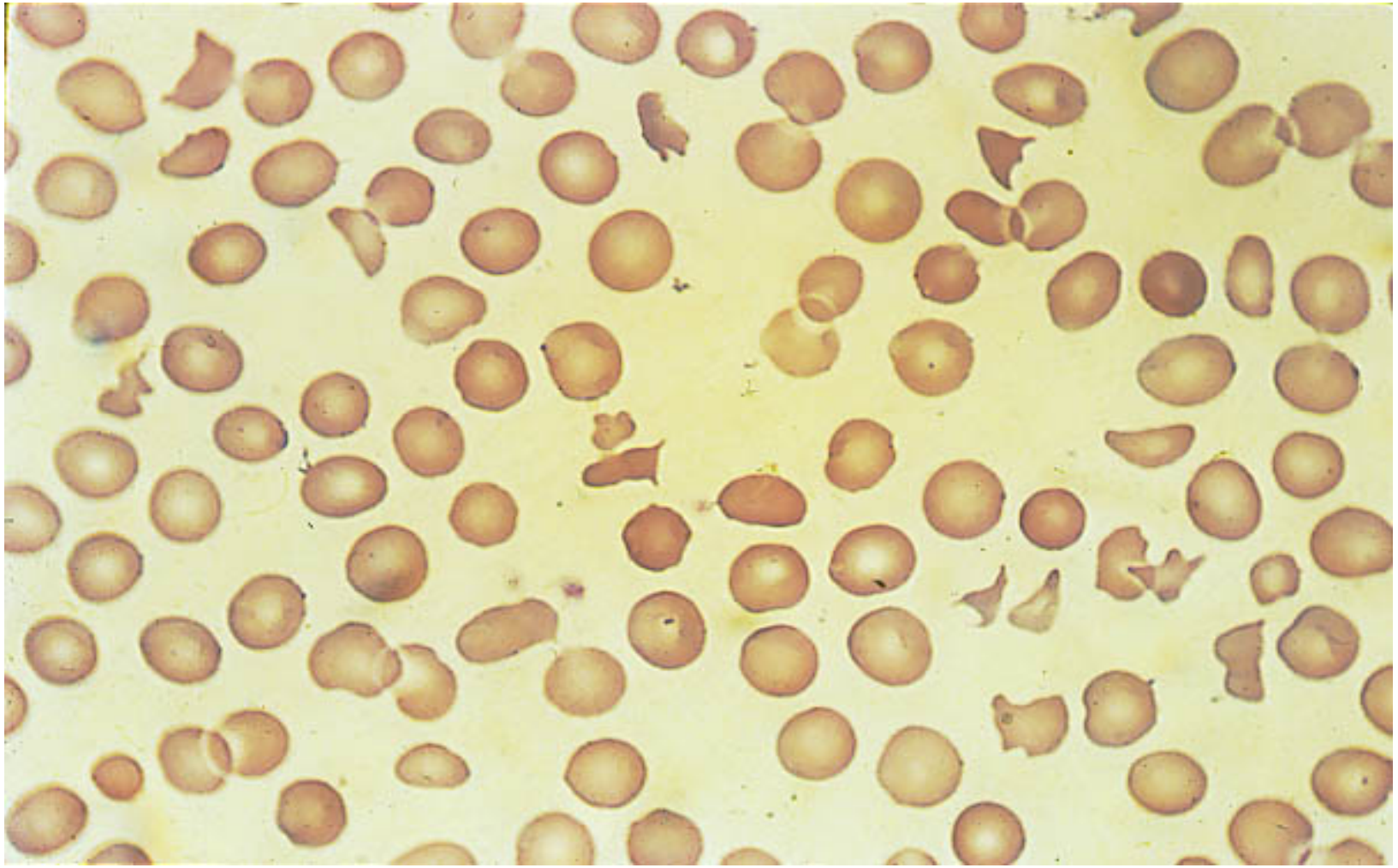
Hemolytic Anemia with Intravascular Hemolysis

- Mechanical damage (Microangiopathic hemolytic anemia)
- Chemical damage (Burns)
- Infection (Malaria or Babesiosis)
- Transfusion reaction (ABO incompatibility)

Differential Diagnosis of Microangiopathic Hemolytic Anemia

- Thrombotic thrombocytopenic purpura (TTP)
- Hemolytic uremic syndrome (HUS)
- Disseminated intravascular coagulation (DIC)
- Vasculitis
- Malignant hypertension
- Metastatic neoplasm with vascular invasion
- Preeclampsia/HELLP syndrome of pregnancy

Schistocytes: Microangiopathic Hemolytic Anemia



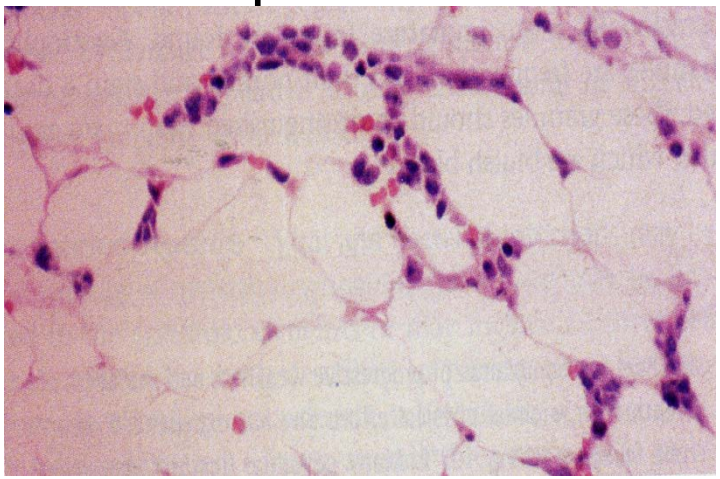
Case 3 B

19 yr old male presented with “anemia syndrome”, fever and easy bruising. No splenomegaly

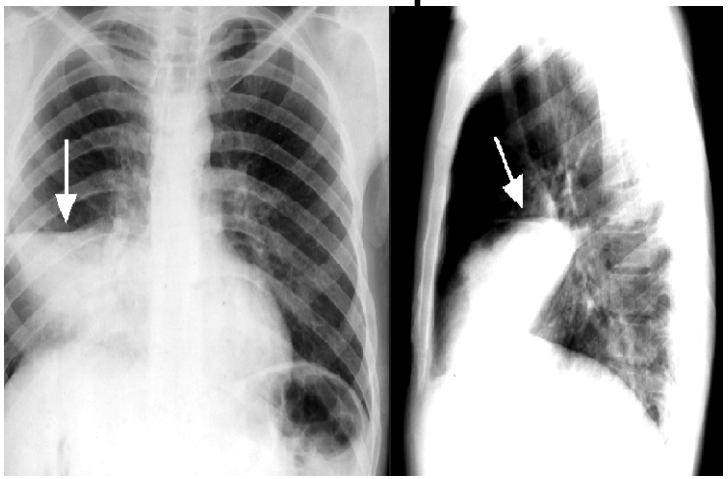
Hb 6 g/dl,WBC 1500 : N10%, L 80%, others 10%.

Retics© 0,001%.MCV 105fl,Plt 20k.

BM/ Trephine



CXR/lobar pneumonia



APLASTIC ANEMIA

- Aplastic anemia is a severe, life threatening syndrome in which production of erythrocytes, WBCs, and platelets has failed.
- Aplastic anemia may occur in all age groups and both genders.
- The disease is characterized by peripheral pancytopenia and accompanied by a hypocellular bone marrow.

APLASTIC ANEMIA

- The primary defect is a reduction in or depletion of hematopoietic precursor stem cells with decreased production of all cell lines
  - 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.

Causes of Bone Marrow Failure

Acquired

- Idiopathic
- PNH

Secondary

- Drugs
- - radiation
- Viruses

Inherited

- Fanconi anemia
- Diamond-Blackfan Anemia
- Other rare conditions

Clinical manifestations of AA

- »Anemia syndrome
- »Neutropenia syndrome
- »Thrombocytopenia syndrome
- »Combination of the above

Presenting Symptoms of Aplastic Anemia

Symptoms	Number of Patients
Bleeding	41
Anemia	27
Bleeding and anemia	14
Bleeding and infection	6
Infection	5
Routine examination	8
Total	101

Classification of aplastic anemia

1. Severe aplastic anemia is defined if at last two of the following criteria are present:

- ANC < 0.5
- PLT < 20
- RTC < 1%

Hypoplastic bone marrow (less than 25%) on biopsy

2. Very severe aplastic anemia

- criteria as above but ANC < 0.2

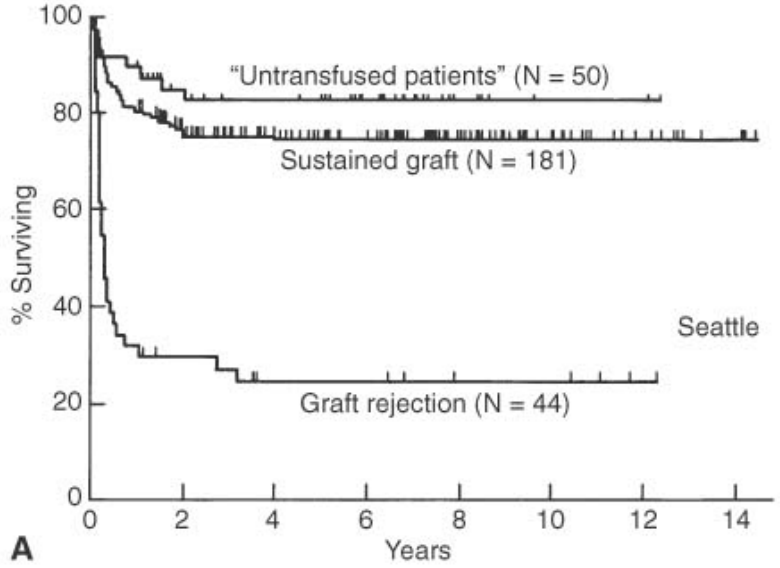
3. Non-severe aplastic anemia.

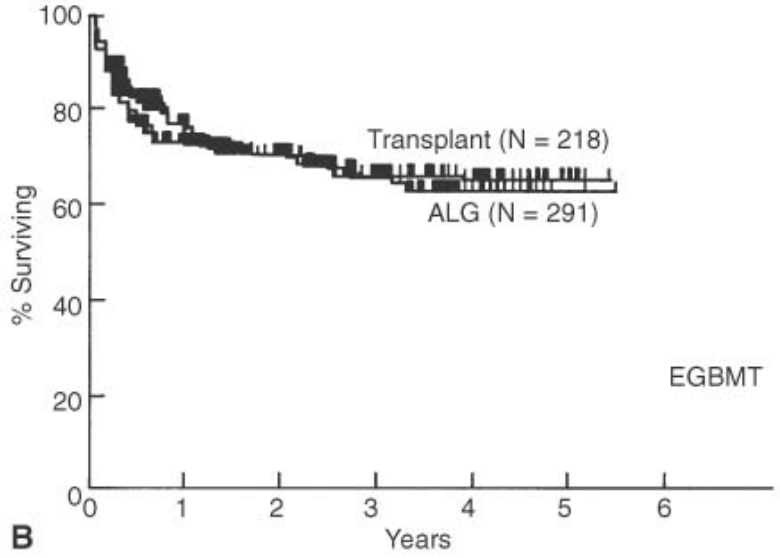
Treatment of AA

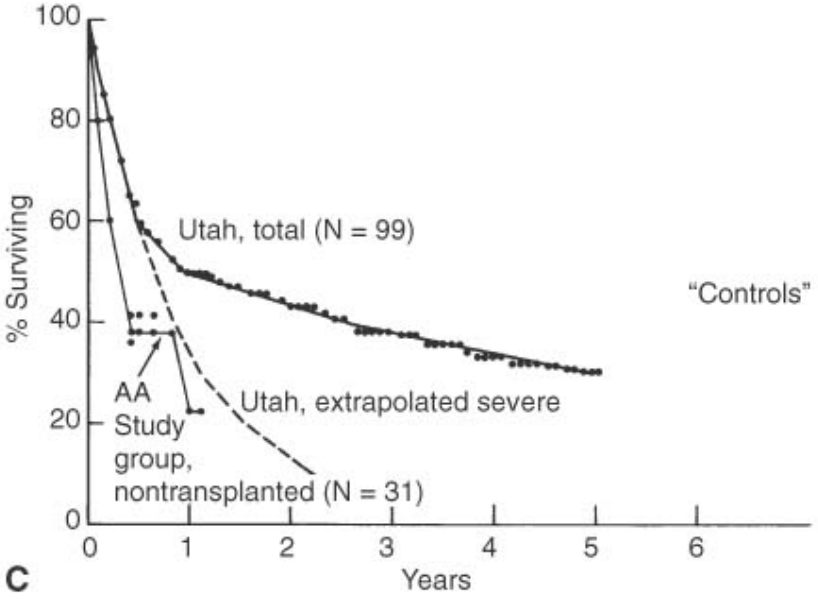
- Remove causative agent, if known
- Supportive care
  - RBC transfusions
  - Treat infections
  - Treat Bleeding
- **Bone marrow transplant**
- Immune suppression
  - CSA
  - ATG

Combination of the above

Survival of Patients with AA







RELATED DISORDERS

1- Disorders in which there is peripheral pancytopenia, but the bone marrow is normocellular, hypercellular, or infiltrated with abnormal cellular elements (Myelopthesic anemia)

- replacement of bone marrow by fibrotic, granulomatous, or neoplastic cells

2- Pure red Cell aplasia

3- Myelodysplastic syndrome (MDS)