

### Thalassemia

**Autosomal recessive** genetic disorder of inadequate production of normal hemoglobin. found in mediterranean ethnic groups.

#### Clinical Manifestation

Asymptomatic | Major retardation | Life threatening

Splenomegaly, Heptomegaly

#### Pathophysiology

Genetic defect, synthesis of one of a or b globin chains, production of abnormal Hgb and RBC, Hemolysis, Anemia

#### Collaborative Care

- No specific drug or diet are effective for treatment
- Minor (hetero-beta) : body adapts to low Hgb
- Major (Homo-beta) : Blood transfusions with IV deferoxamine (used to remove excess iron from the body)

### Polycythemia

A net increase in the total number of Red blood cells. **Overproduction may be due to:**

- myeloproliferative syndrome in bone marrow
- reaction to chronically low oxy lvls or malignancy

#### Pathophysiology

Myeloproliferative due to hypoxia, the stem cell grow uncontrollably, BM becomes Hypercellular, Blood becomes thick to sluggish circulation, BM becomes fibrotic.

#### Complications

- Increased viscosity of blood
- Hemorrhage and thrombosis

#### Treatment

- Phlebotomy
- Myelosuppressive agents : Intron A, Agrylin, Aspirin

### Idiopathic Thrombocytopenia Purpura (ITP)

Disorder of decreased platelets (below 150,000)

#### Causes

- Low production of platelets
- Increased breakdown of platelets

#### Symptoms

- Bruising, Nosebleeds, Petechiae

### Immune Thrombocytopenic Purpura

- Abnormal destruction of circulating platelets
- Autoimmune disorder
- Destroyed in hosts' spleen by macrophages

### Thrombotic Thrombocytopenic Purpura

- Increase agglutination of platelets, form microthrombi

#### Diagnostic Studies

- Platelet count, Hgb/Hct, prothrombin time (PT)
- Activated partial thromboplastin time (aPTT)

#### Treatments

- Corticosteroids (Bethamethasone, prednisone, predlone)
- Plasmapheresis, Splenectomy, platelet transfusion

