**Essential/Primary Thrombocythemia**
- primary thrombocythemia is defined by marked elevation of platelet count of $600 \times 10^9/L$.
- Commonly appearing in older age group.
- It is a diagnosis of exclusion and any known cause of reactive thrombocytosis must be considered.
- Must be differentiated from hemolytic anemia, concurrent chronic inflammation or infection or presence of non-hematogeneous neoplasia.

**Clinical Features**
- Patients with high platelet count complain of headache, dizziness, visual disturbance, syncope, paresthesia, and acrocyanosis.
- Budd-Chiari syndrome (caused by occlusion of hepatic veins) or skin necrosis.
- Splenic enlargement.
- Arterial and deep venous thrombosis.
- Splenic infarction.
- Throbbing and burning pain in hands and feet.
- Hemorrhage complications – from oral and nasal mucus membrane.
- Transient ischemic attacks.

**Diagnosis**
- Differentiation of ET from other MPDs and from all of the many causes of reactive thrombocytosis (hemoglobin less 13 g/dL, platelet count greater than $600 \times 10^9/L$, normal erythrocyte mass, absences of collagen marrow fibrosis and there no splenomegaly).

**Peripheral Blood and Bone Marrow**
- Early phase exhibits thrombocytosis with a greater than normal variation in platelet size and shape, including changes such as giantism, agranularity, pseudopod formation and atypical shape.
- Commonly platelets are in clusters and tend to accumulate in thin edge of blood film.
- Increase in segmented neutrophils.
- Basophils are not increased.
- Erythrocytes are normocytic normochromic unless iron deficiency is present secondary to excessive clinical bleeding.
- Bone marrow -early phase demonstrates megakaryocytic hypercellularity, clustering of megakaryocytes, and increased average megakaryocyte size, associated with nuclear hyperlobulation and density.
- Reticulin fibers may be increased.
- Increased smaller and less mature megakaryocytes.

**Therapy and Prognosis**
- Single-drug chemotherapy is appropriate.
- Since most patients are elderly multi-drug chemotherapy or bone transplant are not feasible.
- Hydroxyurea is used in young patients.

**Agnogenic Myeloid Metaplasia/Chronic Idiopathic myelofibrosis**
- Myelofibrosis refers to deposition of excess collagen in the bone marrow.
- It was first described in 1879 and has more than 30 names.
- It is a clonal MPD in which there is ineffective hematopoiesis.
- There is marrow hypercellularity.
- Increased megakaryocytes.

**Marrow fibrosis**
- Immature granulocytes.
- Erythroblast in peripheral blood.
- Poikilocytosis (including teardrop-shaped erythrocytes).
- Splenomegally.
- Megakaryocytes are enlarged and have pleomorphic nuclei and coarse segmentation and areas of hypochromia.

**Myelofibrosis**
- Comprises three of the five major types of collagen: Types I, III and IV.
- Approximately 30% of patients, biopsies may show no fibrosis.
- Increase in these collagens is secondary to increased release of fibroblastic growth factors.
- Due to increased marrow fibrosis there is expansion of marrow sinuses and vascular volume with increased rate of blood flow.
- Fibrosis of marrow is not sole criterion for diagnosis of AMM. It could be due to reparative response to injury from -benzene, ionizing radiation or immunologic-mediated injury.

**Extramedullary Hematopoiesis**