Myeloproliferative Disorders

- **Myeloproliferative disorders** is the name for a group of conditions that cause blood cells -- platelets, white blood cells, and red blood cells -- to grow abnormally in the bone marrow. [http://www.umm.edu/altmed/articles/myeloproliferative-disorders-000114.htm#ixzz2AdUJETeJ](http://www.umm.edu/altmed/articles/myeloproliferative-disorders-000114.htm#ixzz2AdUJETeJ)

- **Myeloproliferative Disorders** are:
  - chronic diseases
  - caused by proliferation of bone marrow stem cells which leads to excess production of one or more types of blood cells

- **Myeloproliferative Disorders include:**
  - Polycythemia Vera (Pvera)
  - Thrombocytosis
  - Myelofibrosis
  - Chronic Myelogenous Leukemia (CML) – also called Chronic Granulocytic Leukemia (CGL)

- **Splenomegaly and Infarction of the Spleen**
  - Splenomegaly:
    - is an enlargement of the spleen which results from the sequestration of abnormal cells of the blood by the spleen.
  - Infarction of the Spleen:
    - occurs when a thrombus blocks the spleen from its blood supply. The spleen experiences tissue death due to oxygen deprivation.
Polycythemia Vera

- Polycythemia Vera – an excess number of red blood cells are produced. Most people who have PVera carry a mutated gene.
- Symptoms include:
  - Thrombosis (blood clot) caused by hyperviscosity and increased platelets. Thrombosis is the main cause of morbidity (active disease) and mortality (death)
  - Hemorrhage
  - Splenomegaly (enlarged spleen)
- Treatment includes
  - Aspirin to inhibit platelet function
  - Chemotherapy to decrease bone marrow activity
  - Multiple venesections (blood removal) to decrease blood volume

Thrombocytosis

- Thrombocytosis – an excess number of platelets are produced. This can lead to clotting and blockage of blood vessels.
- Symptoms include:
  - Thrombosis (blood clot) - a main cause of morbidity and mortality
  - Excessive hemorrhage after trauma or surgery - a main cause of morbidity and mortality
  - Splenomegaly in 30% of patients; in others the spleen is atrophied because of infarction (blockage).
- Treatment includes
  - Aspirin to inhibit platelet function
  - Chemotherapy to decrease bone marrow activity
**Myelofibrosis**
- Myelofibrosis (primary or idiopathic- also called myelosclerosis) – the bone **marrow produces too much collagen or fibrous tissue**, reducing its ability to produce red blood cells. This can lead to **hematopoiesis in the liver and spleen**.
  - Symptoms include:
    - Massive **spleenomegaly**
    - **Hepatomegaly** (enlarged liver)
  - Treatment includes:
    - **Chemotherapy** to decrease bone marrow activity
    - **Thalidomide** (reduces spleen size in 1/3 of cases)
    - **Transfusions** of red cells and sometimes platelets

**Chronic Myelogenous Leukemia**
- Chronic Myelogenous Leukemia (CML) – is also called **Chronic Granulocytic Leukemia (CGL)**.
  - CML is a cancer of the bone marrow in which **abnormal granulocytes are produced**.
  - Symptoms include:
    - **Spleenomegaly** – often massive
    - Many cases are discovered during routine physicals
  - Treatment includes:
    - **Chemotherapy**
    - **Bone Marrow (Stem Cell) transplantation**
Causes of Myeloproliferative Disorders

- Causes of myeloproliferative disorders are not exactly known, however two factors seem to be involved:
  - Genetics – some people have genetic mutations associated with specific disorders. An example is Philadelphia chromosome associated with CML.
  - Environment – overexposure to radiation, certain chemicals, and electrical wiring are thought to play a part.

Risk Factors

- Risk Factors include:
  - Age – different myeloproliferative disorders are seen more often in certain age groups.
  - Gender - different myeloproliferative disorders are seen more often in one gender over the other.
  - Environment – overexposure to radiation, certain chemicals, and electrical wiring are risk factors.