

- CML  
 - Polycythaemia rubra vera  
 - Central thrombocythopaenia  
 - Myelofibrosis  
 Myeloproliferative Disorders  
 - Myelodysplasia

Myeloproliferative Disorders

- Uncontrolled clonal proliferation of one or more cell lines in the bone marrow
- Normally occur in middle-aged elderly people

THE 4 TYPES of MYELOPROLIFERATIVE DISORDERS:

- ① Chronic Myeloid leukaemia (+WCC)
- ② Polycythaemia rubra vera (+RCC)
- ③ Central thrombocythopaenia (+platelets)
- ④ Myelofibrosis

Polycythaemia rubra vera: <sup>risk of thromboembolic events</sup> - genetic mutation.

Polycythaemia

- An increase in Hb, PCV, or RCC
- Primary polycythaemia is an inherited or acquired mutation leading to an abnormality within the red blood cell progenitors
  - Polycythaemia Vera: a mutation that causes excessive proliferation of RBCs (and possibly of the WBCs and platelets too)
  - Signs and symptoms are the result of hypovolaemia and hypoviscosity and include headache, dizziness, visual disturbance, venous thrombosis, angina, splenomegaly etc
  - Treatment is regular venesection (and possibly things like chemotherapy if there is a high platelet count too)
- Secondary polycythaemia is caused by chronic hypoxia causing an erythropoietin response, or by an erythropoietin-secreting tumour
  - Similar clinical features to those of the above, but usually it is just the RBCs that are elevated in number, and the spleen is a normal size
  - You should treat any tumour and give oxygen

Essential Thrombocythemia

- Where the Hb and WBC count are both normal but the platelet count is elevated (accompanied by abnormal platelet size and functioning)
- Presentation may be with bleeding or thrombosis

Eg: smoking / COPD

Myelofibrosis

- Haemopoietic stem cell proliferation associated with bone marrow fibrosis
- Insidious onset of weakness, weight loss and lethargy, as well as hepatomegaly and splenomegaly and bleeding of thrombocytopaenic.
- Anaemia, with initial high WBC and platelet counts which then fall as the bone marrow fibrosis progresses
- Treatment include splenectomy, transfusions etc

Myelodysplasia

- A group of acquired bone marrow disorders caused by a stem cell defect
- Predominantly a disease of the elderly
- Progressive bone marrow failure which eventually leads to acute myeloid leukaemia