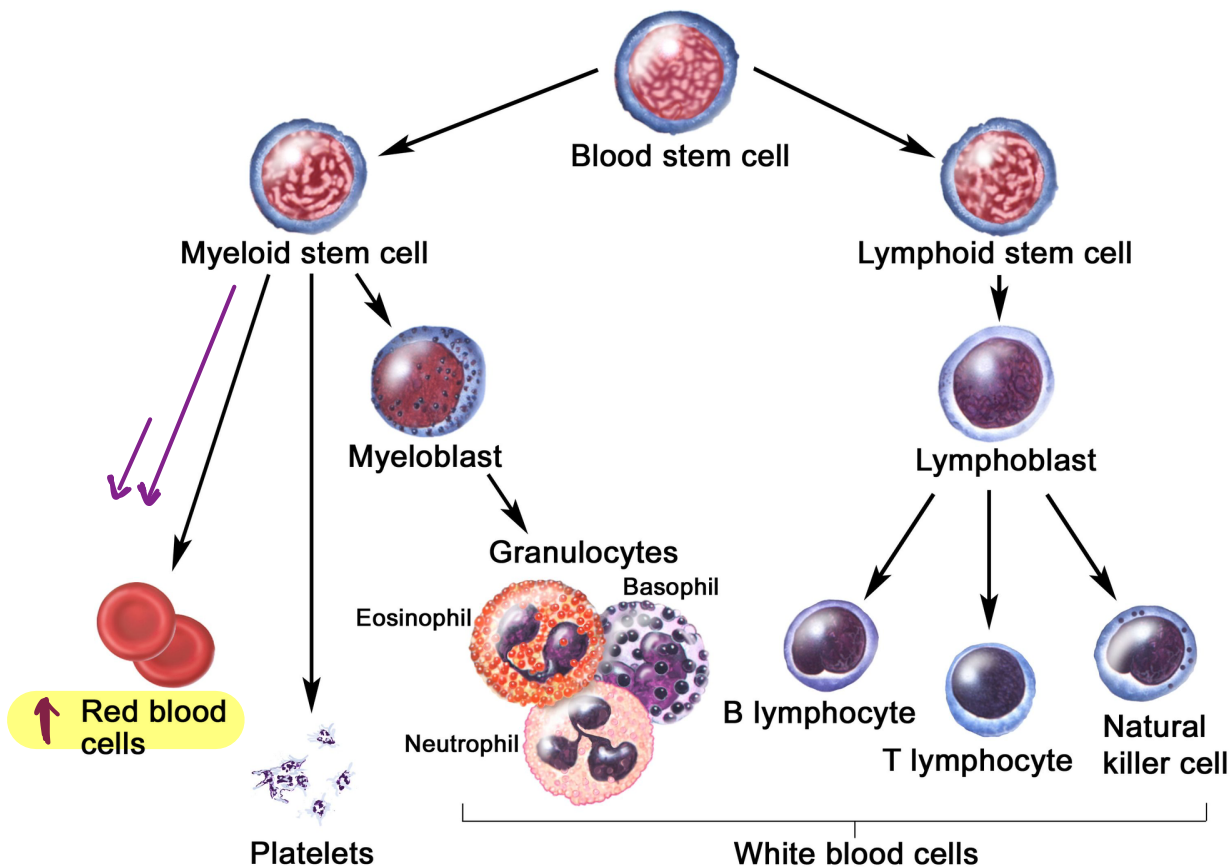
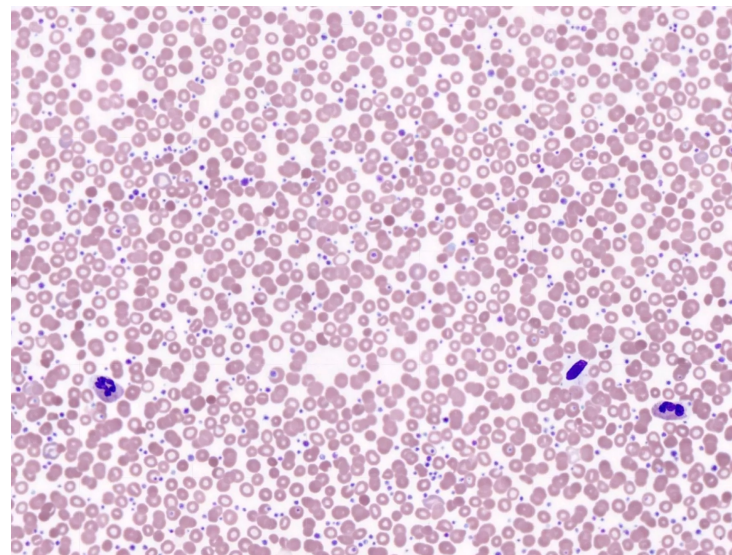


Polycythemia Vera

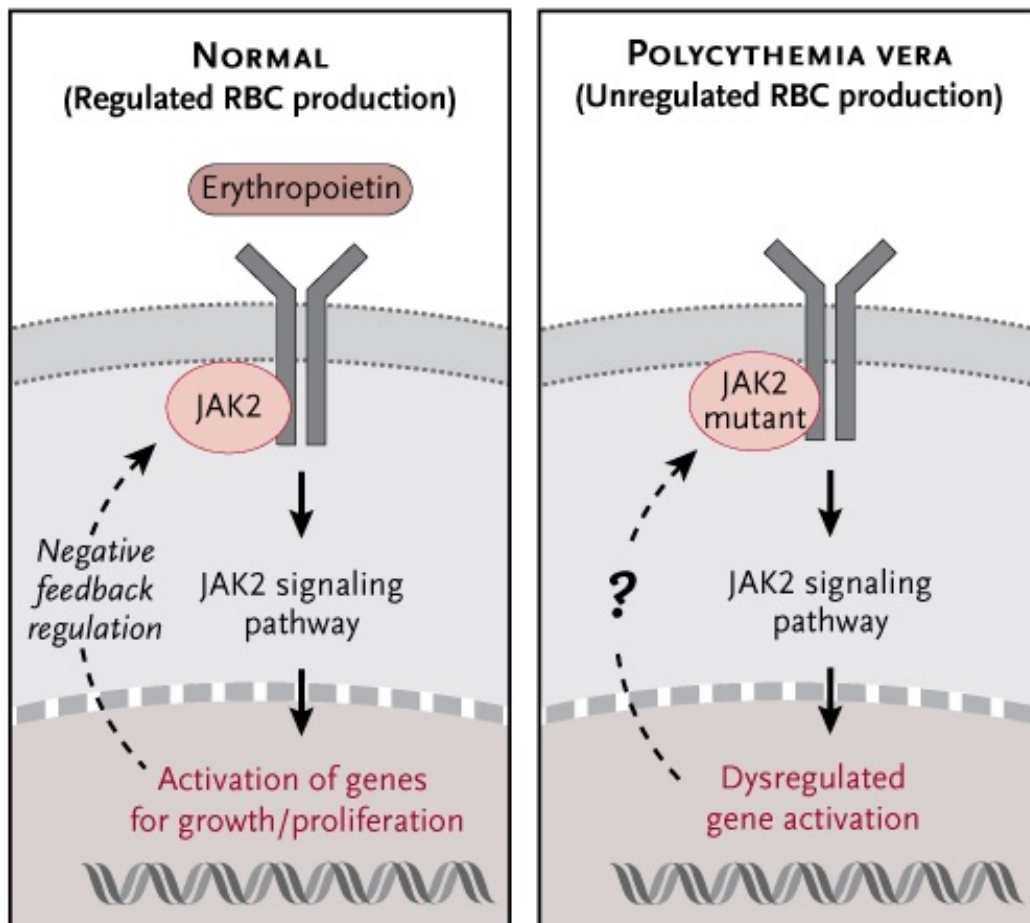


- Neoplastic proliferation of Mature Myeloid cells, esp RBCs.
- Granulocytes and platelets are also increased.
- Associated with JAK2 Kinase mutation.



Pathophysiology:

- Mutation of JAK 2 gene , most commonly resulting in a single amino acid change in its protein product from Valine to Phenylalanine at Position 617.



Clinical Features:

- Mostly due to Hyperviscosity of blood.
- (i) Blurry Vision & Headache
- (ii) Increased Risk of Venous thrombosis
(Hepatic vein , portal vein and dural sinus)
- (iii) Flushed face due to congestion (plethora)
- (iv) Itching , especially after bathing (Histamine ↑)

Signs and Symptoms of Polycythemia Vera

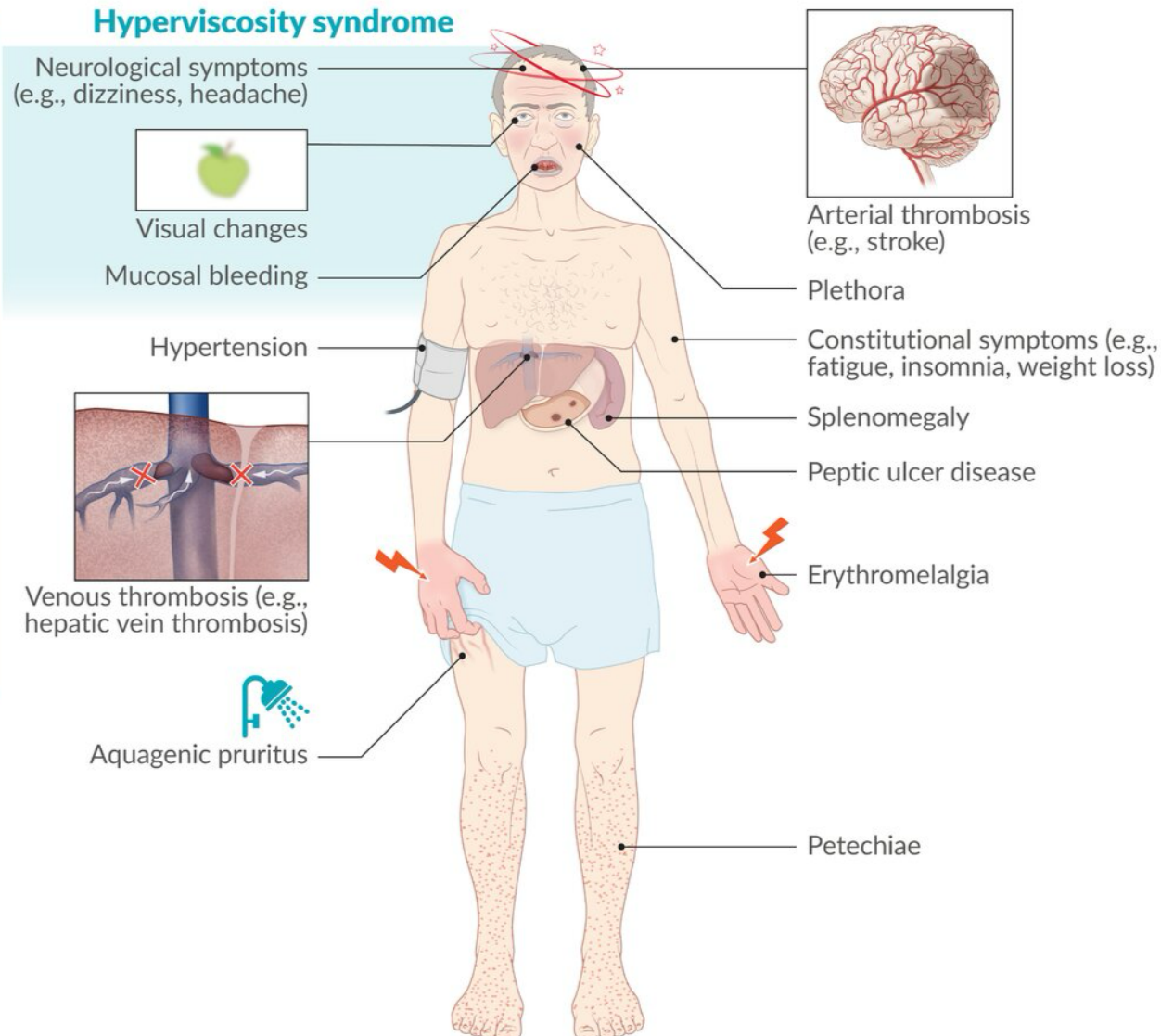
More Common

Hematocrit level >52 percent (0.52) in white men,
 >47 percent (0.47) in blacks and women
Hemoglobin level >18 g per dL (180 g per L) in white men,
 >16 g per dL (160 g per L) in blacks and women
Plethora
Pruritus after bathing
Splenomegaly
Weight loss
Weakness
Sweating

Less Common

Bruising/epistaxis
Budd-Chiari syndrome
Erythromelalgia
Gout
Hemorrhagic events
Hepatomegaly
Ischemic digits
Thrombotic events
Transient neurologic complaints (headache, tinnitus, dizziness, blurred vision, paresthesias)
Atypical chest pain

Hyperviscosity syndrome



Treatment:

- (i) Phlebotomy
- (ii) Second line = Hydroxyurea.
- (iii) Ruxolitinib, a Janus Kinase 2 (JAK2) inhibitor.
- (iv) Ropeginterferon α -2b

* Without treatment, death usually occurs within one year.

Types of Polycythemia:

Primary polycythemia:

- Caused by a mutation in a gene called JAK2: a kind of blood cancer
- **Polycythemia Vera**
- Other types of myeloproliferative neoplasm (MPN) can cause polycythemia, eg, essential thrombocythemia

Secondary polycythemia:

- Caused by high blood erythropoietin (EPO) level
- Physiological response to tissue hypoxia: eg, COPD, obstructive sleep apnea, high altitudes
- **EPO secreting tumors**
- **Testosterone therapy**
- Others

Relative polycythemia:

- Caused by decreased plasma volume
- Diarrhea, vomiting
- Gaisböck's syndrome (stress polycythemia)

Congenital polycythemia



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