

# Polycythemia

Erythrocytosis:  
increased **RBC s**  
**number**

Increase in **total RBC mass** above normal range

## Relative polycythemia

secondary to **decreased plasma volume**  
(water deprivation, severe diarrhea,  
diuretics)

## Absolute polycythemia

true increase in RBC mass, secondary  
to **increased BM production**

## Absolute polycythemia

### Primary (polycythemia vera)

**Autonomous** high bone marrow  
production

Myeloproliferative **neoplasm**

Mutation in **tyrosine kinase JAK2** in bone marrow stem cells

**JAK2** : Normally acts in the signaling pathway of erythropoietin  
receptor and other growth factor receptors

Hematopoietic cells become **less dependent on growth factors**

Excessive proliferation of erythroid, myeloid cells and megakaryocytes  
(**panmyelosis**)

Erythrocytosis is **most prominent**, results in polycythemia

### Secondary

**systemic hypoxia** caused by :

- ? Adaptive: living in high altitude, cyanotic heart disease, chronic pulmonary diseases, sleep apnea.
- ? Paraneoplastic: renal cancer, liver cancer .
- ? Surreptitious (blood doping): endurance athletes .
- ? Alcohol: frequent urination, depressed respiration .
- ? Smoking.

Splenomegaly is common

erythropoietin is low

no splenomegaly

high erythropoietin



increased erythropoiesis

## Symptoms of polycythemia

Similar symptoms :

- ? Plethora/ cyanosis
- ? Headache and dizziness (from hypertension)
- ? Slow circulation and hyperviscosity cause cyanosis, blurred vision, tissue ischemia
- ? Thrombosis, or bleeding (disturbed function of vWF)

In polycythemia Vera (primary) only:

- ? Pruritus (aquagenic)
- ? Peptic ulcer
- ? Secondary gout (arthritis, kidney stones, tophi)
- ? Chronic disease
- ? Spent phase: occurs after an interval of 10 years of symptoms, BM becomes fibrotic, hematopoiesis shifts to spleen
- ? Blast crisis: transformation to acute myeloid leukemia (rare)

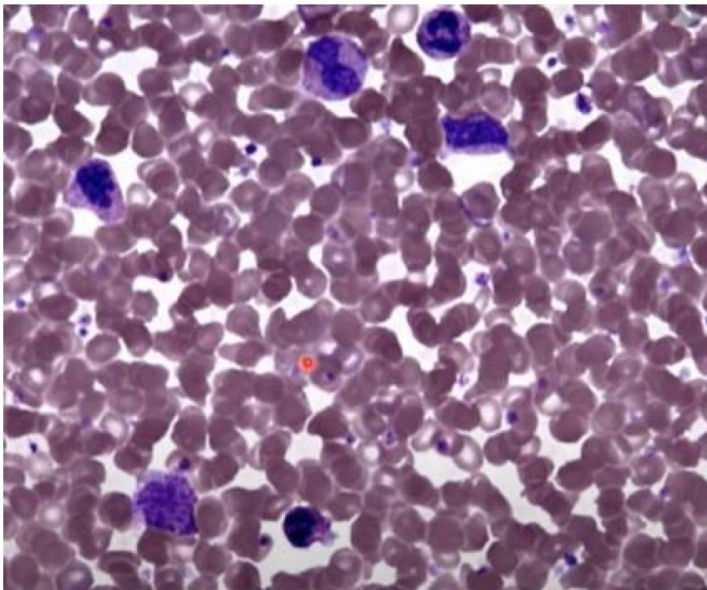
# Laboratory findings of polycythemia

In general:

? **High hemoglobin concentration** (>16.5 g/dL in men, 16 in women) and **high hematocrit** (>49% in men, 48% in women)

? **High RBCs count**

? These tests might be masked **if iron deficiency develops**



In **polycythemia vera**: additional findings:

? Leukocytosis and thrombocytosis are **common**

? **JAK2 mutation**

? **Low erythropoietin level**

? Hypercellular bone marrow with **panmyelosis**



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▪ Peripheral blood smear in polycythemia: packed RBCs