

polycythemia → ↑ RBC Mass  
 → ↑ RBC No. (unless it was empty)

Relative polycythemia  
 ↓ plasma volume due to  
 A- water deprivation  
 B- diuretics  
 C- severe diarrhea

absolute polycythemia

(polycythemia vera)

Primary

- autonomous erythropoiesis by BM
- myeloproliferative (= BM proliferative neoplasm)
- growth factor (erythropoietin) less dependent (due to n-feedback)

low

- 99% due to JAK2\* gene deficiency
- panmyelosis: erythroid, myeloid, megakaryocytes  
 ↓  
 more prominent  
 leading to polycythemia
- splenomegaly is common

\* JAK2 normally plays a role in the signaling pathway of erythropoietin receptor and other growth factor receptors.

Reversible

Secondary

Systemic hypoxia  
 ↓  
 ↑ erythropoietin  
 ↓  
 ↑ erythropoiesis

Causes

- ↓ adaptive
  - ① living in high altitude
  - ② cyanotic H.D. (children)
  - ③ C.O.P.D. (steap apnea)
- ↓ alcohol leads to
  - A- frequent urination
  - B- depressed respiration
- ↓ smoking leads to
  - A- pulm. diseases
  - B- hypoxia
- ↓ paraneoplastic due to
  - A- liver cancer
  - B- kidney cancer
- ↓ surreptitious (hidden)
  - ↓ blood doping due to taking hormones (androgens/growth factor)
  - ex. endurance athletes

Symptoms:

**SYMPTOMS OF POLYCYTHEMIA**

itchiness of skin → if not treated → redness of skin

- 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: similar symptoms plus:**

- Pruritus (aquagenic) → after shower
- Peptic ulcer
- Secondary gout (arthritis, kidney stones, tophi) → tophus → deposition of uric acid crystals in the tissues
- 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)

\* spent phase: ① Anemia → ↓ RBC  
 ② leukopenia ↓ WBC  
 ③ thrombocytopenia ↓ platelets

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↓  
labratory findings

## LABORATORY FINDINGS OF POLYCYTHEMIA

- High hemoglobin concentration (>16.5 g/dL in men, 16 in women) and high hematocrit (>49% in men, 48% in women) *must be persistent*
- High RBCs count
- These tests might be masked if iron deficiency develops

### In polycythemia vera: additional findings:

- Leukocytosis and thrombocytosis are common
- JAK2 mutation *99% D+ → neoplastic*
- Low erythropoietin level
- Hypercellular bone marrow with panmyelosis *fat must be there*  
*Biopsy*

↓  
labratory findings

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