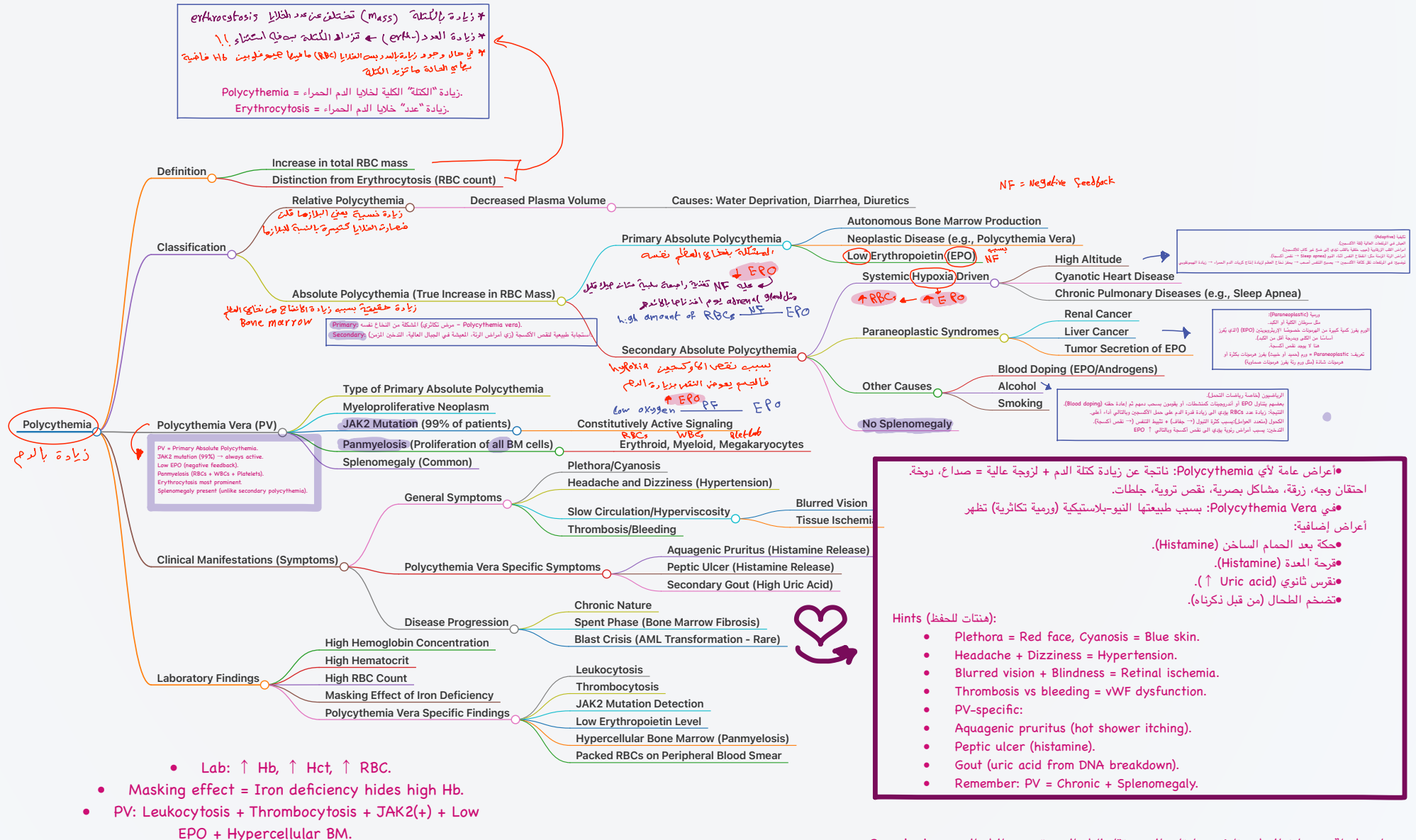




بعد سنوات طويلة - نخاع العظم يتليف - إنتاج الدم ينتقل للطحال - تضخم الطحال Spent phase: لكن نادر (AML) ممكن يتحول لسرطان دم حاد Blast crisis:

Spent Phase = fibrosis + splenomegaly

Polycythemia

📌 Definition

- **Polycythemia** = ↑ RBC mass (↑ Hb, ↑ Hct).
- Can be **Absolute (True)** or **Relative (False)**.

1 Relative Polycythemia (False)

- Cause: ↓ **plasma volume** (dehydration, burns, stress).
- RBC mass = **normal**.
- **No splenomegaly**.
- EPO = **normal**.

🧠 Think: "Looks like polycythemia but actually due to plasma loss."

2 Absolute Polycythemia (True)

A. Primary (Polycythemia Vera – PV)

- **Myeloproliferative neoplasm**.
- **JAK2 mutation (99%)** → constitutive activation → proliferation without EPO.
- **Panmyelosis**: ↑ RBC, ↑ WBC, ↑ Platelets.
- **Splenomegaly common** (extramedullary hematopoiesis).
- **Low EPO** (negative feedback).

📌 Symptoms (PV):

- General: Plethora, cyanosis, headache, dizziness, hyperviscosity, thrombosis/bleeding.
- Specific:
 - **Aquagenic pruritus** (histamine release after hot shower).
 - **Peptic ulcer** (histamine).
 - **Secondary gout** (↑ uric acid from high cell turnover).
 - Chronic → **Spent phase** (BM fibrosis, extramedullary hematopoiesis).
- Rarely → **Blast crisis** → AML.

📌 Lab Findings (PV):

- ↑ Hb (>16.5 M, >16 F), ↑ Hct (>49% M, >48% F).
- ↑ RBC, ↑ WBC, ↑ Platelets.
- **Hypercellular BM (panmyelosis)**.
- **JAK2+** in molecular test.
- **Low EPO**.
- ⚠️ May be "masked" if **iron deficiency** develops → Hb looks normal.

B. Secondary Polycythemia

- Due to ↑ **EPO**.
- **No splenomegaly**.
- Causes:
 - **Adaptive (hypoxia)**: high altitude, cyanotic heart disease, chronic lung disease, sleep apnea.
 - **Paraneoplastic**: renal cell carcinoma, hepatocellular carcinoma → ↑ EPO.
 - **Surreptitious (blood doping, EPO/androgen abuse)**.
 - **Smoking, alcohol** (multifactorial).

🔴 Peripheral Blood Smear

- **Packed RBCs** (high cellularity).

⚡ High-Yield Differences (Exam Must-Know)

Primary (PV)	Secondary	Relative	Feature
↑	↑	Normal	RBC Mass
Normal	Normal	↓	Plasma Volume
↓	↑	Normal	EPO
Yes	No	No	Splenomegaly
JAK2 mutation	Hypoxia / Tumor (↑EPO)	Dehydration	Cause
↑ WBC + ↑ Platelets (panmyelosis)	Normal	Normal	Other Cells

← EPO

🧠 Super Exam Hints

- **Aquagenic pruritus** = PV until proven otherwise.
- **High EPO + no splenomegaly** = **Secondary**.
- **Low EPO + JAK2 mutation + splenomegaly** = **PV**.
- **Relative** = dehydration → normal EPO, no real RBC increase.
- **Spent phase** = marrow fibrosis after ~10 yrs.
- **Blast crisis (AML transformation)** = rare but important.
- **Masking effect**: PV + iron deficiency → Hb may look "normal."