

Polycythemia Vera Disease

Polycythemia is a chronic myeloproliferative disorder where the bone marrow produces too many red blood cells. Patients often have increased WBCs and platelets as well, as this disease is a panmyelosis. Classic symptoms include headache and pruritis, especially after exposure to hot water. This disorder is caused by a mutation in the JAK2 tyrosine kinase, leading to hypersensitivity to erythropoietin (EPO). As polycythemia vera is not curable, treatment for this disorder involves regular phlebotomy to normalize hematocrit levels, as well as low dose aspirin.



PLAY PICMONIC

Pathophysiology

JAK2 Mutation leading to increased red blood cells

[Jack in \(2\) Tutu mutant with Up-arrow RBC](#)

This disorder stems from a mutation in the JAK2 (Janus Kinase) most commonly on chromosome 9 which is a non-receptor tyrosine kinase. This mutation makes cells hypersensitive to EPO (erythropoietin), causing panmyelosis via neoplastic proliferation of erythroid, megakaryocytic and granulocytic cells.

Tyrosine Kinase

[Tire Kite-ace](#)

Tyrosine kinase function as an "on" or "off" switch in many cellular functions, and in this disease these they are responsible for regulating red blood cell production signaling cascades.

Symptoms

Erythromelalgia

[Earth-red-molten-lava](#)

Due to increased blood viscosity in this disorder, the blood vessels of the hands and lower extremities become blocked. This leads to the hands becoming inflamed and hyperemic, and patients often describe swelling with a "burning pain."

Pruritus

[Prairie-dog](#)

A classic symptom in patients is itching, particularly after a hot shower or bath. This is hypothesized to be because of abnormal histamine release or prostaglandin production.

Often After Hot Shower

[Hot Shower](#)

In this disorder, pruritus results from increased histamine levels released from mast cells and basophils. This histamine release is triggered by warm showers or baths.

Headache

[Head-egg lump](#)

The most frequent symptom of this disease is the complaint of headache.

Treatment

Phlebotomy

[Flea-bottom](#)

This condition cannot be cured, and phlebotomy, or bloodletting, is a common form of treatment. Blood is removed from the body to normalize the hematocrit to 42-45. This reduces the risk of blood clots and improves cognitive impairment.

Aspirin

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Low dose aspirin is recommended in this disease, as it reduces the risk of thrombotic complications in patients.