

## Polycythemia Vera

Polycythemia vera (PV) is a chronic blood malignancy caused by an acquired gene mutation of the blood-producing cells in the bone marrow which causes an overproduction of blood cells, particularly red blood cells. White blood cells and platelets are often also increased. An excess amount of red blood cells can increase the blood viscosity (thickness), slowing blood flow. As blood flow is slowed, the risk of blood clotting increases. PV can also cause the spleen to become enlarged. PV is one of a group of rare bone marrow cancers called myeloproliferative neoplasms (MPNs). Conversion to myelofibrosis or, in rare cases, acute leukemia can occur as the disease progresses over time

### Statistics

- This blood disorder affects approximately **44 to 57 in every 100,000 people** in the United States.
- A majority of polycythemia vera cases are diagnosed in people **over the age of 60**.

### Risk Factors

The exact causes of this acquired genetic mutation in polycythemia vera are not yet known. A family history of PV is rarely present; however, occasionally multiple family members will have the disease. Risk factors may include the following:

- **Mutations:** Almost all people with PV have a Janus kinase 2 (JAK2) gene mutation. Of these, 95% have a mutation of JAK2 V617F in exon 14, with most of the remainder having mutations in exon 12.
- **Gender:** Men have a slightly higher risk than women of developing polycythemia vera.
- **Age:** People over age 60 are more at risk.
- **Environment:** Exposure to radiation and toxic substances may increase risk.

### Symptoms

Many patients with PV do not have symptoms when they are diagnosed. Diagnosis often occurs during a routine exam or blood test. However, people may experience persistence of any of the following symptoms.

- Headaches
- Sweating and night sweats
- Ringing in the ears
- Blurred vision or blind spots
- Dizziness or vertigo
- Skin with red or purple hue
- Bleeding or clotting excessively
- Reddened face
- Unexpected weight loss
- Feeling full quickly on eating
- Abnormal discomfort on left side below the ribs
- Itching, especially after a shower or bath
- Burning and redness of the hands or feet
- Fatigue
- Bone pain
- Gout attacks
- Shortness of breath
- Weakness
- Bruising excessively
- Numbness or tingling in hands or feet

### Prevention

PV cannot be prevented. Research is underway to learn more about how the disease develops.

### Treatment

PV is a chronic disease, but it can be effectively managed. PV can cause different symptoms in each patient. In some cases when symptoms may not be present, treatment may not be required for extended periods of time, although routine monitoring with a physician is recommended for all PV patients. One of the main goals of monitoring and treatment is to reduce the risk of blood clotting. Treatment options include phlebotomy (removal of blood), low-dose aspirin, and drug therapies such as hydroxyurea, anagrelide, ruxolitinib, or interferon alfa. Antihistamines and avoidance of hot showers can help with itching. Other treatment options may be available through clinical trials.

## About Texas Oncology

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*Sources: Leukemia and Lymphoma Society, MPN Research Foundation, National Cancer Institute, and National Organization for Rare Disorders*



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