The Blood Project

UNDERSTANDING POLYCYTHEMIA VERA

A brief guide for patients with polycythemia vera

Polycythemia vera (PV) is a chronic blood cancer in which the bone marrow makes too many blood cells, especially red blood cells. This makes the blood thicker than normal and increases the risk of blood clots if not treated. PV is a long-term condition that requires regular monitoring and treatment, but with good care many people live full and active lives.

What are red blood cells?

Blood contains red blood cells that carry oxygen, white blood cells that help fight infection, and platelets that help with clotting.
All of these cells are made in the bone marrow.
In PV, the normal controls on blood cell production do not work properly because of a change in the JAK2 gene, and the marrow makes too many cells.

What is polycythemia vera?

Polycythemia vera is a myeloproliferative neoplasm, a type of chronic blood cancer.
In PV, the bone marrow produces too many red blood cells and sometimes too many platelets and white blood cells.
The excess red blood cells make the blood thicker, which increases the risk of clots in arteries and veins.
PV involves an acquired JAK2 gene change in the great majority of people.
This change develops during life and is not usually inherited.
PV is a lifelong condition and requires ongoing care by a hematologist.

Why polycythemia vera happens

Acquired JAK2 gene change that drives overproduction of blood cells
Excess red blood cell production leading to thickened blood
Increased platelets or white blood cells in some people
Not caused by diet, lifestyle, or activities

Does it cause symptoms?

Some people have no symptoms at diagnosis.
Others may notice headaches, dizziness, or difficulty concentrating.
Common symptoms include itching after warm showers or baths, burning pain or redness in the hands or feet, fatigue, night sweats, or weight loss.
Some people develop fullness or discomfort under the left ribs from an enlarged spleen.
Joint pain or gout can occur because of increased cell turnover.

Is it dangerous?

PV can be dangerous if untreated because thicker blood increases the risk of blood clots.
Clots can cause stroke, heart attack, deep vein thrombosis, pulmonary embolism, or less common clots in the abdomen affecting the liver or intestines.

Doctors consider a person low-risk if they are younger than 60 and have no history of blood clots.
People are considered high-risk if they are 60 or older or have had a clot.
Over many years, a minority of people may develop more advanced marrow problems such as myelofibrosis, and rarely acute leukemia.
These complications are uncommon with modern treatment but reinforce the need for lifelong follow-up.

How is it evaluated?

Complete blood count to measure hemoglobin, hematocrit, platelets, and white blood cells
Testing for the JAK2 mutation
Erythropoietin level to help distinguish PV from other causes of high red blood cells
Physical examination including spleen size
Imaging if the spleen appears enlarged
Bone marrow biopsy in selected cases

Do I need a bone marrow biopsy?

Not everyone with PV needs a bone marrow biopsy.
Many diagnoses are made using blood counts and JAK2 testing alone.
A biopsy may be recommended if the diagnosis is uncertain or if there is concern about disease progression.
Your doctor will explain whether this test is needed in your situation.

How is it treated?

Treatment focuses on lowering the risk of blood clots and keeping blood thickness in a safer range.
Phlebotomy removes blood in a procedure similar to donating blood and lowers the hematocrit.
Early in treatment this may be done more frequently, then less often to maintain safe levels.
Low-dose aspirin is used in most patients unless there is a reason not to take it.
Medicines that slow blood cell production, such as hydroxyurea or interferon, may be used in people at higher risk of clots or those who need frequent phlebotomies.
Treatment choices are individualized based on age, clot history, symptoms, pregnancy plans, and medication tolerance.
Staying well hydrated and managing blood pressure, cholesterol, diabetes, and smoking are also important parts of care.

When should I contact my doctor?

Contact your doctor if you have new or worsening headaches, dizziness, or vision changes.
Chest discomfort, shortness of breath, palpitations, or new leg pain or swelling may signal a blood clot.
Increasing fullness under the left ribs, unusual bleeding, fevers, night sweats, or unexplained weight loss should also be reported.
Sudden weakness, trouble speaking, or severe shortness of breath require emergency care.
Tell emergency staff that you have polycythemia vera.

What is the usual plan going forward?

PV is a lifelong condition.
You will see your hematologist regularly for blood tests and treatment adjustments.
Most people remain stable for many years with phlebotomy, aspirin, and medication when needed.
Regular follow-up helps prevent complications and allows early detection of changes in the disease.

Key points to remember

- **chronic blood cancer**: PV causes the bone marrow to make too many blood cells, thickening the blood
- **JAK2-driven condition**: the JAK2 mutation is acquired during life and is not usually inherited
- **clot prevention matters most**: treatment focuses on reducing the risk of blood clots
- **phlebotomy and aspirin are core therapies**: additional medicines are used when risk is higher
- **lifestyle factors matter**: hydration and cardiovascular risk control are part of PV care
- **lifelong monitoring**: with regular follow-up, most people with PV live many active years