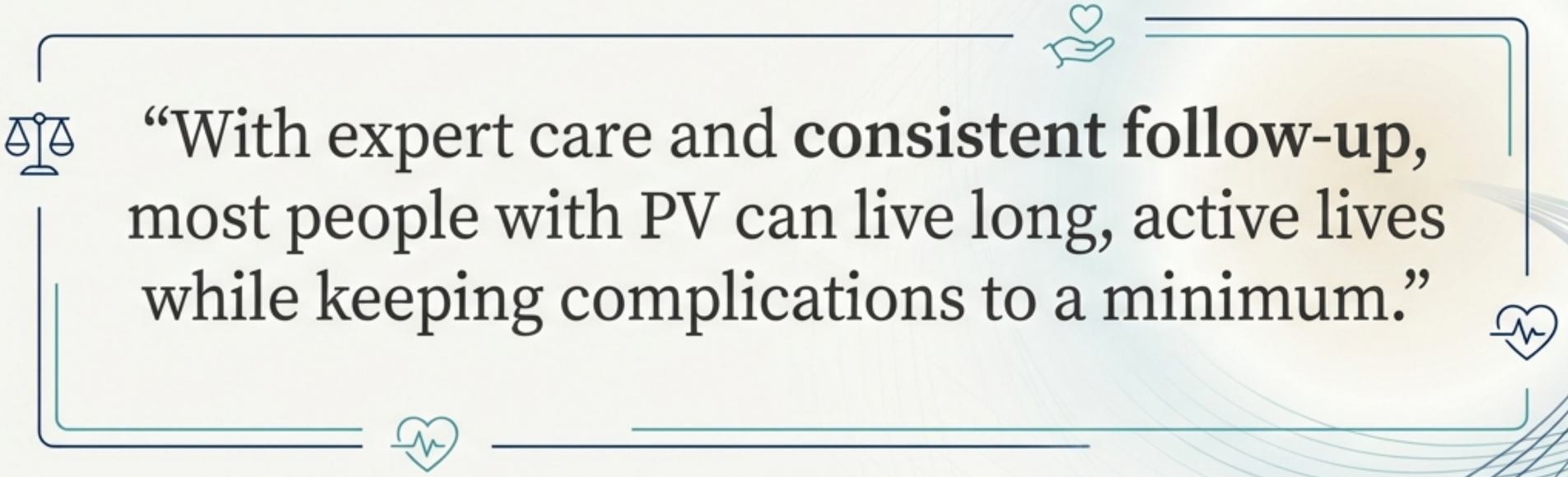
Understanding Polycythemia Vera (PV): A Guide for You and Your Family



> “With expert care and **consistent follow-up**, most people with PV can live long, active lives while keeping complications to a minimum.”

This guide is designed to help you understand your diagnosis, partner with your care team, and take an active role in your health.

What is Polycythemia Vera?

Polycythemia vera (PV) is a myeloproliferative neoplasm, a chronic blood cancer in which the bone marrow makes too many blood cells, especially red blood cells.

This makes the blood thicker and “stickier,” which increases the risk of serious blood clots if PV is not treated.

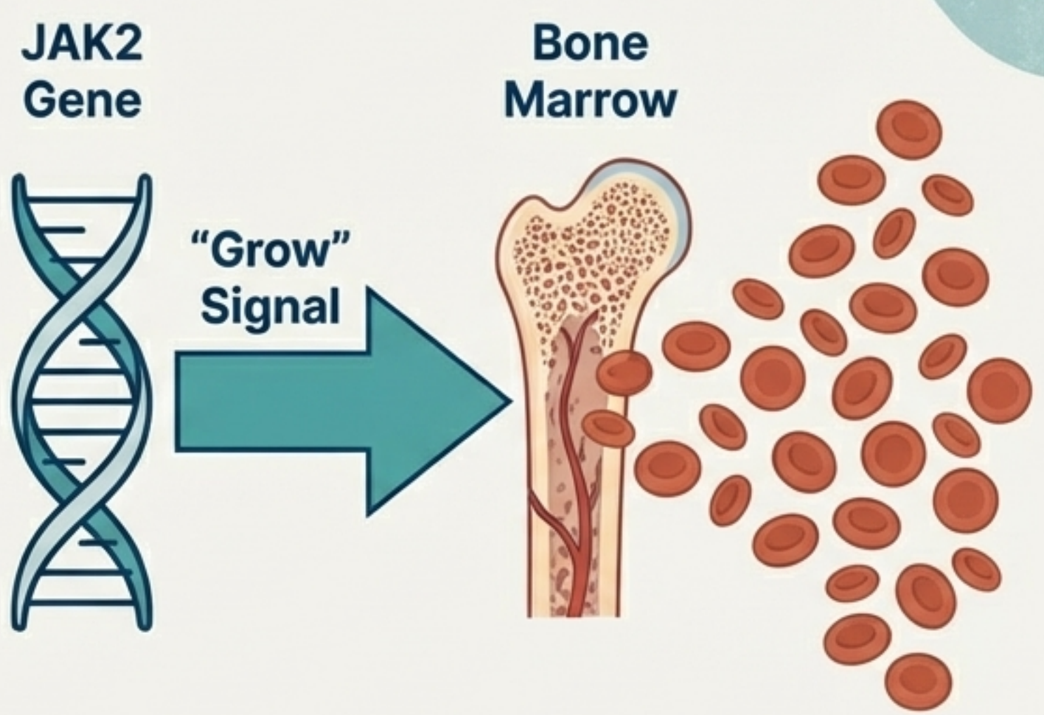
A Note on Diagnosis

Even though many people feel well at the time of diagnosis, PV is never “benign.” It requires lifelong hematology care, regular blood tests, and treatments to to keep blood thickness in a safer range.

What Causes PV? The JAK2 Gene

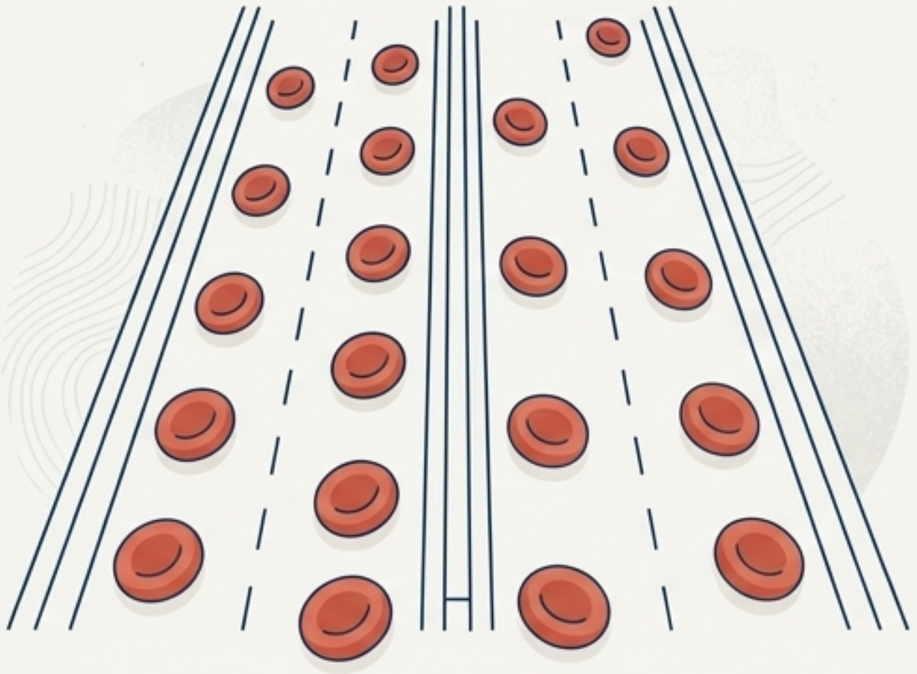
PV begins in the bone marrow, where a change in a gene called JAK2 causes blood-forming cells to grow even when the body does not need more. This sends strong “grow” signals that make the marrow overproduce cells.

- **Acquired**: It is acquired during life, not present at birth.
- **Unpreventable**: It cannot be prevented.
- **Not Your Fault**: It is not caused by anything you did.
- **Not Inherited**: It is not usually inherited or passed on to children.



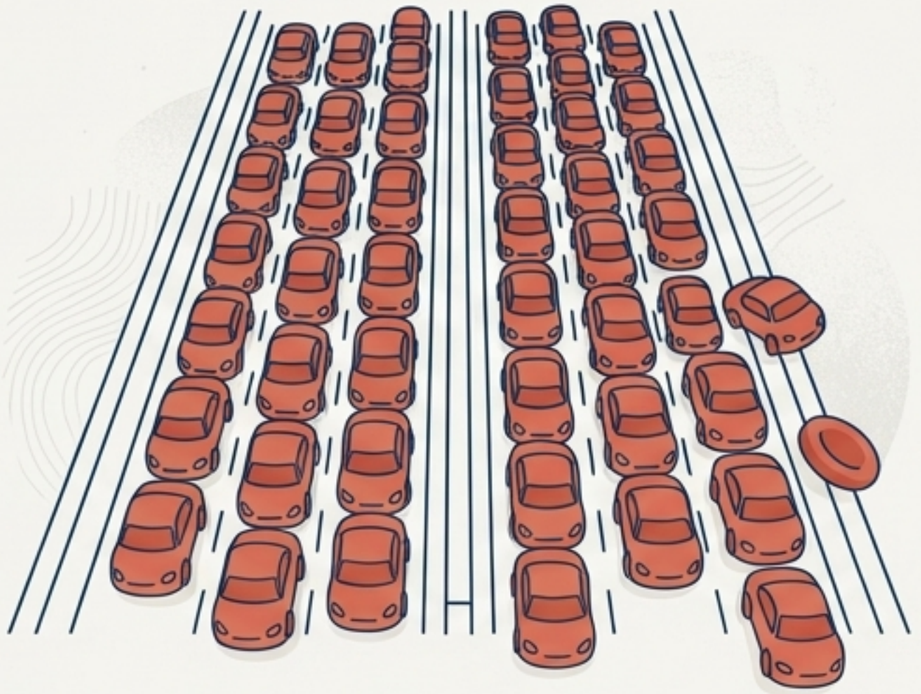
Making Sense of It All: The Busy Highway Analogy

A Normal Bloodstream



Think of your bloodstream as a highway. Normally, traffic flows smoothly and safely.

The Bloodstream in PV



In PV, there are too many ‘cars’ on the road. This slows traffic and raises the chance of ‘accidents’ (blood clots).

Can PV Cause Symptoms?

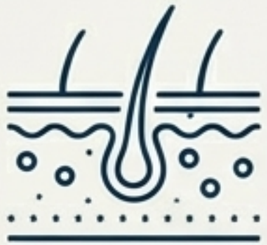
Some people have no symptoms at first, while others notice common patterns. Symptoms often improve when treatment brings blood thickness into a safer range.

Neurological

Headaches, dizziness, or trouble concentrating.

Systemic

Fatigue or low energy, night sweats, unintentional weight loss.



Skin-Related

Itching after warm showers (aquagenic pruritus), burning pain or redness in hands/feet (erythromelalgia), redness or flushing of the face.

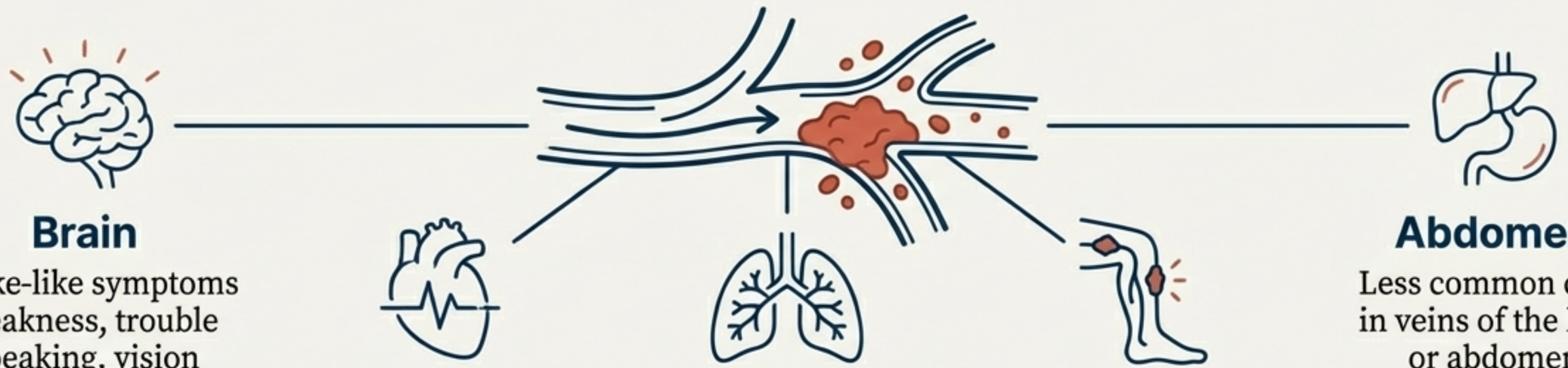
Physical

Fullness or discomfort under the left ribs (from an enlarged spleen), joint pain or gout.

Why is Treatment So Important? The Risk of Blood Clots

The main danger in PV is blood clots.

These can occur in arteries or veins and may be life-threatening.



Brain

Stroke-like symptoms (weakness, trouble speaking, vision changes)

Heart

Heart attack–type symptoms (chest pain, shortness of breath)

Lungs

Clots in the lungs (Pulmonary Embolism or PE)

Legs

Clots in the legs (Deep Vein Thrombosis or DVT)

Abdomen

Less common clots in veins of the liver or abdomen

How Your Doctor Assesses Your Clot Risk

Your doctor classifies PV as low-risk or high-risk to guide your treatment plan. This is based on two key factors:

Low-Risk

Younger than 60 AND no history of blood clots.

High-Risk

Age 60 or older OR a prior history of a blood clot.

Your evaluation also includes a full medical history, blood tests (for hematocrit, platelets, etc.), JAK2 mutation testing, and a review of other cardiovascular risk factors (blood pressure, cholesterol, diabetes, smoking).

Your Treatment Toolkit: A Plan to Control PV

The treatment plan has three main goals: lower your clot risk, reduce blood thickness, and control your symptoms.

1. Phlebotomy

The cornerstone of therapy to directly lower blood thickness.

2. Low-Dose Aspirin

A daily medication to help prevent clots from forming.

3. Cytoreductive Medicines

Medications to slow the bone marrow's overproduction of blood cells.

Your plan is individualized and will be adjusted over time based on your needs.

The Core Tools: Phlebotomy and Low-Dose Aspirin

Phlebotomy

What it is	The process of removing blood, similar to donating blood.
Why it works	It directly lowers the hematocrit (a measure of blood thickness), which is closely tied to clot risk.
What to expect	May be frequent at first to get hematocrit into a safer range, then done periodically for maintenance.

Low-Dose Aspirin

What it is	A daily, low-dose medication taken by most people with PV.
Why it works	Helps prevent platelets from sticking together to form clots.
Added Benefit	Can also ease certain symptoms, like the burning pain in hands and feet (erythromelalgia).

Do not take iron supplements unless your hematologist specifically directs you to. Low iron helps slow red blood cell production.

When More Is Needed: Medicines to Slow Cell Production

These medications (called cytoreductive therapies) are typically used for high-risk patients or for those who need frequent phlebotomies to control their hematocrit.

Hydroxyurea

An oral medicine that effectively slows marrow production.

Interferon

An injectable medication, often used in younger adults or those planning a pregnancy.

Other Treatments

In select situations, other medicines such as JAK inhibitors (e.g., ruxolitinib) may be used.

Your hematologist will work with you to choose the best option for your specific situation. Your treatment plan may change as your needs change.

Your Active Role in Managing PV

Most people with PV can continue to work, exercise, and enjoy daily life. Your lifestyle choices are a powerful part of your treatment plan.

Protect Your Heart

Do not smoke. Work with your care team to manage blood pressure, cholesterol, and diabetes.

Stay Mobile & Hydrated

Drink plenty of water, especially around phlebotomy sessions. Avoid sitting for long periods without moving to stretch your legs.

Be Consistent

Take all medications exactly as prescribed. prescribed. Do not miss scheduled laboratory tests or clinic visits.

Special Consideration: Pregnancy and PV

Pregnancy in PV is considered higher risk and requires specialized, collaborative care.

1. **Advance Planning is Key:**
   Discuss any plans for pregnancy with your hematologist well in advance.

2. **A Joint Care Team:**
   Your care should be managed by both a hematologist and a high-risk obstetrician.

3. **Medication Review:**
   Some PV medicines are not safe during pregnancy and may need to be changed before you try to conceive.

Knowing When to Seek Medical Help

Call Your Hematologist Promptly If...

- ✓ New or worsening headaches/dizziness
- ✓ New chest discomfort
- ✓ New leg pain/swelling
- ✓ Increased fullness under left ribs
- ✓ Unusual bleeding
- ✓ Fevers, night sweats, or unexplained weight loss

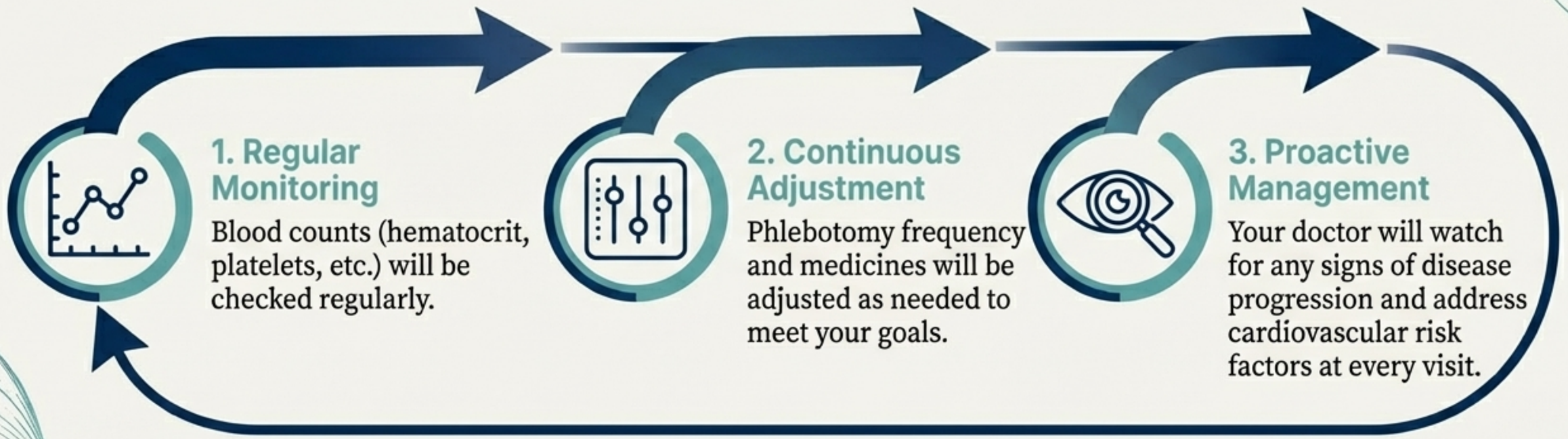
Call 911 or Go to an Emergency Dept. If...

- ↘ Stroke-like symptoms (face drooping, arm weakness, trouble speaking)
- ↘ Sudden severe chest pain or difficulty breathing
- ↘ Coughing up blood
- ↘ Sudden severe abdominal or leg pain
- ↘ Bleeding that will not stop

In an emergency, always tell the medical staff that you have Polycythemia Vera and list the medicines you take.

The Path Forward: Lifelong Partnership in Care

PV is a lifelong condition. The goal of ongoing care is to prevent clots, control symptoms, and detect any changes early.



1. Regular Monitoring

Blood counts (hematocrit, platelets, etc.) will be checked regularly.

2. Continuous Adjustment

Phlebotomy frequency and medicines will be adjusted as needed to meet your goals.

3. Proactive Management

Your doctor will watch for any signs of disease progression and address cardiovascular risk factors at every visit.

With this consistent care, many people with PV remain stable for decades.

Living Well with PV: Your Key Takeaways

- PV is a chronic blood cancer where too many red blood cells make the blood thicker, increasing clot risk.
- It's driven by an acquired JAK2 mutation—it is not your fault and is not inherited.
- The primary goal of treatment (phlebotomy, aspirin, medication) is to prevent dangerous blood clots.
- Your clot risk (age and prior clot history) guides the intensity of your treatment.
- Lifestyle choices—not smoking, staying hydrated, and managing heart health—are a vital part of your care.
- Consistent follow-up is the key to living a long, active life with PV.