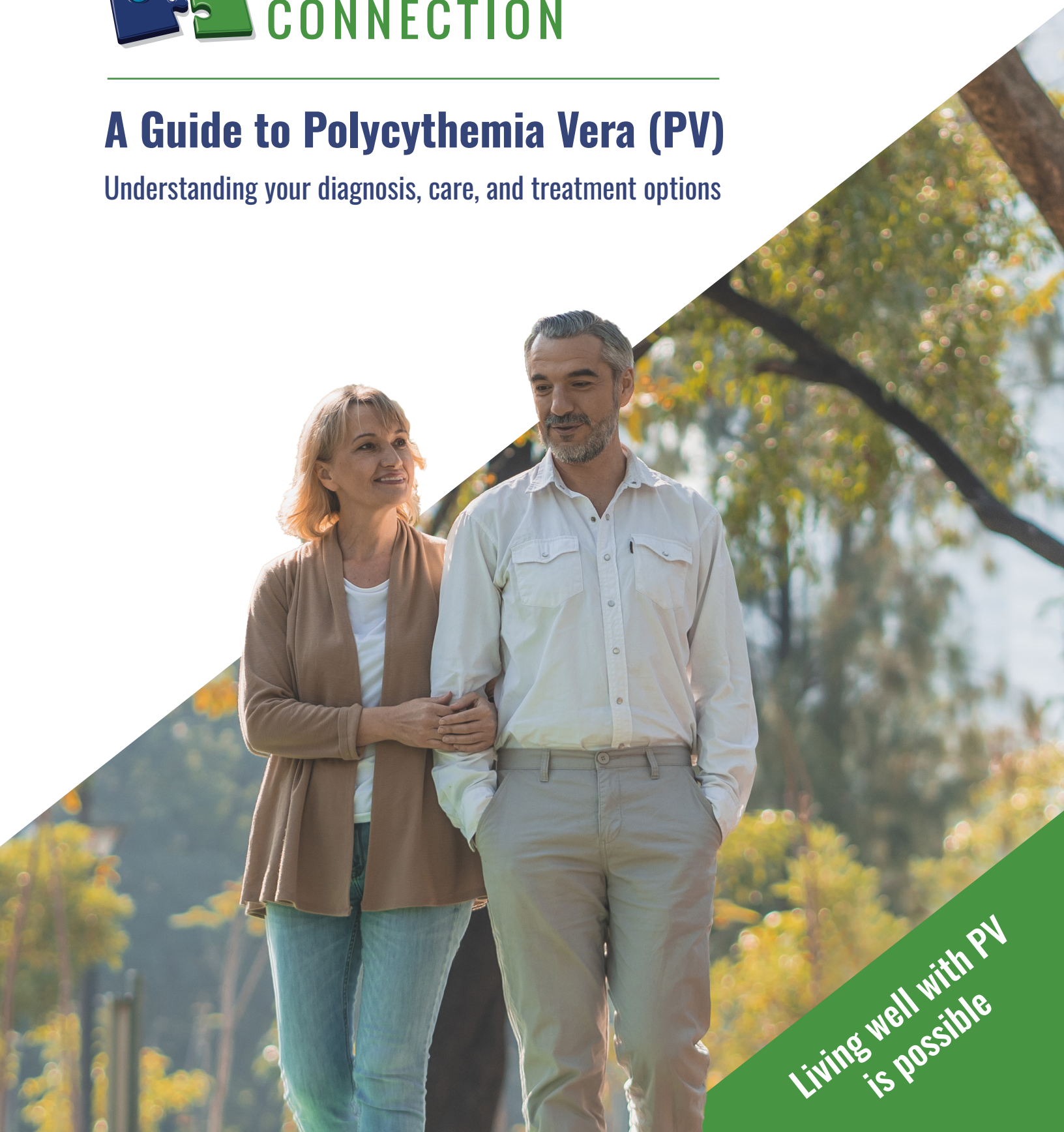
MPN Cancer CONNECTION

# A Guide to Polycythemia Vera (PV)

## Understanding your diagnosis, care, and treatment options



# Understanding PV and Things You Should Know

Learning that you have Polycythemia Vera (PV) often leads to questions about what the diagnosis means and what to expect over time. Because it is a chronic condition that requires ongoing management, learning how PV is monitored and treated can make the diagnosis feel less overwhelming and help patients navigate treatment decisions.

This guide outlines the core features of PV and serves as a reference to support informed discussions with your doctor.

## This Guide Covers:

- Defining PV
- Symptoms and blood counts
- Diagnosis and risk factors
- Treatment options
- Living with PV
- Looking ahead

## Defining PV

Polycythemia vera (PV) is a rare blood cancer that begins in the bone marrow, the soft tissue inside your bones where blood cells are made. In PV, the marrow produces too many blood cells, especially red blood cells. Many people also have elevated white blood cells and platelets.

This overproduction changes how the blood flows and behaves in the body. PV is classified as a myeloproliferative neoplasm (MPN), a group of disorders characterized by excessive blood cell production. Other myeloproliferative neoplasms include essential thrombocythemia (ET) and myelofibrosis (MF).

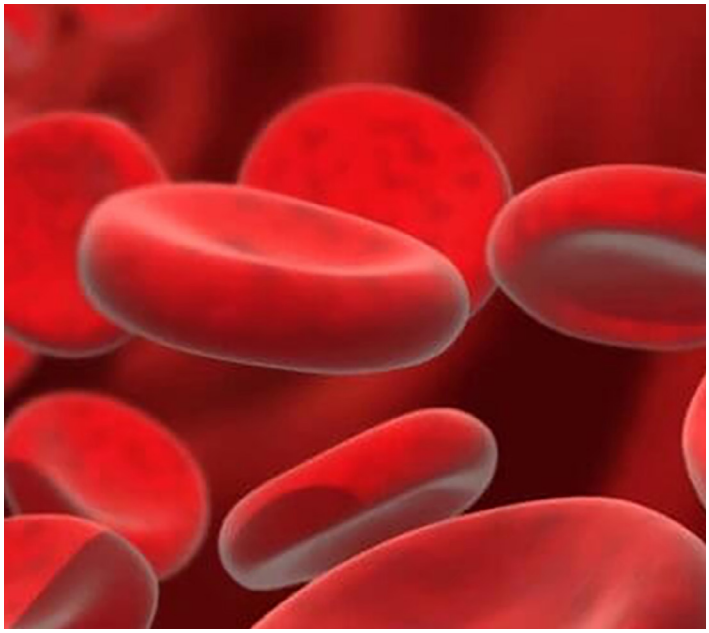
## What Causes PV?

PV is caused by an **acquired genetic mutation**, meaning it develops during a person's lifetime. It is not something you are born with, and it is not typically inherited.

Nearly all people with PV (approximately 95–97%) have a mutation in the **JAK2 gene**. Most have the **JAK2 V617F mutation**, while a small percentage have other JAK2 mutations (such as exon 12). In very rare cases, no mutation is detected, but the disease behaves similarly.

The JAK2 gene helps regulate blood cell production. You can think of it as part of a signaling "switch." In PV, this signaling pathway becomes stuck in the "on" position, telling the bone marrow to continue making blood cells even when the body does not need them.

**JAK2 allele burden** refers to the percentage of blood cells that carry the JAK2 mutation. Your doctor may test for the allele burden as part of your genetic testing. For example, an allele burden of 50% means that about half of the blood cells tested carry the mutation.



## Ask Your Doctor

*"Has my allele burden been measured, and has it changed since I started treatment?"*

# Symptoms & Blood Counts

PV affects each person differently. Some people have very few symptoms at diagnosis, while others notice changes that develop gradually over time. Many symptoms are related to thicker blood, increased cell counts, or the effects of an enlarged spleen. Some symptoms are caused by small blood vessel changes known as microvascular disturbances, which can contribute to headaches, visual changes, and burning pain in the hands or feet.

## Common Symptoms of PV

- **Headache, dizziness, or vision changes**
- **Fatigue**
- **Itching**, especially after a warm bath or shower (aquagenic pruritus)
- **Red or flushed face**
- **Burning pain, redness, or warmth in the hands or feet** (erythromelalgia)
- **Enlarged spleen**, which may cause left-sided fullness or abdominal discomfort
- **Blood clots**, which can lead to stroke, heart attack, deep vein thrombosis (DVT), or pulmonary embolism (PE)
- **Gout or joint pain**, due to increased uric acid levels
- **Brain fog or difficulty concentrating**


## Understanding Your Blood Counts

Polycythemia vera affects how your body makes blood cells, often causing certain counts to rise. This table below shows how those changes compare to normal levels.

## Key Message

The most important blood count in PV is your hematocrit. Keeping it below 45% has been shown to significantly reduce the risk of blood clots.

| Blood Cell Type | Normal Range | What It Looks Like in PV |
|---|---|---|
| **Hematocrit** (HCT) | Men: 41–50%<br>Women: 36–44% | Elevated<br>(often 50–65% or higher at diagnosis) |
| **Hemoglobin** (HGB) | Men: 14–17.5 g/dL<br>Women: 12–16 g/dL | Elevated<br>(often >16.5 g/dL in men, and >16.0 g/dL in women) |
| **Red Blood Cells** (RBCs) | Men: 4.3–5.9 trillion/L<br>Women: 3.5–5.5 trillion/L | Elevated |
| **White Blood Cells** (WBCs) | 4.5–11.0 billion/L | Frequently elevated<br>(often 10–25 billion/L) |
| **Platelets** | 150–400 billion/L | May be elevated<br>(may range from 400–1,000+ billion/L) |

# Diagnosis and Risk Factors

## How Is PV Diagnosed?

Polycythemia vera is often first suspected after a routine complete blood count (CBC) shows elevated blood cell levels, especially red blood cells and hematocrit. Many patients also have elevated white blood cells and platelets, since PV can cause the bone marrow to produce too many of several types of blood cells.

Because other conditions can also cause increased red blood cells (such as chronic low oxygen levels or secondary polycythemia), additional testing is needed to confirm the diagnosis.

### Diagnostic evaluation typically includes:

- **Complete Blood Count (CBC):** Shows elevated red blood cells and hematocrit. White blood cells and platelets may also be increased.
- **Erythropoietin (EPO) level:** This hormone stimulates red blood cell production. In PV, EPO levels are usually low or suppressed.
- **Genetic Testing:** Tests for mutations in the JAK2 gene.
- **Bone marrow biopsy:** May be performed to examine how blood cells are being produced. While helpful in confirming the diagnosis, it is interpreted together with blood tests and genetic findings.

# Your Risk Level Matters

### Risk stratification helps guide treatment decisions.

| LOW RISK | INTERMEDIATE RISK | HIGH RISK |
|---|---|---|
| **CRITERIA**<br>• Age under 60<br>• No history of blood clots | **CRITERIA**<br>• Age 60 or older<br>• History of thrombosis (blood clots) | **CRITERIA**<br>• History of arterial thrombosis (such as stroke or heart attack), OR<br>• A disease that is progressive or difficult to control despite treatment |
| **FOCUS:**<br>• Keeping hematocrit below 45%<br>• Low-dose aspirin (if appropriate)<br>• Regular monitoring | **MAY REQUIRE:**<br>• Cytoreductive therapy (such as interferon or hydroxyurea)<br>• JAK inhibitor therapy in certain cases<br>• Closer monitoring to maintain safe blood counts<br>• Additional strategies to reduce clot risk | **REQUIRES:**<br>• More intensive treatment approaches<br>• Careful monitoring and aggressive clot prevention strategies<br>• Consideration of additional treatment options or clinical trials |
| Treatment may be less intensive, but ongoing management is still required. | | |

## Other Factors That Affect Your Risk

Even within the same risk category, doctors consider additional factors that influence your overall clot risk. These include **high blood pressure, diabetes, high cholesterol, obesity, and smoking**.

These factors do **not change your formal risk category**, but they may affect how closely you are monitored and how aggressively your condition is treated. Managing these conditions is an important part of your overall PV care.

### Key Message

**Your risk level is only part of the picture.** Other health factors can also influence your care and treatment decisions.

### Ask Your Doctor

*"Which risk category am I in, and what other factors might affect my treatment plan?"*

## Pregnancy and PV

Many women with PV have questions about fertility and pregnancy. Managing PV during pregnancy requires careful planning because some treatments may harm a developing baby. **Hydroxyurea and rusfertide should not be used during pregnancy**. If you are considering pregnancy, talk with your doctor so your treatment can be safely adjusted.

When treatment is needed during pregnancy, **interferon is usually the preferred option** because it can control blood counts and has been used safely in pregnancy. Low-dose aspirin is often continued during pregnancy to help reduce the risk of blood clots, unless your doctor advises otherwise. Care is typically coordinated between a hematologist and a high-risk obstetric specialist to help protect both mother and baby.

### Questions to Ask Your Doctor

- What is my risk category?
- Should I consider seeing an MPN specialist?
- Is my hematocrit at goal (below 45%)?
- Would I benefit from cytoreductive therapy?
- Are there newer treatment options I should know about?

### Key Message

If you are of reproductive age and considering pregnancy, ask your hematologist to discuss your treatment plan and monitoring approach well in advance.

# What Treatments Are Available for PV?

Treatment for polycythemia vera aims to reduce the risk of blood clots, control blood counts, and manage symptoms. The primary goal is to keep the hematocrit below 45% and maintain blood counts in a safe range.

Treatment decisions are guided by risk level, age, overall health, and response to prior therapies.

| Treatment | Who Uses This | What It Helps | Possible Side Effects |
|---|---|---|---|
| **Phlebotomy**<br>*Often the foundation of treatment and usually combined with low-dose aspirin.* | Most PV patients, particularly at diagnosis | Lowers hematocrit, reduces blood thickness, and lowers clot risk | Temporary fatigue, dizziness, lightheadedness; iron deficiency with repeated treatment |
| **Low-dose Aspirin**<br>*Typically 81 mg daily.* | Most PV patients, unless their doctor advises otherwise | Reduces the risk of blood clots | May increase bleeding risk; stomach irritation |
| **Ropeginterferon alfa-2b** (Besremi®)<br>*First interferon FDA-approved for PV. A long-acting injection given every few weeks. May target the underlying disease.* | Adults with PV, regardless of treatment history | Normalizes blood counts and may reduce the JAK2 mutation burden over time; some patients need fewer or no phlebotomies | Flu-like symptoms, fatigue, mood changes or depression, injection-site reactions |
| **Peginterferon alfa-2a** (Pegasys®)<br>*Used off-label. A recommended alternative when ropeginterferon is unavailable or not covered by insurance.* | PV patients, especially younger patients | Normalizes blood counts and may reduce the JAK2 mutation burden | Flu-like symptoms, fatigue, mood changes or depression, injection-site reactions |
| **Hydroxyurea** (Hydrea®)<br>*Historically, one of the most commonly used cytoreductive therapies for higher-risk PV. Used off-label. Should not be used during pregnancy.* | Higher-risk PV patients, often older adults | Reduces blood cell production and decreases the risk of blood clots | Low blood counts, fatigue, mouth sores, increased risk of non-melanoma skin cancers |
| **Ruxolitinib** (Jakafi® / Jakafi XR®)<br>*A JAK inhibitor FDA-approved as second-line therapy for PV. Available as twice-daily tablets (Jakafi) or as once-daily extended-release tablets (Jakafi XR), approved in 2026 for more convenient dosing.* | Patients who are intolerant of or no longer respond to hydroxyurea | Reduces spleen size, controls blood counts and hematocrit which may reduce or eliminate the need for phlebotomy, and improves symptoms, including itching | Low blood counts (anemia, low platelets), increased infection risk, dizziness |
| **Rusfertide** (Mimrylo®)<br>*First-in-class hepcidin mimetic that limits iron availability. Weekly self-injection under the skin. FDA-approved for PV in 2026. Should not be used during pregnancy.* | Adults with PV who still need frequent phlebotomy to keep hematocrit controlled; used along with standard care | Controls hematocrit and reduces or eliminates the need for phlebotomy; may also reduce fatigue | Injection-site reactions (common, usually mild and lessen over time), low iron levels or anemia, fatigue |

Note: Some patients develop **hydroxyurea resistance or intolerance**. Resistance means the medication no longer adequately controls blood counts, such as when hematocrit remains too high or frequent phlebotomies are still needed despite the highest safe dose. Intolerance means the drug causes significant side effects. In these cases, doctors may recommend switching to another treatment, such as **interferon-based therapy or ruxolitinib (Jakafi®)**.

## Ask Your Doctor

*"Could I benefit from a clinical trial exploring new treatment options?"*
*Visit clinicaltrials.gov to search for open PV studies.*

# Living With PV & Looking Ahead

Polycythemia vera is a chronic condition, but with appropriate monitoring and treatment, many people live long and active lives. Ongoing care helps reduce complications and maintain quality of life.

## Regular Monitoring

**Regular follow-up is an essential part of managing PV.**

- Blood count checks (frequency depends on disease control and risk level)
- Monitoring hematocrit to keep it below 45%
- Symptom assessments, sometimes using standardized tools such as the MPN-SAF symptom score, to track changes in fatigue, itching, and other symptoms over time.
- Spleen size evaluation
- Watching for treatment side effects and complications

## Complications to Watch For

**Understanding potential complications allows for early intervention.**

- **Thrombosis** (blood clots): The most significant risk in PV
- **Bleeding**: Can occur with very high platelets or from aspirin therapy
- **Progression to myelofibrosis** (MF): Approximately 10–20% over 15–20 years
- **Transformation to acute myeloid leukemia** (AML): Rare but serious (~2–5% over 15–20 years)

## Lifestyle & Symptom Management

**Daily habits can support overall health and reduce risk.**

- Stay well hydrated
- Avoid smoking (increases clot risk)
- Manage cardiovascular risk factors (blood pressure, cholesterol, diabetes)
- Exercise regularly, as tolerated
- Cool or lukewarm showers if itching is problematic
- Seek mental health support when needed, as adjusting to a chronic diagnosis can be challenging

## You Are Not Alone

**Connecting with others and accessing reliable information can make a meaningful difference.**

- MPN Cancer Connection
- PV Reporter
- MPN Research Foundation
- Blood Cancer United (formerly LLS)
- Patient community forums and copay assistance programs

## Caring for someone with PV?

This guide is for you, too. Understanding the diagnosis helps you support the person you care about and take part in conversations with their care team.

## Final Message

PV is a chronic condition that requires ongoing management, but with appropriate treatment and monitoring, many patients live long and active lives. Studies suggest that when PV is well controlled, life expectancy can approach that of the general population. Stay informed, work closely with your healthcare team, and connect with the MPN community for support.

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**Medical disclaimer**. This guide is for general educational purposes only and is not a substitute for professional medical advice, diagnosis, or treatment. Always talk with your hematologist or healthcare team about your individual situation before making any decisions about your care.

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