

# You've been diagnosed with PV

## WHAT'S NEXT?

**Empower yourself with information to help you on your journey with polycythemia vera (PV).**

A PV diagnosis can be shocking. PV is a rare disease and you've likely never heard of it before. With this kit, you can learn more about PV so you are better able to advocate for yourself and manage your health and potential risks—today and tomorrow.

The What's Next PV Self-Advocacy Kit includes:



**KEY FACTS**  
about PV

EXPLORE



**SYMPTOMS**  
and RISKS

LEARN



**QUESTIONS**  
to ask your doctor

ASK



self-advocacy  
**HEALTH**  
**TRACKERS**

GO



**WORDS OF**  
**WISDOM**  
from PV patients

READ

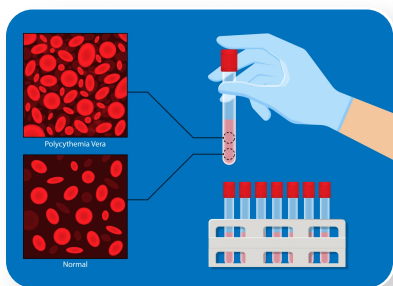


**RESOURCES**  
related to PV  
and other  
blood disorders

MORE



## KEY FACTS ABOUT PV



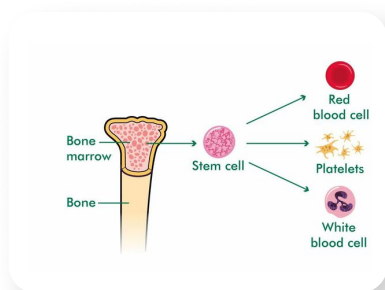
### What is PV?<sup>1-4</sup>

- PV is a rare, chronic blood cancer involving the production of too many blood cells (red blood cells, white blood cells, and platelets) by stem cells in the bone marrow (called myeloproliferation)
- Having too many blood cells causes blood to thicken and increase in volume (which is called hyperviscosity)

PV affects  
between  
**44 and 57 out of 100,000**  
people in the US<sup>1</sup>

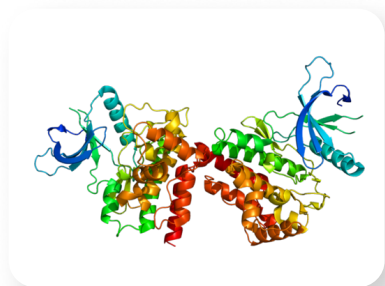
Approximately  
**160,000 people**  
in the US  
may be living with PV<sup>5</sup>

PV most often occurs in people  
**over 60 years old.**  
It's extremely rare  
in people **under 20**<sup>1</sup>



### What causes PV?<sup>1-8</sup>

- Stem cells in the bone marrow are responsible for producing different blood cells (red blood cells, white blood cells, and platelets)
- PV develops when stem cells in the bone marrow develop a mutation (or DNA change) that leads to an overproduction of blood cells



### About the mutation<sup>1-8</sup>

- Typically, the mutation in the stem cells is not inherited. It is not known why it happens
- Researchers have found that over 90% of people with PV have a mutation in the JAK2 gene
- JAK2 is a gene that makes a protein that sends signals to cells to promote cell growth and helps control the number of blood cells that are made in the bone marrow
- Because PV is caused by a mutation of stem cells in the bone marrow, it is considered a blood cancer



## SYMPTOMS AND RISKS

### What are the symptoms?<sup>1-3</sup>

It's common to get diagnosed with PV during an annual checkup without experiencing any symptoms. However, symptoms of PV usually develop slowly over many years. Many people have a variety of general symptoms (headaches, fatigue, weakness, etc.), that may progress to more serious issues.

### Common symptoms include<sup>1-3</sup>:



Headaches



Dizziness



Gastrointestinal  
issues



Abnormal redness of skin  
(especially on face)



Weakness



Itchy skin - pruritis  
(especially after hot shower)



Blurred vision



Burning sensation  
in feet



Fatigue



Unusual bleeding  
(eg, nose, gums)



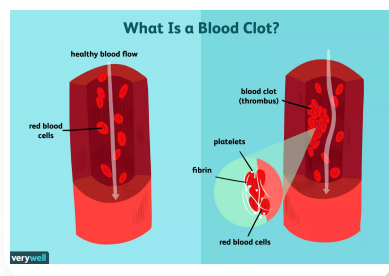
Ringing in ears  
(tinnitus)



## SYMPTOMS AND RISKS

### What short- and long-term risks should I be aware of?

Over time, as PV progresses, there are a number of potential issues to be aware of:



#### Blood Clots<sup>1,3</sup>

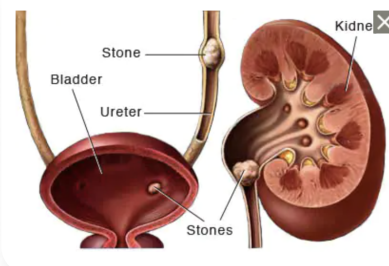
Reduced blood flow (due to thickening of the blood) can increase a person's risk of developing blood clots.

- Sometimes a blood clot may be the first obvious sign of PV
- Blood clots occur in about 30% of patients even before the PV diagnosis
- is made
- During the first 10 years after a diagnosis, 40%-60% of untreated PV patients may develop blood clots

A blood clot can cause:

- Stroke
- Chest pain (angina)
- Heart attack
- Deep vein thrombosis (DVT) (in the legs)
- Pulmonary embolism (in the lungs)

For older patients with PV who may also suffer from vascular disease, there may be an increased risk for stroke, heart attack, and pulmonary embolisms.



#### Kidney Stones<sup>1,9</sup>

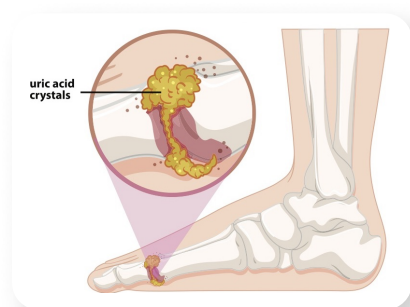
- The high turnover of red blood cells with PV leads to higher-than-normal uric acid production
- Kidney stones are associated with higher-than-normal uric acid production
- Passing kidney stones can be extremely painful, but the stones usually cause no permanent damage
- In many cases, treatment involves pain medication and drinking plenty of water
- If stones become lodged in the urinary tract, surgery may be needed





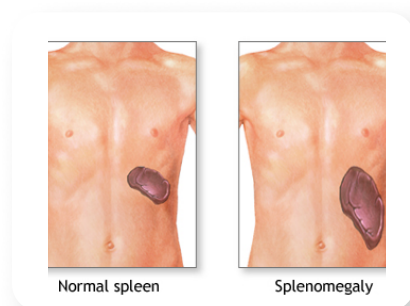
## SYMPTOMS AND RISKS

### Short- and long-term risks (cont'd)



#### Gout<sup>1,10</sup>

- Gout is an inflammation of the joints caused by a build up of uric acid
- Gout can cause intense joint pain, most commonly in the big toe, ankles, knees, elbows, wrists, and fingers



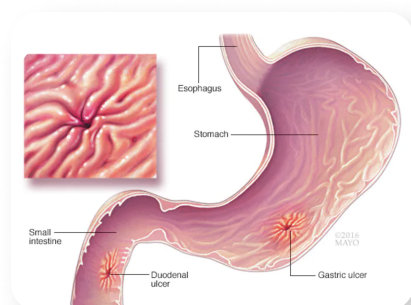
#### Enlarged Spleen<sup>1,11</sup>

- The spleen is an organ in the abdomen that filters out blood cells
- The spleen often becomes enlarged in people with PV because it's working overtime to clear a greater number of blood cells than normal
- An enlarged spleen (splenomegaly) may cause a bloated or full feeling in the abdomen
- It can also lead to more frequent infections, anemia, and/or increased bleeding
- An enlarged spleen is also much more likely to rupture, which can cause life-threatening bleeding in the abdominal cavity



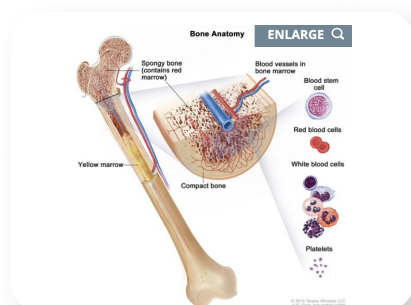
## SYMPTOMS AND RISKS

### Short- and long-term risks (cont'd)



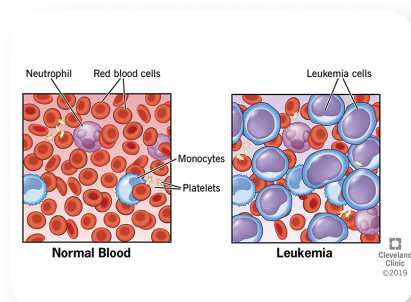
#### Peptic Ulcers<sup>1,12</sup>

- Peptic ulcers are open sores on the lining of the gastrointestinal tract
- Left untreated, they can cause internal bleeding, a hole in the stomach wall, or an obstruction in the digestive tract causing you to become full easily, vomit, and/or lose weight



#### Scarring of Bone Marrow<sup>1,3,8</sup>

- Bone marrow is the soft, spongy tissue that fills the center of most bones
- When PV is uncontrolled, it may transform into MF (myelofibrosis), which is scarring in the bone marrow
- Between 10%-21% of PV patients may develop MF within 10 years
- When this scarring occurs, the marrow can no longer produce blood cells, which results in low levels of healthy, functioning red blood cells (anemia), platelets (thrombocytopenia), and white blood cells (leukopenia)



#### Leukemia<sup>2,3,8,13,14</sup>

- When PV is uncontrolled and scar tissue begins to replace bone marrow, abnormal bone marrow cells may begin to grow out of control
- This can lead to a cancer of the blood and bone marrow called acute myeloid leukemia (AML)
- Up to 10% of PV patients may develop AML within 10 years
- AML often quickly moves from the bone marrow into the blood, and can sometimes spread to other parts of the body, including the lymph nodes, liver, spleen, and central nervous system (brain and spinal cord)



## SELF-ADVOCACY HEALTH TRACKERS

Now that you understand more about polycythemia vera (PV), what can you do to take charge of your health? You play a huge part in how you manage your PV. Together, with your doctor, you can address your symptoms and develop a treatment strategy that's right for you.

### Your Blood Tests

A complete blood count (CBC) measures several components and features of your blood (listed below<sup>15</sup>). Enter your results to track your levels over time.


TEST	WHAT IT IS <sup>15</sup>	DATE	RESULT
Red blood cells (RBC)	These blood cells carry oxygen	00 / 00 / 1900	
Hematocrit (HCT)	The proportion of red blood cells to the fluid component, or plasma, in your blood	00 / 00 / 1900	
Hemoglobin (HGB)	The oxygen-carrying protein in red blood cells	00 / 00 / 1900	
White blood cells (WBC)	These cells help fight infection	00 / 00 / 1900	
Platelets	Help with blood clotting	00 / 00 / 1900	



## SELF-ADVOCACY IN ACTION

### Symptom Tracker

Place a checkmark next to the symptom you're experiencing, then specify whether it is a new symptom or if it has worsened. Use the notes section for when you talk with your doctor about how to manage your symptoms.

SYMPTOM	NEW SYMPTOM?	WORSENERD SYMPTOM?	NOTES
Headache 			
Fatigue 			
Weakness 			
Dizziness 			
Blurred vision 			
Itchy skin 			
Unusual bleeding (eg, nose, gums) 			
Gastrointestinal issues 			
Ringing in the ears 			
Abnormal redness of the skin 			
Burning sensation in the feet 			
Other			



## QUESTIONS TO ASK YOUR DOCTOR

Here are questions to ask your doctor as you work together on managing your disease:

- 1 How often should I come in for a PV-related visit?**
- 2 How will you monitor my PV? How should I monitor it?**
- 3 How often will I be seeing you and getting complete blood counts (CBCs)?**
- 4 At what levels do you want to keep my hematocrit (HCT), hemoglobin (HGB), white blood cells (WBC), and platelets?**
- 5 How do you treat various symptoms associated with PV (eg, itching)?**
- 6 What types of symptoms or changes should I notify you about?**
- 7 Have you treated people who have PV with anything other than phlebotomies and aspirin or hydroxyurea?**
- 8 What can I do to proactively manage my PV?**
- 9 Will you order a bone marrow biopsy at some point? If so, what will trigger it to be done?**
- 10 Will you test me for JAK2 mutations and other variants implicated in research?**
- 11 If my PV is secondary, what tests will you be performing to find the cause?**
- 12 If I consult with a specialist in myeloproliferative neoplasms (MPNs), are you willing to coordinate with them?**

### Is your doctor an MPN specialist?

Many PV patients seek help from an MPN specialist. Here's how to know if your doctor is a true MPN specialist:

- They have attended the American Society of Hematology (ASH) conference
- They have published peer-reviewed research papers about MPNs
- They have participated in clinical trials related to MPNs
- They stay abreast of the latest MPN research, including international studies
- They see a minimum of 50 MPN patients a year



## WORDS OF WISDOM FROM PV PATIENTS

We talked with real patients about their experience living with and managing PV. Here is some of their advice for newly diagnosed patients.



Start with an expert  
and go from there."



Ask a lot of questions to make sure that  
you understand exactly what's going on  
and what the newest options are available  
to you."



Learn to live with  
it and take  
responsibility."



Try to keep  
stress out of  
your life. Enjoy  
the little things."



It does get better.  
Learn to live with  
the new normal  
that is your life  
now. Life goes on."



Enjoy life. Rest when  
you're able to."



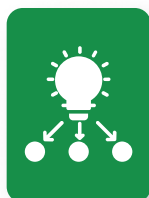
I joined some Facebook  
groups of people that have  
PV. I didn't feel so alone, and I  
learned a lot from some of  
these people."



Learn about it,  
understand it.  
You've got it."



I started seeking a second opinion because the  
more I started educating myself, I realized this is  
pretty serious and it's progressive."



## RESOURCES

related to PV and other blood disorders



**PV Reporter**  
**PVReporter.com**



**Patient Power**  
**PatientPower.info**



**MPN Advocacy and Education**  
**MPNAdvocacy.com**



**MPN Research Foundation**  
**MPNResearchFoundation.org**



**MPN Education Foundation**  
**MPNInfo.org**



**Leukemia and Lymphoma Society**  
**LLS.org**



**MPN Cancer Connection**  
**MPNCancerConnection.org**

### Additional Organizations with PV information

- National Institutes of Health: National Heart, Lung, and Blood Institute – **nhlbi.nih.gov**
- Mayo Clinic – **mayoclinic.org**
- Merck Manual – **merckmanuals.com**

### Interested in Clinical Trials?

You can search for clinical trials related to PV and other MPNs at **clinicaltrials.gov**



## REFERENCES

1. Polycythemia vera. National Organization for Rare Disorders (NORD). <https://rarediseases.org/rare-diseases/polycythemia-vera/>
2. Polycythemia vera. National Heart, Lung, and Blood Institute, National Institutes of Health. <https://www.nhlbi.nih.gov/health-topics/polycythemia-vera>
3. Polycythemia vera facts. Leukemia and Lymphoma Society. [https://www.lls.org/sites/default/files/file\\_assets/FS13\\_PolycythemiaVera\\_FactSheet\\_final5.1.15.pdf](https://www.lls.org/sites/default/files/file_assets/FS13_PolycythemiaVera_FactSheet_final5.1.15.pdf)
4. Polycythemia vera. Mayo Clinic. <https://www.mayoclinic.org/diseases-conditions/polycythemia-vera/symptoms-causes/syc-20355850>
5. United States Census Bureau. US and world population clock. Accessed June 15, 2021. <https://www.census.gov/popclock/>
6. Hematopoietic stem cell. National Cancer Institute. <https://www.cancer.gov/publications/dictionaries/cancer-terms/def/hematopoietic-stem-cell>
7. JAK2 gene. National Cancer Institute. <https://www.cancer.gov/publications/dictionaries/cancer-terms/def/jak2-gene>
8. Tefferi A. *Am J Hematol*. 2008;83:491-497.
9. Kidney Stones. Mayo Clinic. <https://www.mayoclinic.org/diseases-conditions/kidney-stones/symptoms-causes/syc-20355755>
10. Gout. Mayo Clinic. <https://www.mayoclinic.org/diseases-conditions/gout/symptoms-causes/syc-20372897>
11. Enlarged spleen (splenomegaly). Mayo Clinic. <https://www.mayoclinic.org/diseases-conditions/enlarged-spleen/symptoms-causes/syc-20354326>
12. Peptic ulcer. Mayo Clinic. <https://www.mayoclinic.org/diseases-conditions/peptic-ulcer/symptoms-causes/syc-20354223>
13. Finazzi G, et al. *Blood*. 2005;105(7):2664-2670
14. What is acute myeloid leukemia (AML)? American Cancer Society. <https://www.cancer.org/cancer/acute-myeloid-leukemia/about/what-is-aml.html>
15. Complete blood count (CBC). Mayo Clinic. <https://www.mayoclinic.org/tests-procedures/complete-blood-count/about/pac-20384919>