



The Myeloproliferative neoplasms (MPNs) are a group of blood cancers. MPNs are a chronic condition and tend to worsen over time. The term includes three diseases: essential thrombocythemia (ET), polycythemia vera (PV), and myelofibrosis (MF). This fact sheet provides information on polycythemia vera.

People living with PV and their caregivers often feel isolated and are unsure where to go for help and facts about the disease. Talk with your health care team about the

best sources of information on living with PV. The good news is that the number of resources for people with MPNs is growing.

If you or a loved one has been diagnosed with PV, you may have many questions about the disease, treatment options, and how the diagnosis affects your quality of life.



# What is Polycythemia Vera?

Polycythemia vera (PV) is a rare type of blood cancer. It is a type of cancer known as myeloproliferative neoplasms (MPNs). PV is usually a result of a mutation in blood-forming stem cells, which decreases the body's ability to limit the production of blood cells (particularly red blood cells). Higher-than-normal numbers of red blood cells can cause the blood to be thicker than normal. This slows down the movement of blood through the veins and arteries, increasing the risk for blood clots in veins, as well as heart attacks and strokes.

Researchers can't yet identify what causes PV. They do know, however, that in nearly all cases, people who are diagnosed with PV have a specific genetic mutation of the JAK2 gene in their bone marrow stem cells.

Although polycythemia vera is associated with a genetic mutation, it is not an inherited disease. This is because the mutation is generally only found in the stem cells, not in the rest of the body's cells, such as the sperm or eggs. Scientists are unsure

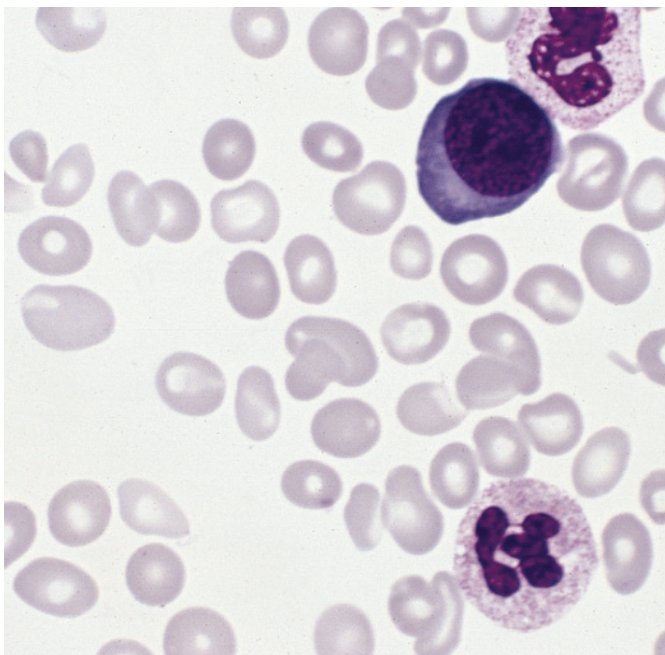
what leads to the mutation that causes PV. Research is currently underway to determine the exact cause.

Scientists do know that the disease affects slightly more men than women and is more commonly found in people in their 60s, 70s, and 80s.

There is no cure for polycythemia vera. It is considered a chronic and progressive condition, but the effects of the disease can be managed with appropriate medical care. Over decades, there is some risk that the disease will transform into a more aggressive form of blood cancer.

Occasionally people with PV develop acute leukemia or a condition called secondary myelofibrosis. Scientists are seeking ways to preventing or limit PV from transforming into these conditions.

In general, however, PV develops and progresses very slowly. Some people may live with PV for many years without knowing it because they do not experience any symptoms. In many cases, once PV is diagnosed it can be controlled for many years without causing significant changes in a person's life expectancy or quality of life.



BLOOD SMEAR FROM A PATIENT WITH POLYCYTHEMIA VERA

*“One of the things about polycythemia vera, and all the MPNs, is that the symptoms that folks have don’t always correspond to blood counts—so be sure to mention symptoms to your doctor.”*

*—Dr. Laura C. Michaelis*

## Symptoms

When polycythemia vera develops, and there is an increase in the quantity of red blood cells, patients can have a number of symptoms. Some are due to the thickness of the blood, and some due to abnormal chemicals made in the bone marrow. Some people have few symptoms. Others may experience one or more of the following at some point in their disease:

- Blood clots in the legs, lungs, or other areas
- Headaches
- Excessive sweating
- Blurred vision or blind spots
- Dizziness
- Itchiness, especially after a warm shower or bath
- Reddened appearance of skin
- Peptic ulcers
- Bloating or feeling of fullness due to enlarged spleen
- Congestive heart failure or angina
- Gout/inflammation of joints

## Your Health Care Team

An important decision you will make is choosing a doctor and team that has the expertise to treat PV. It's critical to feel confident in the people who are treating you and to be able to communicate with them. You can always seek a second opinion or change doctors if your situation changes and you need a different level of care.

Questions to consider as you choose your team:

- Is your doctor board certified in hematology or only oncology?
- Does your doctor specialize in treating myeloproliferative neoplasms?
- Does your doctor coordinate your treatment plan with a range of other health care professionals?
- Does your treatment center offer clinical trials?
- What is the best way to reach your doctor and health care team? Do they have an oncology social worker or navigator available?
- Do they use a patient portal where you can track your medical information?

## DIAGNOSIS AND STAGING

**Blood tests are used to diagnose polycythemia vera. If you have PV, bloods tests may indicate:**

- An increase in red blood cells, platelets, and/or white blood cells
- An elevated hematocrit (the proportion of red blood cells in the blood)
- An increase in hemoglobin concentration (a protein found in red blood cells)
- Low levels of erythropoietin (a hormone that controls red blood cell production)

**Additional tests may include:**

- A bone marrow biopsy and aspirate
- Testing for the presence of JAK2 mutation or other genetic mutations

# Treating Polycythemia Vera

There may be several health care providers involved in your care. These can include a hematologist/oncologist, nurse, nurse navigator, oncology social worker, and pharmacist. Each of these professionals can help guide you to information and resources that may help you both during and after your treatment. If you do not meet these professionals upfront, ask your doctor how you can arrange an appointment with

them. You may also want to ask your insurance carrier to ask for a case manager to be assigned to you to help you manage your treatment coverage options.

There are several approaches to treatment depending on your PV diagnosis and its progression. There are various drugs used in the treatment of PV. Many of them have very different ways of slowing the disease process and/or controlling the effects of the disease.

## TREATMENT OPTIONS

**PHLEBOTOMY**—Some people can safely avoid taking any cancer medication for their PV, but nearly all patients need to have blood drawn to keep their red blood cell count under control. This procedure is called phlebotomy, which means removing blood from a vein in order to reduce the amount of overall red blood cells in the body. This procedure can reduce symptoms, and prevents blood clots, heart attacks, and strokes from occurring. Your doctor will talk to you about how frequently you should have phlebotomy.

**ASPIRIN**—Nearly all people with PV should take a baby aspirin or its equivalent, unless there are specific medical reasons why they shouldn't, or if they are on another blood thinner.

While these are not anti-cancer therapies, they are very helpful in preventing some of the common complications of PV.

**HYDROXYUREA**—This therapy is used with phlebotomy to reduce the blood counts. It is not necessary for everyone, but it very helpful when a person has certain risk factors. The medicine is given in the form of a tablet.

**INTERFERON**—This medicine is also used to reduce blood counts. It is not necessary for everyone, but can be helpful in selected cases. This medicine is a shot the patient administers to themselves on a weekly or every-other-week schedule.

**JAK INHIBITORS**—These targeted therapies reduce symptoms. Ruxolitinib (Jakafi®) can be used to treat PV when hydroxyurea does not work.

# Clinical Trials

Ask about clinical trials. Clinical trials are research studies to test new treatments or learn how to use existing treatments better. Today's clinical trials may become tomorrow's newest treatments.

## KEY THINGS TO KNOW

■ A clinical trial may be the only way to get some of the newest, most promising treatments.

- The U.S. Food and Drug Administration and local review boards oversee all U.S. clinical trials to keep patients safe.
- If you join a clinical trial, you can leave at any time.
- Every doctor does not have the same trials.
- Most often, the trial pays the costs of the drug being studied and your health insurance only has to pay for “standard” treatment costs. However, your health insurance may not pay for everything. Be sure to ask.

Learn more about Clinical Trials by visiting [www.CancerSupportCommunity.org/ClinicalTrials](http://www.CancerSupportCommunity.org/ClinicalTrials).

## QUESTIONS TO ASK YOUR HEALTH CARE TEAM:

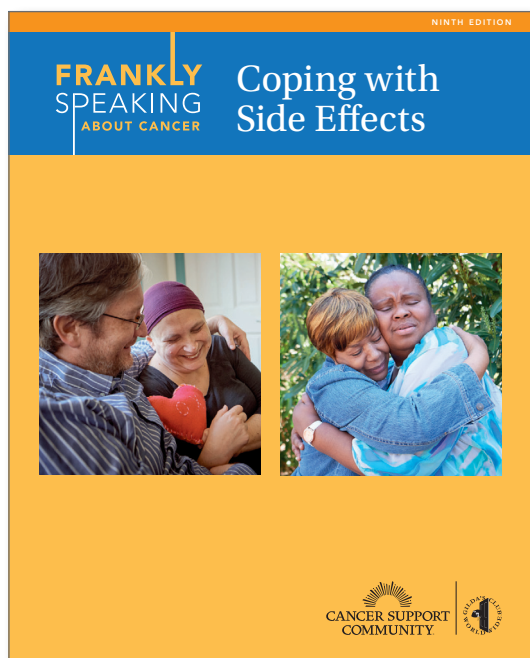
- Which treatment(s) do you recommend? What is the goal of the treatment? How will we know if the treatment is working?
- How often will I need phlebotomy sessions? How can we keep my hematocrit in a safe range? Will there be side effects to the phlebotomy?
- Are there dangers to taking aspirin?
- Is stem-cell transplant ever used in this disease?
- When will I need to start chemotherapy, like hydroxyurea? What side effects should I expect? Can I prevent or treat any of these side effects?
- Will there be long-lasting side effects? What can I do about them?
- How will I know if my treatment is working?
- How often will I need check-ups?
- What information do you have so I can learn more about this treatment?
- What if my cancer progresses or transforms to leukemia or myelofibrosis?

## Side Effect Management

It helps to learn more about the side effects from your treatment(s) before you begin, so you will know what to expect. When you know more, you can work with your health care team to manage your quality of life during and after treatment.

Tell your health care team about any side effects you may have so they can help you manage symptoms. Everyone reacts differently to treatment and experiences side effects differently.

Side effects from treatment can be short-term and go away even without stopping the treatment. Other side effects only go away when the therapy is stopped. Side effects of treatment may be long term. Ask your doctor if you might have any of these side effects and how to manage them.



An educational booklet and personal planner combination designed to help people learn about cancer treatments and manage the potential physical side effects from treatment, as well as the emotional burden. Find *Frankly Speaking About Cancer: Coping with Side Effects* at [www.CancerSupportCommunity.org/SideEffects](http://www.CancerSupportCommunity.org/SideEffects).



## Challenges of Living with Polycythemia Vera

People can live with polycythemia vera for many years without having any symptoms. This means that for some people who have been diagnosed with PV, but who don't have symptoms, doctors may advise against starting any kind of chemotherapy, but will start phlebotomy and aspirin therapy.

People living with PV can have feelings of worry and fear about living with a chronic condition. They may have concerns about whether their cancer will transform into more aggressive disease like acute leukemia or myelofibrosis.

Living with this uncertainty can be stressful. It is important to bring up any concerns you have with your doctor. By working with your health care team, you can take charge and maintain your quality of life.

## Here are some tips to help you manage your disease and your life:

**DON'T TRY TO DO EVERYTHING AT ONCE.** Stay in the moment, as much as possible. Focus on what is happening now, and what you need to do to address that situation.

**ASK FOR HELP.** Many of your family and friends will want to be there for you. Be open about what they can do to support you. Be specific. Ask someone for a meal, to watch your children, give you a ride, or just be there on a tough day.

**TAKE SOMEONE WITH YOU** to medical appointments and procedures. An extra set of eyes and ears often helps to remember and interpret the information you are given.

**MAKE TIME FOR YOURSELF** and the things you enjoy. It is important to continue to do what you love as much as possible. For some people, that is as simple as taking a walk or planting some flowers. For others, it may mean continuing to work. You may have to adjust your activities to fit your energy level and physical condition, but it's important to keep doing them.

**TRY MIND/BODY ACTIVITIES,** such as yoga or meditation. Whether it's breathing exercises to help reduce stress and anxiety or guided imagery techniques to focus the mind away from current concerns, mind/body exercises can benefit your overall well-being.

**GET SUPPORT.** Although polycythemia vera is a rare condition, there are support groups that focus on people who have been diagnosed with MPNs. These groups let you meet and learn from other people who have the same feelings and worries, compare situations, and seek opinions. Since PV is a chronic condition, talking to others who have similar side effects may be useful. Talking can help relieve concerns and feelings of isolation and loneliness that people living with a rare disease may often experience. Whether that support is found in-person, through online support groups, or telephone support groups, the benefits are the same: Receiving support helps lower anxiety.

# MPN Information, Survivorship & Support

**Cancer Support Community** • 888-793-9355 • [www.CancerSupportCommunity.org](http://www.CancerSupportCommunity.org)

**CancerCare** • 800-813-4673 • [www.cancercare.org](http://www.cancercare.org)

**The Leukemia & Lymphoma Society** • 800-955-4572 • [www.lls.org](http://www.lls.org)

**MPN Education Foundation** • [www.mpninfo.org](http://www.mpninfo.org)

**MPN Research Foundation** • 312-683-7249 • [www.mpnresearchfoundation.org](http://www.mpnresearchfoundation.org)

**Patient Advocate Foundation** • 800-532-5274 • [www.patientadvocate.org](http://www.patientadvocate.org)

**Unlocking Answers in MPN** • [www.mpnunlocked.org](http://www.mpnunlocked.org)

## CANCER SUPPORT COMMUNITY RESOURCES

The Cancer Support Community's (CSC) resources and programs are available free of charge. To access any of these resources below call 888-793-9355 or visit [www.CancerSupportCommunity.org](http://www.CancerSupportCommunity.org).

**Cancer Support Helpline®**—Have questions, concerns or looking for resources? Call CSC's toll-free Cancer Support Helpline (888-793-9355), available in 200 languages Mon–Fri, 9 am–9 pm ET.

**Open to Options®**—Preparing for your next appointment? Our trained specialists can help you create a list of questions to share with your doctor. Make an appointment by calling 888-793-9355 or by contacting your local CSC or Gilda's Club.

**Frankly Speaking About Cancer®**—Trusted information for cancer patients and their loved ones is available through publications, online, and in-person programs.

[www.CancerSupportCommunity.org/FranklySpeakingAboutCancer](http://www.CancerSupportCommunity.org/FranklySpeakingAboutCancer).

**Services at Local CSCs and Gilda's Clubs**—With the help of 170 locations, CSC and Gilda's Club affiliates provide services free of charge to people touched by cancer. Attend support groups, educational sessions, wellness programs, and more at a location near you. [www.CancerSupportCommunity.org/FindLocation](http://www.CancerSupportCommunity.org/FindLocation).

**Cancer Experience Registry®**—Help others by sharing your cancer patient or cancer caregiver experience via survey at [www.CancerExperienceRegistry.org](http://www.CancerExperienceRegistry.org).

**MyLifeLine**—CSC's private, online community allows patients and caregivers to easily connect with friends and family to receive social, emotional, and practical support throughout the cancer journey and beyond. Connect with other caregivers by joining the Caregiver Support online discussion board. Sign up at [www.MyLifeLine.org](http://www.MyLifeLine.org).

**Grassroots Network**—Make sure your voice is heard by federal and state policy makers on issues affecting cancer patients and survivors by joining our Network at [www.CancerSupportCommunity.org/become-advocate](http://www.CancerSupportCommunity.org/become-advocate).

**THIS PROGRAM WAS MADE POSSIBLE WITH GENEROUS SUPPORT FROM:**



This booklet is available to download and print yourself at [www.CancerSupportCommunity.org/MPNs](http://www.CancerSupportCommunity.org/MPNs). For print copies of this booklet or other information about coping with cancer, visit [Orders.CancerSupportCommunity.org](http://Orders.CancerSupportCommunity.org).

The Cancer Support Community and its partners provide this information as a service. This publication is not intended to take the place of medical care or the advice of your doctor. We strongly suggest consulting your doctor or other health care professionals to answer questions and learn more.

© June 2020. Cancer Support Community. All rights reserved. Photography by Photography by Jared Zagha.