

The Myeloproliferative neoplasms (MPNs) are a group of blood cancers. MPNs are a chronic condition and are prone to worsen over time. The term typically 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 information. It is important to talk with your health care team to find the correct information about living with PV and support to address long-term care issues.

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 questions about how the diagnosis affects your quality of life.



SUPPORT FOR FINDING INFORMATION ABOUT MPNs

Cancer Support Community 888-793-9355 www.cancersupportcommunity.org

CancerCare 800-813-4673 www.cancercares.org

The Leukemia & Lymphoma Society 800-955-4572 www.lls.org

MPN Education Foundation www.mpninfo.org

MPN Research Foundation 312-683-7249 www.mpnresearchfoundation.org

Patient Advocate Foundation 800-532-5274 www.patientadvocate.org

What is Polycythemia Vera?

Polycythemia vera (PV) is a rare type of blood cancer and typically included in the category of cancers 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, slowing down the movement of blood through the veins and arteries, increasing the risk for venous blood clots, heart attacks or strokes.

Researchers can't yet identify what events lead to a person developing PV. They do know, however, that in nearly all cases, individuals who are diagnosed with PV can be shown to have a specific genetic mutation of the JAK2 gene in their bone marrow stem cells.

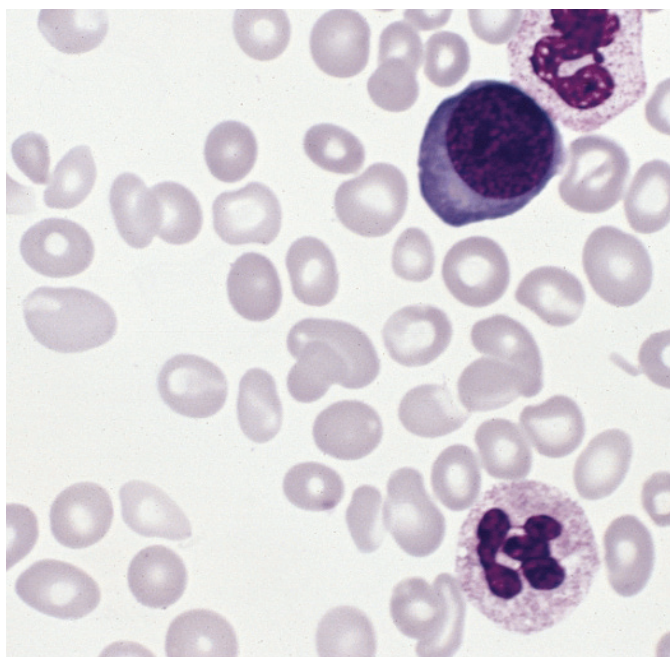
Although polycythemia vera is associated with a genetic mutation, it is not an inherited disease, since it is generally only found in the stem cells, not in the rest of the body's cells, such as the sperm or eggs. At this time, scientists are unsure what leads to the mutation to cause PV, however more 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, rather than younger people.

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 and interventions. Over decades, there is some risk that the disease will transform into a more aggressive form of blood cancer.

Occasionally individuals develop acute leukemia or a condition called secondary myelofibrosis. Preventing or limiting the risk for these transformations to more aggressive disease is one of the most important research priorities in this condition.

In general, however, PV develops and progresses very slowly. In fact, 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 and managed 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 due to the viscosity, or 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 about your care 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. Remember 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 use a multidisciplinary team approach in coordinating your treatment plan?
- 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 including 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 might arrange an appointment with them. In addition, you may want to contact your insurance carrier to ask for a case manager to be assigned

to you to assist in managing 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, and many of them have very different approaches to slowing the disease process and/or controlling the effects of the disease. With any drugs, side effects may occur. Your doctor or nurse should review these and what you can expect from the drug before initiating the medication.

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 just 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, but more importantly prevents blood clots, heart attacks and strokes from occurring. Your doctor will talk to you about how frequently phlebotomy should occur.

ASPIRIN Nearly all people with PV, unless there are contraindications or the patient is on another blood thinner, should take a baby aspirin or its equivalent. 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 in conjunction with phlebotomy to reduce the blood counts. It is not necessary for everyone, but it very helpful when a person has certain higher risk factors. The medicine is given in the form of a tablet.

INTERFERON This medicine is not officially chemotherapy, but is also used to reduce blood counts. It is again 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 A new class of drugs that target the JAK2 mutation has been approved by the FDA (Food & Drug Administration) for the treatment of PV and can be used occasionally when patients have intolerable side effects to other treatments or when the other treatments fail to work.

“I went to a doctor who treats people with MPNs. She gave me information and then I also did my research before deciding on my treatment.”

— Olivia, living with an MPN

CLINICAL TRIALS

Clinical trials are the engine that drives progress in cancer treatment. These studies compare the existing standard of care—the best known available treatment for any cancer—with new drugs, agents or procedures that may offer improved outcomes. Never hesitate to ask your doctor whether there is a clinical trial that might benefit you.

If you participate in a clinical trial, you will:

- Receive either the standard of care or the new treatment being studied. Be fully informed before entering the trial of any potential side effects. Be fully informed of the goals and aims of the trial.
- Receive a very high level of ongoing care and monitoring for your cancer.
- Your doctor should be your partner in working with you to find a trial that fits your cancer and your goals. For more information or to find a clinical trial, visit www.cancersupportcommunity.org.

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 hydrea®? What side effects should I anticipate? 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.

Communicate with your health care team about any side effects you may be experiencing so they can help you manage symptoms and help you cope. Keep in mind that 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 resolve when the therapy is stopped. Talk with your health care team about any side effects you experience, so they can help you manage them.

There are side effects of treatment that may be long term. It is important to talk with your doctor to know about these side effects from treatment to help you cope and manage.



Challenges of Living with Polycythemia Vera

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

People living with PV can experience feelings of worry and fear about living with a chronic condition and experience 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 talking and working with your health care team, you can take charge and maintain your quality of life.

“PV is a rare condition. That makes connecting with others who have it all the more useful. Informed and engaged patients improve not only their own care, but can help contribute to improving the care of the larger community of people with rare medical disorders.”

— Dr. Laura C. Michaelis

You will find your own path and develop your own strategies to manage your disease and your life, but here are some more tips that can help:

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. It is useful to 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. This is important because 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 don't lose sight of the importance of continuing to do what you like to do in your life.

TRY MIND/BODY ACTIVITIES, such as yoga or meditation. These activities can be beneficial to a person's overall well-being. 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 provide benefit in reducing stress for people living with PV.

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 offer an opportunity to meet and learn from other people who are experiencing the same feelings and worries, compare situations and seek opinions. Since PV is a chronic condition, talking to others who are experiencing similar side effects may help a person living with PV to feel support and care. Talking is helpful in relieving concerns and any feeling 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.

General Cancer Information, Survivorship & Support

CancerCare 800-813-4673 www.cancer.org

Clinicaltrials.gov www.clinicaltrials.gov

The Leukemia & Lymphoma Society 800-955-4572 www.lls.org

MPN Education Foundation www.mpninfo.org

MPN Research Foundation 312-683-7249 www.mpnresearchfoundation.org

Patient Advocate Foundation 800-532-5274 www.patientadvocate.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 1-888-793-9355 or visit www.cancersupportcommunity.org

CANCER SUPPORT HELPLINE ®

Whether you are newly diagnosed with cancer, a long-time cancer survivor, caring for someone with cancer, or a health care professional looking for resources, CSC's toll-free Cancer Support Helpline (1-888-793-9355) is staffed by licensed CSC Helpline Counselors available to assist you Mon-Fri 9 am- 9 pm ET.

OPEN TO OPTIONS ®

If you are facing a cancer treatment decision, this research-proven program can help you. In less than an hour, our trained specialists can help you create a written list of specific questions about your concerns for your doctor.

FRANKLY SPEAKING ABOUT CANCER ®

CSC's landmark cancer education series provides trusted information for cancer patients and their loved ones. Information is available through publications, online, and in-person programs.

AFFILIATE NETWORK SERVICES

Over 50 locations plus more than 100 satellite locations around the country offer on-site support groups, educational workshops, and healthy lifestyle programs specifically designed for people affected by cancer at no cost to the member.

CANCER EXPERIENCE REGISTRY®

The Registry is a community of people touched by cancer. The primary focus of the Registry is on collecting, analyzing and sharing information about the experience and needs of patients and their families. To join, go to www.CancerExperienceRegistry.org.

A special thanks to our contributors and reviewers: Laura C. Michaelis, MD, Linda Zignego, RN, BSN, Froedert Hospital and Medical College of Wisconsin; and Rebecca Fritz, LCSW, Gilda's Club Chicago.

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.

THIS PROGRAM WAS MADE POSSIBLE THROUGH A CHARITABLE GRANT FROM:



© August 2015 Cancer Support Community. All rights reserved.

Design by Yolanda Cuomo Design, NYC; Cover Illustration © Anthony Russo