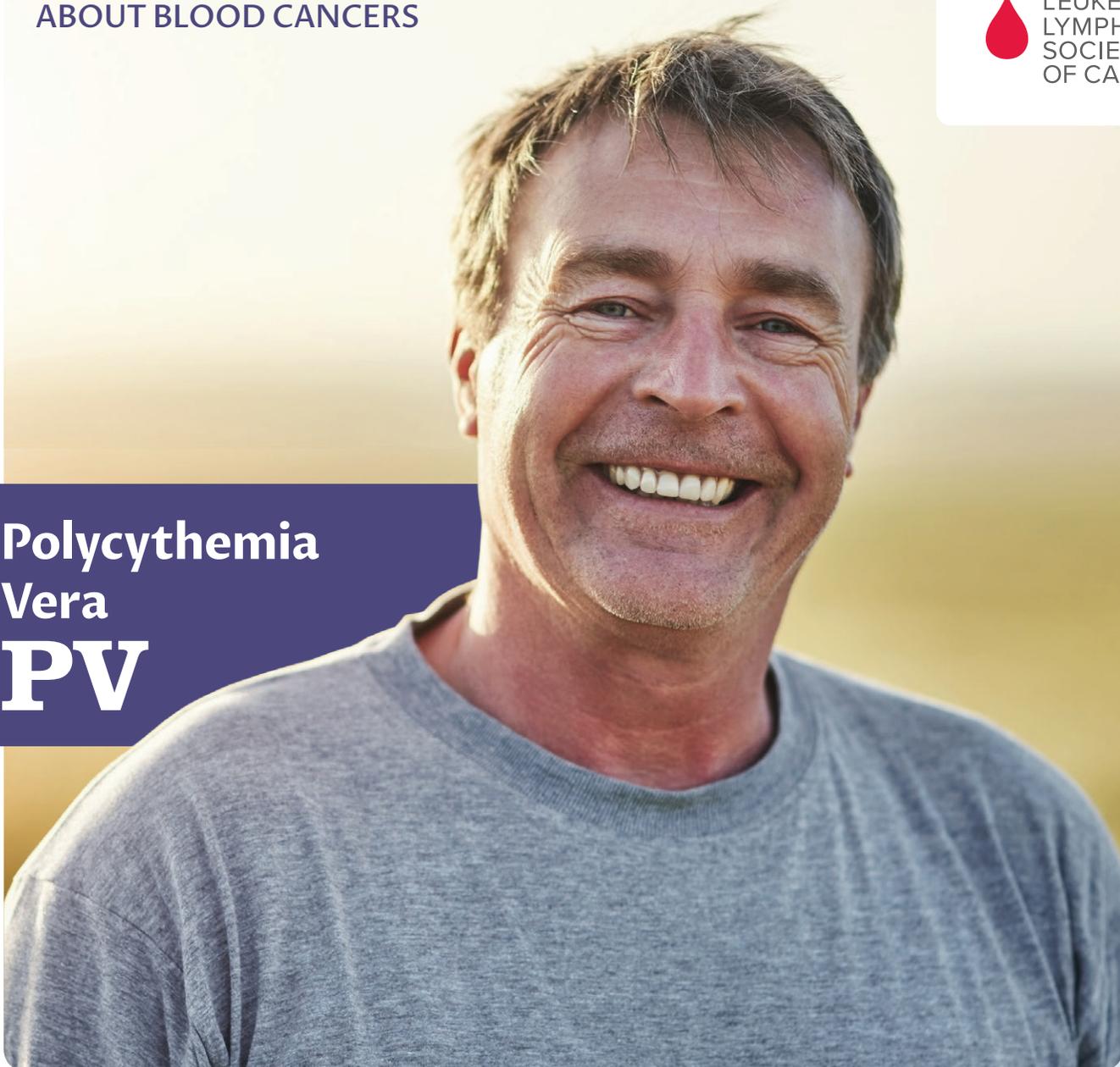


Polycythemia
Vera
PV



WHAT YOU NEED TO KNOW

You or your loved one has been diagnosed with polycythemia vera (PV). What does it mean and how will it affect you?

This fact sheet will help you:

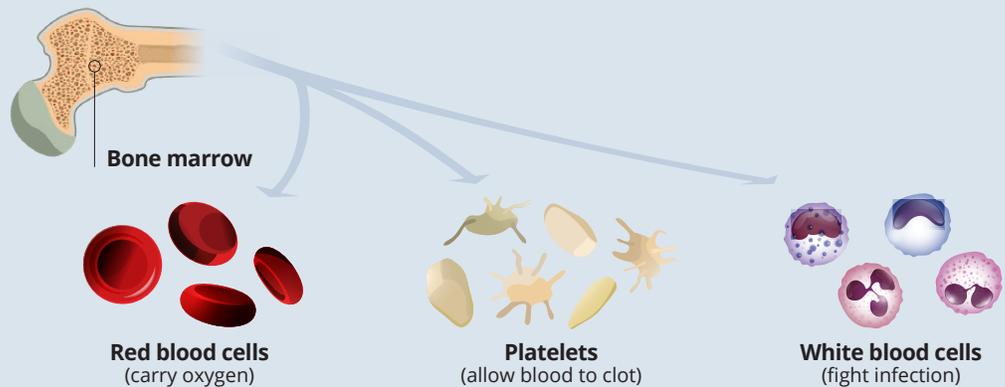
Learn about PV
and how it is
diagnosed

Get an overview
of treatment
options

Understand what
happens next

What is polycythemia vera (PV)?

PV is a type of myeloproliferative neoplasm (MPN). This group of blood cancers features too many blood cells of a particular type made in the bone marrow. MPNs begin with one or more changes to the DNA of a single stem cell in the bone marrow. These changes cause the stem cell to create more and more abnormal stem cells.



PV is a rare and chronic blood disease. With good medical care, you can manage your PV well for many years.

People with PV have too many oxygen-carrying red blood cells in the bone marrow. In many cases, the numbers of white blood cells and platelets are also high. White blood cells help your body fight infection, while platelets help your blood to clot (stop bleeding).

About PV

- 95% of people with PV have a genetic mutation of the JAK2 gene.
- It is more common in men and most often found in adults 60 and older.
- People with PV are at increased risk of heart attack, stroke, and blood clots in the legs, lungs, and abdomen (treatments help to reduce this risk).
- For a small number of people, PV may progress to a more aggressive blood disease over many years.

Signs and symptoms

PV develops slowly and may not cause symptoms for many years. It is often detected during a routine blood test. The signs and symptoms can be similar to other less serious diseases. You may experience:

- Itchy skin, especially after warm baths or showers
- Headaches, reddened skin, and excessive sweating
- Ringing in the ears, blurred vision, double vision, or seeing dark or blind spots that come and go
- Weakness, dizziness, fatigue, and feeling out of breath
- Numbness, tingling, or burning sensation in the feet
- Excessive bleeding or bruising
 - Caused by dysfunctional platelets
- Feeling of fullness or bloating in the left upper abdomen
 - When you have an enlarged spleen
- Weight loss
 - When you are eating less or using more energy because of PV
- Painful inflammation in the joints, known as gout
 - Caused by high levels of uric acid

After your diagnosis

With your diagnosis, your doctor can determine the right treatment for you. Your test results help your doctor predict how PV will likely progress and how you may respond to treatment.

Name of test	Description
Medical history and physical exam	The doctor reviews past illnesses, injuries, and symptoms. They examine your lungs, heart, and other organs.
Complete blood count (CBC)	This test measures the number of red blood cells, white blood cells, and platelets in your blood. If you have PV, you will have an abnormally high number of blood cells.
Comprehensive metabolic panel	This group of blood tests, or blood chemistry profile, measures the levels of certain substances released into your blood. With PV, it is important to measure your level of serum erythropoietin, the hormone that helps produce red blood cells.
Bone marrow aspiration and biopsy	These two tests examine bone marrow cells for abnormalities and look for fibrosis (scar tissue). They are usually done at the same time.
Molecular testing	This genetic test looks for a specific mutation of the gene known as JAK2 when PV is suspected.



PV treatment

PV is a chronic disease that can be well managed with treatment. Your treatment is focused on easing your symptoms. It also aims to reduce the risk of thrombosis (a blood clot inside a blood vessel).

Factors that affect treatment

Discuss your treatment options with your doctor to make sure you understand the benefits and risks of each approach. Your treatment plan is based on:

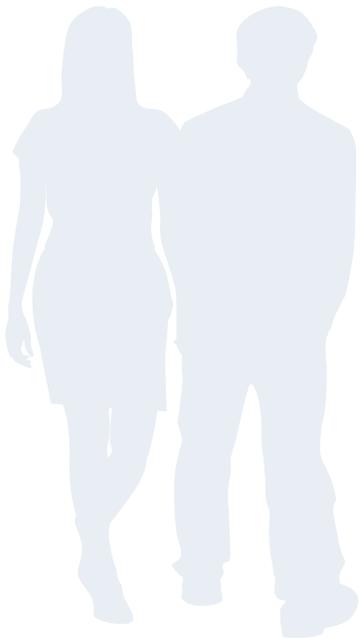
- Age and overall health status
- Risk for complications of thrombosis
- Whether you are considered low risk or high risk

Types of treatment

- **Low-dose aspirin** is used to reduce the risk of a blood clot.
- **Therapeutic phlebotomy** removes blood from a vein to reduce the number of blood cells and decrease blood volume. The blood becomes thinner and the red blood cells are less likely to become a mass along the walls of your blood vessels.
- **Drug therapy** is used for high-risk people with PV to reduce the number of red blood cells. These drugs may include one or a combination of:
 - **Hydroxyurea** decreases the number of blood cells in your bone marrow.
 - **Interferon alpha** lowers your platelet count.
 - **Ruxolitinib** stops cancer cells from multiplying.

If you are **low risk**, you may be prescribed low-dose aspirin and therapeutic phlebotomy.

If you are **high risk**, you may be prescribed low-dose aspirin, therapeutic phlebotomy, and drug therapy.



Treatment side effects

When you begin your treatment for PV, you may experience mild to severe side effects, depending on your age, your overall health, and your treatment plan. New drugs and therapies may help control side effects. Speak to your doctor if you are having side effects.

Common side effects

You may experience side effects such as:

- Upset stomach and heartburn from low-dose aspirin
- Iron deficiency (low iron) from repeated therapeutic phlebotomy treatments
- Mouth ulcers (open sores), changes in your sense of taste, skin ulcers, or rash from hydroxyurea treatments
- Confusion, depression, and flu-like symptoms from interferon alpha treatments
- A slight increase in the risk of infections from ruxolitinib treatments

Living with PV

Medical follow-up is important with PV. Your medical team should provide you with a care plan listing the frequency of follow-up visits and the tests you will have at those visits.

If you need elective surgery for something unrelated to PV, you have an increased risk of bleeding and clotting complications after surgery.

- Your doctor should check your platelet and red blood cell counts before surgery to make sure they are normal.
- They will also plan for possible excessive bleeding and deep vein thrombosis after your surgery.



Living with PV can be overwhelming. Seek medical help if you are feeling “down” or “blue” or don’t want to do anything – and your mood does not improve over time. These could be signs of depression, an illness that should be treated even when you’re undergoing treatment for PV. Treatment for depression has important benefits for people living with cancer. Remember, you are not alone.

This fact sheet was reviewed by:

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