Newly diagnosed with polycythaemia vera (PV)

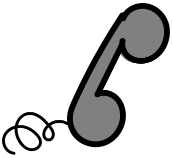
A Guide for Patients

Leukaemia Care
YOUR Blood Cancer Charity

About Leukaemia Care

Leukaemia Care is the UK's leading leukaemia charity. For over 50 years, we have been dedicated to ensuring that everyone affected receives the best possible diagnosis, information, advice, treatment and support.

Our services

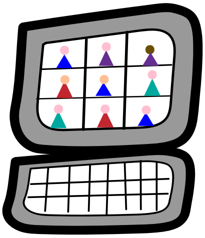


Helpline

Our helpline is available 9am to 4:30pm Monday to Friday. If you need someone to talk to, call **08088 010 444**.

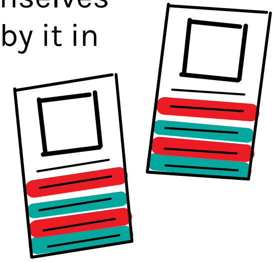
Alternatively, you can send a message via WhatsApp on **07500 068065** on weekdays 9am to 5pm.

Support groups



Our nationwide support groups are a chance to meet and talk to other people who have been affected by a PV diagnosis. For more information, scan this QR code:

Buddy support



We offer one-to-one phone support with volunteers who have had PV themselves or been affected by it in some way. You can speak to someone who knows what you are going through. For more information on how to get a buddy call **08088 010 444** or email support@leukaemiacare.org.uk

Counselling service

Our counselling service helps PV patients and their loved ones access up to six sessions of counselling. To apply, scan this QR code:

Advocacy and welfare

Our advocacy and welfare officers are here to help you find the support you need for many issues surrounding a PV diagnosis. These include insurance, benefits and clinical trials. If you would like support from our advocacy or welfare officer, email advocacy@leukaemiacare.org.uk or call **08088 010 444**.

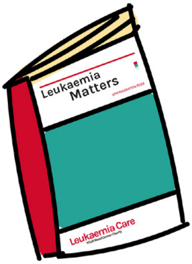
Cost of living fund

This fund provides grants to patients and families affected by PV, to help with essential living costs. All applications must be made via the form which can be found by scanning the QR code:

Write a Will

Using our complimentary service, you can write a simple Will so you know what happens to your estate when you die. To start writing your Will today, scan this QR code:

Patient magazine



Our magazine includes inspirational patient and carer stories as well as informative articles by medical professionals. To subscribe to our magazine, scan this QR code:

In this booklet

There is a lot of information about cancer on the internet. Some of it may not be reliable or up-to-date, and much of it will not be applicable to you. Your haematology team is best placed to give you information that is specific to you because they know your individual circumstances. If you want to search for information yourself, look for reputable organisations like the NHS or national charities. Look for a quality mark, such as the Patient Information Forum (PIF) tick.

Introduction

Polycythaemia vera (PV) is a type of blood cancer where your bone marrow makes too many red blood cells. Sometimes other blood cells such as white blood cells and platelets are affected too. In this booklet we cover what PV is, including how it is diagnosed and what treatments you may have. We also include practical information about living with PV.

This booklet is only a guide of what you might experience. Your haematology team will give you a copy of your specific treatment plan.

A patient information writer researched and wrote this booklet. We used an AI tool, with human oversight, to fine-tune the readability of the content.

In compiling this booklet, we are grateful to, Professor Claire Harrison, Professor of Myeloproliferative Neoplasms. This booklet was also reviewed by patient reviewers, Andrew and Eleanor. We thank them all for their valuable contribution.

Throughout this booklet, you will see URLs and QR codes that link to webpages for further support. If you are not able to access the webpages, please email information@leukaemiacare.org.uk or call 08088 010 444.

About polycythaemia vera (PV)

Summary

- Polycythaemia vera (PV) is a slow-growing blood cancer.
- It happens when your bone marrow makes too many red blood cells. Sometimes other blood cells such as white blood cells and platelets are affected too.
- This makes your blood thicker than normal, which can lead to blood clots. Blood clots can stop your blood from flowing as it should do.
- PV is rare.
- It can affect people of any age, but it is more common in people over 60.
- We do not know the exact cause of PV. It is not usually inherited, and you cannot usually pass it on to any children you may have.

What is polycythaemia vera?

Polycythaemia vera (PV) is a slow-growing blood cancer. It belongs to a group of conditions called **myeloproliferative neoplasms** (MPNs).

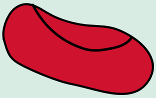
PV happens when your bone marrow makes too many red blood cells. Sometimes other blood cells such as white blood cells and platelets are affected too. This makes your blood thicker than usual. This can cause blood clots to form, which can stop your blood from flowing as it should do.

Sometimes PV can develop or transform into faster-growing types of blood cancer called myelofibrosis or acute myeloid leukaemia (page 16).

Bone marrow and types of blood cells

Blood cells develop from immature cells called stem cells in your bone marrow. Bone marrow is the soft spongy substance found in the middle of some large bones in your body. These stem cells can turn into any of the following types of blood cell:

- Red blood cells – these carry oxygen around your body
- White blood cells – these help you fight infection
- Platelets – these help to stop bleeding by causing blood clots to form



Red blood cell

White blood cell

Platelet

What is a myeloproliferative neoplasm?

MPNs are blood cancers that develop when cells in your bone marrow grow out of control and make too many blood cells. There are different types of MPN depending on which type of blood cell is being over-produced.

- In PV, your bone marrow makes too many red blood cells. But the affected bone marrow cells may also develop into other types of blood cells. This means your number of platelets and white blood cells may increase as well.
- In essential thrombocythaemia (ET) your bone marrow makes too many platelets.
- In myelofibrosis (MF) your bone marrow becomes filled with scar tissue, which stops it making enough healthy blood cells. It can develop from PV and ET.

We have separate information about essential thrombocythaemia and myelofibrosis. Follow the link, scan the QR code or search for 'essential thrombocythaemia' or 'myelofibrosis' at leukaemiacare.org.uk.

Who gets PV?

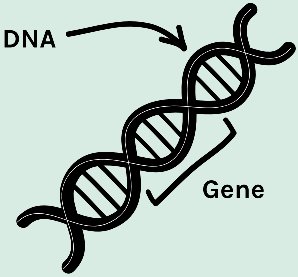
PV is a rare disease. Around 1,140 people are diagnosed with it each year in the UK. This is why you might not have heard of PV or met anyone with it before.

PV can affect people of any age, but it is more common in people over 60. It is slightly more common in men than in women.

What causes PV?

We do not know the exact cause of PV. Research shows that 95 in 100 people with PV have a change in a protein called JAK2. This protein helps regulate blood cell production. But we don't know why the change happens.

Understanding DNA and proteins



- DNA is like a thread of code in each cell of your body. Your DNA contains lots of different genes.
- Genes contain instructions for your cells on how to make proteins needed by your body. These proteins are important in the normal growth, development, and function of your cells.
- Sometimes changes occur in a gene. These are known as variants. These variants can cause a different protein to be made.

In most people with PV, the genetic changes that change JAK2 happen by chance. PV is not usually inherited, and you cannot usually pass it on to any children you may have.

Symptoms and complications of PV

Summary

- Around half of people with PV do not have symptoms when they are diagnosed. But you might get some signs or symptoms over time. These can include:

Blood clots

Bleeding or bruising

Fatigue

Night sweats

Fever

Tummy pain or bloating

A feeling of fullness

Headaches

Dizziness or light headedness

Itching

- In some cases, PV can develop or transform into faster-growing types of blood cancer called myelofibrosis or acute myeloid leukaemia. If this happens, you will need different treatment.

Symptoms of PV

Around half of people with PV do not have symptoms when they are diagnosed. Most people are diagnosed after a routine blood test. But you might get some signs or symptoms over time. These might include:

- Blood clots (page 12)
- Bleeding (page 15)
- Fatigue
- Night sweats
- Headaches
- Dizziness or light headedness
- Blurred vision
- Breathlessness
- Itching, especially after your skin has been in contact with water
- Reddish or purple skin (this may look different in different skin tones)
- Fever
- Swollen spleen, which can cause symptoms like:
    - Tummy pain
    - Bloating
    - Discomfort under your ribs on the left-hand side
    - Feeling full quickly when you eat

- Bone pain
- A painful or swollen joint due to a build-up of uric acid (gout)
- Weight loss

> **"Being a young cancer patient, I hear all the time how well I look. People assume you can't have cancer or be unwell if you still have all your hair and aren't stick thin. This is such a myth."**
>
> Emma, living with PV

Blood clots

Around 4 in 10 people with PV get a blood clot at some point. They can occur anywhere in your body.

The risk of getting a blood clot varies from person to person.

You have a higher risk of getting a blood clot if:	You have a lower risk of getting a blood clot if:
● You are over 60 years of age ● You have had a blood clot before	● You are under 60 years of age ● You do not have high blood pressure or high cholesterol ● You do not have diabetes ● You do not smoke ● You are a healthy weight

Symptoms of a blood clot depend on where it is. If you get any of the following symptoms seek medical help straight away.

Symptoms of a blood clot in your heart, which can cause a heart attack, such as:

- Pain or discomfort in your chest, back, arm or shoulder
- Shortness of breath
- Feeling sick
- Sweating and light headedness

Symptoms of a blood clot in your brain, which can cause a stroke, such as:

- Drooping of your face on one side
- Not being able to lift your arms and keep them raised
- Slurred or garbled speech
- Not being able to understand what is being said to you

Symptoms of a blood clot in your limbs (arm or leg), known as deep vein thrombosis, such as:

- Throbbing or cramping pain
- Swelling, redness or warmth in the area

Symptoms of a blood clot in your lungs, known as pulmonary embolism, such as:

- Difficulty breathing that comes on suddenly
- Chest pain, which can be worse when breathing in
- Coughing up blood

Symptoms of a blood clot in your eye, such as blurred vision or loss of vision

If you have PV, you could get blood clots in other, more unusual sites. This could include:

Clots in the veins that drain blood from your gut, which could cause:

- Pain in your tummy
- Bloating
- Bleeding in your gut

Clots in the vein that drains blood from your brain, which could cause:

- Headaches
- Fits (seizures)
- Fainting or loss of consciousness

Bleeding

PV can cause abnormal bleeding and any bleeding may be heavier than usual. Around 8 in 100 people with PV will suffer from a bleeding event at some point.

Contact your medical team if you get any of the following:

- Nosebleeds
- Bruising easily
- Bleeding gums
- Heavier or longer periods than usual
- Blood in your wee or poo

Seek urgent medical help if:

- You are coughing up blood
- You have blood in your sick (sometimes this can look like coffee granules)

If you cut yourself, you may find that you bleed longer than usual. Applying pressure to any small cut or wound with a sterile bandage will help stop the bleeding.

Swollen spleen

Around 3 in every 4 people with PV get a swollen spleen. This may cause symptoms like tummy pain, bloating and feeling full quickly when you eat. The swollen spleen may cause some discomfort under your ribs on the left-hand side. More rarely, your liver might become enlarged too.

Risk of transformation

PV is a slow-growing, long-term type of blood cancer. But in some cases, it can develop or **transform** into another, usually faster-growing, type of blood cancer. This could be either:

- Myelofibrosis (MF) – this is a type of MPN where your bone marrow becomes filled with scar tissue, your spleen enlarges and your blood counts drop
- Acute myeloid leukaemia (AML) – this is a fast-growing blood cancer where immature white blood cells in your bone marrow multiply

If your PV transforms you may notice that certain symptoms may get worse, such as fatigue, joint pain and the swelling of your spleen increases. Your haematology team will closely monitor your blood tests to check if your PV has transformed.

The risk of PV transforming into either MF or AML varies from person to person. It depends on lots of factors, including your age, your blood counts and the particular pattern of genetic changes in your cancer cells.

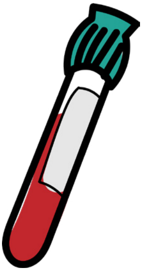
The chance of PV transforming increases with time. In general:

- Between 5 and 6 in every 100 people with PV develop MF within 10 years of diagnosis. This means 94 to 95 in every 100 do not.
- Between 2 and 3 in every 100 people with PV develop AML within 10 years of diagnosis. So 97 to 98 in every 100 do not.

We have separate information about acute myeloid leukaemia (AML) and myelofibrosis (MF). Follow the link, scan the QR code, or search for 'AML' or 'myelofibrosis' at leukaemiacare.org.uk.

Diagnosis of PV

Summary



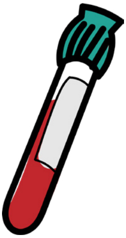
- Your haematology team will diagnose PV based on:
  - Blood tests
  - Bone marrow tests
- Depending on your symptoms, you might also have other tests or scans.
- It can be difficult waiting for test results. We are here for you if you need support. You can email us at support@leukaemiacare.org.uk, message us on WhatsApp at **07500 068065** or call our freephone helpline at **08088 010 444**.

Diagnosis of PV

Many people with PV do not have symptoms when they are diagnosed. You may be diagnosed after having a blood test for something else. You will have further tests to confirm a PV diagnosis. Some of these tests might be repeated throughout your treatment to measure your response to the treatment.

> **"I remember hearing the words when I was told I had PV, even now 11 years later. Denial struck through me. I was diagnosed late into my pregnancy, and I didn't (nor want to) spend too much time researching what PV was or how it affected the body. I just wanted to ignore it."**
>
> Andrea, diagnosed with PV age 33

Blood tests



You are likely to have blood tests to check:

- Your blood cell counts
    - This measures the number of red blood cells, white blood cells and platelets in your blood
- What your blood cells look like under a microscope
- How well your kidneys and liver are working
    - This can help your medical team rule out other causes of high red blood cells
- Your iron and calcium levels
    - These tend to be low in people with PV

- Your levels of a hormone called erythropoietin (EPO)
  - This hormone boosts red blood cell production in your bone marrow
  - Measuring your EPO levels can help rule out other causes of a high red blood cell count
- The levels of oxygen in your blood
  - Your haematology team usually measure this using a device that clips on to your finger

Blood tests to measure red blood cells

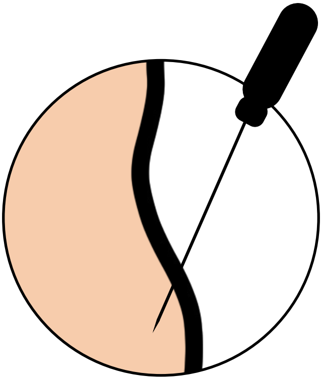
There are different blood tests that can measure your red blood cells.

A full blood count can measure a number of things including:

- Your red blood cell count
- Your **haemoglobin** level (Hb): This is the amount of a protein called haemoglobin in your blood. It is the protein your red blood cells use to transport oxygen around your body. It is a good indicator of your red blood cell count.
- Your **haematocrit** level (Hct): This is the percentage of red blood cells in your blood. This is also a good indicator of your red blood cell count.

If you have PV your blood tests usually show high numbers of red blood cells. This is because the affected bone marrow makes too many red blood cells. Sometimes the affected bone marrow might also make other types of blood cells. This means that your number of white blood cells and platelets may be high too.

Bone marrow tests

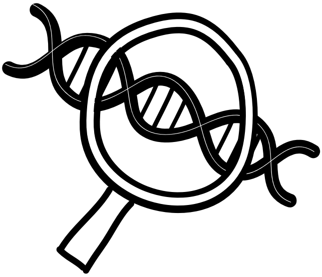


Your haematologist might recommend having a bone marrow test to confirm your diagnosis, but this is not always needed. They will be looking for signs that your bone marrow has been affected by PV.

To have a bone marrow test:

- You have a local anaesthetic to numb the area where you're having the test. This is usually your hip bone (pelvis).
- Your haematologist then uses a special needle to suck out a sample of liquid bone marrow. This is called a bone marrow aspiration.
- They will also need a small sample of harder bone marrow. They use a larger needle for this. It is called a bone marrow trephine biopsy.
- A bone marrow test can be uncomfortable both during and after the procedure. Ask your doctor or nurse what painkillers you can use if you need them.
- You should avoid vigorous exercise 24 to 48 hours afterwards.

Genetic tests



Your doctors will send your blood samples, and occasionally your bone marrow samples, for testing for genetic changes. They will test for a change in a gene called *JAK2*, which can be used to diagnose PV.

Around 5 in every 100 people with PV do not have this change. In this case, your haematology team will diagnose PV based upon your signs and symptoms and the results of your other blood tests and bone marrow tests.

Other tests

Your haematology team may also arrange for you to have an ultrasound to check if your spleen is swollen. They may feel the area instead, to see if they can feel any obvious signs of your spleen being enlarged.

Your test results may take a little while, which can be a worrying time for you. It is important for your haematology team to have all the results so they can make an accurate diagnosis. It can also help them work out the most suitable treatment options for you.

We are here for you if you need support whilst you are waiting for your results. You can email us at support@leukaemiacare.org.uk, message us on WhatsApp at **07500 068065** or call our freephone helpline at **08088 010 444**.

Treatment of PV

Summary

- The aim of PV treatment is to manage your symptoms, prevent blood clots, reduce your risk of transformation, and improve your quality of life.
- Your haematology team will suggest the most suitable treatment for you based on your individual circumstances.
- Treatment options include:
    - Venesection – a simple procedure to remove some of your blood and help keep your blood count stable
    - Aspirin, to reduce your risk of blood clots
    - Medicines to reduce your blood counts, such as chemotherapy tablets or targeted medicines
- If you get a blood clot or experience heavy bleeding, you will need treatment for these too.
- Keeping active and eating a healthy balanced diet can help reduce your risk of blood clots, reduce side effects, and improve your health in general.
- You may also have treatments to help relieve symptoms like itching.
- If you have any side effects, it is important to tell your haematologist.

Treatment of PV

Overview of treatment

The aim of PV treatment is to manage your symptoms, prevent blood clots, reduce your risk of transformation, and improve your quality of life. As well as treatment, there are things you can do yourself that can help.

Your haematology team will suggest the most suitable treatment for you based on:

- Your risk of getting blood clots. This risk is higher if:
    - You have had a blood clot before
    - You have high blood pressure or high cholesterol
    - You have diabetes
    - You smoke
    - You have obesity
- Your personal characteristics such as:
    - Your age
    - Your overall health
    - Your family history
    - Your symptoms
    - Your blood test results
    - Your preferences on how you wish to be treated
    - Any other health conditions that you may have
- Previous treatment
    - Whether or not it is your first treatment
    - Any side effects that you may have had with other treatments

Treatment if you have a low risk of getting clots

These treatments are recommended for all people with PV to help reduce the risk of blood clots and to keep your blood volume under control. If you have a low risk of getting clots, these might be the only treatments that you need.

Venesection

Venesection is a simple procedure to remove some of your blood. It is sometimes called phlebotomy. It is similar to donating blood. It aims to reduce the thickness of your blood and keep your blood counts stable, which reduces your risk of blood clots. It is recommended for all people with PV.

- You have venesection as an outpatient, usually at the hospital.
- A nurse then puts a needle attached to a thin plastic tube into a vein in your arm.
- They then take out around 200ml to 500ml of blood.
- It takes around 30 minutes.

You may need to have blood taken every few weeks or months to keep your haematocrit level stable. This can help improve symptoms such as headaches, dizziness and ringing in your ears.

Venesection might be the only treatment that you need to keep your red blood cell count at the level it should be.

> **"I remember first being on the ward for a venesection; I was surrounded by people who were visibly unwell, and yet I was going home that day to be a mum and cook the tea or go to work the next day."**
>
> Andrea, diagnosed with PV at age 33

Possible side effects

Sometimes venesection can lead to low iron levels. This may cause symptoms like:

- Restless legs
- Concentration problems
- Impaired cognitive function
- Dizziness, fatigue and headaches

If you have any of these symptoms, it is important to tell your haematologist so they can consider other treatment options that may be better suited to you. Also, if you need to have venesection often, your haematology team might recommend another treatment for you.

Do not take iron supplements without talking to your haematology team first.

Aspirin

You might have used aspirin before to help reduce pain or lower your temperature if you have had a fever. Aspirin also helps stop your platelets sticking together, which prevents blood clots.

- Low-dose aspirin is recommended in all people with PV.
- It comes as tablets or soluble tablets, which you take by mouth.
- Most people with PV take around 100mg every day. Your medical team will tell you what dose is best for you based on your own individual situation.

Possible side effects

Most people don’t have any problems when taking aspirin. But, like all medicines, it can cause side effects. Seek medical help if you experience any of the following serious side effects:

- Unusual bleeding, or bleeding that doesn’t stop
- Trouble breathing, or worsened asthma
- Bleeding in your stomach, or irritation in your tummy
- Stroke or mini-stroke (also known as transient ischaemic attack or TIA)
- Cramps in your lower chest
- Heavier or longer periods than usual
- Bruises with purple spots (this may look different on different skin tones)

The Electronic medicines compendium has more information about aspirin. Follow the link, scan the QR code or search ‘aspirin’ at www.medicines.org.uk/emc and select the brand of medicine you are taking.

Treatment if you have a higher risk of getting clots

If you have a higher risk of blood clots you will be offered treatment in addition to having venesection and aspirin. Your medical team may recommend a medicine to help lower your blood cell counts. This is sometimes called cytoreductive therapy.

Hydroxycarbamide (also known as hydroxyurea)

Hydroxycarbamide is a chemotherapy medicine that lowers your blood cell counts. It comes as tablets or capsules, which you take by mouth. Usually, you will take either a smaller dose every day, or a higher dose every 3 days. Your medical team will tell you what dose is best for you based on your own individual situation. Your haematologist might suggest it for you as a first treatment if you have a higher risk of getting clots. It may not be suitable if you are pregnant, breastfeeding or currently trying for children.

Possible side effects

Hydroxycarbamide can cause a number of different side effects. Different people may get different side effects. If you experience any of the following serious side effects, contact your haematology team:

- Frequent infections
- Bruising or mild bleeding
- Changes to your skin, including ulcers or sores
- Signs of possible skin cancer, which can include:
  - A growth or unusual patch on your skin
  - A new mole or a change in an existing mole
- Mouth ulcers and sores in your mouth

Hydroxycarbamide might also increase the risk of your PV transforming to MF or AML if you are taking it for a long time.

We have separate information about hydroxycarbamide. Follow the link, scan the QR code or search for ‘hydroxycarbamide’ at shop.leukaemiacare.org.uk.

Peginterferon

Peginterferon (also called pegylated interferon) is a medicine that alters the way your immune system works. It helps stop cancer cells growing and multiplying. Your team might recommend it for you if your white blood cell count is increasing, the swelling of your spleen is growing or if hydroxycarbamide doesn't suit you.

You have peginterferon as an injection, typically once a week. You can be taught to take it yourself, or a carer can give it to you. Otherwise, a GP or nurse can give you the injection. Your medical team will tell you what dose is best for you based on your own individual circumstances.

Possible side effects

Peginterferon can cause side effects, but not everyone will get them. Seek medical help if you experience any of the following serious side effects:

- Changes in your heartbeat (when the heart beats very fast and unevenly)
- Attempted suicide, suicidal thoughts
- Loss of vision or blindness
- Blood spots or bleeding in your eye
- Breathing problems (such as shortness of breath, coughing, chest pain)

Cancer Research UK have more information about peginterferon. Follow the link, scan the QR code or visit www.cancerresearchuk.org/ and search 'peginterferon'.

Ruxolitinib

Ruxolitinib is a type of targeted therapy. Targeted therapies are drugs designed to block specific proteins in cancer cells. This means they kill cancer cells with as few effects on healthy cells as possible.

Ruxolitinib blocks the JAK1 and JAK2 proteins. It is the JAK2 protein that encourages your bone marrow to make too many red blood cells.

Your haematology team might recommend ruxolitinib for you if:

- You have not responded to hydroxycarbamide or
- Hydroxycarbamide caused side effects that were difficult to cope with

Ruxolitinib comes as tablets that you take by mouth. Most people with PV start on a dose of 10mg twice a day. You will have blood tests throughout your treatment to monitor your response. Your doctor might increase or decrease your dose, depending on your blood counts and haemoglobin levels. They will tell you what dose is best for you based on your own individual circumstances.

Possible side effects

Ruxolitinib can cause side effects but not everyone will get them. Seek medical help if you experience any of the following serious side effects:

- Blood in your poo or vomiting blood
- Unexpected bruising or bleeding
- Painful skin rash with blisters or unusually pale skin
- Unusual tiredness
- Shortness of breath
- Fever, chills, or other signs of infection
- Persistent headaches, numbness, tingling, weakness or paralysis

Taking ruxolitinib can increase your risk of skin cancer. If you have any of the following signs of skin cancer, you should inform your medical team:

- A growth or unusual patch on your skin (can vary in size, colour and texture)
- A new mole or a change in an existing mole

Do not stop taking ruxolitinib without talking to your haematology team first.

Cancer Research UK have more information about ruxolitinib. Follow the link, scan the QR code or visit www.cancerresearchuk.org/ and search 'ruxolitinib'.

Other treatments your team might recommend

Depending on your circumstances, your team might suggest other treatment options, such as busulfan.

Busulfan is a chemotherapy medicine that stops cancer cells in your bone marrow multiplying. This can help lower your blood cell counts. But busulfan has a higher risk than some other treatments of your PV developing into MF or AML, so it is not used routinely.

If you need to have busulfan, your medical team will tell you how you have it and what you can expect from treatment.

If you have a high platelet count that has not responded to treatment, your team might recommend a medicine called anagrelide alongside other medicines.

Lifestyle measures to help prevent blood clots

There are things you can do to reduce your risk of getting blood clots, and help you cope with fatigue and other effects of PV. These lifestyle measures can also help improve your quality of life.

Having a healthier lifestyle

It is important to look after yourself well. Making some small changes to your lifestyle can help you stay as well as possible after diagnosis and during treatment.

It is important to not change too much at once. Adopting a healthier way of living is about making small, manageable changes to your lifestyle.

Your doctor might recommend compression stockings to help improve blood flow and prevent blood clots in your legs.

> **"I strongly advise people with an MPN to take good care of their general health, watch their weight, exercise, don’t smoke, monitor their blood pressure and cholesterol - this also reduces their risk of blood clots including stroke and heart disease."**
>
> Professor Claire Harrison

Heart health

It is important to look after your heart health to reduce your risk of blood clots. Some changes you can make to help reduce this risk include:

- Not smoking
- Maintaining a healthy weight
- Eating a healthy diet
- Exercise
- Getting your blood pressure checked regularly
- Having regular tests to check your cholesterol and blood sugar levels

> **"I have now also had to start taking cholesterol tablets and blood pressure tablets to help reduce my risk of stroke or heart attack."**
>
> Jayne, living with PV

Diet

If you can, try to eat a well-balanced diet. This will help you:

- Feel stronger
- Have more energy
- Cope better with your treatment
- Improve your heart health

Side effects from treatment, such as sickness and diarrhoea can make it difficult to eat a healthy diet. If you are struggling, ask your haematology team for advice.

The NHS website has information and guidance on eating a healthy, balanced diet. Scan the QR code, or go to the 'live well' section at www.nhs.uk

Exercise

Exercise can improve your heart health, quality of life and wellbeing. It can also help to reduce some of the side effects and symptoms you may be experiencing. You may not feel like being active, especially with some of the side effects you may have. Remember, even a gentle walk can help. Choose a level of exercise that works for you and how you are feeling.

Treatment if you get a blood clot

If you have a blood clot, your doctor might start you on medicine to help keep your blood counts under control (page 26), if you are not already on it. If your haematocrit is high when you have the blood clot, you might also have venesection (page 24).

You will also have treatments to treat your clot and prevent future clots. This might be:

- Lifestyle measures to reduce your risk of getting another clot (page 31)
- Injected anti-clotting medicines like heparin or low molecular weight heparin at first
- Longer-term treatment with anti-clotting medicines that you take by mouth, such as:
    - Warfarin
    - Rivaroxaban
    - Dabigatran
    - Apixaban
    - Edoxaban

For more serious blood clots, you may also have:

- Injections to help break down the clot and restore blood flow
- A procedure to remove the clot or allow the blood to flow around the clot

Your doctor will tell you what they recommend for you and what to expect from treatment. For some treatments, you need regular blood tests to check how easily your blood clots.

How long you need to take anti-clotting medicines depends on where the blood clot is and your risk of further blood clots.

If you have high blood pressure or high cholesterol you will also have medication to lower these, which reduces your risk of further clots. You may also be advised to make some lifestyle changes to help you manage your risk of blood clots (page 31).

Treatment of bleeding

Bleeding (haemorrhage) is a possible complication of PV. If you have heavy bleeding, you may receive the following:

- Replacement fluids and a blood transfusion
  - You may need these if you have lost a lot of blood
  - They will help bring your blood circulation back to normal
- An injection of tranexamic acid
  - This stops your body from breaking down blood clots too soon
  - It can help your body stop the bleeding

Your medical team will also look at what medications you are on for PV. They may adjust them to help improve your blood counts and reduce your risk of bleeding. This can also include any anticoagulation medication you are on, such as aspirin. You may also have some blood tests to check your risk of future clots, and if you've developed any other problems with your clotting.

Treatment to help with symptoms

You might receive treatment known as supportive treatment to help you deal with the symptoms of your PV, such as itching. Itching can be a side effect that impacts your quality of life, so the following medications can help provide some relief:

- Antihistamines
  - These are usually tablets that you take by mouth
- Corticosteroids
  - Available as creams, foams, gels, lotions and ointments that you apply directly to your skin to help soothe and relieve itching
- Antidepressants
  - A particular type of anti-depressants, known as selective serotonin re-uptake inhibitors, can help with itching
- Phototherapy
  - You will be given medicine first known as psolarens to make your skin more sensitive to light
  - Your skin is then exposed to UV light, which can help relieve itching

Outcomes of PV

Summary

- People with PV who do not develop another type of cancer usually have excellent outcomes, and only a slightly reduced life expectancy.
- Outcomes vary from person-to-person. They depend on lots of different factors. Your haematology team are best placed to discuss what they expect for you because they know your individual circumstances.

Outcomes of PV

People with PV who do not develop MF or AML usually have excellent outcomes, and only a slightly reduced life expectancy.

Your outcome can vary depending on your individual situation, your health history, and the way you respond to treatment. PV is a life-long condition, so it is important to go to all your follow-up appointments and tell your team if you get any new, different or worsening symptoms.

As with most cancers, outcomes vary from person-to-person. They depend on lots of different factors, including:

- Your age
- Your blood cell counts when you were diagnosed
- Your heart health
- Whether or not you have had clots in the past
- The size of your spleen
- The genetic changes in your blood cells

Your haematology team are best placed to discuss what they expect for you because they know your individual circumstances.

If you are interested in general survival numbers for PV, we include some figures on the next page. You may prefer not to look at these.

It's important to remember that survival numbers cannot tell you what will happen in individual situations. They look at what happened to groups of people with a similar diagnosis in the past. They are based on data collected over many years, when people may not have received treatments that are available now.

5-year survival rates are commonly quoted. This is the proportion of people with a particular condition who are still alive 5 years after diagnosis.

Survival rates do not tell us anything about what people who are not alive 5 years after diagnosis died from. It may have been the condition, or it may have been from another cause.

People with PV that does not transform to MF or AML usually have excellent outcomes. On average around 94 in 100 people with PV are still alive 5 years after their diagnosis.

If you develop another blood cancer such as AML or MF, it may be harder to treat and outcomes are less favourable.

We have separate information about acute myeloid leukaemia (AML) and myelofibrosis (MF). Follow the link, scan the QR code, or search for 'AML' or 'myelofibrosis' at leukaemiacare.org.uk.

Living with PV

Summary

- Having PV can affect your day-to-day life in a number of ways.
- You may experience a variety of emotions. There is no right or wrong way to feel. But if you think you may be depressed, contact your GP.
- If you are struggling, don't be afraid to ask for help from friends, family, your haematology team or Leukaemia Care.
- You might want to talk to your friends and family about your condition. Remember, this is your choice. You can choose when to tell them and how much.
- You might experience fatigue. Planning, pacing yourself and saving energy for things that are important to you can help. Your haematology team can suggest things to help you or refer you for support if you need it.
- Having PV can affect your work or finances. You are entitled to reasonable adjustments to help you cope with work. You may also be eligible for financial support.
- You probably have a lot of questions. Make a list of them so you don't forget to ask them when you see your medical team.

Living with PV

Being diagnosed with PV can be overwhelming. It is something that can affect you both physically and emotionally. Symptoms, the effects of treatment, and hospital appointments, may all impact your day-to-day life. Here, we cover some practical information about living with PV and where to get support if you need it.

Managing your emotions

Being diagnosed with cancer can be very upsetting, so you may need emotional and practical support. It is likely that you will experience a range of complex thoughts and emotions. These thoughts and emotions may feel strange or unfamiliar to you. You may feel a range of emotions like:

- Shock or disbelief
- Uncertainty, anxiety or fear about the future
- Sadness or depression
- A sense of loss of the person you used to be, and how safe you felt
- Worry about other people's reactions
- Isolation, or a feeling that other people don't understand what you're going through
- Guilt, anger, frustration or irritability
- A loss of self-confidence

Everybody reacts differently. You may experience some of these emotions but not others. You might have different feelings at different times. There is no right or wrong way to feel.

> **"With everything I have going on, my anxiety was on overdrive, and I was very down, so I'm also taking sertraline to help with that."**
>
> Jayne, living with PV

Sadness and depression

You might be feeling low, which is a natural effect of your diagnosis and treatment. However, you may have depression if:

- Your low mood persists for several weeks
- You feel hopeless
- You lose interest and pleasure in life

If you think you may be depressed, it is important to contact your GP. They can help you access the support and treatment that you need.

If you are in crisis, the NHS has urgent mental health helplines that offer 24-hour advice and support. Scan the QR code for details or or go to the 'mental health' section at www.nhs.uk

Where can I get support?

There are several places where you can seek emotional support, such as:

- Care givers and loved ones
- Your clinical nurse specialist, if you have one
- Your consultant
- Charities and other organisations

Talking to other people can help. It can be difficult to talk to loved ones, so you might prefer to speak to someone independent. It can also help to talk to other people who have gone through a similar experience.

We're here for you if you need support. Scan the QR code, follow the link or search 'support for you' at leukaemiacare.org.uk to find out how we can help you. Our support services include:

- Helpline and advocacy services to provide information and support
- Support groups and a buddy service, where you can connect with people who understand your experience
- A counselling service where you can apply for a grant for up to six sessions of counselling

If you'd like to talk to someone who understands what you're going through:

- Call our freephone helpline on **08088 010 444**
- Message us through WhatsApp on **07500 068065**
- Email support@leukaemiacare.org.uk

Telling other people

When you are first diagnosed with PV, there is a lot to take in. You may need to give yourself time to adjust before you decide when and how you tell others about it. People may be anxious to know what is happening which can make you feel under pressure to tell them. Let them know you need time to process the information yourself before you are ready to talk about it. There will be some people that you want to tell and others you prefer not to. It is up to you who you tell and how much you tell them.

> **"Sometimes all we need is to just talk about our health. I don't need people to get a violin out for me, but I do want empathy and understanding."**
>
> Andrea, diagnosed with PV age 33

In the conversation with your loved ones, you might want to:

- Explain that you have a condition that means your bone marrow doesn't work properly and this affects the number of blood cells it produces.
- Explain what symptoms you get.
- Explain your needs. Your family and friends may be happy to know they can help support you by helping around the house or doing the food shop.
- Be open and honest about how you feel. People who care about you will help you as best as they can.
- Have a print-out or factsheet handy so you don't have to remember everything your friends and family might want to know. Or you could give them a copy of this booklet.

If you are struggling to come to terms with your diagnosis, you can speak to us on our helpline on **08088 010 444**.

Coping with fatigue

One of the main symptoms of PV is fatigue. This is a feeling of physical or mental exhaustion that doesn't improve with sleep. Fatigue can be frustrating as it cannot be treated with medicines. It is common for fatigue to continue after your treatment, but it does often get better over time.

Tips for coping with fatigue

- Make plans and pace yourself
    - Prioritise things that are important to you and reserve energy for these
    - Build time to rest around planned busy periods
    - Allow yourself to rest as and when you need
- Keep to a regular sleep schedule
    - Try to go to bed and wake up at around the same time each day
    - Keep your bedroom quiet and at a comfortable temperature
    - Avoid eating or drinking alcohol, coffee, tea, or chocolate before bedtime
    - Avoid using laptops, tablets, or mobile phones before going to bed
- Take regular, gentle exercise to maintain your fitness levels
- Accept help with household tasks

Make sure to tell your haematology team if you are experiencing fatigue. They may be able to suggest things to help or refer you for support if you need it.

We have more resources to help you cope with fatigue on our website. Scan the QR code to find out more, or search 'fatigue' at leukaemiacare.org.uk

Work and education

Being diagnosed with PV and having to juggle work or education with hospital or GP appointments can be challenging. Your diagnosis, managing symptoms or side effects, or going to appointments may mean that you need time off from work.

Your consultant or GP can write letters to your employer to confirm your diagnosis and how this may affect your work life. This can help your employer have arrangements in place for when you need time off and discuss how else they can support you.

If you are diagnosed with PV while you are at school or university, you should contact them. They might be able to offer you extra support or defer your attendance for a while if you need it.

Money and financial help

Your PV diagnosis may also affect your finances, whether or not you are working. Being diagnosed with PV can come with extra costs, such as travel to and from hospital, childcare costs or parking charges.

You should be able to get free NHS prescriptions as a person with cancer. Your haematology team or GP can tell you how to apply.

Charities and financial support

We have a range of services that can help you when living with PV, including a welfare service and cost of living hub. Follow the link, scan the QR code or search 'support for you' at leukaemiacare.org.uk

Macmillan have information on benefits and financial support you might be able to access. Follow the link, scan the QR code or search for 'benefits and financial support' on www.macmillan.org.uk for more information. Or contact them on **0808 808 0000** to discuss your eligibility for benefits, grants and support available.

Travelling with PV

Having PV shouldn't stop you living a fulfilling life. But, if you're planning to travel, there are a few things to think about. It's a good idea to plan ahead.

Tips for planning long-distance journeys

- Have a chat with your GP, haematologist or medical team about your plans.
    - They can tell you if it is safe and discuss how to manage your PV while you are away.
    - They might assess your aspirin dose and discuss other medicines to help prevent clots.
    - You could also ask them for a letter explaining your condition and the medicines you take, in case you need it for any security checks.
- It's important to consider getting travel insurance, especially if you are travelling to a country with high medical costs.
    - You will need to declare you have PV when buying your insurance policy. Otherwise, it won't cover any costs relating to your illness.

- Take all your PV medicines with you, and some extra in case you lose any or need to stay away longer than planned.
  - Check the medicine information leaflet to see how you need to store your medicine while you are travelling.
  - Make a list of all the medicines you take, when you take them, and the dose you take.
- Be prepared for emergencies.
  - Check for a doctor or health centre near your destination, so you know where to go if you need medical help.
  - Take contact numbers of your healthcare professionals.
- Make sure your vaccinations are up to date and check if you need any extra vaccinations for where you are travelling to.
- Take water, snacks, pillows and anything else that helps you be comfortable and manage any fatigue.

Preventing blood clots when you travel

Travelling long distance increases your risk of getting a blood clot in your leg. This is called a deep vein thrombosis (DVT). Your risk is higher however you travel – by car, coach, bus, train – but especially if you travel by air.

It's important to try and reduce this risk as much as possible. You can do this by:

- Walking around or standing up fairly often.
    - If you can, try and walk around the cabin or carriage every 2 to 3 hours. It helps if you're able to get an aisle seat.
    - If you are travelling by car, plan in rest stops so you have time to stretch your legs and walk around.
- Exercising your leg muscles and stretching during your journey. You could try:
    - Raising and lowering your heels while keeping your toes on the floor.
    - Raising and lowering your toes while keeping your heels on the floor.
    - Squeezing and relaxing your leg muscles.
- Asking your doctor or medical team about compression stockings. These are tight-fitting socks that help prevent blood clots. You can buy them from high street chemists, airports or online.

If you get symptoms of a blood clot (page 12) whilst travelling, seek urgent medical help.

Going to appointments

Hospital and GP appointments often involve a lot of waiting around. You could download a podcast or TV programme to keep you occupied while you wait. Or take something physical like a book or travel game.

Once you're in your appointment, it can be hard to take in everything your doctor or nurse tells you. Below are some tips to help you get the most from your appointment:

- It can help to take a family member or friend with you for support.
- Ask any questions that you may have. If you don't understand something, ask your medical team to explain. They will be used to this.
- You can take notes on your phone or paper to help you remember. You can always ask your doctor or nurse to make notes for you or ask if they are happy for you to record the conversation.
- Be open and honest when discussing your symptoms and how you are coping.

> **"The confidence to push back or ask questions doesn't always come naturally or quickly; it takes time to build. But when you do, you know you're advocating for your best care, and it is empowering to be able to do that."**
>
> Andrea, diagnosed with PV age 33

Questions for appointments

Sometimes it can be overwhelming to know what to ask in your appointments. Some questions you might want to ask include:

- What tests will I need?
- What might these tests show?
- How long will it take to get the results back?
- What treatment will I need and how long for?
- How will I know if my treatment has worked?
- What might the side effects be?
- Are there any foods or medications I need to avoid?
- Will I be able to go back to work?
- Where can I get help claiming benefits and grants?
- Where can I get help dealing with my feelings?

Glossary of medical terms

Anaesthetic: A medicine or substance that causes a loss of feeling or numbness.

Anagrelide: A medicine that helps treat the over production of platelets.

Bone marrow: Spongy tissue in the cavities of bones where blood cells are made.

Busulfan: A type of chemotherapy medicine that stops cancer cells in your bone marrow multiplying.

Cancer: The uncontrolled growth of abnormal cells.

Chemotherapy: Drugs that kill cancer cells or stop them dividing and multiplying.

Cytoreductive: A medicine or treatment that aims to reduce the number of cancer cells.

DNA (deoxyribonuclease acid): The genetic code that tells your cells how to grow and behave.

Essential thrombocythaemia (ET): A type of myeloproliferative neoplasm where your body produces too many platelets.

Gene: A section of DNA that tells your cells how to make a particular protein.

Haematocrit: The percentage of red blood cells in your blood.

Haematologist: A doctor that specialises in diseases of the blood and bone marrow.

Haemoglobin: The protein your red blood cells use to transport oxygen around your body.

Hydroxycarbamide: A chemotherapy medicine that helps lower your blood cell counts.

JAK2: A protein that helps regulate blood cell production.

***JAK2*:** The gene that codes for the JAK2 protein.

Myelofibrosis: A type of cancer where your bone marrow becomes filled with scar tissue, which stops it making enough healthy blood cells.

Myeloproliferative neoplasms: Blood cancers that develop when cells in your bone marrow grow out of control and make too many blood cells.

Peginterferon: A type of medicine that alters the way your immune system works and helps stop cancer cells growing and multiplying.

Platelets: Small cell fragments in your blood that help clots form and stop or prevent bleeding.

Polycythaemia vera: A type of cancer where your bone marrow makes too many red blood cells.

Red blood cell: A type of cell in your blood that carries oxygen around your body.

Ruxolitinib: A type of targeted therapy used to treat polycythaemia vera.

Spleen: An organ that sits under your ribs on the left side. It filters and stores blood and makes some blood cells.

Stem cells: Immature cells that can develop into many different types of cells in the body.

Transformation: When one type of blood cancer develops into another type, usually a faster-growing one.

Ultrasound: A scan that uses sound waves to look at the inside of your body.

Venesection: A simple procedure to remove some of your blood, similar to donating blood.

White blood cells: Cells in your blood that help your body fight infections.

Useful contacts and further support

There are a number of helpful sources to support you during your diagnosis, treatment and beyond, including:

- Your haematologist and healthcare team
- Your family and friends
- Your psychologist (ask your haematologist or CNS for a referral)
- Reliable online sources, such as Leukaemia Care
- Charitable organisations

Leukaemia Care

Helpline: 08088 010 444 (Monday to Friday, 9am to 5pm)

WhatsApp: 07500 068065 (Monday to Friday, 9am to 5pm)

www.leukaemiacare.org.uk
support@leukaemiacare.org.uk

Blood Cancer UK

Leading charity into the research of blood cancers.

0808 2080 888

www.bloodcancer.org.uk

MPN voice

Provide information, community and advocacy for MPN patients, their families and carers.

07934 689 354

www.mpnvoice.org.uk

Macmillan

Provides free practical, medical and financial support for people facing cancer.

0808 808 0000

www.macmillan.org.uk

Cancer Research UK

Leading charity dedicated to cancer research.

0808 800 4040

www.cancerresearchuk.org

Maggie's Centres

Offer free practical, emotional and social support to people with cancer and their loved ones.

0300 123 1801

www.maggies.org

Citizens Advice

Offers advice on benefits and financial assistance.

0800 144 8848 (England)

0800 702 2020 (Wales)

0800 028 1456 (Scotland)

www.citizensadvice.org.uk

The Citizens Advice service does not cover Northern Ireland but their website lists contact details for local community advice agencies, depending on where you live.

How you can help us

If you've been affected by PV, sharing your story can help others going through a similar situation and help the public to better understand.

Scan the QR to share your story:

Alternatively, you can email our Communications team at communications@leukaemiacare.org.uk.

Tell us what you think of this booklet

We aim to provide information that's reliable, up-to-date, and covers what matters to you. We want you to feel supported and able to be involved in decisions about your care. Please follow the link or scan the QR code to complete our short survey to help us improve our information and make sure it meets your needs. Or get in touch with us by email, phone or post.

You can also contact us if you'd like a list of the references we used to compile this booklet.

- Email our Information team at information@leukaemiacare.org.uk
- Call our Head Office on **01905 755 977**
- Write to us at Leukaemia Care, One Birch Court, Blackpole East, Worcester, WR3 8SG
- Leave us a review if you've ordered a booklet online

If we've helped you - here's how you can give back

Fundraising is at the core of what we do here at Leukaemia Care, and without it we wouldn't be able to provide the support we do.

Fundraising isn't all about running a marathon, and there are plenty of ways to give thanks and show your support.

You could:

- Ask your local shop or workplace to host a collection tin
- Ask your place of work about charity of the year partnerships or grants
- Take on one of our more accessible walking challenges
- Host a quiz night or get your friends together for a catch-up and a meal
- Host a bake sale at work or school, or even a coffee morning with friends
- Share information about the activities we have going on to get friends and family joining in
- Stream online from the comfort of your own home

However, if you can run a marathon or want to do a thrilling skydive, we've got you covered!

Whatever you want to do, we can support you to raise money for Leukaemia Care. Get in touch with the fundraising team by email fundraising@leukaemiacare.org.uk or calling **08088 010 444**.

You can also find out more about how to get involved by scanning the QR code.

Plenty of ways to give

There are so many ways you can give in support of those affected by a leukaemia diagnosis, the possibilities are endless - find one that fits you and let's get giving!

By bank transfer

You can transfer your donation straight from your account to ours. Our bank details are:

Sort code: **20-98-61**

Account number: **80823805**

Account name: **Leukaemia Care**

By cheque

Please make your cheque payable to Leukaemia Care, and then pop it in the post to: **Leukaemia Care, One Birch Court, Blackpole East, Worcester, WR3 8SG**

Online

Simply pop onto our website at www.leukaemiacare.org.uk/donate or scan the QR code to donate.

By phone

You can call us to pay by debit or credit card over the phone. Simply call **01905 755977**.

Leukaemia Care is the UK's leading leukaemia charity. For over 50 years, we have been dedicated to ensuring that everyone affected receives the best possible diagnosis, information, advice, treatment and support.

Every year, 10,000 people are diagnosed with leukaemia in the UK. We are here to support you, whether you're a patient, carer or family member.

Want to talk?

Helpline: **08088 010 444**
(free from landlines and all major mobile networks)

WhatsApp: **07500 068065**

Office Line: **01905 755977**

www.leukaemiacare.org.uk

support@leukaemiacare.org.uk

Leukaemia Care,
One Birch Court,
Blackpole East,
Worcester,
WR3 8SG

Leukaemia Care is registered as a charity in England and Wales (no. 1183890) and Scotland (no. SC049802).

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Leukaemia Care
YOUR Blood Cancer Charity

Trusted Information Creator
Patient Information Forum

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