Polycythaemia vera (PV)

Polycythaemia vera (PV) is a rare, slow-growing type of cancer that occurs when your bone marrow makes too many red blood cells. Find out what it is, signs and symptoms of PV, tests you might have to diagnose PV and what treatment you may have.

Summary

- Polycythaemia vera (PV) is a rare slow-growing blood cancer which happens when your bone marrow makes too many red blood cells.
- 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.
- Around half of people with PV do not have symptoms when they are diagnosed. But you might get signs and symptoms over time. These include fatigue, a swollen spleen and itching.
- People with PV have a higher risk of blood clots and bleeding than other people.
- Your haematology team will usually diagnose PV using blood tests and sometimes bone marrow tests.
- PV treatment focuses on managing your symptoms and preventing blood clots. Your haematology team will suggest the most suitable treatment for you based on your individual circumstances.
- Making some lifestyle changes like stopping smoking, maintaining a healthy weight, eating a healthy diet and exercising can reduce your risk of blood clots.
- 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.
- People with PV who do not develop another type of blood cancer usually have excellent outcomes, and only a slightly reduced life expectancy.
- **We are here for you if you need support.**

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What is PV?

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.

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Who gets PV?

PV is rare. Around 1,140 people are diagnosed with it each year in the UK.

- 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.

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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, which helps regulate blood cell production. But we don't know why this change happens.

PV is not usually inherited, and you cannot usually pass it on to any children that you may have.

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Signs and symptoms of PV

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

- Blood clots

- Unusual bleeding or bruising more easily than usual
    - These might mean you get nosebleeds, heavier or longer periods, blood in your wee or poo, or more seriously, coughing up blood
- A swollen spleen
    - This can cause tummy pain, bloating, feeling full quickly after you eat or discomfort under your ribs on the left-hand side
- Fatigue
- Itching, especially after your skin has been in contact with water
- Night sweats
- Fever
- Joint or bone pain
- Weight loss
- Headaches, dizziness or light headedness

Blood clots

Blood clots can cause different symptoms depending on where they are. They include:

- **Heart**: chest pain, breathlessness or discomfort in your back, arm, shoulder or neck.
- **Brain**: drooping of your face on one side, slurred or garbled speech or difficulty keeping both arms raised. You may also get headaches, fits (seizures), fainting or loss of consciousness.
- **Lung**: chest pain, sudden breathlessness or coughing up blood.
- **Arm or leg**: painful, red, hot or swollen arm or leg.
- **Eye**: blurred vision or loss of vision.
- **Gut**: tummy pain, bloating or bleeding in your gut.

Risk of transformation

PV is a slow-growing blood cancer. Sometimes it can transform into a faster-growing type of blood cancer such as myelofibrosis (MF) or acute myeloid leukaemia (AML).

The risk of PV transforming varies from person to person. 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. This means 97 to 98 in every 100 do not.

If your PV transforms, you may notice that certain symptoms may get worse such as fatigue, joint pain and the swelling of your spleen.

Your haematology team will regularly monitor your blood tests for signs that your PV has transformed. If it happens, you'll need different treatment.

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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.

Your medical team will usually diagnose PV using:

- Blood tests
- Bone marrow tests
- Genetic tests on your blood and bone marrow samples
- Other tests, such as an ultrasound to check if your spleen is swollen

You might not need all of these tests.

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Treatment for PV

PV is a life-long condition. The aim of 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 risk of getting blood clots
- Your symptoms and blood test results
- Your age, overall fitness and family history
- Your personal preference
- Any other health conditions that you may have
- Whether or not you have had treatment before

- Any side effects that you may have had with other treatments

Some treatments, such as venesection and aspirin, are recommended for all people with PV. If you have a higher risk of getting blood clots then you may also need medicines that will lower your red blood cell counts.

Venesection

Venesection is a simple procedure to remove some of your blood, 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.

- You have venesection as an outpatient, usually at the hospital.
- A nurse puts a needle attached to a thin plastic tube into a vein in your arm.
- They 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 blood counts stable.

Sometimes venesection can lead to low iron levels. This may cause symptoms like restless legs, concentration problems, dizziness, fatigue and headaches. Tell your haematologist if you have any side effects.

Do not take iron supplements without discussing it with your haematology team first.

Aspirin

Aspirin helps stop your platelets sticking together, which prevents blood clots.

- 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.

Most people don't have any problems when taking aspirin. But if you experience serious side effects such as unusual bleeding, trouble breathing, stroke, cramps in your lower chest or bruising you should seek medical help.

Treatment if you have a higher risk of getting blood clots

If you have a higher risk of getting 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 take 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.

Hydroxycarbamide can cause a number of different side effects. If you experience serious side effects such as frequent infections, bruising, mild bleeding, changes to your skin or mouth ulcers, you should contact your haematology team.

Hydroxycarbamide might also increase the risk of your PV transforming to a faster-growing blood cancer if you are taking it for a long time.

Peginterferon (also known as pegylated interferon)

Peginterferon is a medicine that alters the way that your immune system works. It helps stop cancer cells growing and multiplying.

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

Peginterferon can cause side effects, but not everyone will get them. If you experience serious side effects such as changes in your heartbeat, suicidal thoughts, loss of vision, bleeding in your eye or breathing problems, you should seek medical help.

Ruxolitinib

Ruxolitinib is a type of targeted therapy. It blocks a protein that encourages your bone marrow to make too many red blood cells.

- Ruxolitinib comes as tablets that you take by mouth.

- Usually, you start on a dose of 10mg twice a day. You have blood tests during your treatment to monitor your response.
- Your haematology team will tell you what dose is best for you based on your own individual situation.

Ruxolitinib can cause side effects but not everyone will get them. Seek medical help if you get any serious side effects such as:

- Unexpected bruising or bleeding
- Painful skin rash with blisters
- Shortness of breath
- Infection
- Numbness, tingling or weakness

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

- A growth or unusual patch on the 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.

Other treatments

Depending on your circumstances, your team might suggest other treatment options. They will tell you what treatment they recommend and what you can expect from it.

Treatment for blood clots or bleeding

If you get a blood clot, you will have anti-clotting medicines to treat your clot and prevent future clots.

If you experience heavy bleeding, you will have treatment to stop the bleed and help replace the fluids you have lost.

Treatment to help with symptoms

Supportive treatment can help you deal with the symptoms of your PV, such as itching, which can impact your quality of life. Treatments that may help relieve itching include:

- Antihistamine tablets
- Corticosteroid creams, foams, gels, lotions or ointments

- Some antidepressants
- Light therapy

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Lifestyle measures to help prevent blood clots

There are things you can do yourself 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.

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.

Looking after your heart health can help 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
- Having regular tests to check your cholesterol and blood sugar levels
- Getting your blood pressure checked regularly

If you can, try to eat a well-balanced diet. 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.

Exercise can improve your heart health, quality of life and wellbeing. You may not feel like being active, especially with some of the side effects you may have. Choose a level of exercise that works for you and how you are feeling.

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Outcomes of PV

People with PV who do not develop myelofibrosis (MF) or acute myeloid leukaemia (AML) usually have excellent outcomes, and only a slightly reduced life expectancy.

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.

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Need support?

You are not alone. We're here for you whether you have a diagnosis yourself or know someone who has. If you'd like advice, support, or a listening ear, call our freephone helpline on 08088 010 444 or send a WhatsApp message to 07500 068 065.

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