Polycythaemia Vera (PV)

A Guide for Patients

Leukaemia Care
YOUR Blood Cancer Charity

Introduction

Being diagnosed with polycythaemia vera (PV) can be upsetting, particularly when you may never have heard of it before, and may even have had no obvious symptoms. If you have questions about PV – what causes it, who it affects, how it affects your body, what symptoms to expect and likely treatments – this booklet covers the basics for you.

You'll also find useful advice about how to get the best from your haematologist, plus practical advice on how to help important people in your life understand such a rare condition. For more information, talk to your haematologist, clinical nurse specialist or hospital pharmacist.

This booklet was originally compiled by Professor Claire Harrison, Consultant Haematologist at Guy's and St Thomas' NHS Foundation Trust. Subsequently, it was peer reviewed by Manos Nikolousis, Consultant Haematologist at Heart of England NHS Trust. The rewrite was done by Lisa Lovelidge and reviewed by Claire Harrison. This booklet has since been updated by our Patient Information Writer, Isabelle Leach, and reviewed by Dr. Mallika Sekhar, UCLH. We are also grateful to Nicki Payne, Clare White, Tracy Broadley, Andrew Norman, Joe Thomas, Andrew Walker and Daigon North for their valuable contributions as patient reviewers.

If you would like any information on the sources used for this booklet, please email **communications@leukaemiacare.org.uk** for a list of references.

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In this booklet

Introduction	2
In this booklet	3
About Leukaemia Care	4
What is polycythaemia vera (PV)?	6
Symptoms of PV	8
How is PV diagnosed?	10
What is the prognosis of PV?	12
Treating PV	13
Living with PV	18
Talking about PV	22
Glossary	25
Useful contacts and further support	27

About Leukaemia Care

Leukaemia Care is a national charity dedicated to ensuring that people affected by blood cancer have access to the right information, advice and support.

Our services

Helpline

Our helpline is available 8:30am – 5:00pm Monday - Friday and 7:00pm – 10:00pm on Thursdays and Fridays. If you need someone to talk to, call **08088 010 444**.

Alternatively, you can send a message via WhatsApp on **07500068065** on weekdays 9:00am – 5:00pm.

Nurse service

We have two trained nurses on hand to answer your questions and offer advice and support, whether it be through emailing **nurse@leukaemiacare.org.uk** or over the phone on **08088 010 444**.

Patient Information Booklets

We have a number of patient information booklets like this available to anyone who has been affected by a blood cancer. A full list of titles – both disease specific and general information titles – can be found on our website at **www.leukaemiacare.org.uk/support-and-information/help-and-resources/information-booklets/**

Support Groups

Our nationwide support groups are a chance to meet and talk to other people who are going through a similar experience. For more information about a support group local to your area, go to **www.leukaemiacare.org.uk/support-and-information/support-for-you/find-a-support-group/**

Buddy Support

We offer one-to-one phone support with volunteers who have had blood cancer 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**

Online Forum

Our online forum, **www.healthunlocked.com/leukaemia-care**, is a place for people to ask questions anonymously or to join in the discussion with other people in a similar situation.

Patient and carer conferences

Our nationwide conferences provide an opportunity to ask questions and listen to patient speakers and medical professionals who can provide valuable information and support.

Website

You can access up-to-date information on our website, **www.leukaemiacare.org.uk**.

Campaigning and Advocacy

Leukaemia Care is involved in campaigning for patient well-being, NHS funding and drug and treatment availability. If you would like an update on any of the work we are currently doing or want to know how to get involved, email **advocacy@leukaemiacare.org.uk**

Patient magazine

Our magazine includes inspirational patient and carer stories as well as informative articles by medical professionals: **www.leukaemiacare.org.uk/communication-preferences/**

What is polycythaemia vera (PV)?

PV belongs to a group of conditions called myeloproliferative neoplasms (MPNs), which also includes essential thrombocythaemia (ET) and myelofibrosis (MF). PV is a chronic condition characterised by too many red blood cells, and sometimes too many platelets and white blood cells, in the blood.

If you would like more information about ET or MF, you can download our dedicated booklets from our website **www.leukaemiacare.org.uk** or request a copy by emailing **support@leukaemiacare.org.uk** or calling our helpline on **08088 010 444.**

There has been some debate about whether or not MPNs are types of cancer. This is because the word neoplasm (to mean new growth) is a term used both for cancers (malignant neoplasms) and noncancerous tumours (benign neoplasms). In PV there is an uncontrolled increase in cells; therefore, many haematologists and cancer organisations do consider MPNs as blood cancer. Nevertheless, the symptoms and prognosis of patients with PV can vary widely.

Blood cells which are produced from stem cells in the bone marrow include red blood cells (to carry oxygen to the tissues in your body), white blood cells (to fight infection and disease) and platelets (to help prevent bleeding by causing blood clots to form).

Your body closely controls the production of new blood cells so that it is balanced with the loss of worn-out cells or cells lost by bleeding or damage. About one in 5000 cells in the bone marrow is a blood-forming stem cell which either divides to produce more stem cells or develops into one of the working blood cells. An average adult produces about one trillion new blood cells each day.

In patients with PV, the bone marrow makes too many red blood cells (although white blood cell counts and platelet

counts may also increase in number), making blood thicker than normal. In 30% of patients with PV, this excess of red blood cells may cause blood clots to form more easily. Clots can block blood flow through the arteries and veins, potentially leading to heart attacks or strokes. In addition, as thicker blood doesn't flow as quickly as normal blood, your organs may not get enough oxygen.

Who is affected by PV?

PV is considered to be a rare disease. The number of people diagnosed each year is around two to three per 100,000 people. This may explain why you might not have heard of PV or met anyone with the condition before. PV is rarely diagnosed before the age of 40 years, and is most common in those aged over 60. It is more commonly seen in men than women.

What causes PV?

While the exact cause of PV is not known, research has found that about 95% of people who have PV have a change (commonly referred to as a mutation) in a protein that regulates blood cell production. This protein is known as JAK2 – short for Janus Kinase 2 – and the mutation in the chromosome is known as JAK2 V617F.

It is also important to be aware that, although some families seem to develop the disease more readily than others, PV is not usually inherited nor passed on from parent to child. Some researchers believe MPNs may also be triggered by past exposure to ionising radiation (a type of radiation that has very high energy, like medical x-rays or nuclear fallout) or to some chemical substances such as benzene and toluene.

Symptoms of PV

Approximately 50% of patients with PV do not have any symptoms when they are diagnosed. Typically, these patients are identified following abnormal results during a routine full blood test for something else. If symptoms do develop, they tend to do so over time. If you have persistent symptoms of PV as listed below, make an appointment to see your GP.

The most common symptoms of PV result from blood clots in the arteries, which may lead to heart attacks, strokes, or damage to the gastro-intestinal tract, and in the veins, leading to venous thrombosis or pulmonary embolism, where a clot in a vein travels through the blood stream and causes a blockage in one of the arteries of the lungs.

Enlargement of the spleen affects up to 75% of patients with PV because it overcompensates for the bone marrow not functioning correctly and producing more red blood cells. More rarely, the liver may also be affected and become enlarged.

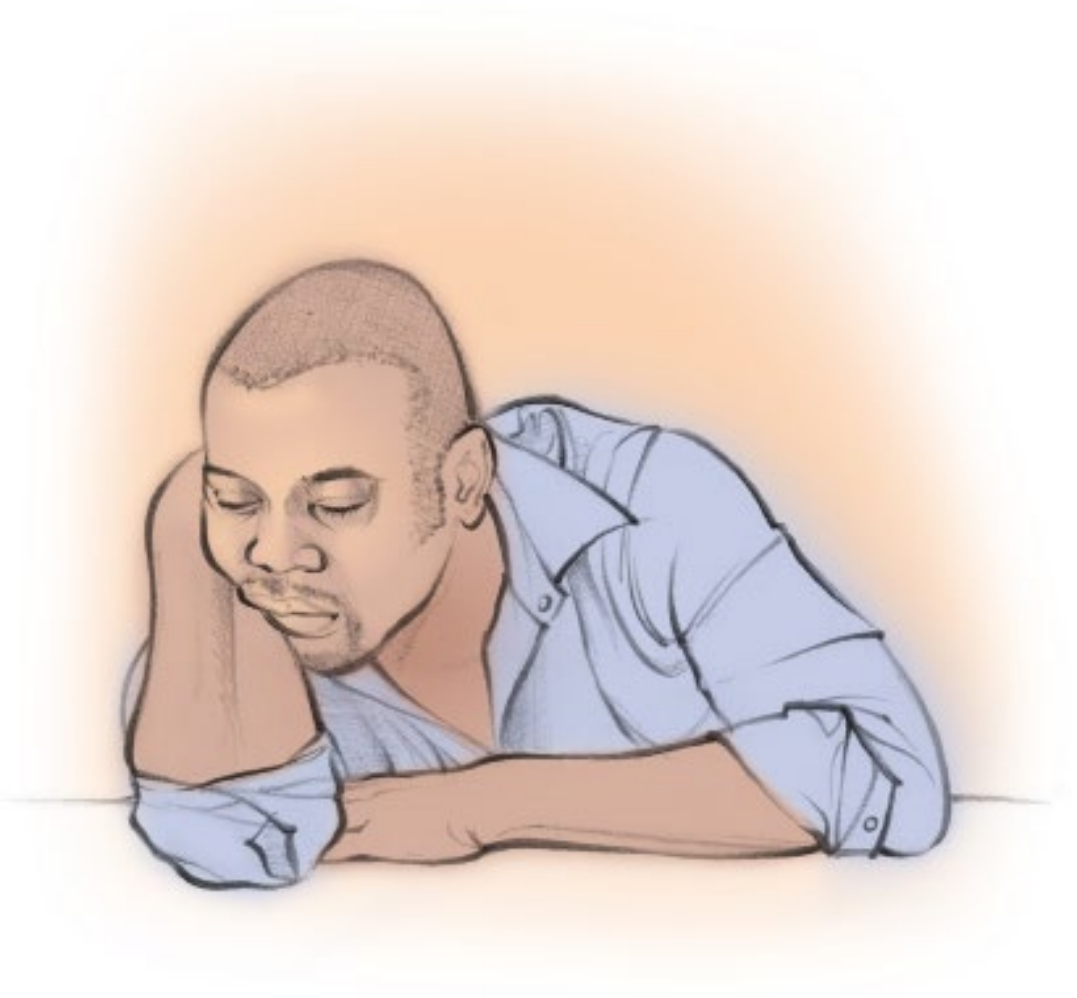
Common symptoms include:

- Fatigue
- Night sweats
- Headaches
- Itching
- Fever
- Swollen spleen
- Bone pain
- Gout
- Weight loss
- Dizziness or light headedness
- Reddish or purple skin
- Bleeding or clotting

Not everyone will have the same symptoms or to the same degree of severity.

Blood clots and bleeding in patients with PV are more likely to occur in people aged over 60 years or those who have a history of thrombosis.

People considered to be at low risk of developing thrombosis are those who are younger than 60 years, with no previous history of thrombosis, and without any other risk factors for cardiac disease, such as high blood pressure, diabetes, high cholesterol or smoking.



How is PV diagnosed?

A diagnosis of PV requires both of the following:

- A haematocrit of 52% (0.52) or more for men and 48% (0.48) or more for women (normal range is 41% to 51% (0.41 to 0.51) for men, and 37% to 47% (0.37 to 0.47) for women). A haematocrit is the volume of red blood cells expressed as a percentage of the total volume of blood cells.
- The presence of a JAK2 mutation.

The JAK2 V617F mutation can be found in the majority of patients with PV, but in those where there is an absence of a JAK2 mutation, there are a number of further characteristics which must be present to confirm a diagnosis of PV:

- A haematocrit of 60% or more in men and 56% or more in women.
- No secondary cause of increase in red blood cells, such as disorders that cause oxygen deprivation in tissues or abnormal increase of, or sensitivity to, erythropoietin, a hormone which stimulates the bone marrow to produce red blood cells.
- A microscopic structure of the bone marrow consistent with PV.

In addition, at least one of the following must also be present to confirm a diagnosis of PV:

- An enlarged spleen which can be felt on examination or seen on a scan/x-ray.
- An acquired gene mutation, other than BCR-ABL1, in the bone marrow cells.
- Platelet count greater than $450x10^9$/L (normal range is $150x10^9$/L and $450x10^9$/L).
- A neutrophil white blood cell count greater than $10x10^9$/L in non-smokers and $12.5x10^9$/L or more in smokers. Neutrophils are white blood cells which protect the body against bacterial infections and inflammation.
- Low serum erythropoietin levels. Erythropoietin is a hormone released when the oxygen level in the blood is low to encourage stem cells to become more red blood cells.

PV is often suspected if a routine blood test shows that a patient has a high red blood cell count, sometimes in conjunction with raised white blood cell and platelet counts.

PV is diagnosed using laboratory tests including:

- **Blood tests** – Blood tests can identify an increase in blood cells and exclude other causes of a high blood cell count.
- **Bone marrow investigations** – You may have either a bone marrow aspiration or both an aspiration and a bone marrow biopsy. These will be done to look for classic signs of PV. During a bone marrow aspiration, the doctor or nurse takes some bone marrow cells up into a syringe. A bone marrow biopsy is when they remove a one to two-centimetre core of bone marrow in one piece using a trephine. The sample is then sent to the laboratory for testing.
- **Gene mutation analysis** – You may also be tested for gene mutations, including JAK2.

What is the prognosis of PV?

When properly monitored and treated, patients with PV have an excellent prognosis and life expectancy. Your prognosis can be influenced by a number of factors including your individual situation, your health history and the way you respond to treatment. PV is a chronic condition, so it is important for you to see your haematologist regularly and report any new or different symptoms.

For the majority of patients whose PV does not progress to MF or acute myeloid leukaemia (AML), a normal or very slightly reduced life expectancy can be anticipated.

In some patients, PV can develop into MF or AML. Ten years after diagnosis, the risk of your PV transforming to MF is 10%, and the transformation risk to AML is between 5% and 21%. The prognosis of patients with PV can vary widely. Your haematologist is the best person to advise you based on your individual circumstances.

While the presence of a JAK2 gene mutation is associated with the possible development of MF or AML, up to 15% of patients with PV have more than one mutation, such as ASXL1 or SRSF2. Both of these are also associated with a reduced rate of overall survival.

Other factors which contribute towards your prognosis and survival are your age, blood cell counts and cardiovascular risk factors for thrombosis:

- Older age, a high white blood cell count and previous history of venous thrombosis are associated with a poor survival.
- An enlarged spleen and an increased risk of transformation into MF or AML have been associated with shorter overall survival.
- High platelet counts and itching are factors associated with a better survival.

So far, drug treatment for patients with PV has not increased survival or prevented transformation into MF and AML. Treatment is focussed mainly on preventing any complications from thrombosis which is associated with PV. Low-risk patients are best managed with low dose aspirin therapy or venesection, and patients with high-risk PV will benefit mainly with cytoreductive therapy (a treatment to control your blood counts).

Treating PV

Overview of treatment

Most treatments for PV are intended to manage your symptoms and prevent any associated problems, particularly the thrombotic complications of PV, in order to maintain your quality of life.

Treatments for PV are intended to lower the production of blood cells and help to maintain a normal blood volume. This is called cytoreductive therapy. There are a number of medications as well as a procedure called venesection (drawing blood), also called phlebotomy, which can achieve this. Your medical team will give all the information about the treatment which is best for you.

While cytoreductive therapy is fundamental in treating PV, managing any potential thrombosis or haemorrhage is equally important.

Treatment will be based on an assessment of your risk factors for thrombosis which is a major cause for troublesome symptoms and shortened survival. These risks include:

- Your history of clotting
- Advanced age (over 60 years)
- Cardiovascular risk factors (such as high cholesterol levels, diabetes, smoking, obesity or high blood pressure)

Their management is important as up to a third of patients with PV will present with a thrombotic complication.

Venesection

Venesection is the removal of blood from a vein. It is also known as phlebotomy. It is the usual starting point of treatment for most patients. A volume of blood is drawn at regular intervals and the haematocrit concentration is brought down to normal values within a period of weeks to months. The procedure is similar to that used for blood tests or donating blood. The immediate effect of venesection is to reduce the haematocrit, which usually results in the improvement of certain symptoms such as headaches, ringing in the ears and dizziness.

Venesection may be the only form of treatment required for many patients. Acceptable disease control may be achieved with the withdrawal of a volume of blood every few months. However, patients may feel tired afterwards and need to rest for a short time.

Treating PV (cont.)

Medications

Management of thrombosis

Aspirin

Along with venesection, daily low-dose aspirin will usually be prescribed for patients with PV when treatment has started. As well as being used for reducing pain and lowering temperature, aspirin is known to prevent platelets sticking together and may reduce your risk of developing a blood clot. Low-dose aspirin for patients with PV reduces the risk of:

- Non-fatal heart attacks
- Non-fatal strokes
- Pulmonary embolisms (a blood vessel in your lungs being blocked by a blood clot)
- Major venous thrombosis (a blood clot that forms in the vein)
- Death from cardiovascular causes

Low-dose aspirin can have some side effects such as bleeding and indigestion. It can also cause gastric irritation and bleeding in the stomach and ulcers. You may find that you bruise more easily and that you bleed for a long time if you cut yourself. Applying pressure to any small cut or wound with a sterile bandage will stop the bleeding. Aspirin can make the symptoms of asthma worse for those who suffer with it. In addition, some people can be allergic to aspirin.

Your haematologist will tell you if aspirin is safe for your particular situation and may suggest other similar medications, such as dipyridamole or clopidogrel, if necessary.

Anticoagulation treatment

Long-term anticoagulation treatment can be started if you have had a venous thromboembolism that has occurred with no identifiable risk factor. This will be done after your bleeding risk has been assessed. Patients with PV and other MPNs more commonly have unusual sites of venous thrombosis:

- **Splanchnic vein thrombosis** - The splanchnic venous circulation drains the blood from the stomach, pancreas, spleen and the intestines.
- **Cerebral vein thrombosis** - The cerebral vein drains blood from the brain.

The use of anticoagulants will be carefully monitored to balance the

increased risk of bleeding with the need to prevent thrombosis recurrence.

Management of haemorrhage

Haemorrhage (bleeding) is a less common complication of PV occurring in approximately 8% of patients. In addition, while bleeding is a less severe complication of PV than thrombosis, it can still affect your mucous membranes, gastrointestinal tract and skin. Mucous membranes are thin skin membranes that line the body's cavities such as the mouth, eyes and gastrointestinal tract.

Bleeding in patients with PV is more common if they have a high platelet count (greater than $1500x10^9$/L) or associated acquired von Willebrand syndrome. If you have experienced some bleeding, you will be screened for acquired von Willebrand syndrome, and if the test is negative, you will have your platelet function tested. Your haematologist will closely monitor any antiplatelet drugs or anticoagulant drugs you may be receiving.

Any significant bleeding episodes will be managed with tranexamic acid, which stops bleeding in the short term, or a transfusion of platelets.

Cytoreductive therapy

Cytoreductive therapy is crucial in enabling PV and its complications to be controlled. It allows the blood cell counts to be maintained at the right levels.

If venesection is not sufficient to achieve acceptable control of your PV, there are a number of drugs which can help achieve this.

Hydroxycarbamide (also known as hydroxyurea)

This is the most commonly used chemotherapy drug to treat PV and is available as a tablet/capsule to be taken orally.

It can cause mild side effects such as:

- Increased risk of infection
- Bruising or mild bleeding
- Anaemia (as it reduces all the types of blood cells, including red blood cells)
- Fatigue
- Diarrhoea or constipation
- Sore mouth
- Changes to the skin, including ulceration and increased risk of

Treating PV (cont.)

developing skin cancer

Hydroxycarbamide may also affect fertility. If you are taking it, you will be advised not to get pregnant or father a child, as there may be a risk of harming the developing baby. It is advisable to use effective contraception while taking the drug and also for a few months afterwards.

Being treated with hydroxycarbamide over a long period of time, either alone or in combination with other chemotherapy drugs, may increase the chance of the PV developing into AML.

Interferon alpha

Interferon alpha is a substance which occurs naturally in the body and reduces the production of bone marrow cells. It can be made into a medicine to be given as an injection under the skin which reduces the rate at which blood cells are made. It does have side effects such as:

- Flu-like symptoms
- Headaches
- Vision disturbances
- Depression
- Liver and thyroid disease

However, it does not increase the risk of leukaemia and can be used in pregnancy.

Another form of this medicine, Pegylated interferon alpha-2a, where a polyethylene glycol is attached to the interferon drug, slows its breakdown in the body, enabling it to act for longer, and have fewer side effects.

JAK2 inhibitors

JAK2 inhibitors were developed as a result of the discovery of the link between the JAK2 gene mutation and the incidence of PV. Approximately 95% of patients with PV are found to have a JAK2 mutation.

The JAK2 gene makes a protein called JAK2, which stimulates cell growth, but also helps control the number of cells made in the bone marrow. The JAK2 V617F mutation results in the JAK2 protein being constantly 'on', leading to uncontrolled blood cell production. JAK2 inhibitors block the function of JAK2 mutation which slows down blood cell production, reduces spleen size and improves symptoms.

Other treatments

The following treatments have been found to increase the risk of PV progressing to acute

leukaemia and are therefore reserved for patients with PV who have a limited life expectancy:

- **Busulfan** – This is usually given to patients who have side effects when taking hydroxycarbamide. It can be given as a tablet. Like hydroxycarbamide, busulfan affects the bone marrow directly and can lead to a fall in the blood count. Its main side effects are nausea and low platelet counts. Your haematologist can advise you if busulfan is suitable for you, and will monitor your progress carefully during your treatment. There is evidence that busulfan can increase the risk of leukaemia and can cause lung scarring.
- **Radioactive phosphorus (P-32)** - P-32 is given as an injection which irradiates the bone marrow with the result of lowering the number of blood cells being produced. The effects of a single injection can last from months to years and may be used in some situations. Treatment with radioactive phosphorus over many years can cause an increased risk of leukaemia, but this is rare.

Relief from itching

Other than ruxolitinib which is recommended for severe itching, the following treatments can also provide relief from itching:

- **Antihistamines** – Antihistamines or related drugs may be prescribed to relieve itching. Side effects may include a dry mouth, drowsiness and dizziness.
- **Phototherapy** – The use of light can be used to treat itching. It involves exposing the skin to ultraviolet light on a regular basis under medical supervision. It works temporarily to ease generalised itching.

Summary of treatment for PV

In summary, treatments for patients with PV according to their risk are:

- **Low-risk PV:** Venesection to a haematocrit target of 45% and low-dose aspirin.
- **High-risk PV:** Venesection to a haematocrit target of 45%, low-dose aspirin and cytoreductive therapy (hydroxycarbamide, pegylated interferon alpha or a JAK2 inhibitor in the event of prolonged itching or a very enlarged spleen).

Living with PV

After a diagnosis of PV, you may find that it affects you both physically and emotionally. This chapter will talk about both of these aspects.

Emotional impact of PV

Being told you have cancer can be very upsetting. Although the outlook for many PV patients is a positive one, it is a blood cancer and a rare condition and, because of this, you may need emotional, as well as practical, support. Being diagnosed with a rare disease can affect the whole of you, not just your body, and can impact you emotionally at any point of your journey. It is likely that you will experience a range of complex thoughts and emotions, some of which may feel strange or unfamiliar to you. It is important to know that these feelings are all valid and a normal response to your illness.

It is important to remember that with current treatments, you can expect a good response and to live a long, normal life.

If you would like some emotional support, there are a number of places that you can seek it from:

- Caregivers and loved ones can be there for you at home. They can also attend hospital appointments with you. Having someone else there to take in all the information can help things seem less vast.
- You may be assigned a clinical nurse specialist. They will be there to assist you with your treatment pathway. They can be a great source of medical and supportive information relating to your PV.
- Charities and organisations, including Leukaemia Care, offer a number of services that aim to offer some comfort. You can call our helpline on **08088 010 444** to speak to a nurse or a trained member of our Patient Services team. Sometimes, speaking to someone you don't know or who isn't directly involved in your situation can be useful. For a full list of our other services, you can ask about them on our helpline, or find more information on our website **www.leukaemiacare.org.uk**
- It is also important to form a positive working relationship with your consultant. This

should be based on good patient-doctor communication. You should feel comfortable to seek as much information from them as you would like, but also be able to question something if it doesn't feel quite right. Having this type of working relationship with them will help you to feel supported.

Looking after you

To help live a long and normal life with PV, you may want to make changes to your lifestyle to try to stay as well as possible after your diagnosis and during treatment. Don't try to change too much at once. Adopting a healthy way of living is about making small, manageable changes to your lifestyle.

A healthy lifestyle includes having a well-balanced diet and being physically active. With some of the side effects you may be experiencing, the idea of getting out and being active may be the last thing you want to do. However, it is important to try and stay as active as possible to make you feel better and reduce some of the symptoms or side effects you may be experiencing.

"People with an MPN should not smoke and should exercise to maintain their general fitness and improve their vitality and cardiovascular performance – this also reduces their increased risk of cardiovascular disease." – Professor Claire Harrison

One of the most commonly reported side effects of the treatment of PV is fatigue. This isn't normal tiredness and doesn't improve with sleep.

Some general tips on how to deal with fatigue include:

- Have a regular lifestyle – try going to bed and waking up approximately the same time every day and try to avoid lying in.
- Take part in regular, gentle exercise to maintain your fitness levels as much as possible.
- Reserve your energy for what you find important and build

Living with PV (cont.)

rest periods around those times.

- Before going to bed avoid stimulants such as alcohol, coffee, tea or chocolate, or using laptops, tablets or mobile phones.
- Keep your bedroom quiet and at a comfortable temperature.
- Talk about your worries with family, friends or your doctor or nurse, or patient support groups.
- Discuss your fatigue with your doctor or nurse.

Practical support

Work and finances

Being diagnosed with PV can sometimes lead to difficulties relating to your work life. Your diagnosis may lead to temporary sick leave or a reduction in working hours but it can also mean that you have to stop work altogether. You may need to make an arrangement with your employer for times when you may need to go into hospital or for those times when you may not be well enough to go into work.

Your consultant or your GP can arrange letters to your employers to confirm your diagnosis and the effects it may have on your work life. It is often worth taking time to explain PV to your employer, as it is likely they will never have heard of the disease.

It is important for you to know that people with any form of cancer are covered by law by the Equality Act. This means that legally your employer cannot discriminate against you and must make reasonable arrangements and adjustments relating to your disease.

If you would like advice about some of the financial help available to you, then you can speak to our Patient Advocacy team on **08088 010 444**. Alternatively, Macmillan has published a booklet about financial support following a diagnosis of cancer that might be useful to you. They can also give you personal advice over the phone via their helpline at **0808 808 0000** and you can discuss which benefits you are eligible for. Some Macmillan centres can arrange face-to-face meetings with a benefits advisor. They can also provide financial assistance

in the form of grants – ask your nurse in the hospital how to apply.

As PV can be regarded as a cancer, you will also be entitled to apply for a medical exemption certificate which means that you are entitled to free NHS prescriptions. Your GP or specialist nurse at the hospital can provide you with the details on how to apply for this.

Talking about PV

Talking to your GP

Arranging an appointment with your GP will be one of the first things you will need to do. You will then be referred to the hospital who will be able to confirm this through tests.

Be honest with your GP; there is no need to feel embarrassed about anything. Although some GPs may not have heard of PV before your diagnosis, you should not feel disheartened by this. Your GP and your medical team at the hospital will do everything they can to ensure you receive the best treatment plan for you.

It is important to know exactly what you would like to ask your GP so try to think of all the questions you would like to ask before your appointment. Always ensure that you leave the GP surgery or the hospital having shared everything you know about your condition, with all of your questions answered, and knowing exactly what the next steps are.

Talking to your haematologist

PV is a rare condition. It is important for you to develop a good working relationship with your haematologist so you are given the best treatment possible for you.

The following gives advice on working well with your haematologist:

- If it's an initial consultation, take along a list of your current medications and doses, and a list of any allergies you may have.
- If you have a complicated medical history, take a list of diagnoses, previous procedures and/or complications.
- Make a list of questions to take to your appointment. This will help the discussion with your haematologist.
- It can be useful to repeat back what you have heard so that you can be sure you have fully understood.
- Note information down to help you remember what was said.
- Be open when you discuss your symptoms and how you are coping. Good patient-doctor communication tends to improve outcomes for patients.

Other tips:

- Bring someone along to your appointment. They can provide support, ask questions and take notes.
- Don't be afraid to ask for a second opinion – most haematologists are happy for you to ask.

You need to tell your haematologist if...

You're having any medical treatment or taking any products such as prescribed medicines, over the counter treatments or vitamins. It is important to understand that treatments, including complementary therapies, which are perfectly safe for most people, may not be safe if you are being treated for PV. Remember, if you choose to start any form of complementary therapy outside of your medical treatment, consult your haematology consultant or CNS prior to beginning it. It is important to understand the difference between complementary therapies, used alongside standard treatment, and alternative therapies, used instead of standard treatment.

There is no evidence that any form of alternative therapy can treat PV.

Talking to other people

Telling people you have a rare condition like PV can be hard to explain. You might find it useful to let your close family and friends, as well as your employer, know about your health condition. It might be easier to provide people with basic information and give them information leaflets about PV if they want to know more in-depth details.

It is probably best to focus conversations on the symptoms that you are experiencing, how the condition affects you and how you feel about it. Often people misunderstand and, unfortunately, it will mostly fall to you to educate them as best as you can. Where possible, it's advisable to let people know what you find helpful and unhelpful, in terms of what others say and do. Often people make assumptions and do what they think helps. For example, saying you look well, recounting stories of others they know with a similar diagnosis as well as encouraging you to look ahead and stay positive isn't always what people really want

Talking about PV (cont.)

to hear. In many ways, the more you communicate with them the better.

These points may help you:

- Explain that you have a condition that means your bone marrow does not function properly, and this affects the number of blood cells it produces.
- Explain your symptoms (maybe you are tired, or have a lot of pain).
- Explain what you need (maybe more to help day-to-day, or someone to talk to).

You could also consider the following when telling people about your diagnosis:

- Find out more – Try to find out as much as you can about your condition, from reliable internet sources, charitable organisations or your consultant haematologist. The more you know, the more you can share.
- **Have a print-out to hand** – It may help to have a factsheet to hand to share with family and friends. This will take the pressure off you having to remember everything they may want to know. Alternatively, you can give them this booklet to read when you're finished with it.
- **Explain your needs** – Try and be clear about what your needs may be. Perhaps you need help with the weekly food shop, help with cooking dinner, or someone to drive you to and from appointments. You may find that friends and family are pleased that they can do something to help you.
- **Be open about how you feel** – Don't be afraid of opening up about how you feel, as people who care will want to help you as best they can. Talk as and when you feel comfortable, so those around you will know when you need them most.

If you're struggling to come to terms with your diagnosis and prognosis, you can speak to us on our helpline. Call us on **08088 010 444**.

Glossary

Amino Acids

Organic molecules which are the building blocks for making proteins.

Anaemia

A condition where the number of red blood cells are reduced. Red blood cells contain haemoglobin and transport oxygen to body cells. This may be due to a lack of iron, leukaemia, or sickle cell disease.

Anticoagulation

A process of administering anticoagulants which are drugs to prevent the blood from clotting unnecessarily.

BCR-ABL1 Mutation (also called the Philadelphia chromosome)

The breakpoint Cluster Region-Abelson Murine Leukaemia Viral proto oncogene 1 (BCR-ABL1) is a cancer gene formed by the fusion of a portion of chromosome 9 on chromosome 22 to form the BCR-ABL fusion gene.

Bone Marrow

The soft blood-forming tissue that fills the cavities of bones and contains fat, immature and mature blood cells, including white blood cells, red blood cells and platelets.

DNA (Deoxyribonucleic Acid)

A thread-like chain of amino acids found in the nucleus of each cell in the body which carries genetic instructions used in the growth, development and functioning of the individual's cells.

Eosinophil

A type of white blood cell which has a protective immunity role against parasites and allergens.

Genes

Genes are made up of DNA which stores the genetic information required to make human proteins.

Monocyte

A white blood cell that attacks invading organisms and helps combat infections.

Myeloproliferative Neoplasms (MPNs)

A disease of the bone marrow in which excess cells are produced.

Myeloid

Relates to the bone marrow.

Platelets

One of the types of blood cell which helps to stop bleeding.

Prognosis

An indication of how well a

Glossary (cont.)

patient is expected to respond to treatment based on their individual characteristics at the time of diagnosis or other timepoint in the disease.

Pulmonary Embolism

A blockage of a blood vessel in the lung. It can be secondary to a clot elsewhere in the body which then travels up to the lung.

Red Blood Cell

A small blood cell that contains haemoglobin and carries oxygen and other substances to all tissues of the body.

Spleen

The largest organ of the lymphatic system whose function is to help rid the body of toxins, waste and other unwanted materials. The spleen is located under the ribs on the left of the abdomen.

Stem Cell

The most basic cell in the body that has the ability to develop into any of the body's specialised cell types, from muscle cells to brain cells. However, what makes these stem cells reproduce, uncontrollably, as in cancer, is thought to be linked to chromosome abnormalities.

Thrombosis

Clotting or coagulation of the blood in a part of the circulation in both arteries and veins.

von Willebrand Syndrome

A common blood-clotting disorder which causes varied degrees of bleeding. It can be hereditary or acquired. In acquired von Willebrand syndrome, the patient has auto-antibodies to the clotting factor affected by the syndrome.

White Blood Cells

White blood cells are one of the types of cells found in the blood and bone marrow, along with red blood cells and platelets. White blood cells create an immune response against both infectious disease and foreign invaders. Granulocyte white blood cells include the neutrophils (protect against bacterial infections and inflammation), eosinophils (protect against parasites and allergens) and basophils (create the inflammatory reactions during an immune response). Other white blood cells include lymphocytes (recognise bacteria, viruses and toxins, to which they produce antibodies) and monocytes (clear infection products from the body).

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

There are a number of organisations, including ourselves, who provide expert advice and information.

Leukaemia Care

We are a charity dedicated to supporting anyone affected by the diagnosis of any blood cancer. We provide emotional support through a range of support services including a helpline, patient and carer conferences, support group, informative website, one-to-one buddy service and high-quality patient information. We also have a nurse on our help line for any medical queries relating to your diagnosis.

Helpline: **08088 010 444**
www.leukaemiacare.org.uk
support@leukaemiacare.org.uk

Blood Cancer UK

Blood Cancer UK is the leading charity into the research of blood cancers. They offer support to patients, their family and friends through patient services.

0808 2080 888
www.bloodcancer.org.uk

Cancer Research UK

Cancer Research UK is a leading charity dedicated to cancer research.

0808 800 4040
www.cancerresearchuk.org

Macmillan

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

0808 808 0000
www.macmillan.org.uk

Maggie's Centres

Maggie's offers free practical, emotional and social support to people with cancer and their families and friends.

0300 123 1801
www.maggiescentres.org

Citizens Advice Bureau (CAB)

Offers advice on benefits and financial assistance.

08444 111 444
www.adviceguide.org.uk

Leukaemia Care is a national charity dedicated to providing information, advice and support to anyone affected by a blood cancer.

Around 34,000 new cases of blood cancer are diagnosed in the UK each year. 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)

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).
Company number: 11911752 (England and Wales).
Registered office address: One Birch Court, Blackpole East, Worcester, WR3 8SG

Leukaemia Care
YOUR Blood Cancer Charity