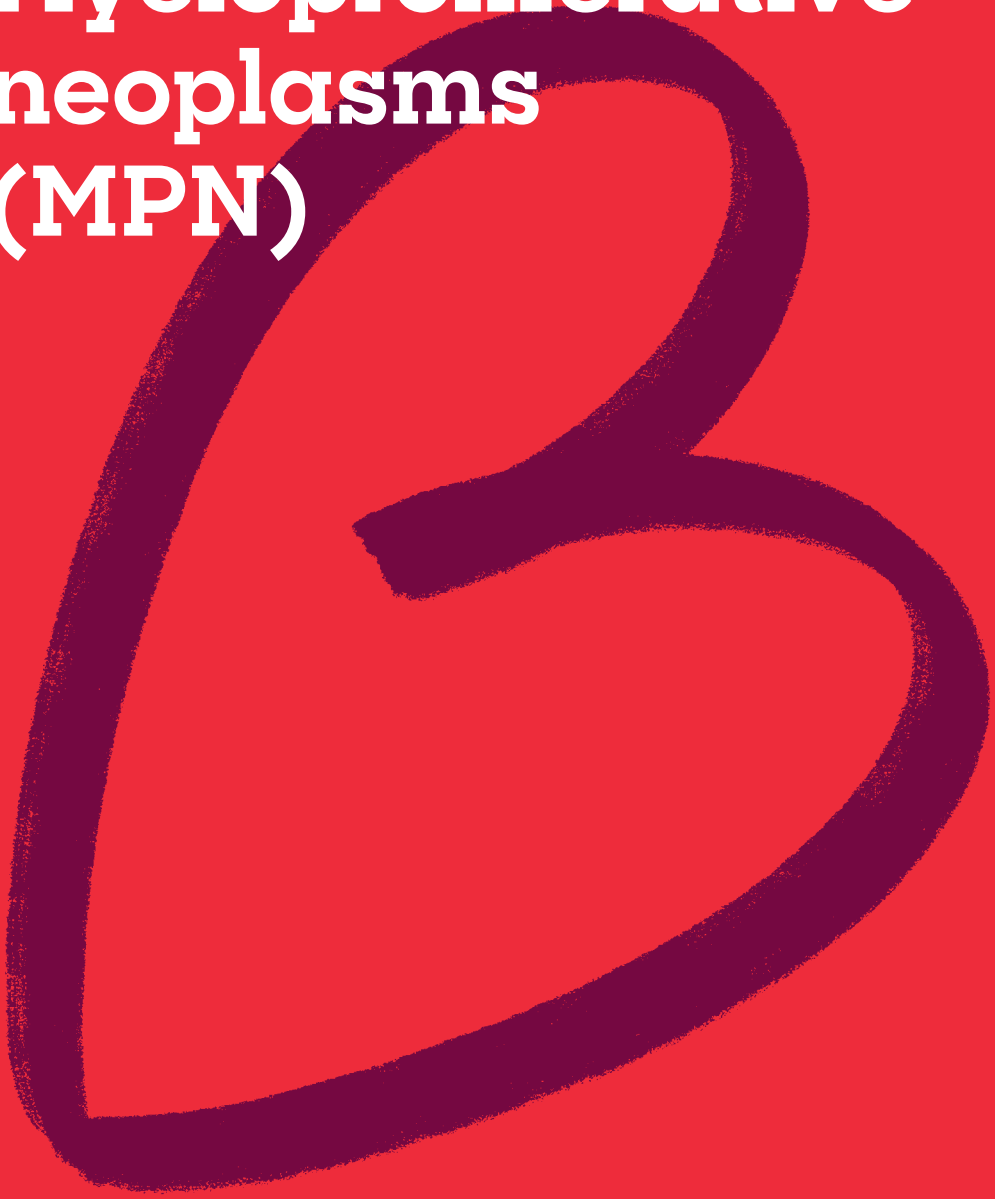
Myeloproliferative neoplasms (MPN)



Blood cancer UK

About this booklet

We have produced this booklet in collaboration with expert medical professionals and people affected by blood cancer. Thank you to Professor Tony Green, Dr Jacob Grinfeld, Dr Jyoti Evans, Dr Will Thomas and Clinical Nurse Specialists Vashti Ragoonanan, Yvonne Francis, Siobban McGuckin and Millicent Blake-McCoy for their support checking the content of this booklet.

We're a community dedicated to beating blood cancer by funding research and supporting those affected. Since 1960, we've invested over £500 million in blood cancer research, transforming treatments and saving lives. To find out more about what we do, see page 113.

bloodcancer.org.uk

0808 2080 888

(Mon–Fri, 10am–4pm, Sat–Sun, 10am–1pm)

support@bloodcancer.org.uk

A list of references used in this booklet is available on request. Please email us at **information@bloodcancer.org.uk**

Disclaimer

We make every effort to make sure that the information in this booklet is accurate, but you shouldn't rely on it instead of a fully trained clinician. It's important to always listen to your specialist and seek advice if you have any concerns or questions about your health. Blood Cancer UK can't accept any loss or damage resulting from any inaccuracy in this information, or from external information that we link to.

The information in this booklet is correct at the time it was printed (March 2015). In January 2019 we made some edits, which included updated information on busulfan, ruxolitinib and hydroxycarbamide. Date of next full review 2020.
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Contents

Introduction

This is a booklet for anyone with a myeloproliferative neoplasm (MPN), and their family and friends.

Being told that you, or a loved one, has MPN can be a tough thing to hear. We hope this booklet will help you understand your condition and feel more in control. It covers symptoms, tests, treatments, living with MPN and where you can get support.

Everyone is different, with a different medical history. So when you're deciding what's right for you, talk to your specialist and get information from this booklet and other trustworthy places.

For the most up-to-date information about MPNs and treatment, visit **bloodcancer.org.uk**

Our Support Services Team are here for anyone affected by blood cancer. Contact us on **0808 2080 888** or **support@bloodcancer.org.uk**

We have more information and personal stories about living with and beyond blood cancer on our website: **bloodcancer.org.uk/living-well**

Stick to trustworthy information from cancer charities or the NHS. Your healthcare team will tell you the best place to find it

Wendy, diagnosed with blood cancer aged 46

Myeloproliferative neoplasms (MPN) at a glance

Myeloproliferative neoplasms (MPN) are a group of conditions that affect the blood. In MPN, your body produces too many of a particular type of blood cell.

MPNs are classed by the World Health Organisation as types of blood cancer. This means your care and treatment follows national cancer guidelines and you should have access to cancer support services.

Although MPN is classed as a cancer, many people with MPN feel quite well and the condition doesn't interfere too much with everyday life. Often, MPNs can remain stable or progress quite slowly.

There are around 520 cases of MPN in the UK per year.

There are three main types of MPN:

– polycythaemia vera (PV)
– essential thrombocythaemia (ET)
– myelofibrosis (MF).

The type of MPN you have depends on the type of blood cell your body is making too many of.

For detailed information about PV, see **page 19**.

For detailed information about ET, see **page 35**.

For detailed information about MF, see **page 49**.

Polycythaemia vera (PV) at a glance

What is PV?

If you've got polycythaemia vera (PV), too many red blood cells are made in your bone marrow (the soft material inside your bones).

Who gets PV?

Your chance of getting PV increases with age. PV is very rare in children and around 95% of people with PV are aged 40 years or older.

What's the outlook?

As PV is generally diagnosed in later life, there's a good chance that you'll live a normal lifespan, if your condition is carefully monitored and treated as necessary.

Although PV isn't a curable condition, many people with the disease will have a good quality of life.

What are the treatments for PV?

The treatment of PV may vary from person to person. You may not need treatment at first – especially if you don't have any symptoms.

The main aim of treatment for PV is to reduce your symptoms and risk of complications, such as thrombosis or blood clots, by reducing the number of red blood cells in your blood. If you have more severe symptoms, you may be treated with a mild form of chemotherapy.

Can PV lead to any other conditions?

In a small number of people with PV (less than 5%), PV can transform into a faster-growing cancer known as myelofibrosis, or into acute myeloid leukaemia. If this happens, your medical team will explain these conditions to you in more detail.

For more information about myelofibrosis, turn to **page 49**.

To find out more about acute myeloid leukaemia, order or download our booklet: **Acute myeloid leukaemia (AML)**. See **page 110**.

Essential thrombocythaemia (ET) at a glance

What is ET?

In essential thrombocythaemia (ET), too many platelets are made in your bone marrow (the soft material inside your bones). This is the type of blood cell involved in blood clotting.

Who gets ET?

Your chance of getting ET broadly increases with age, but younger people can get ET too. The condition is rare in children.

What's the outlook?

As ET is generally diagnosed in later life, there's a very good chance of living a normal lifespan if your condition is carefully monitored and treated as necessary.

Although ET isn't a curable condition, many people with the disease will have a good quality of life.

What are the treatments for ET?

Treatment for ET aims to reduce the risk of complications such as blood clots. Your doctor will check your condition with regular blood tests.

In some cases, you may have mild chemotherapy to reduce the number of platelets in your blood.

Can ET lead to any other conditions?

In a small number of people with ET (less than 5%) it can transform into a faster-growing cancer known as myelofibrosis, or into acute myeloid leukaemia.

If this happens, your medical team will explain these conditions to you in more detail.

For more information about myelofibrosis, turn to **page 49**.

To find out more about acute myeloid leukaemia, order or download our booklet: **Acute myeloid leukaemia (AML)**. See **page 110**.

Myelofibrosis (MF) at a glance

What is MF?

In myelofibrosis (MF), scar tissue forms in your bone marrow (the soft material inside your bones). As this builds up, your blood cells can no longer develop properly inside your bone marrow.

Who gets MF?

Your chance of getting MF increases with age. The condition is rare in children. Sometimes people with other blood cancers such as PV or ET can develop myelofibrosis; this is known as secondary MF.

What's the outlook?

MF can progress in different ways in different people. Some people have a mild form of MF and the condition doesn't progress rapidly.

In these cases, the condition doesn't interfere too much with everyday life.

In other cases, MF may progress more quickly. If this is the case, you may need regular blood transfusions or other medication.

What are the treatments for MF?

Treatment aims to control any symptoms you have. If you don't have any symptoms you may not need treatment for a while. In this case, your doctor will monitor your condition regularly.

If you do have symptoms, you may have what's known as active treatment. Active treatment for MF may include blood transfusions, mild chemotherapy or other medication. Some people with MF may have a stem cell transplant, but this is rare.

Can MF lead to any other conditions?

In around a quarter of people, MF may progress to acute myeloid leukaemia (AML). If this happens, your team will explain the condition to you in more detail.

To find out more about acute myeloid leukaemia, order or download our booklet: **Acute myeloid leukaemia (AML)**. See **page 110**.

Knowing the basics about blood, bone marrow and your immune system is useful.

If you're worried, get in touch on **0808 2080 888**
or email **support@bloodcancer.org.uk**

Blood, bone marrow and your immune system

It’s a good idea to know a bit about blood,bone marrow and your immune system, as your healthcare team will talk to you about them.

Blood

The blood has many important functions.

Transport system

It carries food, oxygen and proteins to different parts of your body. It also carries waste chemicals to the kidneys and lungs so they can get rid of them.

Defence system

White blood cells are part of your immune system, which fights infections.

Communication system
Organs in the body release hormones into the blood which send messages to other organs.

Repair system
It contains cells and chemicals which can seal off damaged blood vessels and control blood loss.

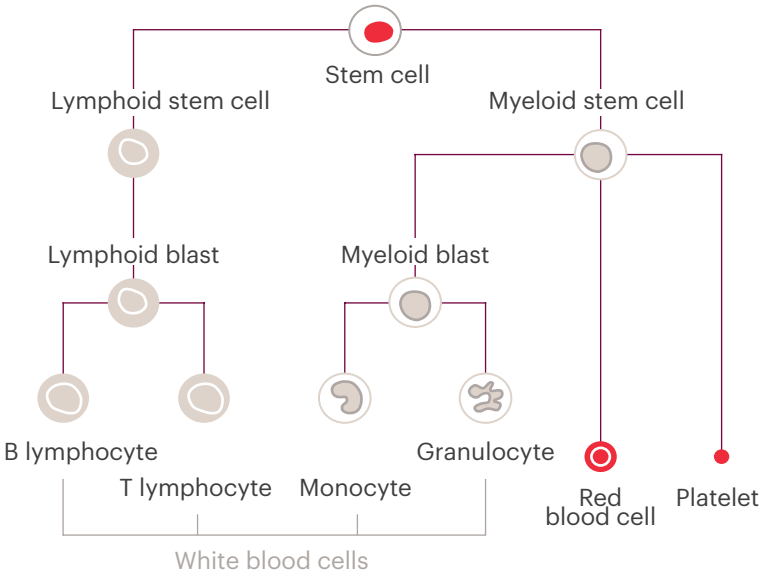
Blood cells
Blood contains three types of cells: red blood cells, white blood cells and platelets.

Red blood cells, platelets and some white blood cells (neutrophils, monocytes, eosinophils and basophils) are myeloid cells, made from myeloid stem cells. Other white blood cells, known as lymphocytes, are lymphoid cells, made from lymphoid stem cells.

Red blood cells (erythrocytes)
These contain a chemical called haemoglobin which carries oxygen to all the tissues of your body. Muscles and other tissues need oxygen to use the energy from your food.

White blood cells
These fight and prevent infection. There are five different types of white blood cell: lymphocytes, monocytes, eosinophils, neutrophils and basophils. (The final three are types of white blood cells called granulocytes).

Blood cell production



Platelets (thrombocytes)

These stick together at the site of any tissue damage and stop bleeding.

How many of each type of blood cell should you have?

Everyone has slightly different amounts of each type of blood cell depending on age, sex and ethnicity. If you're healthy, the amount you have of each normally stays the same, with slight changes up and down over time.

Here's a table which shows how many of each type of blood cell a healthy person has:

Blood cell or substance	Levels found in a healthy person
Haemoglobin (Hb) level (for red blood cells)	130–180 g/l (men) 115–165 g/l (women)
Platelets	150–400 x 10^9/l
White blood cells (WBC)	4.0–11.0 x 10^9/l
Neutrophils	2.0–7.5 x 10^9/l
Lymphocytes	1.5–4.5 x 10^9/l

Your blood values depend on a number of different things, including sex, age, and ethnicity. Also, different laboratories will use different equipment and testing methods, so normal values can vary slightly from hospital to hospital. So this table should only be used as a rough guide. Your healthcare team can explain what your results mean for you.

Bone marrow and stem cells

Blood cells all start off in the soft material inside your bones (bone marrow), from a type of cell called a stem cell. Like all cells, stem cells divide and eventually form mature, fully-formed blood cells.

A lot of blood cells are made in the bone marrow every second because your body needs them. If everything's working normally, your body makes the right number of each type of cell to keep you healthy.

The production of different blood cell types by the stem cells is controlled by particular genes. MPN usually develop because of a fault (mutation) in a particular gene.

Your immune system

Your immune system is a network of cells, tissues and organs which protect your body against infection. It's able to react quickly to infections it has seen before: white blood cells play an important role in this. They circulate around your body in your blood, and fight infections.

You can read more about the genes involved in each type of MPN on **pages 20**, **36** and **50**.

In PV, too many red blood cells are made in your bone marrow (the soft material inside your bones).

If you're worried, get in touch on **0808 2080 888**
or email **support@bloodcancer.org.uk**

Polycythaemia vera (PV)

For the most up-to-date information about PV and its treatment, visit **bloodcancer.org.uk**

PV is a rare blood cancer, with the majority of people diagnosed over 40 years old. PV generally develops slowly, and for most people doesn't affect their normal lifespan.

For more information on bone marrow and how blood cells are produced see **page 17**.

What is PV?

PV is a myeloproliferative neoplasm. If you have it, you will have too many red blood cells in your bone marrow.

As a result, your blood becomes thicker than normal. Some people may also have increased numbers of white cells and platelets.

What causes PV?

All cells in your body contain a set of instructions which tell the cell what to do and when to do it, stored inside the cells in structures called chromosomes. The chromosomes are made up of a chemical known as DNA.

The DNA is arranged in sections called genes. There are 23 pairs of chromosomes in each cell in your body. When cells divide to form new cells, normally the chromosomes stay the same in each new cell.

However with PV, something goes wrong and causes a genetic fault to occur: you may hear your doctor talk about a fault, or mutation, in the JAK2 gene. This may happen because you've been exposed to hazardous chemicals, but more usually because of copying mistake when a cell was dividing. Around 95% of people with PV have this genetic fault.

The JAK2 gene is involved in the response of bone marrow stem cells to different growth factors. A growth factor is a substance which sends signals to your stem cells, so they can produce the right number of blood cells to keep you healthy.

When you have a fault with your JAK2 gene, the stem cells can start producing red blood cells even when they've not be 'told' to do so by growth factors. This results in too many red cells being produced.

It's important to note that the genetic fault happens during a person's lifetime. As you're not born with it, you can't pass it onto your children.

Who gets PV?

As well as the presence of the faulty JAK2 gene, there are some other factors which mean you might be more at risk of getting PV.

Age

Your chance of getting PV increases with age. Around 95% of people diagnosed with PV are aged 40 years or older. The disease is rare in people under the age of 15 years.

Sex

PV is slightly more common in men than women; we're not sure why.

Symptoms

You may not have any symptoms before or when you're diagnosed. That's why many people with PV are diagnosed following a routine blood test.

The increase in red blood cells makes it hard for blood to flow smoothly through your blood vessels. This is known as hyperviscosity and may mean you get some of these symptoms:

- headaches
- confusion
- blurred vision
- skin reddening (plethora)
- itchy skin (pruritus).

A less common complication of PV is gout, which can cause inflammation of the joints.

Thrombosis

People with PV are more at risk of thrombosis (blood clots). Thrombosis is a serious condition and may occur in the blood vessels of your:

- brain (causing a stroke or mini-stroke/TIA [transient ischaemic attack])

- eyes (causing blurred vision or loss of vision)

- heart (causing a heart attack).

Blood clots can also form in the veins of your legs. This is known as deep vein thrombosis or DVT. Clots can also form in the vessels in your abdomen (stomach area). If a clot dislodges and travels to the lung, it may cause a pulmonary embolism (or PE). This usually results in low oxygen levels, sharp chest pain and shortness of breath. In some cases this may be fatal. The risk of clots is highest if the PV isn't treated.

You'll have regular blood tests so your healthcare team can monitor your condition and spot any early signs of a blood clot.

Symptoms of a blood clot

If you have any of these symptoms, you should get urgent medical attention.

- sudden chest pain or shortness of breath
- swelling and/or pain in your calf on one side
- slurred or abnormal speech, weakness in your arms or legs, or drooping on one side of your face
- swelling in your abdomen or jaundice (your skin turning a yellow colour)
- sudden loss of vision in one eye.

Diagnosis

Most people are suspected of having PV after a routine blood test or by going to their GP with symptoms. You'd then have a set of tests to confirm the exact diagnosis of PV.

Full blood count

Polycythaemia (rather than polycythaemia vera) is defined as a persistent increase in the proportion of your blood that's made up of red blood cells. Your doctor may refer to this as your 'packed cell volume' (or PCV). This is usually checked using a test called a full blood count.

For this test a small sample of your blood will be taken, then the cells will be studied under a microscope in a laboratory.

Bone marrow biopsy

Some people need tests on their bone marrow before their doctors can make a diagnosis. This helps to rule out any other bone marrow problems. During the test, a small amount of bone marrow is taken from the hip bone using a needle.

You don't need to stay overnight in hospital for this. You have the procedure as an outpatient with a local anaesthetic or mild sedation. It's usually quite quick but it will be uncomfortable while the sample is being taken.

You can take painkillers if you need to. Your doctors will then look at the bone marrow sample under a microscope to assess it for any disease.

The results of these tests alone aren't enough for your doctor to diagnose PV, as there are a number of other reasons and conditions that can mean you have too many red blood cells.

These include:

- smoking
- lung diseases
- sleep apnoea (where your normal breathing is disrupted while you're asleep)
- living at high altitudes
- some kinds of tumours
- anabolic steroids (prescription performance-enhancing drugs)
- testosterone treatment
- some rare inherited genetic disorders.

JAK2 blood test

The discovery of the JAK2 genetic fault in 2005 has made it easier to diagnose PV. Some of your DNA will be taken from a sample of your blood, and tested. If you have this genetic fault, doctors will be able to confirm you have PV.

Further tests

If there's no clear cause for your polycythaemia and you don't have the JAK2 genetic fault, your doctor will do more tests to confirm a diagnosis.
These may include:

- further blood tests for erythropoietin (a hormone that increases red blood cell production) levels and other genetic tests
- red cell mass studies (using radioisotopes) to distinguish 'apparent' polycythaemia from actual polycythaemia
- tests on blood samples taken from an artery (instead of vein) to measure your oxygen levels
- tests on samples of your bone marrow
- lung function tests
- scans to see if your spleen is swollen or other possible causes, for example a tumour which is releasing erythropoietin.

Your doctor will be able to talk to you about any of these tests and explain how they're done and what they're looking for. Be sure to ask them any questions about these tests if you're unsure.

Treatment

The aim of your treatment is to reduce the risk of getting thrombosis by reducing the number of red blood cells in your blood.

You should also be monitored for cardiovascular risk factors such as diabetes, high cholesterol, high blood pressure and smoking, and these should be addressed as effectively as possible.

Although currently PV can't be cured, it can be kept under control to reduce the symptoms and complications it may cause.

The treatment you receive for PV will depend on the following factors:

- how high the packed cell volume (PCV) is
- your age
- the type of blood cell most affected.

If you've been diagnosed with PV and don't have any symptoms, you may not need to start treatment for a while. While this might seem strange, there's no evidence to show that treating people with no symptoms has any impact on their outcome. It also means you don't get any side effects from unnecessary treatment.

If you do have symptoms, your treatment will include some or all of the following options.

Venesection

This is one of the simplest and quickest ways to reduce the number of red blood cells in your blood and make your blood thinner. This is also known as blood-letting or phlebotomy. It involves taking around a pint (half a litre) of blood from you. This may be done once a week initially and then repeated as often as needed.

You may feel faint after the blood is taken, so replacement fluid can be given at the same time to help with this.

You can read about the experiences of other people with blood cancer on our online community forum: **forum.bloodcancer.org.uk**

Low-dose aspirin

If you have PV and other risk factors such as previous clots, diabetes and high blood pressure, you may be at an increased risk of blood clots so your doctor may recommend you take low-dose aspirin regularly. If you need to take painkillers for any other reason at the same time, ask your doctor what you can safely take with the aspirin.

If you are already taking an anticoagulant such as warfarin, your doctor may decide you do not need aspirin as well.

Hydroxycarbamide

If your platelet count is high or you have other symptoms such as weight loss or sweats, you may be given tablets called hydroxycarbamide (or hydroxyurea) to take.

Hydroxycarbamide is a mild form of chemotherapy and works by directly preventing the production of red blood cells. Hydroxycarbamide is the most common chemotherapy drug used to treat PV.

You might get some side effects from this treatment. These might include more infections than normal, diarrhoea or constipation. Your healthcare team will be able to help you manage side effects like this.

Hydroxycarbamide is a very safe treatment. However, there is a theoretical risk that it may increase the risk of PV transforming into acute myeloid leukaemia if it is used as a long-term treatment. For many people, the benefits of the treatment usually outweigh any potential small risk.

Interferon

Interferon is another drug you may take if your platelet count is high or you have symptoms such as weight loss or sweating.

Interferon is an injection that slows down the production of blood cells. It is not thought to carry the same risk of leukaemia as hydrodroxycarbamide when used in the long-term, so it's often the preferred choice in younger patients. However, many people find the short-term side effects unpleasant and not all patients can tolerate interferon therapy.

Possible side effects you may get while being treated with interferon include:

- flu-like symptoms
- headaches
- dizziness
- mood swings
- tiredness.

Ruxolitinib

Ruxolitinib is a type of drug known as a JAK2 inhibitor. In Scotland, it is available for people with PV if hydroxycarbamide is not working for them. In the rest of the UK, it is not routinely available, but it may be a possibility for some patients.

Busulfan

Busulfan is another chemotherapy drug. It may be used when hydroxycarbamide isn't appropriate or isn't working. It's given as a tablet. Side effects can include lung tissue damage or reduced numbers of red blood cells, white blood cells or platelets in the blood. Busulfan can also increase the risk of leukaemia developing, so it's only used when doctors believe the benefits of treatment outweigh the risks.

Outlook

As PV is generally diagnosed in later life, there's a very good chance of having a normal lifespan and a good quality of life if the condition is carefully monitored and treated as needed.

Around 5% of people with PV go on to develop a more aggressive cancer. This may include progressing to myelofibrosis, where the bone marrow becomes scarred and less able to produce cells, or to acute myeloid leukaemia (AML).

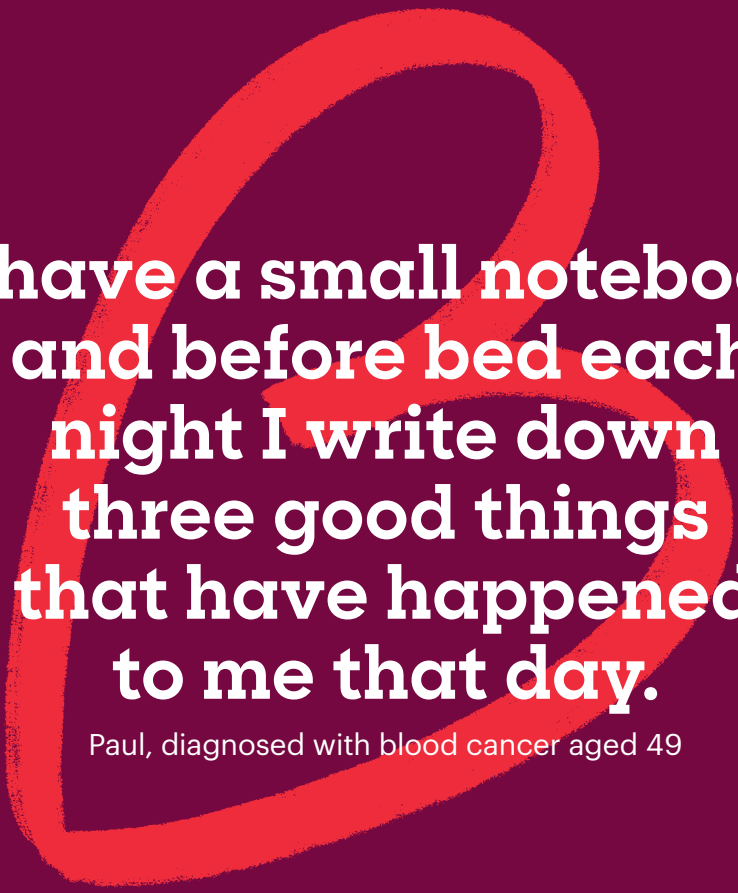
You may find it hard to ask or talk about your prognosis. Sometimes those close to you might want to know your prognosis even if you don't. However, your healthcare team aren't allowed to give this or any other information to anyone – not even family members – without your permission.

Try to decide early on who you want to know about your condition, then tell your healthcare team – you can change your mind at any time.

If you're worried about your outlook, contact our Support Services Team on **0808 2080 888** or **support@bloodcancer.org.uk**

For more information about myelofibrosis, turn to **page 49**.

To find out more about acute myeloid leukaemia, order or download our booklet: **Acute myeloid leukaemia (AML)**. See **page 110**.



I have a small notebook and before bed each night I write down three good things that have happened to me that day.

Paul, diagnosed with blood cancer aged 49

With essential thrombocythaemia, there are too many platelets produced in your bone marrow.

If you're worried, get in touch on **0808 2080 888**
or email **support@bloodcancer.org.uk**

Essential thrombocythaemia (ET)

For the most up-to-date information about ET and its treatment, visit **bloodcancer.org.uk**

Essential thrombocythaemia (ET) is most common in people over 50. It usually develops very slowly and for the majority of people it doesn't affect their normal lifespan.

For more information on bone marrow and how blood cells are produced see **page 17**.

What is ET?

ET occurs when the cells which produce your platelets in your bone marrow are abnormal. Because of this, you have too many platelets in the blood.

What causes ET?

All cells in your body contain a set of instructions which tell the cell what to do and when to do it, stored inside the cells in structures called chromosomes. The chromosomes are made up of a chemical known as DNA.

The DNA is arranged in sections called genes. There are 23 pairs of chromosomes in each cell in your body. When cells divide to form new cells, normally the chromosomes stay the same in each new cell.

However with ET, something goes wrong and causes a genetic fault to occur: you may hear your doctor talk about a fault, or mutation, in the JAK2, CALR or MPL genes. Genetic faults may happen because you've been exposed to hazardous chemicals, but more usually because of copying mistake when a cell was dividing. Around 60% of people with ET have the JAK2 genetic fault, 30% have the CALR fault and 5% have the MPL fault.

The JAK2 and MPL gene are involved in the response of bone marrow stem cells to different growth factors. A growth factor is a substance

which sends signals to your stem cells, so they can produce the right number of blood cells to keep you healthy. A growth factor is released when there are low levels of platelets in the blood.

When you have a fault with your JAK2 or MPL gene, the stem cells can start producing platelets even when they've not be 'told' to do so by the growth factor. This results in too many platelets being produced.

The CALR gene was discovered in 2013. We don't fully understand it yet, but we know it causes signals that lead to too many platelets being produced.

The presence of one of these genes in your blood might lead doctors diagnosing you with MPN. However we 're learning about these faulty genes all the time, and their impact on treatment options and outlook.

If you'd like to know more about those genes, your consultant will be happy to talk to you about them.

It's important to note that these genetic faults happen during a person's lifetime. As you're not born with these faults, you can't pass these on to your children.

Who gets ET?

People who get ET are usually between 50 and 70 years old. The condition is rare in children, but can occur at any age.

Symptoms

It's likely that you won't have any symptoms at all before or when you're diagnosed. That's why so many people with ET are diagnosed after a routine blood test. Older people and people with very high platelet counts get symptoms more often.

Here are some symptoms you may experience:

- persistent or repeated headaches
- disturbed vision (described by some people as light shows or silent migraines)
- dizziness or ringing in your ears
- bruising or bleeding easily (including heavy periods in women or nose bleeds)
- erythromelalgia (pain and redness in some or all of your hands, feet, arms, legs, ears and face)
- your fingers or toes being blue, or feeling cold.

Thrombosis

People with ET are at an increased risk of thrombosis (blood clots). Thrombosis is a serious condition and may occur in blood vessels in your:

- brain (causing a stroke or mini-stroke/TIA [transient ischaemic attack])
- eyes (causing blurred vision or loss of vision)
- heart (causing a heart attack).

Blood clots can also form in the veins of your legs. This is known as deep vein thrombosis or DVT. Clots can also form in the vessels in your abdomen (stomach area). If a clot dislodges and travels to the lung it may cause a pulmonary embolism (or PE). This usually results in low oxygen levels, sharp chest pain and shortness of breath. In some cases this may be fatal.

The risk of clots is higher in older people who also have other medical conditions such as diabetes or heart problems, or in people who have had clots in the past. However, the risk of thrombosis is reduced if your ET is treated appropriately.

You'll have regular blood tests so your healthcare team can monitor your condition and spot any early signs of a blood clot.

Symptoms of a blood clot

If you have any of these symptoms, you should get urgent medical attention.

- sudden chest pain or shortness of breath
- swelling and/or pain in your calf on one side
- slurred or abnormal speech, weakness in your arms or legs, or drooping on one side of your face
- swelling in your abdomen or jaundice (your skin turning a yellow colour)
- sudden loss of vision in one eye.

Diagnosis

Most people are suspected of having ET after a routine blood test or by going to their GP with symptoms.

You'd then have a set of tests to confirm the diagnosis of ET.

Full blood count

In ET there's an abnormally high level of platelets in the blood. A blood test known as a full blood count will detect if your platelet count is higher than normal.

For this test, a small sample of your blood will be taken, then the cells will be studied under a microscope in a laboratory.

Tests for genetic faults

DNA from one of your blood samples will be used to test for genetic faults to the JAK2, CALR and MPL genes. Around 60% of people with ET have a fault in the JAK2 gene. However, some people won't have one of these faults, so a diagnosis can't always be confirmed after these tests.

Bone marrow biopsy

Some people need tests on their bone marrow before their doctors can make a diagnosis. This helps to rule out any other bone marrow problems such as myelofibrosis (MF).

A small amount of bone marrow is taken using a needle from the hip bone. You don't need to stay overnight in hospital for this; you can have it as an outpatient using local anaesthetic or mild sedation. It's usually quite quick but will be uncomfortable while the sample's being taken from the marrow; you can take painkillers if you need to. Your doctors will then look at the bone marrow sample under a microscope to assess it and look for any disease which might be in it.

Other causes of high platelet counts

If tests show that you don't have any of the genetic faults linked with ET, your doctor will need to rule out other possible causes of a high platelet count before confirming a diagnosis. Other causes of a high platelet count can include:

- unusual bleeding
- iron deficiency
- infections
- inflammatory diseases such as arthritis
- other blood cancers such as polycythaemia vera (PV), primary myelofibrosis (PMF) and chronic myeloid leukaemia (CML)
- other types of cancer
- your spleen not functioning normally.

You can read about the experiences of other people with blood cancer on our online community forum **forum.bloodcancer.org.uk**

Treatment

The treatment you receive for ET depends in part on your risk of developing complications. Patients are generally divided into low, intermediate or high-risk categories.

This is based on a combination of the following factors:

- your age
- the symptoms you have
- your medical history (including risk factors for blood clots)
- your platelet count.

You should be monitored for cardiovascular risk factors such as diabetes, high cholesterol, high blood pressure and smoking, as these should be addressed as effectively as possible.

Aspirin

If you have a low risk of complications, you may be treated with aspirin. Aspirin can help prevent clots because it affects the way platelets stick together. This doesn't affect your platelet count, but helps reduce the risk of clots. If you need to take painkillers for any other reason, ask your doctor which ones are safe.

Chemotherapy

You may have mild chemotherapy to treat your ET. Whether you have chemotherapy depends on a number of things, including:

- your risk of thrombosis
- how well you'll be able to cope with the side effects
- your personal preference.

Hydroxycarbamide

You may be prescribed a tablet called hydroxycarbamide (or hydroxyurea). This is a mild form of chemotherapy and works by reducing the number of platelets in your blood. Hydroxycarbamide is the most common chemotherapy drug used to treat ET. It does have some side effects: you may get diarrhoea, constipation or get more infections than usual.

Hydroxycarbamide is a very safe treatment. However, there is a theoretical risk that it may increase the risk of ET transforming into acute myeloid leukaemia if it is used as a long-term treatment. For many patients, the benefits of the treatment usually outweigh any potential small risk.

Busulfan

Busulfan is another chemotherapy drug. It may be used when hydroxycarbamide isn't appropriate or isn't working. It's given as a tablet.

Side effects can include lung tissue damage or reduced numbers of red blood cells, white blood cells or platelets in the blood. Busulfan can also increase the risk of leukaemia developing, so it's only used when doctors believe the benefits of treatment outweigh the risks.

Interferon

Interferon is an injection that slows down the production of platelets. It is not thought to carry the same risk of leukaemia as hydrodroxycarbamide when used in the long-term, so it's often the preferred choice in younger patients. However, many people find the short-term side effects unpleasant and not all patients can tolerate interferon therapy.

Possible side effects you may get while being treated with interferon include:

- flu-like symptoms
- headaches
- dizziness
- mood swings
- tiredness.

Anagrelide

Anagrelide is usually given only when other treatments have already been tried. It's taken as a capsule but may increase the risk of developing myelofibrosis.

Outlook

As ET is generally diagnosed in later life, for most people there's a very good chance of living a normal lifespan if the condition is carefully monitored and treated. It's important to note that less than 5% of people with ET progress to a more aggressive disease such as myelofibrosis, where the bone marrow becomes scarred and less able to produce cells, or to acute myeloid leukaemia (AML).

You may find it hard to ask or talk about your prognosis. Sometimes those close to you might want to know your prognosis even if you don't. However, your healthcare team aren't allowed to give this or any other information to anyone – not even family members – without your permission. Try to decide early on who you want to know about your condition, then tell your healthcare team – you can change your mind any time

If you're worried about your outlook, contact our Support Services Team on **0808 2080 888** or **support@bloodcancer.org.uk**.

For more information about myelofibrosis, turn to **page 49**.

To find out more about acute myeloid leukaemia, order or download our booklet: **Acute myeloid leukaemia (AML)**. See **page 110**.

The Blood Cancer UK Online Community Forum means I never feel alone because there's always someone there to talk to, who really understands.

Carina, diagnosed with blood cancer aged 43

In myelofibrosis, scar tissue builds up inside the bone marrow and blood cells aren't able to develop properly.

If you're worried, get in touch on **0808 2080 888**
or email **support@bloodcancer.org.uk**

Myelofibrosis (MF)

For the most up-to-date information about MF and its treatment, visit **bloodcancer.org.uk**

Myelofibrosis (MF) is a rare type of blood cancer affecting the bone marrow. It is most common in people over 50.

What is MF?

In MF, your bone marrow is overactive and then develops scar tissue (known as fibrosis). The scar tissue builds up inside your bone marrow and blood cells can't develop properly.

When blood cell production is reduced in the bone marrow, it starts to take place in the liver and spleen instead. As the liver and spleen aren't as good at

producing blood cells, people with MF may develop anaemia (not enough red blood cells in your blood).

The spleen may also become enlarged, as it 'holds on' to red blood cells instead of releasing them into the blood.

People who have no history of problems with their bone marrow can get MF. This is known as primary myelofibrosis (PMF). Secondary MF is where the condition develops as a result of other blood cancers such as polycythaemia vera (PV) and essential thrombocythaemia (ET).

The information in this booklet is relevant for both primary and secondary MF.

What causes MF?

The underlying causes of MF are still not fully understood, but you may hear your doctor talking about a genetic fault in your JAK2, CALR or MPL gene which they think is involved in causing MF. Around 65% of people with MF will have the JAK2 gene. 25% of people will have the CALR gene and up to 8% will have the MPL one.

All cells in your body contain a set of instructions which tell the cell what to do and when to do it, stored inside the cells in structures called chromosomes. The chromosomes are made up of a chemical known as DNA.

DNA is arranged in sections called genes. There are 23 pairs of chromosomes in each cell in your body. When cells divide to form new cells, normally the chromosomes stay the same in each new cell.

A small genetic change to DNA can cause a genetic fault. These changes can be caused by exposure to hazardous chemicals or copying mistakes when a cell was dividing. When the JAK2, CALR or MPL gene becomes mutated, your bone marrow may not function correctly and scar tissue can build up in your bone marrow.

It's important to note that these genetic faults happen during a person's lifetime. As you're not born with these faults, you can't pass these onto your children.

The presence of one of these genes in your blood might lead doctors diagnosing you with MPN. However we're learning about these faulty genes all the time, and their impact on treatment options and outlook. If you'd like to know more, your consultant will be happy to talk to you about this.

For more information on bone marrow and how blood cells are produced see **page 17**. For more about PV, see **page 19**. For more about ET, see **page 35**.

Who gets MF?

People who get MF are usually between 50 and 70 years old. The condition is rare in children, but can occur at any age.

Previous diagnosis of an MPN

A previous diagnosis of PV or ET may increase the risk of getting MF, in which case it's known as secondary MF.

Symptoms

Of those diagnosed with MF, 80% of people showed symptoms of MF which led them to go to their doctor. In the remaining 20%, MF was usually picked up by chance, after a routine blood test.

The symptoms you get when you have MF are because not enough blood cells are being made in your bone marrow:

- a lack of red blood cells may lead to breathlessness, fatigue, chest pains, headaches and tinnitus

- a lack of platelets might mean you bruise easily or have unusual bleeding

- a lack of white blood cells might mean you get more infections than normal.

You may also have pain in your abdomen due to the enlargement of your liver and spleen.

Other symptoms may include night sweats, fevers and weight loss.

Thrombosis

People with MF are at an increased risk of thrombosis (blood clots). Thrombosis is a serious condition and may occur in the blood vessels of your:

- brain (causing a stroke or mini-stroke/TIA [transient ischaemic attack])
- eyes (causing blurred vision or loss of vision)
- heart (causing a heart attack).

Blood clots can also form in the veins of your legs. This is known as deep vein thrombosis or DVT. Clots can also form in the vessels in your abdomen (stomach area). If a clot dislodges and travels to the lung, it may cause a pulmonary embolism (or PE). This usually results in low oxygen levels, sharp chest pain and shortness of breath. In some cases this may be fatal.

You'll have regular blood tests so your healthcare team can monitor your condition and spot any early signs of a blood clot.

Symptoms of a blood clot

If you have any of these symptoms, you should get urgent medical attention.

- sudden chest pain or shortness of breath
- swelling and/or pain in your calf on one side
- slurred or abnormal speech, weakness in your arms or legs, or drooping on one side of your face
- swelling in your abdomen or jaundice (your skin turning a yellow colour)
- sudden loss of vision in one eye.

Diagnosis

Most people are diagnosed withMF through a routine blood test or following a visit to their GP with symptoms. You would then have a set of tests to confirm the exact diagnosis of MF.

Full blood count

If your doctor thinks you might have MF, you'll firstly have a simple blood test to measure the numbers of the various types of blood cells. This is called a full blood count. Your blood cells will also be examined under a microscope in a laboratory: red blood cells with a distinctive, abnormal shape can suggest to your doctor that you might have MF.

Your doctor will also examine you to check whether you have an enlarged spleen.

Tests for genetic faults

DNA from one of your blood samples will be used to test for genetic faults to the JAK2, CALR and MPL genes. Around 50% of people with MF have a fault in the JAK2 gene and 30% have a fault in the CALR gene. However, some people won't have one of these faults, so a diagnosis can't always be confirmed after these tests.

Bone marrow biopsy

Some people need tests on their bone marrow before their doctors can make a diagnosis. This helps to rule out any other bone marrow problems and lets doctors assess your fibrous tissue.

During the test, a small amount of bone marrow is taken using a needle from the hip bone. You don't need to stay overnight in hospital for this; you can have it as an outpatient using local anaesthetic or mild sedation. It's usually quite quick but will be uncomfortable while the sample's being taken from the marrow. You can take painkillers if you need to.

You can read about the experiences of other people with blood cancer on our online community forum: **forum.bloodcancer.org.uk**

Your doctors will look at the bone marrow sample under a microscope to assess it for any disease which might be there.

Bone marrow trephine

As someone with MF will have a large amount of fibrous tissue in their bone marrow, it may be hard to get a sample using a needle and syringe – this is known as a 'dry tap'. This means you may need a trephine biopsy.

This is where a 'core' of bone marrow from the hip bone is taken, under local anaesthetic or mild sedation. It provides information about the structure of your bone marrow and the number and distribution of the different blood cell types – and cancer cells, if present.

How MF develops

Myelofibrosis is a chronic (long-term) and progressive condition. Once a diagnosis has been confirmed, your doctor will do tests to find out how much the MF has progressed.

In the earlier stages of the disease, the bone marrow can be overactive and produces too many blood cells. Usually as the condition develops, your bone marrow becomes more and more scarred (fibrosis) and less able to produce normal red blood cells, white blood cells or platelets.

This begins to cause symptoms such as anaemia, fatigue and often an enlarged spleen.

As myelofibrosis is usually not curable, treatment is mainly focussed on relieving these symptoms. Your healthcare team will monitor your blood counts and symptoms for signs of progression, to make sure you are getting all the treatments you need.

In advanced stages of myelofibrosis, the bone marrow scarring worsens, and you may need to have more blood transfusions. Treatments may eventually become less effective. In some people, myelofibrosis can progress to acute myeloid leukaemia.

Some people remain in the early stages of myelofibrosis for some time and may not need treatment, or can manage well with regular blood transfusions or drug treatments. However, for many people the condition will eventually progress over time.

To find out more about acute myeloid leukaemia, order or download our booklet: **Acute myeloid leukaemia (AML)**. See **page 110**.

Treatment

The treatment you have will depend on a variety of factors, including:

- your blood counts
- the symptoms you have
- your overall fitness.

You should be monitored for cardiovascular risk factors such as diabetes, high cholesterol, high blood pressure and smoking, as these should be addressed as effectively as possible.

The main aim of treatment is to control any symptoms you have. If you've been diagnosed with MF and don't have any symptoms, you may not need to start treatment for a while. While this might seem strange, there's no evidence to show that treating people with no symptoms has any impact on their outcome. It also means you don't get any side effects from unnecessary treatment.

Blood transfusion

If you have severe anaemia you'll need regular blood transfusions, usually every one to three months.

These can be carried out during a single day and you would need to stay in hospital overnight. You might also have platelet transfusions if you have low platelets and you're having unusual bleeding or bruising.

Ruxolitinib

Ruxolitinib is a type of drug known as a JAK2 inhibitor. It is recommended for people with an enlarged spleen or troublesome symptoms of myelofibrosis. It can reduce the size of your spleen, improve symptoms and improve your overall outlook. It is taken as tablets, twice a day, for as long as it is working.

Possible side effects include anaemia, a low platelet count, a low white blood cell count and increased risk of infections. Your doctors will monitor your response, and may need to change the dose or stop ruxolitinib if it's not working well for you.

Other treatments for enlarged spleen

An enlarged spleen may cause you problems by becoming painful. It may also cause anaemia.

Radiation

To treat an enlarged spleen, you may receive local radiation. This would usually reduce the size of your spleen from anywhere between a few months to a couple of years.

Splenectomy

Another option is have an operation to remove your spleen (a splenectomy). There can be complications after a splenectomy: your doctor will discuss the pros and cons with you to help you decide if the procedure is right for you.

Hydroxycarbamide

If your platelet count is high or you have other symptoms such as weight loss or sweats, you may be given tablets called hydroxycarbamide (or hydroxyurea) to take. This is a mild form of chemotherapy and works by directly preventing the production of red blood cells. Hydroxycarbamide is the most common chemotherapy drug used to treat MF.

You might get some side effects from this treatment. These might include more infections than normal, diarrhoea or constipation. Your healthcare team will be able to help you manage side effects like this.

Hydroxycarbamide is a very safe treatment. However, there is a theoretical risk that it may increase the risk of myelofibrosis transforming into acute myeloid leukaemia if it is used as a long-term treatment. For many patients, the benefits of the treatment usually outweigh any potential small risk.

JAK2 inhibitors

Drugs called JAK2 inhibitors are now available to treat MF. These can reduce the size of your spleen, improve symptoms and your overall outlook. However, JAK inhibitors may worsen anaemia.

The length of time you take the drug for depends on how well it works and any side effects you get from it. Your doctor can discuss if these medications would be appropriate treatment for you.

Thalidomide

Thalidomide is a targeted therapy to treat MF. Thalidomide can cause birth defects, so it shouldn't be given to pregnant women. People taking thalidomide who are sexually active should use a barrier form of contraception as some hormonal methods of birth control (such as the pill) can be made less effective by thalidomide.

You may also get side effects from this treatment such as feeling tired or drowsy, constipation and numbness or tingling in your hands and feet.

The length of time you take the drug for depends on how well it works and any side effects you get from it. The use of thalidomide is becoming less common in the UK.

Danazol

You may be given danazol to help improve anaemia (low number of red blood cells). The length of time you take the drug for depends on how well it works and any side effects you get from it.

Donor stem cell transplant

A stem cell transplant is what used to be called a bone marrow transplant. It aims to give you healthy stem cells, which then produce normal blood cells.

It isn't a suitable treatment for most people with MF due to the risks associated with the procedure. For a minority of people – especially those whose disease is progressing more quickly – a transplant may provide a cure, but there isn't enough evidence to be sure of this yet.

There are two main types of stem cell transplant:

- Autologous or autograft – this uses your own stem cells

- Allogeneic or allograft – this uses donor stem cells and is a high-risk procedure.

You can find out more about stem cell transplants in our booklet: **Blood stem cell and bone marrow transplants - the seven steps**. See **page 110** for how to order or download a copy.

Outlook

The outlook for people with MF can vary. Some people may have a mild form of MF that doesn't progress rapidly. For others, MF progresses more quickly and requires regular blood transfusions or drug treatments.

Around 20% of people with MF go on to develop acute myeloid leukaemia. Sometimes it's possible to treat MF with a stem cell transplant.

Although it's impossible to say what will happen to an individual person, your healthcare team will think about prognosis to help them make the best treatment decisions with you. They will consider various factors such as:

- your age
- levels of blood cells in the blood
- severity of symptoms
- the particular genetic mutations you have.

All of these things can have an impact on your prognosis.

You may find it hard to ask or talk about your prognosis. Sometimes those close to you might want to know your prognosis even if you don't. However, your healthcare team aren't allowed to give this or any other information to anyone – not even family members – without your permission.

Try to decide early on who you want to know about your condition, then tell your healthcare team – you can change your mind any time.

Remember that your outlook might change, for example if you respond well to treatment. If there's a change in your condition, or if you've finished all or part of your treatment, you might want to consider asking if your prognosis is still the same.

If you're worried about your outlook, contact our Support Services Team on **0808 2080 888** or **support@bloodcancer.org.uk**.

I learned to live day by day. If I felt panicked by something, I would try to concentrate on the next 24 hours.

Louise, diagnosed with blood cancer aged 51

Everyone is different, so listening to the advice of your specialist and your healthcare team is really important.

If you're worried, get in touch on **0808 2080 888** or email **support@bloodcancer.org.uk**

Your healthcare team

If you're diagnosed with an MPN, your hospital should give you the names and contact details of your consultant, clinical nurse specialist and other members of your healthcare team.

There's space to write them in the back of this booklet if you want to. You can then use these details to contact your team if you have any questions you want to ask when you're not in the hospital.

Your consultant

Because MPN are so rare, you shouldn't be surprised if your GP hasn't seen a previous case. Your GP will refer you to a consultant. It's likely you'll be treated by a haematologist – a doctor who specialises in treating people with blood diseases. Your consultant will be an expert in treating your specific disease.

Your clinical nurse specialist

People with blood cancers are usually assigned a clinical nurse specialist (CNS). They are your key point of contact with the rest of your healthcare team. You may like to have a meeting with your clinical nurse specialist when you're first diagnosed, to discuss your condition.

It's likely that your clinical nurse specialist will become important to you and your family, as they'll be with you right through your treatment.

Your multidisciplinary team

When you're diagnosed with an MPN, your condition is discussed at a multidisciplinary team (MDT) meeting. An MDT brings together doctors, nurses and any other specialist staff who'll be looking after you. A senior consultant usually leads the meetings, which are held regularly. They'll discuss the best treatment for you and every aspect of your care, including any changes in your condition.

Talking to other people

You might like to ask your consultant or key worker if they can recommend someone you can talk to who's had the same diagnosis and treatment as you. If you do this, remember that someone else's experience won't always be the same as yours. For example, some people have side effects from a drug and others don't.

You may also want to contact a support organisation – many provide meetings or further online support.

Your other healthcare professionals

It's definitely worth telling other healthcare professionals you see – like your dentist or optician – about your diagnosis and any medication you're taking.

You can read about the experiences of other people with blood cancer on our online community forum: **forum.bloodcancer.org.uk**

It's important to know and understand your diagnosis. You can ask your team to write this in a booklet, so you have it to hand.

If you're worried, get in touch on **0808 2080 888**
or email **support@bloodcancer.org.uk**

Finding out more

After you've been diagnosed, it's worth taking some time to think about what information you want to know, when and how. For some people, this is a way to have some control over what's happening.

Let your consultant and clinical nurse specialist know how much information you'd like, and in what form. You can always ask for more information later.

Write down any questions you have and keep them handy for when you see your consultant or key worker. If they can't answer your questions, they'll be able to tell you who to speak to.

You might prefer to ask your clinical nurse specialist questions rather than your consultant, but do whatever works for you.

Most people say they find it useful taking someone with them to consultations. If you'd find it helpful, you could ask them to take notes while you listen. You can choose who to take; it doesn't have to be a family member.

If you're staying in hospital it might be harder to have someone with you when you speak to your consultant. It might be useful to ask in advance what time the consultant is likely to speak to you, so you can try to arrange for someone to be with you at that time.

Some people find that joining a support group is helpful. It may be easier to talk to someone outside of your family about your situation and being able to share similar experiences might also help you.

You can find a list of questions you might want to ask on **page 100**.

A large part of my job is listening and talking to patients about their fears and other emotions. It's as important as the clinical care.

Darren, Clinical Nurse Specialist

You may only want to tell those closest to you about your diagnosis. It is your choice and completely up to you.

If you're worried, get in touch on **0808 2080 888**
or email **support@bloodcancer.org.uk**

Telling people

Many people tell us that keeping in touch with loved ones throughout their illness keeps them going. However, some people may find it stressful having to discuss their condition lots of times with family, friends and colleagues.

You might find it easier to ask a trusted family member or friend to be your ‘information person’ and ask them to keep people updated on your behalf. Another idea is setting up a blog or Facebook page, so you or different people can post information on it that everyone can read.

Talking to children and teenagers

Talking to children and teenagers about your condition can be a difficult thing to do. There are many agencies available to support you and offer you advice about how to explain it to children of different ages. You might not want to tell many people – or anyone at all – about your condition. This is ok too, whatever works for you.

Telling your GP

Your team at the hospital will keep your GP informed about your condition and any treatment you're having. They'll usually send your GP a letter with this information.

As the patient, you'll often be sent a copy too. These letters can have a lot of medical terms in them which you might not have heard before, or there might be something in it which worries you. If this is the case, let your hospital or GP know – a quick chat with them might help to reassure you.

Telling your work

Consider telling someone at work about your diagnosis. It can be hard asking for time off at short notice if no one knows about your illness, and your colleagues and human resources department might be able to offer support.

For support with telling other people about your diagnosis, contact our Support Services Team on **0808 2080 888** or **support@bloodcancer.org.uk**

We have more information and personal stories about cancer and work on our website: **bloodcancer.org.uk/living-well**

Your healthcare team should look after your emotional needs as well as your physical ones.

If you're worried, get in touch on **0808 2080 888**
or email **support@bloodcancer.org.uk**

Everyday life and MPN

If you've been diagnosed with MPN you might experience a range of emotions at different times. There can be physical impacts on your day-to-day life too. This section will guide you through both aspects.

Looking after yourself emotionally

Being told that you have an MPN can be very upsetting and will almost certainly bring many different emotions. If you were diagnosed by chance, it can come as even more of a shock. Friends and family often offer a great deal of support, but it can be harder for them to understand the long term emotional impact that you might experience.

Your healthcare team look at your emotional, as well as physical, needs – this is called a holistic needs assessment. You'll have one a few times throughout the course of your treatment and beyond, as your emotional needs might change.

Looking after yourself physically

You might need to live with symptoms for a long time – your healthcare team will be able to give you advice on how to cope with them.

Keeping active

You might feel tired a lot (fatigue). This might be caused by your condition and isn't the same as normal tiredness which improves with rest and sleep.

While even the idea of doing something can be tiring if you've got fatigue, try to keep as active as you can because evidence shows that this could help to make your symptoms less severe.

We have information and personal stories about coping with your emotions and keeping active on our website: **bloodcancer.org.uk/living-well**

Although staying active may help, there's no evidence of any particular exercise programme improving your condition or how you respond to treatment.

Diet

Similarly, there's no evidence that any special diet will improve your condition or how you respond to treatment. However, you're likely to feel fitter and healthier if you follow general advice on good diet from your hospital or GP.

Some treatments can mean your immune system may not be working as normal, in which case you'll need to take extra care to avoid infections that you might get from food. Your body won't be able to destroy germs and resist infection as easily, so be careful about food 'use by' dates and things like keeping cooked and raw meat separate in the fridge.

You may hear healthcare professionals talk about a 'neutropenic diet'. This means a diet for people with a weakened immune system.

Some patients, particularly those with MF who have an enlarged spleen, may have difficulty eating a lot. This is because of your spleen pressing against your stomach. Your healthcare team can advise you on a diet to help you put on weight.

Complementary therapies

Complementary therapies are treatments like massage, meditation or acupuncture that are used alongside standard medical treatments with the aim of making you feel better.

There's no evidence to suggest that these therapies can treat or cure blood cancer, but there's some that suggests some of them may help you manage your symptoms or the side effects of your treatment. Other therapies may just help you relax or improve your general sense of wellbeing.

Alternative therapies

There's an important difference between complementary therapies, which are used alongside standard medical treatments (like chemotherapy and radiotherapy), and alternative therapies, which are offered instead of these treatments.

We don't recommend that you use any alternative therapy in place of proven medical care, but you may be interested in using complementary therapies alongside your treatment.

Keeping yourself safe

If you're thinking about using complementary therapies, you should let your healthcare team know, so you can discuss what's safe for you. They may advise you to avoid certain therapies because of specific risks to do with your condition or the

treatments you're receiving. In other cases, they may say a therapy is OK as long as you take specific precautions, like visiting a complementary therapist who's a member of the relevant professional association or register. Your healthcare team can explain how to check this.

Some hospitals will have a complementary therapies team that offers sessions free of charge, while others might have a specialist who visits once or twice a week. Sometimes these therapies are there for your partner or close relatives, too. Your healthcare team will be able to tell you what's on offer.

If your hospital doesn't offer complementary therapies, there may be a local cancer centre or charity that you could visit instead. Speak to your healthcare team to see if they can recommend anywhere nearby.

Some people choose to see an independent complementary therapist. If you do this, it's important to make sure they will keep you safe. Speak to your healthcare team about what you need to keep in mind when finding a therapist.

Practical support

Work, education and home life

If you work or are studying you might want to talk to your employer or college about your condition, or ask someone to do it for you. Most employers will do all they can to help.

You might need to make a short term arrangement with your employer or college at the time when you're diagnosed, so you can have time off when you need to be at the hospital. If you have to stay in hospital for your treatment, or you're not well enough to go to work or college, you'll probably need to make a more formal agreement.

You might need to bring in written proof of your diagnosis from your healthcare team, which makes clear the effect that your condition could have on your ability to work or study.

If you're a parent or a carer, you may need support during your treatment. You might have unplanned stays in hospital because of infection, for example – it's helpful to have plans in place just in case.

Cancer and the law

People with a disability are protected by the Equality Act 2010 in England, Scotland and Wales, and the Disability Discrimination Act 1995 in Northern

Ireland. For the purposes of these Acts, cancer is considered a disability.

This means that employers and places of study are required by law to makereasonable adjustments for people with cancer and can't discriminate against them. An example of a reasonable adjustment would be allowing you time off to go to hospital for treatment.

Getting to hospital

If you're being treated as an outpatient (not staying in overnight) you might need to be visiting the hospital a lot over a long period of time.

If you find this hard because of transport or any other reason, you can ask your consultant if you can have any of your treatment nearer to where you live. It might not always be possible but sometimes it is – it depends on the healthcare facilities close to your home and the type of treatment you're having.

If this isn't possible and transport is a problem, you can ask about hospital transport. You might also be able to claim a refund from the hospital for what it costs you to travel to your appointments. If you'd like to find out more about this support, you can speak to your team at the hospital or a benefits advisor.

Financial support

Your finances might be the last thing on your mind if you've just been diagnosed with a blood cancer, but there are lots of places you can get help and advice.

Your hospital will normally have medical social workers or welfare rights (benefits) advisors who can advise on which benefits you might be able to receive. These might be especially useful if you're on a low income or unemployed.

If you're worried you can ask to speak with an advisor as soon as possible after your diagnosis. Alternatively, your hospital might be able to arrange for an advisor from somewhere else to visit you.

If you normally pay for your prescriptions but are being treated for cancer (including the effects of cancer or the treatment) you can apply for a medical exemption certificate for any drugs you need for these reasons. Application forms are available from your GP surgery or hospital clinic.

For information about help with travel and other costs relating to your treatment, go to **citizensadvice.org.uk** or **macmillan.org.uk** and search 'help with health costs'.

My colleague took an information booklet into work, so people could understand what I was going through.

Hinna, diagnosed with blood cancer aged 23

The treatment you decide on with your healthcare team will depend on your health, your individual condition and your wishes.

If you're worried, get in touch on **0808 2080 888**
or email **support@bloodcancer.org.uk**

Clinical trials

If there's a clinical trial (study) available, your consultant might recommend that you consider taking part.

Clinical trials are done for several reasons, including to look for new treatment options and to improve existing treatments.

Taking part in a clinical trial has many advantages, such as the opportunity to have the newest available treatment which may not be offered outside of the trial. You'll also be very closely monitored and have detailed follow-up.

In a clinical trial, the best current treatment available is compared to one that could be better – so as a patient you'll either have the best available current treatment, or the new one which could be better. Your treatment in a trial won't differ too much from the best current treatment, and your safety and wellbeing is always the first priority.

Taking part in a clinical trial does come with uncertainties, and you may prefer not to take part in one. If you don't want to be in a trial, or there isn't a suitable trial available, you'll be offered the best treatment available at that time which is suitable for your individual condition.

Blood Cancer UK offers information and support to anyone affected by blood cancer. You'll find other useful organisations listed here as well.

If you're worried, get in touch on **0808 2080 888**
or email **support@bloodcancer.org.uk**

Places you can get help and support

Blood Cancer UK

We are here for anyone affected by blood cancer, whether it's you who's been diagnosed or someone you know.

We offer free and confidential support by phone or email. We provide information about blood cancer and life after a diagnosis. And we have an online forum where you can talk to others affected by blood cancer.

- bloodcancer.org.uk
- 0808 2080 888 (Mon–Fri 10am–7pm, Sat–Sun 10am–1pm)
- support@bloodcancer.org.uk
- forum.bloodcancer.org.uk

General information and support

Macmillan Cancer Support

Offers practical, medical, financial and emotional support.

- 0808 808 0000
- macmillan.org.uk

Cancer Research UK

Offers information about different conditions, current research and practical support.

- 0808 800 4040
- cancerresearchuk.org

MPN Voice

A specialist charity for people with MPN. They offer newsletters and patient events.

- mpnvoice.org.uk

Leukaemia Care

Offers information, a support line and support groups for people affected by leukaemia, lymphoma, myeloma and other blood disorders.

- 08088 010 444
- support@leukaemiacare.org.uk
- leukaemiacare.org.uk

African Caribbean Leukaemia Trust (ACLT)

Aims to increase the number of black, mixed race and ethnic minority people on UK stem cell registries by raising awareness and running donor recruitment drives.

- 020 3757 7700
- info@aclt.org
- aclt.org

Anthony Nolan

Runs the UK's largest stem cell register, matching donors to people with leukaemia and other blood-related disorders who need a stem cell transplant.

- 0303 303 0303
- anthonynolan.org

Maggie's

Has centres across the UK, run by specialist staff who provide information, benefits advice and psychological support.

- 0300 123 1801
- enquiries@maggies.org
- maggies.org

Marie Curie

Runs nine hospices throughout the UK and offers end-of-life support to patients in their own homes, free of charge.

- 0800 090 2309
- mariecurie.org.uk

MedicAlert

Offers personalised jewellery which provides vital medical information to emergency professionals.

- 01908 951045
- info@medicalert.org.uk
- medicalert.org.uk

Tenovus (Wales)

Provides an information service on all aspects of cancer, plus practical and emotional support for people with cancer and their families living in Wales.

- 0808 808 1010
- tenovuscancercare.org.uk

Financial advice

Citizens Advice

Offers advice on benefits and help with filling out benefits forms.

- 03444 111 444 (England), 03444 77 20 20 (Wales)
- in Scotland or Northern Ireland, contact your local Citizens Advice
- citizensadvice.org.uk

Department for Work & Pensions (DWP)

Responsible for social security benefits. Provides information and advice about financial support, rights and employment.

- gov.uk/government/organisations/department-for-work-pensions

Travel insurance

Macmillan Cancer Support

Provides information about what to consider when looking for travel insurance, along with recommendations from the Macmillan online community.

- 0808 808 0000
- macmillan.org.uk

British Insurance Broker's Association (BIBA)

Offers advice on finding an appropriate BIBA-registered insurance broker.

- 0370 950 1790
- enquiries@biba.org.uk
- biba.org.uk

Listen to your body and take time out when you need it. Don't set a time limit on when you should feel better.

Nichola, diagnosed with blood cancer aged 41

Questions for your healthcare team

It can be a good idea to write down any questions you have before your next appointment. Here are some things you might want to ask while you're waiting to receive your diagnosis, or once you've been diagnosed.

Tests

What tests will I have?

What will these tests show?

Where will I have the tests done?

Are there any risks associated with the tests?

Will any of the tests be painful?

Do I need to know anything about preparing for the tests, for example not eating beforehand?

How long will it take to get the results?

Treatment - general

Will I need to have treatment?

What does the treatment do?

Is there a choice of treatments?

Is there a clinical trial that I could join?

What's likely to happen if I decide not to have the treatment my healthcare team recommended?

If I don't need to start treatment straight away, how will I know when I need to start it?

Who do I contact if I take a turn for the worse?

Type of treatment

What type of drugs will I have?

Will I have to stay in hospital?

If I don't stay in hospital, how often will I need to go to hospital as an outpatient?

What chemotherapy regimen will I be given?

Will I be given it by mouth, injection or drip (into a vein)?

Will my treatment be continuous or in blocks of treatment (with a break in between)?

How long will my treatment last?

What side effects could I get from my treatment?

Can side effects be treated or prevented?

Will side effects affect me all the time or only while I'm taking certain drugs?

What effect is the treatment likely to have on my daily life?

Will I be able to carry on working/studying?

Will I need to take special precautions, for example against infection?

Will I need to change my meal times or work my drugs around these?

Stem cell transplant

Is a transplant an option for me?

How long will I be in hospital for?

Do I have to be in isolation?

How long will it be before I get back to normal?

Choosing the right treatment for you

If you're asked to choose between treatments, you might like to ask your consultant these questions about each one:

What's the best outcome I can hope for?

How might the treatment affect my quality of life?

Glossary

Anaemia
A condition where you either have fewer red blood cells than normal, or you have less haemoglobin (a protein in your red blood cells which carries oxygen) than normal. In either case, this means less oxygen is carried around in your bloodstream.

Blood count, full blood count or FBC
A blood test that counts the different types of cells in your blood.

Bone marrow
A spongy material inside long bones, which produces your blood cells.

Central nervous system (CNS)
The system of nerve tissues which control your body's responses. The CNS includes the brain and the spinal cord. Your peripheral nervous system is related to your CNS; this includes all of the nerves outside of your brain and spinal cord that carry messages to the CNS.

Chemotherapy

Treatment using anti-cancer drugs; it can be a single drug or a combination of drugs. Chemotherapy is used to kill cells or stop them growing and dividing. Although it's aimed at the cancer cells, the treatment also affects normal cells which divide quickly, such as those in your hair and your gut.

Clinical nurse specialist

A qualified nurse who specialises in a particular clinical area. Some deal with all blood cancers while others may specialise in leukaemia, myeloma, lymphoma or another specific area. Your nurse specialist can provide information and expert advice about your condition and treatment.

Clinical trial

A planned medical research study involving patients. They can be small trials involving only a few patients or large national trials. Clinical trials are always aimed at improving treatments and reducing any side effects they cause. You'll always be told if your treatment is part of a trial.

Chromosome

Structures in all of your cells which carry your genetic information and contain your genes.

Cytogenetics

The study of the structure of chromosomes. Cytogenetic tests (or FISH tests) are carried out on samples of blood and bone marrow taken from leukaemia patients. They aim to find any changes which could be linked to the disease. They can also help doctors to decide on the treatment you'll have.

Fatigue

A feeling of extreme tiredness which doesn't go away after rest or sleep. It might be caused by the AML itself or might be a side effect of treatment. It's one of the most common problems for people with cancer.

Fitness for treatment

Factors which determine how well your body will be able to cope with different treatments. These include things like age, medical fitness and any other conditions you might have.

Leukaemia

A type of blood cancer that's divided into many different types – some which develop faster (acute), and others which develop more slowly (chronic). People with leukaemia have large numbers of abnormal blood cells, usually types of white blood cell, which take over the bone marrow and often spill out into the blood stream.

Other areas that may also be affected are lymph nodes, spleen, liver, testes, the membranes surrounding the brain and spinal cord (meninges), gums and skin.

Medical fitness

How healthy your vital organs are before treatment. This refers to organs such as the heart, lungs, kidney and liver. Age, along with other factors, can affect this.

Myeloid cells

The term for the group of cells including red blood cells, platelets and all white blood cells except lymphocytes.

Progenitor cell

An immature cell in the bone marrow that produces mature blood cells. It can only divide a certain number of times, whereas a stem cell can divide again and again.

Radiotherapy

The use of radiation in treatment. Radiotherapy kills cancer cells in the area of the body being treated.

Secondary AML

AML which has developed as a result of you having another condition which affects your bone marrow.

Stem cells

Cells that are able to develop into other cell types. Stem cells act as a repair system for your body and replenish other cells. They're found in embryos and some organs in adults.

Treatment-related AML (tAML)

AML which develops after you've been treated for another type of disease. It can be caused by certain types of chemotherapy or radiotherapy.

Our health information

Find out more about blood cancer, its treatments and living with blood cancer. All our information is produced with expert medical professionals and people affected by blood cancer. It's available to anyone to download or order for free.

Symptoms guide

A credit-card sized guide that folds out to explain the symptoms of blood cancer.

Booklets

Leukaemia

- Acute lymphoblastic leukaemia (ALL)
- Acute myeloid leukaemia (AML)
- Acute promyelocytic leukaemia (APL)
- Chronic lymphocytic leukaemia (CLL)
- Chronic myeloid leukaemia (CML)

Lymphoma

- Hodgkin lymphoma (HL)
- High-grade non-Hodgkin lymphoma (NHL)
- Low-grade non-Hodgkin lymphoma (NHL)

Other blood cancers

- Myeloma
- Myelodysplastic syndromes (MDS)
- Myeloproliferative neoplasms (MPN)

Treatment and beyond

- Blood stem cell and bone marrow transplants: the seven steps
- Eating well with neutropenia
- Diary for anyone affected by blood cancer

My information folder

An A5 folder to keep your information in.

Fact sheets

Conditions

- Burkitt lymphoma
- Monoclonal gammopathy of undetermined significance (MGUS)
- Waldenström macroglobulinaemia (WM)

Treatments

- Blood transfusions
- Donating stem cells
- What is CAR-T therapy?
- Watch and wait - What you need to know
- Watch and wait - My blood counts
- Watch and wait - A quick guide for partners, carers, family and friends
- Watch and wait - A quick guide for employers
- Watch and wait - A toolkit for GPs and practice nurses

Side effects

- Managing sickness and vomiting
- Sore mouth or gut (mucositis)
- Understanding infection

Living with blood cancer

- If your employee or colleague has blood cancer
- My activity planner

To order or download information

Visit **bloodcancer.org.uk/information**
Call **0808 2080 888** (Mon–Fri 10am–4pm, Sat–Sun 10am–1pm)
Email **support@bloodcancer.org.uk**
Or use the order form on **page 120**

We're a community dedicated to beating blood cancer.

About us

We're the scientists who dedicate our careers to finding cures.

We're the nurses who find the right words in the darkest moments.

We're the campaigners and volunteers standing up for the people we love.

We're the bucket-collectors, race-runners and cake-bakers who make our research possible.

We're the friends, parents, children and grandparents affected by blood cancer.

Why?

Because we've invested over £500 million in life-saving research.

Because the finish line's in sight.

Because it's time to beat blood cancer.

Because we give people the support they need

People with blood cancer and their family and friends have unique support needs.

We offer free and confidential support by phone or email, provide information about blood cancer and life after a diagnosis, and have an online forum where you can talk to others affected by blood cancer.

bloodcancer.org.uk
0808 2080 888
(Mon–Fri, 10am–7pm, Sat–Sun, 10am–1pm)
support@bloodcancer.org.uk
forum.bloodcancer.org.uk

Because our research is saving lives

The money raised by our community has meant we've been able to invest £500 million in research, which has changed the outlook for people with blood cancer. Our research has led to better treatments that have dramatically increased survival rates. Right now, we're funding research projects across the UK that are finding out more about blood cancer and the best way to treat it.

Find out more: **bloodcancer.org.uk/research**

Because we campaign for better treatment and care

We work to make sure people affected by blood cancer are at the heart of Government and NHS decision-making.

We're campaigning to end delays to diagnosis, improve access to the latest treatments and help people with blood cancer get the mental health support they need.

Find out more: **bloodcancer.org.uk/campaigns**

Because we'll beat it together

Donate

A quick way to help. Every pound brings us one step closer to beating blood cancer:
bloodcancer.org.uk/donate

Fundraise

Sign up to one of our events, or do something you enjoy with family and friends – there are lots of ways to fundraise: **bloodcancer.org.uk/fundraise**

Join your local community group

Local community groups raise money and awareness in their local area. Volunteer for yours to meet new people and get involved in local activities:
bloodcancer.org.uk/local-community-groups

Volunteer

Give your time, meet new people and experience new things by volunteering with us. There are lots of ways you can help, from your own home or in your local community: **bloodcancer.org.uk/volunteer**

Get your organisation involved

From funding a project, to becoming a corporate partner, find out how your organisation can help us:
bloodcancer.org.uk/corporate-partnerships

Or call us on **0808 169 5155**

I love being part of the Blood Cancer UK family. Being involved has helped me and my family cope with my diagnosis.

Anna, diagnosed with blood cancer aged 39

Notes

Blood cancer UK

Go to **bloodcancer.org.uk/donate**, call us on **0808 169 5155** or complete and send this form to us freepost using the address: **FREEPOST PLUS RTSU-XAYE-XZYK, Blood Cancer UK, 111 George Street, Edinburgh, EH2 4JN**

Full Name .....

Address .....

.....

Email ..... Phone .....

As a supporter, you're at the heart of everything we do. We'd love to keep you updated about our exciting work and the ways you can help, including campaigns and events that you might be interested in. We promise to respect your privacy and we will never sell or swap your details.

I'm happy for Blood Cancer UK to contact me by: ☐ Email ☐ Phone ☐ SMS
Don't contact me by post: ☐

You can change how we communicate with you at any time.
Contact us on **0808 169 5155** or email **hello@bloodcancer.org.uk**

I'd like to donate ☐ £10 ☐ £25 ☐ £50 ☐ Other .....
☐ I enclose a cheque/CAF voucher made payable to Blood Cancer UK
OR please debit my ☐ Visa ☐ Maestro ☐ Mastercard ☐ CAF card
Cardholder's name .....
Card number ☐☐☐☐ ☐☐☐☐ ☐☐☐☐ ☐☐☐☐ (Maestro only) ☐☐☐
Start date ☐☐☐☐ Expiry date ☐☐☐☐ Issue number ☐☐

Make your donation worth an extra 25p for every £1 at no extra cost to you!

giftaid it

I'd like Blood Cancer UK to claim Gift Aid on this donation and any donations I make in the future or have made in the past 4 years.

☐ *By ticking this box I confirm that I'm a UK taxpayer and I understand that if I pay less Income Tax and/or Capital Gains Tax than the amount of Gift Aid claimed on all my donations in that tax year, it's my responsibility to pay any difference.

*Today's date .....

If you stop paying tax, change your name or address, or if you have any further questions about Gift Aid, please contact our Supporter Relations Team on **0808 169 5155**.

*Information required for Gift Aid declaration to be valid.

Order information from Blood Cancer UK

All of our information is free to people affected by blood cancer, but if you would like to include a donation with your order, please fill in the donation form over the page.

You can order more information by:

- visiting **bloodcancer.org.uk/information**
- emailing **support@bloodcancer.org.uk**
- calling **0808 2080 888**
- or completing and sending this form to us freepost using the address: **FREEPOST PLUS RTSU-XAYE-XZYK, Blood Cancer UK, 111 George St, Edinburgh, EH2 4JN**

Please send me some information

Full Name .....

Address .....

.....

Email .....

Phone .....

Please tell us the publications you would like us to send you, free of charge (see page 110)

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Keep in touch

We'd love to keep you updated about our exciting work and the ways you can help, including campaigns and events that you might be interested in. We promise to respect your privacy and we will never sell or swap your details.

I'm happy for Blood Cancer UK to contact me by: ☐ Email ☐ Phone ☐ SMS

Don't contact me by post: ☐

You can change how we communicate with you at any time.

Contact us on **0808 169 5155** or email **hello@bloodcancer.org.uk**

My details

My name and hospital number

My NHS number

My condition

My contacts

My consultant

My key worker (usually CNS)

Haematology ward

Haematology clinic

Out of hours

Other contacts

Because we face it together

We're a community dedicated to beating blood cancer by funding research and supporting those affected.

Get in touch for:

- Free and confidential support by phone or email
- Information about blood cancer and life after diagnosis
- An online forum for people affected by blood cancer

bloodcancer.org.uk
0808 2080 888
(Mon–Fri, 10am–7pm, Sat–Sun, 10am–1pm)
support@bloodcancer.org.uk
forum.bloodcancer.org.uk

Your feedback on this booklet can help us improve – please send any comments to **information@bloodcancer.org.uk**

Blood cancer UK