Myeloproliferative neoplasms (MPN)
My details

This is a place to put important information about you, your condition and key contacts.

Name and hospital number

My NHS number

My condition

My contacts

My consultant

My key worker (usually your CNS)

Haematology ward

Haematology clinic

Out of hours

Notes
Our patient information is for you and those close to you to use whenever, wherever and however you need it. You’ll probably have lots of questions; this booklet aims to help you answer as many of them as possible.

Our information is developed for and with patients. It’s written in line with national guidelines and created with health professionals from our dedicated Medical Advisory Panel, so you know it’s accurate and up to date.

This booklet is one of many we make – you can find a list of our other booklets on pages 102–103. For the very latest information, visit our website.

Our booklets contain general information. Always listen to the advice of your specialist about your individual treatment – because every person is different.

When you see the symbols below in the booklet, it’s a sign that we think the websites and other organisations mentioned will also give you good information and support.

A team of people helped produce this booklet. We'd like to thank a member of our Medical Advisory Panel, Professor Tony Green (Addenbrookes Hospital and University of Cambridge), for his help and support in developing the content and checking for clinical accuracy. The draft was also assessed by Dr Jacob Grinfeld, Dr Jyoti Evans, Dr Will Thomas and Clinical Nurse Specialists Vashti Ragoonanan, Yvonne Francis, Siobban McGuckin and Millicent Blake-McCoy.

Bloodwise staff revised the text to make it easy to read, and a non-medical panel, including patients and relevant societies, checked it for understanding. A member of Bloodwise’s Medical Advisory Panel, Professor Tony Green, is responsible for the content overall.

A list of references used in this booklet is available on request. Please email us at patientinformation@bloodwise.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. Bloodwise 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).
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Everyone is different, so listening to the advice of your specialist and your healthcare team is really important.

Introduction

This is a booklet for people with a blood disorder, and for people who may know someone with a blood disorder.

Being told that you, or a loved one, have a blood disorder can be a tough thing to hear, especially as you may also hear MPN being referred to as a blood cancer.

There’s sure to be a lot of information to take in right now.

We hope this booklet will help you to understand your condition and feel in control throughout this time. We’ll try to answer as many of the questions you might have along the way – from symptoms through to tests, treatment and living with MPN, and where you can get support.

Every person is different, with a different medical history. So when you’re deciding what’s right for you, discuss your situation with your specialist as well as getting information from this booklet and other places.

We also produce a diary which you can order online. It’s yours to use however you like – for practical information or to record thoughts or sketches > bloodwise.org.uk/patient-diary
Myeloproliferative neoplasms are a group of rare conditions related to leukaemia. In these blood disorders, your body produces too much of a particular type of blood cell.

In 2008 the World Health Organization defined MPNs as cancers, but this is a matter of definition and doesn’t have any specific impact on your treatment or prognosis.

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.

Some doctors describe chronic myeloid leukaemia (CML) as an MPN, but it’s more usual to treat this as a separate condition.

You can find more information about chronic myeloid leukaemia in our booklet > Chronic myeloid leukaemia
Polycythaemia vera 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 patients 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 patients (less than 5%), PV can transform into a faster growing disease known as myelofibrosis, or into acute myeloid leukaemia. If this happens, your medical team will explain these conditions to you in more detail.
Essential thrombocythaemia 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 patients (less than 5%) ET can transform into a faster growing disease known as myelofibrosis, or into acute myeloid leukaemia. If this happens, your medical team will explain these conditions to you in more detail.

You can find more information about acute myeloid leukaemia in our booklet > Acute myeloid leukaemia
Myelofibrosis 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 disorders such as PV or ET develop myelofibrosis; this is known as secondary MF.

What's the outlook?
MF can progress in different ways in different people. Some patients 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 patients may have a stem cell transplant, but this is rare.

Can MF lead to any other conditions?
In around a quarter of patients, MF may progress to acute myeloid leukaemia (AML). If this happens, your team will explain the condition to you in more detail.
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 four 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).

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, gender and ethnicity. If you’re healthy, the amount you have of each normally stays the same. Here’s a table which shows how many of each type a healthy person has.

<table>
<thead>
<tr>
<th></th>
<th>WHITE BLOOD CELLS</th>
<th>RED BLOOD CELLS</th>
<th>HAEMOGLOBIN</th>
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<td>(10^9/l)</td>
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<td>0.9 to 4.2</td>
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(Some data unavailable for Afro-Caribbean and African patients)

Adapted with permission from A Beginners Guide to Blood Cells
Dr Barbara Bain; Blackwell Oxford; 1996
Bone marrow, blasts and blood 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 go on to 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 22, 34 and 44.

You can find more information about how to manage infections in our booklet > Supportive Care.
Polycythaemia vera

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

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 patients may also have increased numbers of white cells and platelets.

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

For more information on bone marrow and how blood cells are produced see page 18.
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 patients diagnosed with PV are aged 40 years or older. The disease is rare in people under the age of 15 years.

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

Symptoms
You may not have any symptoms at all 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
Patients with PV 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. 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

- 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.

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

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.

I didn’t have any symptoms, so I wasn’t really expecting my blood test to come back with a diagnosis of PV.

When I was diagnosed I asked my consultant to write it down and I’m really glad I did. I could then go away and do my own research and also tell people accurate information about my condition.
The result of this test alone isn’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.
Polycythaemia vera

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 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’s a theoretical risk that it may increase the risk of PV transforming into acute myeloid leukaemia if it’s used as a long term treatment, and because of this it doesn’t tend to be given to people under 50. For older patients, it’s felt that the benefits of the treatment outweigh any potential small risk involved.

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 reduces the rate at which blood cells are made. It’s not thought to carry the same risk of developing leukaemia and is the preferred choice for younger patients. However, many patients 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.

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

› how high the packed cell volume (PVC) 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.

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.

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

For more information about treatment decisions, see our booklet > Treatment decisions

For more information about chemotherapy see another one of our booklets > Chemotherapy: what do I need to know?
Polycythaemia vera

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

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

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.

Outlook

Since my diagnosis, my healthcare team have been there supporting me every step of the way.

For more information on myelofibrosis see page 43

You can find more information about acute myeloid leukaemia in our booklet > Acute myeloid leukaemia
With essential thrombocythaemia, there are too many platelets produced in your bone marrow.

**Essential thrombocythaemia**

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.

**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.

For more information on bone marrow and how blood cells are produced see page 18.
Essential thrombocythaemia

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 chromosomes. 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.

Who gets ET?
As well as the presence of certain genetic faults, there are some other factors which mean you might be more at risk of getting ET.

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

Gender
Men and women are at equal risk of developing ET.

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 patients 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
Patients with ET 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.

The risk of clots is higher in older patients who also have other medical conditions such as diabetes or heart problems, or in patients 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
› 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.

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

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 ET patients 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
› some types of cancer
› other diseases of the bone marrow such as polycythaemia vera (PV), primary myelofibrosis (PMF) and chronic myeloid leukaemia (CML)
› if your spleen isn’t functioning normally.

For a list of organisations who can offer support, see page 81

You can read about the experiences of other people who are going through, or have been through, the same thing on our website > bloodwise.org.uk/patient-support

My specialists were really helpful after I was diagnosed with ET. They explained what my likely outlook was, and took the time to answer all the questions I had.

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.

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 to reduce the risk of blood clots. If you need to take painkillers for any other reason, ask your doctor which ones you can take safely at the same time.

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.

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’s a theoretical risk that it may increase the risk of ET transforming into acute myeloid leukaemia if it’s used as a long term treatment, and because of this it doesn’t tend to be given to people under 50. For older patients, it’s felt that the benefits of the treatment outweigh any potential small risk involved.

Interferon
Interferon is an injection that slows down the production of platelets. It’s not thought to carry the same risk as hydroxyurea so is the preferred choice in younger patients. However, many patients 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.

For more information about treatment decisions, see our booklet > Treatment decisions

For more information about chemotherapy see another one of our booklets > Chemotherapy: what do I need to know?

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 ET patients 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.

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.
Myelofibrosis

Myelofibrosis (MF) is a rare condition of the bone marrow and is most common in people over 50 years.

What is MF?
MF is a myeloproliferative neoplasm where 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, patients 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 bone marrow disorders 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 patients 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.

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.

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 about those genes, your consultant will be happy to talk to you about them.
Who gets MF?
As well as the presence of certain genetic faults, there are some other factors which mean you might be more at risk of getting MF.

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

Gender
Men and women are at equal risk of developing MF.

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
Patients 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
› 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.

If you have any of the following symptoms, you should get urgent medical attention.
Most people are diagnosed with MF 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 MF patients 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. Your doctors will then look at the bone marrow sample under a microscope to assess it for any disease which might be in it.

Bone marrow trephine
As a patient 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. This 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.

When I was diagnosed with myelofibrosis, one of the first things I thought was: why me? But then I came to understand that with MF it’s not something you can clearly point to – nothing I did caused it.

For a list of organisations who can offer support, see page 81

You can read about the experiences of other people who are going through, or have been through, the same thing on our website > bloodwise.org.uk/patient-support
Staging MF
Once a diagnosis has been confirmed, your doctor will do tests to find out how much the MF has progressed.

Some patients will be in the early proliferative (or cellular) phase.

At this stage:

› Your bone marrow will be overactive with increased numbers of platelet-producing stem cells (megakaryocytes).

› There will be a slight increase of fibrin in your blood (the protein that forms when the blood clots, and disrupts the flow of blood through the fibrous network).

Fibrotic phase
At this stage, there is:

› An increase in the amount of fibrosis in your bone marrow.

› A decrease in the amount of normal blood-forming tissue.

The structure of the bone marrow changes which in turn leads to a reduction in the mature cells in your blood (red cells, platelets and/or white cells).

Acute myeloid leukaemia
In a proportion of patients, MF can progress to acute myeloid leukaemia.

Blood counts will be very high at this stage. Your doctor will conduct detailed tests to distinguish MF from leukaemia. Your doctor will explain these tests to you if you need to have them.
Treatment
The treatment you have will depend on a variety of factors, including:

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

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.

Treatment 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 may be to 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’s a theoretical risk that it may increase the risk of MF transforming into acute myeloid leukaemia if it’s used as a long term treatment, and because of this it doesn’t tend to be given to people under 50. For older patients, it’s felt that the benefits of the treatment outweigh any potential small risk involved.

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 patients healthy stem cells, which then produce normal blood cells.

It isn’t a suitable treatment for most MF patients due to the risks associated with the procedure. For a minority of patients – 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 the patients’ own stem cells

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

Outlook
The outlook for people with MF can be very varied. Some patients may have a mild form of MF that doesn’t progress rapidly. In these cases MF shouldn’t interfere significantly with your everyday life.

In other cases, MF progresses more quickly and patients need regular blood transfusions.

In some patients, MF can progress very quickly and needs treating with a stem cell transplant.

It’s estimated that around a quarter of patients with MF may develop acute myeloid leukaemia.

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

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 patients with blood diseases. Your consultant will be an expert in treating your specific disease.

Your clinical nurse specialist

People with blood disorders 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 something like 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 patients

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 patients have side effects from a drug and others don’t.

You may also want to contact a support organisation – many provide patient 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 who are going through – or have been through – through the same thing on our website > bloodwise.org.uk/patient-support
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.

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 patients 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 patient 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.
Your questions

Your questions

You can find a list of questions you might want to ask on page 87 and room to write more questions on page 90.
Telling people

Many patients 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.

Macmillan make a booklet about talking to children about cancer > go to macmillan.org.uk then search for ‘talking to children and teenagers when an adult has cancer’
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.

While MPN aren’t typical cancers, some of the information from organisations about cancer and work may still be useful.

"I didn’t know whether to tell people I had this condition and at first only told people on a need-to-know basis – after all, I looked well physically so didn’t think people would understand. But I’ve found since telling people that I’ve largely continued to be treated as ‘me’ rather than ‘me with MPN’.

Macmillan have some useful advice about cancer and work online; you can also order a booklet > go to macmillan.org.uk then search for ‘work’

There’s more information about cancer and how it can affect your work or study on page 72"
Your healthcare team should look after your emotional needs, as well as your physical ones.

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.

You can read about the experiences of other people who are going through, or have been through, the same thing on our website > bloodwise.org.uk/patient-support

You might like to get in touch with an organisation that can offer support for you and people close to you > see page 81
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.

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.

Alternative and complementary therapies
There’s an important difference between alternative therapies, which are offered in place of medical treatment, and complementary therapies, which are used alongside standard treatment.

We don’t recommend that you use any alternative therapy in place of proven medical care.

There’s some evidence that some complementary therapies may help, particularly with the side effects of standard treatment. If you’re considering using any form of complementary therapy it’s very important to tell your healthcare team, as some treatments may interfere with the standard treatments you’re having.

There’s also a risk that some treatments, for example deep massage or acupuncture, may be unsafe if you have a weakened immune system.

We have a booklet on dietary advice > Dietary advice for patients with neutropenia

We have a booklet on complementary and alternative therapies (CAM)
Practical support

Your work, education and domestic arrangements

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.

MPN and the law

People with MPN, or any other serious disease, are covered by a law called the Equality Act – for the purposes of the act, MPN and other cancers are considered a disability. This means that employers and places of study are required by law to make reasonable arrangements for ‘people with disabilities’ and cannot discriminate against you.

An example of a reasonable arrangement would be if you need time off to go to hospital for treatment. Your employer or college has to allow this and isn’t allowed to reduce your pay or make you take the time as unpaid leave.

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 disorder, 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.

While having an MPN can be different from having many forms of cancer, some of the information Macmillan Cancer Support have online about finances may be useful for you; you can also order a booklet > go to macmillan.org.uk then search for ‘financial support’

For a list of other organisations with useful information about your finances, see page 84
The treatment you decide on with your healthcare team will depend on your health, your individual condition and your wishes.

Clinical trials

If there’s a clinical trial (study) available, your consultant might recommend that you consider this.

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.

For more information on clinical trials, read our booklet >

Clinical trials

If you don’t need treating immediately for your condition, we have a booklet on ‘watch and wait’ which you might like to read >

Watch and wait: monitoring while treatment isn’t necessary
Our researchers are making discoveries that will have a positive impact for people with MPN.

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Research and new developments

Each year we invest a large part of the money we raise in research which aims to stop people dying from blood cancer; make patients’ lives better; and stop people getting blood cancer and related disorders in the first place. We have a number of research projects going on around MPN. Here’s an overview of some of the things our researchers are looking into.

“The research that’s going on is truly amazing. It really gives me and my family hope to know that so much time, energy and wisdom is being devoted to finding new and better treatments.”
Understanding the biology
Our researchers in Cambridge and Southampton are investigating the biology of myeloproliferative neoplasms (MPN), to discover the genetic changes that drive the formation of these diseases. By better understanding the underlying biology, we’ll be able to develop and improve diagnostic tools, prognostic predictions and new treatments.

Our Cambridge researchers, for example, have discovered that the order in which genetic errors happen can influence how severe each person’s disease is and how they’ll respond to treatment. The aim is to help doctors tailor individual treatment plans for individual patients.

Better, kinder treatments
We’re also supporting a number of clinical trials to improve the treatment of MPN. These trials are all based on a new targeted drug called ruxolitinib, which interferes with growth signals used by cancer cells.

One trial, which aims to improve treatments for high risk polycythaemia vera and high risk essential thrombocythaemia, is being run through our national Trials Acceleration Programme.

Some patients can’t tolerate the side effects of the standard treatment, hydroxycarbamide, and others stop responding to it. The researchers are testing whether ruxolitinib is safer, better tolerated and more effective than the next best option for these patients.

Myelofibrosis: working to improve outcomes for patients
The only current cure for myelofibrosis is a stem cell transplant, but not every patient can have one and a transplant may bring complications.

One of our trials in Oxford is looking first at whether pre-treatment with ruxolitinib is safe and whether it can improve the likelihood of successful transplants. Another study in our Trials Acceleration Programme is testing whether combining ruxolitinib with chemotherapy is safe for patients whose myelofibrosis has progressed, and who can’t have a stem cell transplant.
Places you can get help and support

Many people affected by MPN find it useful to call on the expert information, advice and support offered by a variety of organisations, including ourselves. As MPN are blood disorders that are closely linked to blood cancers, many cancer charities will offer you support with your condition. Here are some we recommend.

Bloodwise
We offer patient information online and in free printed booklets, and have an online community you may like to join.

› 020 7504 2200  › patientinformation@bloodwise.org.uk
› bloodwise.org.uk

We can help with practical and emotional support and signpost you to other available services.

› 0808 2080 888  › patientservices@bloodwise.org.uk

Macmillan Cancer Support
Offers practical, medical, financial and emotional support.

› 0808 808 0000  › macmillan.org.uk
CancerHelp UK
(Cancer Research UK’s patient support service)
Offers information about different conditions, current research and practical support.
› 0808 800 4040 › cancerresearchuk.org/cancer-help

MPN Voice
A specialist charity for people with MPN. They offer newsletters and patient events.
› mpnvoice.org.uk

Leukaemia Care
Offers patient information, a 24 hour care line and support groups for people affected by leukaemia, lymphoma, myeloma, myelodysplastic syndromes, myeloproliferative neoplasms and aplastic anaemia.
› 01905 755 977 (general enquiries) or 08088 010 444 (Care Line)
› care@leukaemia-care.org.uk › leukaemia-care.org.uk

African Caribbean Leukaemia Trust (ACLT)
The ACLT aims to increase the number of black, mixed race and ethnic minority people on the UK Bone Marrow Register by raising awareness and running donor recruitment drives.
› 020 8240 4480 › info@aclt.org › aclt.org

Anthony Nolan
Runs the UK’s largest stem cell register, matching donors to patients with leukaemia and other blood related disorders who need a stem cell transplant.
› 0303 303 0303 › anthony-nolan.org

Maggie’s Cancer Caring Centres
Centres across the UK, run by specialist staff who provide information, benefits advice and psychological support.
› 0300 123 1801 › enquiries@maggiescentres.org
› maggiescentres.org

Marie Curie Cancer Care
Nine hospices throughout the UK and offers end of life support to patients in their own homes, free of charge.
› 0800 716 146 › supporter.services@mariecurie.org.uk
› mariecurie.org.uk

MedicAlert Foundation
Provides an identification system for individuals with hidden medical conditions and allergies, in the form of emblems you wear on your body and necklaces or wrist bands.
› 0800 581 420 › info@medicalert.org.uk
› medicalert.org.uk
Financial advice

Citizens Advice Bureau (CAB)
Offers advice on benefits and help with filling out benefits forms.
› 08444 111 444 (England) or 0844 477 2020 (Wales)
› adviceguide.org.uk

Department for Work & Pensions (DWP)
Responsible for social security benefits. Provides information and advice about financial support, rights and employment.
› gov.uk

Macmillan Cancer Support Grants
A Macmillan grant is a one-off payment for adults, young people or children with cancer, to cover a wide range of practical needs.
› macmillan.org.uk/HowWeCanHelp/FinancialSupport/MacmillanGrants

Travel insurance

Macmillan Cancer Support
Provides information about what to consider when looking for travel insurance. It also has a list of insurance companies recommended by people affected by cancer.
› 0808 808 0000  macmillan.org.uk

Association of British Insurers (ABI)
Provides information about getting travel insurance and contact details for specialist travel companies.
› 020 7600 3333  abi.org.uk

British Insurance Broker’s Association (BIBA)
Offers advice on finding an appropriate BIBA-registered insurance broker.
› 0870 950 1790  enquiries@biba.org.uk  biba.org.uk
Questions to ask

It’s easy to forget the questions you wanted to ask when you’re sitting with your healthcare team and trying to take in lots of new information. Some patients find it useful to write down the questions they want to ask before they get there.

Here are some questions you might like to ask at different times.

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?

It can be a good idea to write down the questions you want to ask before each appointment.
Questions to ask

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?
If you're diagnosed with blood disorder you need to know that there are people who can help. You want to know what it means, what’s going to happen, what the treatment is like and what your chances of living a normal life are. You need to know someone is there for you.

Blood cancers represent one in 10 of all new cancer diagnoses – this means that each year 38,000 people are diagnosed with blood cancers and closely related conditions.

We play a vital role, working in collaboration with the NHS, health professionals, government, pharmaceutical companies and other charities to ensure that the needs of blood cancer patients are addressed. We take a leading role in research into blood cancers; we ensure patients have access to innovative clinical trials where possible; we provide information; and we’re a voice of influence when it really counts.

This means no blood cancer patient ever needs to feel alone. We have more than 1,000 researchers, clinicians and nurses making sure that our research has a clear line of sight to improving patients’ lives.

We ensure that when our expert knowledge counts, we speak to the people in the right places to influence decisions.

We support a community of thousands of individuals, families and friends who have their own experience of blood cancer and we create a safe space for patients to share their worries and also see that there can be light at the end of the tunnel.

As one of the UK’s leading blood cancer charities, we feel both the responsibility and the opportunity that we have to make patients’ lives better.
Our patient services

We put the patient at the heart of everything we do. We strive to help everyone affected by a blood cancer to live the best possible quality of life, for life.

Alongside our ongoing commitment to support vital medical research into the cause and cure of blood cancers, we’re developing and delivering quality support and services for patients, their family, friends and carers to help with the emotional and practical impact of blood cancer.

Through our dedicated Patient Support area on our website, people can share their experiences of blood cancer, connect with each other and access our wide range of patient information. We provide support, information and advice over the phone and online, and we’re constantly developing new ways to support our patients based on what they tell us they want and need.

How we raise money

We don’t get any government funding: it’s the money raised by our incredible supporters that lets us continue our life-saving work.

It’s because of them that we can offer our patient information free of charge to blood cancer patients, so we’d like to say a big thank you to everyone who gives so generously to us.

If you – or anyone close to you – ever feels able to make any kind of donation, large or small, it will help us continue our life-saving work: bloodwise.org.uk/give

Our patient information is available to download, order or read online. You can also blog about your journey there and read about other people’s blood cancer experiences: bloodwise.org.uk/patient-support
How you can get involved

There's lots of other ways you can get involved that will help us achieve our vision of beating blood cancer.

**Patient Support**

Patient Support is your space to find information, share knowledge and experiences and connect with others affected by blood cancer.

bloodwise.org.uk/patient-support

**Patient focus groups**

Patients are at the heart of everything we do. That's why we consult patients at every opportunity to get their views about the services we provide. Check our website for upcoming events.

bloodwise.org.uk/focus-groups

**Cycling**

We like to think we're the UK's premier cycling charity! From short family rides through to our flagship London and Birmingham Bikeathons and epic London | Paris challenge, if you've got a bike you can cycle with us to beat blood cancer.

bloodwise.org.uk/cycling

**Running**

All around the country, right throughout the year, our unstoppable runners take to the streets to help us beat blood cancer. Whether it's at the London Marathon, one of the Great Run series or a junior run, it's a case of every step counts.

bloodwise.org.uk/running

**Triathlons**

You won't know what you can do until you tri! Triathlons are ever popular with sporty types who want to conquer swimming, cycling and running, all on the same day! We're the proud title sponsor of the wonderful Blenheim Palace Triathlon but there are so many more on offer.

bloodwise.org.uk/triathlon

**Challenges**

And then there are some who want to climb mountains, trek through jungles and canoe rivers for us! Get in touch to tell us about your challenge and we'll support you all the way.

bloodwise.org.uk/challenges
How you can get involved

We're always looking for companies who share our vision of a future without blood cancer and we recognise the benefits that partnering with us can bring your business. We know we can achieve more together than we ever could alone and our dedicated corporate team will work with you to build an innovative, mutually beneficial partnership.

[Link to corporate section]

We were formed back in 1960 by some brave parents in Middlesbrough whose daughter sadly died from leukaemia. Fast forward all these years and our local fundraisers are still right at the heart of our organisation. Our friendly regional teams can support you in every aspect of your fundraising and are always on hand for a chat.

[Link to local fundraising section]

To give gifts that give back, visit our online shop. We have a great range of ethically sourced products and 100% of profits go towards beating blood cancer.

[Link to shop section]

Meet the likes of Billy Connolly and Miranda Hart at our annual ‘Audience with’ events or frock up for our star-studded ‘Christmas with the Stars’ concert at the Royal Albert Hall. To see our very latest events check out our website.

[Link to special events section]

Short on time? Join our Facebook and Twitter communities. Every quick share could mean a new supporter, a new donation or a new patient finding out about our support services.

[Links to Facebook and Twitter]

Shop with us

Corpor**ate fundraising**

Local fundraising

Special events

Being part of our online community
Your feedback

We're always looking for ways to improve the information we provide for people with blood cancer.

We welcome your feedback on this booklet and our other patient information. Any improvements you suggest mean we can make better information for other blood cancer patients and people close to them.

To fill in a short survey about our patient information online, please go to > bloodwise.org.uk/bookletsurvey

Other booklets

Leukaemia
- Acute lymphoblastic leukaemia (ALL in children up to 16 years)
- Adult acute lymphoblastic leukaemia (ALL) in children and young adults up to 16 years
- Acute myeloid leukaemia (AML) in children and young adults up to 16 years
- Acute promyelocytic leukaemia (APL)
- Adult acute myeloid leukaemia (AML)
- Childhood acute myeloid leukaemia (AML)
- Chronic lymphocytic leukaemia (CLL)
- Chronic myeloid leukaemia (CML)

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

Myeloma
- Myeloma

Related conditions
- Myelodysplastic syndromes (MDS)
- Myeloproliferative neoplasms (MPN)

Treatment
- Bone marrow and stem cell transplantation – for children and adults
- Chemotherapy
- Clinical trials
- Donating stem cells
- Donor lymphocyte infusion
- The seven steps – blood & bone marrow transplantation
- Treatment decisions
- Undergoing high dose therapy and autologous stem cell transplant

General
- Complementary and alternative medicine
- Dietary advice for patients with neutropenia
- Newly diagnosed with a blood cancer
- Supportive care
- Watch and wait

For children and young adults
- Jack’s diary
- Wiggly’s world
- Young adults with a blood cancer – what do I need to know?
It can sometimes seem that blood disorders have their own language. Here are some of the most common words you might hear.

**Glossary**

**Anaemia**
Anaemia is where you don’t have enough red blood cells in your blood. This can mean that your muscles don’t get as much energy as they need, most commonly leading to tiredness or shortness of breath.

**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.

**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 the hair and 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 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.

**Cytogenetics**
The study of the structure of chromosomes. Cytogenetic tests (or FISH tests) are carried out on samples of blood and bone marrow. 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.
Gene
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. Your DNA is arranged in sections called genes.

Fatigue
Fatigue is a feeling of extreme tiredness which doesn’t go away after rest or sleep. It might be caused by the MPN itself or might be a side effect of treatment.

Lymph vessels
Small tubes which make up a network which runs around your body. They carry a fluid called lymph.

Spleen
An organ that filters the blood. It sits under your ribs on the left hand side of your body. The spleen has two main jobs: to remove old red blood cells and to help protect your body from infections.

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.