Your guide to

Myelofibrosis

Information and advice about causes, symptoms and management



MPN voice

MPN voice

Supporting patients and families affected by MPN

Established by a group of MPN patients over 17 years ago, MPN Voice, registered under the auspices of Guy's & St Thomas' Foundation, has a website with a lively online community and up to date information on essential thrombocythaemia (ET), polycythamia vera(PV), and myelofibrosis (MF).

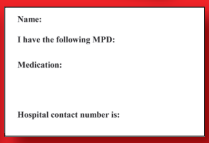
Backed by leading healthcare professionals, MPN Voice also publishes a newsletter, MPN disease leaflets and drug leaflets, runs regular regional patient forums, funds MPN research and offers a buddy scheme.

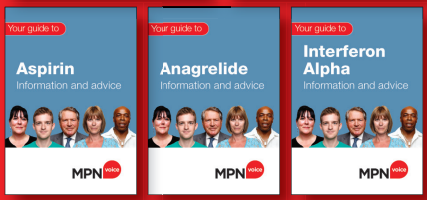
For more information visit the website **www.mpnvoice.org.uk**

Email **info@mpnvoice.org.uk**

MPN Voice

previously known as MPD Voice





Introduction

Being diagnosed with Myelofibrosis (MF) can be a shock, particularly when you may never have heard of it.

If you have questions about MF – what causes it, who gets it, how it affects your body, what symptoms to expect and likely treatments – this leaflet covers the basics for you, but for more information talk to your haematologist or pharmacist.

You'll also find useful advice about how to get the best from your haematologist, plus practical advice on how to help important people in your life understand such a rare condition.

Confused by all the terminology linked to your condition?

Or perhaps you've come across some medical terms you don't recognise? If so, our glossary – an A-Z list of words often associated with MF – at the back of this brochure – might come in handy.

What's inside this brochure

With special thanks to Professor Claire Harrison (DM, FRCP, FRCPath); Consultant Haematologist and Dr Rachel Davis (PhD, MSc, BSc (Hons), CPsychol); Chartered Health Specialist

Visit www.mpnvoice.org.uk

MPN voice

What is myelofibrosis?

Myelofibrosis (MF) is a disorder of the bone marrow. It happens when the marrow – the soft, fatty tissue inside your bones that produces blood cells – is replaced by fibrous (or scar) tissue. Scarring of the bone marrow means the marrow is not able to make enough blood cells, which leads to a set of debilitating symptoms.

If you have MF that occurs on its own, you have what's known as 'primary myelofibrosis'. This is most commonly seen in people over the age of 50.

Or you may have previously been diagnosed with another bone marrow disorder such as essential thrombocythaemia (ET) or polycythaemia vera (PV) – in this case your condition is known as post-essential thrombocythaemia MF or post-polycythaemia vera MF.

ET, PV and MF are closely related diseases that belong to a group of conditions called myeloproliferative neoplasms (MPNs).

How common is MF?

MF is considered to be a rare disease. A disease is generally regarded as rare if it affects fewer than

50 people in **100,000** per year

MF affects men and women in relatively equal numbers.

1:1

That might explain why you probably haven't heard of it, or met anyone with the condition before. MF is virtually unheard of in children and is very rare in young adults. It is most commonly diagnosed in patients between 60 and 70 years of age.

Is MF a type of cancer?

In the early part of this decade there was some debate about whether MPNs such as MF were types of cancer. This is because the word 'neoplasm' (new growth) is a term that has been used both for cancers (malignant neoplasms) and non-cancerous tumours (benign neoplasms). Now, because MF is characterised by uncontrolled cell growth, haematologists and cancer organisations do classify it as a blood cancer.

"There is unprecedented dialogue in the MPN community in trying to pull all our ideas together and better understand MF."

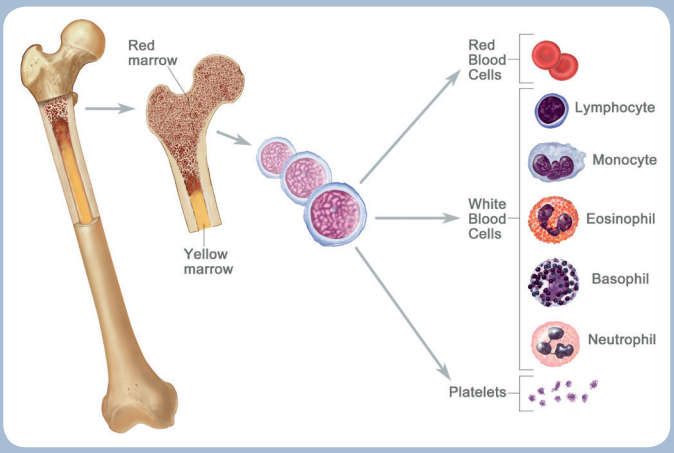
For new patients this can be alarming terminology but it is important to remember that the symptoms and prognosis can vary widely. Your specialist will advise you depending on your individual circumstances – find out more about treatments on page 10.

MPN voice

What causes MF?

What causes MF is not fully understood. The majority of patients with MF have what is commonly referred to as a 'driver mutation'. This is an acquired mutation (change) in the genetic code within a proportion of the stem cells in the bone marrow that are associated with the development of the disease. The most common in Myelofibrosis are JAK2 V617F, CALR (Calreticulin) and MPL. In a small number of patients there is no detectable JAK2, CALR or MPL mutation present so the disease is denoted as 'Triple-negative'. Some patients may in fact have very low-level mutations that are not 'picked up' by the commonly used tests or have another as yet unknown mutation. Some clinical departments have access to specialised tests known as 'myeloid-disease gene panels', whereby targeted sequencing of commonly mutated genes can be performed to aid detection of other key mutations. For MF, these are particularly important for a number of genes and p53 mutations. Dependent on the circumstances, such testing can help refine discussions about prognosis and their overall treatment plan in the longer term'. These mutations may also be triggered by past exposure to ionizing radiation (a type of radiation that has very high energy, like medical X-rays) or to some chemical substances such as benzene and toluene. It is also important to note that MF is rarely inherited and is not passed on from parents to child, although some families do seem to develop the disease more readily than others.

What are stem cells?



Stem cells are 'master cells' found in many organs and tissues of the body which can divide and develop into many other types of cell – such as blood cells, muscle cells and brain cells – to replenish those lost or damaged. Blood stem cells (called haematopoietic stem cells) circulate in our blood and bone marrow, as well as in the umbilical cords of newborn babies. In the bone marrow, they have the potential to develop into mature blood cells.

How does MF affect your body?

In a person without MF...

…bone marrow (the soft, fatty tissue inside your bones) contains blood stem cells that in time develop into mature blood cells – red blood cells (to carry oxygen to the tissues of your body); white blood cells (to fight infection and disease); or platelets (to help prevent bleeding by causing blood clots to form).

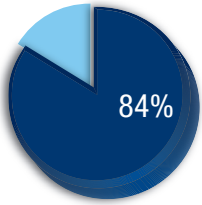
In someone with MF...

…abnormal stem cells take over the bone marrow, leading to fibrosis (scarring) and chronic inflammation. The consequence is that the marrow is not able to make enough normal blood cells. The spleen and then the liver try to compensate by producing red blood cells and sometimes – but not always – this causes the spleen to become enlarged. Due to the inability of the bone marrow to make enough blood cells, MF patients often have low numbers of red blood cells, white blood cells and/or platelets, resulting in anaemia, neutropenia and thrombocytopenia respectively. These changes lead to some of the symptoms of myelofibrosis (read more about them over the page).

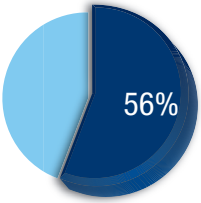
> **"Even though MPNs are caused by a genetic change, they are rarely inherited."**

What are the most common symptoms of MF?

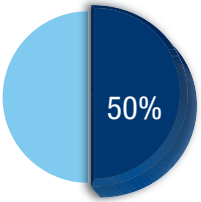
While most patients are diagnosed having presented with symptoms, many people experience few or no symptoms at all in the early stages of MF. In fact, patients are often diagnosed after having tests for an unrelated condition. Regardless, not every person has the same combination of symptoms to the same degree of severity. One study asked 456 patients with MF to identify the symptoms that affected their quality of life. See below for the results:



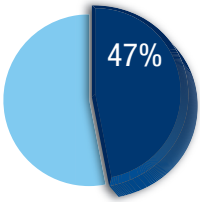
Fatigue



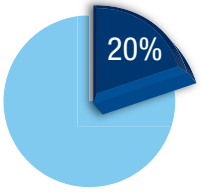
Night sweats



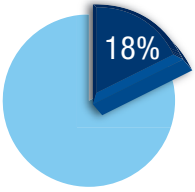
Itching



Bone pain



Undesired weight loss



Fevers

Track your symptoms

It's sensible to monitor how you're feeling and what symptoms you're experiencing. If you notice them changing or getting worse, you can tell your haematologist and explain how they are affecting you on a day-to-day basis.

What happens next?

Your individual situation and health history, as well as the ways you respond to treatment, can all affect your prognosis (the predicted outcome of the disease). MF affects people differently, and an individual's prognosis may vary depending on a number of factors, such as age, blood count and symptoms.

Life expectancy varies from person to person. Your clinician will discuss these factors and your prognostic score which will be based on the result of various tests that will be undertaken at the beginning of your diagnosis.

"It can take time to adjust to the news that your condition could reduce your life expectancy," says Dr Rachel Davis, Health Psychology Specialist. "But with good monitoring and regular updates of your treatment plan, you and your haematologist can work together to manage your symptoms in the best way possible."

About 10–20% of MF cases develop into acute myeloid leukaemia, a blood and bone marrow cancer that can progress rapidly – your haematologist will be monitoring you to check for changes in your condition.

Bear in mind when you're reading up about the condition, that in some patients the disease may remain stable or gradually progress over time. This means that many of the symptoms and outcomes you read about may not happen to you.

"It is not always true that an increase in symptoms heralds an advancing or changing disease."

Treating MF

Usually, the goal of treatment is to reduce any symptoms and improve quality of life.

The following key factors will help your haematologist to work out your best treatment options:

- The number of abnormal red blood cells and white blood cells
- The number of blast cells in the blood (blast cells are cells which are in the early stages of development and do not yet carry out any function)
- Your age
- Certain genetic changes
- Symptoms such as fever, night sweats, or weight loss
- Health complications such as spleen and liver enlargement, gastrointestinal bleeding, infections and malnutrition.

Options that are available to your haematologist include:

Watch and wait

If you have no symptoms when you are first diagnosed with MF, your haematologist may suggest a 'watch and wait' approach. This usually involves regular check-ups and blood counts, as well as your haematologist advising you on ways to live a healthy lifestyle. If your symptoms develop or the disease progresses, you may then start a suitable treatment.

Clinical trials

You may be invited to participate in a clinical trial. All new treatments must be tested thoroughly in different phases of a trial. Through trials, researchers find out if new drugs are effective and safe in patients. Your haematologist will discuss this with you and your decision will not affect any current treatment options that you already receive.

Bone marrow or stem cell transplant

A bone marrow or stem cell transplant offers the only curative treatment for MF. Patients are individually assessed as transplants are not commonly recommended. This is due to a high risk of life-threatening side effects, as well as a risk that the new stem cells

will react against your body's healthy tissues. Transplants are often only considered an option for fit patients with advanced disease. The first step involves very high levels of chemotherapy or radiation therapy.

Drug treatment

Your haematologist will discuss the options tailored to your needs.

Antiplatelet drugs

Such as low dose aspirin may be suitable if the platelet count is high and there is a risk of blood clots (thrombosis). Aspirin can cause bleeding and ulcers in the stomach and gut. Sometimes alternative drugs may be used similar to aspirin, such as dipyridamole and clopidogrel.

Anticoagulant

Warfarin or other anticoagulant drugs such as the injections of heparin or low molecular weight heparin are used to treat blood clots and often used to prevent them in high risk situations e.g. after surgery.

We have provided summary information here. For full details refer to either product insert sheet provided with the medicine or the drug information leaflet/s we provide.

Azacitidine

This drug works by switching off a protein called DNA methyltransferase. In turn this switches on genes to stop the growth and division of abnormal cells and helps to control blood cell growth. It is administered at the hospital as an injection, usually given under the skin (subcutaneously) for 7 days every 4 weeks.

Busulfan

This usually is used when there are side effects with Hydroxycarbamide, or when it is difficult to take Hydroxycarbamide tablets. Like Hydroxycarbamide, Busulfan also affects the bone marrow directly and can lead to a fall in the blood count. This tablet is prescribed in short courses and monitored regularly. Busulfan can cause lung problems, damage to fertility and the bone marrow, and is known to increase the risk of the disease changing into leukaemia.

Cytarabine

This is an injectable drug used to control the disease. It directly affects the bone marrow by interfering with the production of cells. This is given if the disease is progressing quickly or the spleen is very large and causing discomfort. It is a short course needing regular monitoring since blood counts may fall quickly, skin rashes, allergy and fever and potential complications of this agent may occur.

Danazol

This mimics a hormone that is naturally made by the body and can stimulate the production of red cells. It is a tablet and can sometimes make the haemoglobin level rise so blood transfusions will not be necessary. It can cause weight gain by fluid retention and can affect the liver or kidneys. In women it can lead to a slight increase in facial hair growth. Rarely this drug can increase the risk of liver and prostate cancer.

Erythropoiesis-stimulating agents (ESAs)

Erythropoietin (EPO) is a natural substance made in your body by the kidneys, which tells stem cells in the bone marrow to make more red blood cells. Synthetic ESA drugs (sometimes called epoetins) act like the natural erythropoietin, triggering the production of more red blood cells. They can be injected under the skin and may sometimes be given as an alternative to a blood transfusion. However, due to the cause of the disease and the fibrotic bone marrow, ESAs have limited activity in MF.

Hydroxycarbamide (Hydroxyurea)

This drug interferes with cell metabolism. In MF this drug is used to control high blood counts and to reduce the size of the spleen or the liver, it may also improve symptoms. Side effects can include changes in skin colour, mouth and leg ulcers, and stomach problems. This drug can damage cells and may affect fertility therefore women who are pregnant or those trying to conceive, should not use it. There is a small chance that Hydroxycarbamide can increase the risk of acute leukaemia.

Interferon

This is a natural agent that reduces the production of bone marrow cells. Interferon is taken by injection at a frequency that is recommended by your haematologist. Side effects are mainly flu-like symptoms, hair loss, depression, liver and thyroid changes. It does not increase the risk of leukaemia and can be used in pregnancy. Sometimes, in early cases of MF, Interferon can reverse some of the fibrosis and scarring.

JAK inhibitors

Jakavi® (Ruxolitinib)

Is a tablet which inhibits JAK1 and JAK2. In clinical trials called the COMFORT studies this drug strikingly improved symptoms due to disease, lowered blood counts and reduced spleen size. There are early signs that some patients may benefit from prolonged survival after therapy with Jakavi®. Common side effects are anaemia, low platelets and increased risk of infections. Other JAK inhibitors are in clinical trials.

Thalidomide +/- Prednisolone

Thalidomide combined with steroids can help with both anaemia and low platelet counts. Thalidomide is a medication that is thought to work by reducing the blood flow to the marrow and stopping the fibroblast cells from causing the scarring. It can cause nerve problems, sleep disturbance and constipation. Low doses are used for MF and it is usually well tolerated. New more potent agents derived from thalidomide are currently being used in trials to see if they produce better results. Side effects of the steroids are weight gain, increased appetite, mood swings and gastric irritation. In long-term use it can lead to thin bones (osteoporosis) and a risk of diabetes.

Management of an enlarged spleen

An enlarged spleen is a common symptom of MF, often leading to pain, discomfort and a feeling of fullness or a loss of appetite. Treatment options include:

Splenectomy (surgical removal of the spleen)

This is considered if your enlarged spleen is painful and causing complications. However, as all surgical procedures carry some risks other treatment options are often tried first.

> "It is quite uncommon to use splenectomy for the management of myelofibrosis nowadays."

Radiotherapy

Radiotherapy or radiation of the spleen is an option if splenectomy is ruled out. Performed in hospital, radiotherapy helps to reduce the size of the spleen and can also relieve other related symptoms, such as bone pain. Radiation kills cells using high-strength laser-beams such as x-rays. It usually provides temporary relief that lasts between three and six months.

Management of anaemia

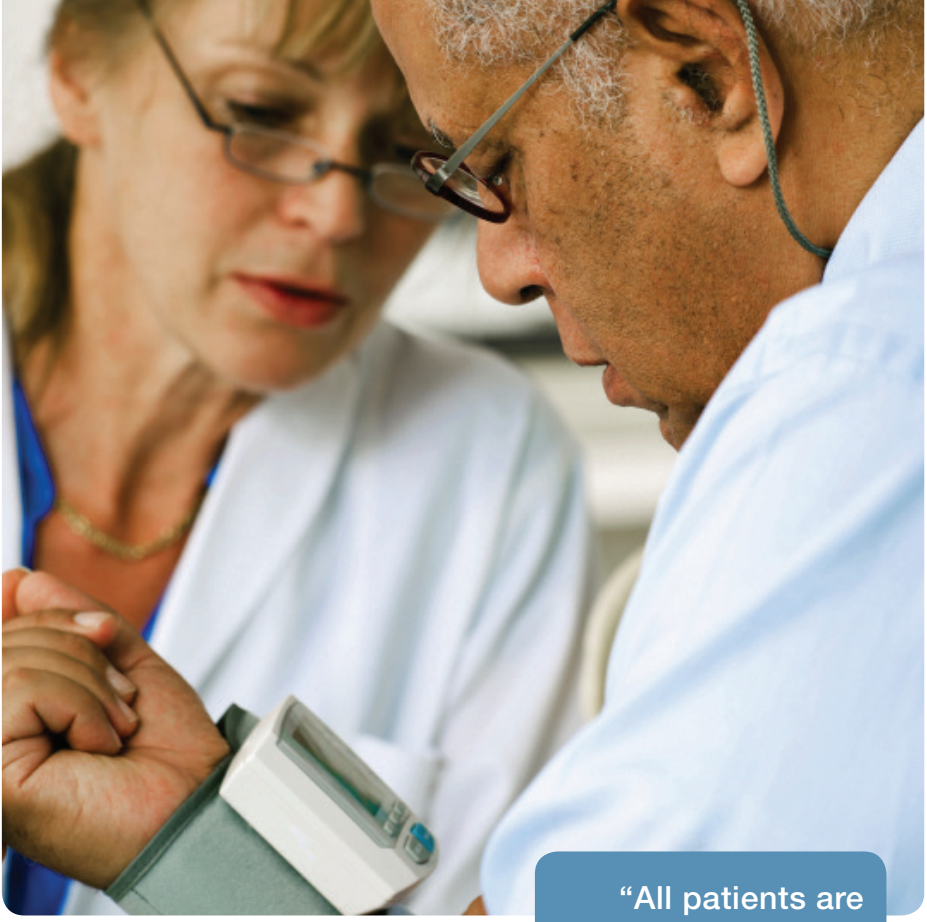
Anaemia is common in people with MF. Symptoms include excessive tiredness, weakness and shortness of breath. If these symptoms are causing you difficulties, your haematologist may suggest:

Blood transfusions

A blood transfusion – usually done in an out-patients' clinic – involves the transfer of red blood cells from a compatible donor into your body. This can increase red blood cell count and quickly reduce symptoms of anaemia, often within 24 hours. Blood transfusions are a relatively safe procedure and don't usually cause serious complications, however there is an increased risk of iron overload if you receive a series of transfusions, usually over a number of years, or after a total of 10-20 transfusions have been given. Always call the nurse if you feel hot, cold, shivery or in any way unwell during or after the procedure, as this might be a sign that you are having a reaction.

Medical exemptions for treatment

MPN patients in England are entitled to an exemption certificate and should obtain a FP92A form from their GP or haematologist. Further details can be found on our website at www.mpnvoice.org.uk in the living with MPNs, everyday challenges/cost of medication section.



> "All patients are entitled to a second opinion and most doctors would encourage this."

Talking to your haematologist

MF is a very rare condition. "For this reason, it's important to form a good working partnership with your haematologist to ensure you're getting the best treatment possible," says consultant haematologist Professor Claire Harrison. Her advice?

- If it's an initial consultation, take along a list of your medications and a list of any allergies
- If you have a complicated medical history, take a list of diagnoses, previous procedures and/or complications
- It can be useful to repeat back what you have heard so that you can be sure that you have understood fully

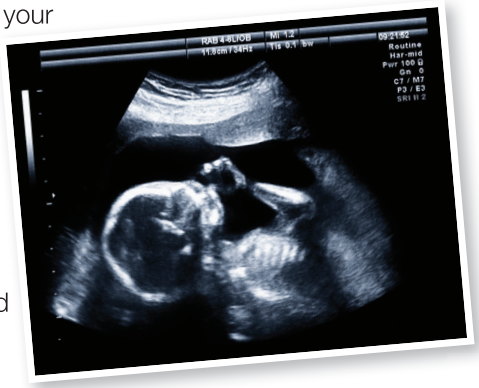
Other tips:

- Keep a record of symptoms to discuss with your medical team
- Make a list of questions to take to your appointment. This will help the discussion with your haematologist
- Bring someone along to your appointment. They can provide support, ask questions and take notes
- Note information down to help you to remember later what was said
- Be open when you discuss your symptoms and how you are coping. Good patient-doctor communication tends to improve outcomes for patients
- Don't be afraid to ask for a second opinion – most doctors are happy for you to ask and, as your condition is rare, it is usually seen as the standard practice

You need to tell your haematologist if...

You're having any medical treatments or taking any products, such as prescribed medicines, over-the-counter treatments or vitamins

You're pregnant or planning to become pregnant, as many treatments can affect your baby's development in the womb



Your partner is planning to become pregnant, as some therapies can affect male fertility and sperm quality

Talking to other people

If you tell someone you've got diabetes or breast cancer, they'll probably have heard of it. Having a rare condition like MF can be harder to explain. This simple summary may help:

"I have a condition where my bone marrow is not working properly and this affects the number of blood cells it produces. This affects my body in several ways. For example, my spleen and liver work extra hard to produce blood cells and this can cause them to become bigger. It also makes me (list your symptoms here e.g. I feel tired, I experience a lot of pain etc). I have good days and bad days and sometimes I may need (explain what you need here e.g. I need a bit more support with my day-to-day activities; someone to talk to; someone to come to appointments etc.)."

There are also other things you can do to make it easier to explain what you're experiencing, how the condition affects you and the support that you need:

1 Find out more

Try to find out more about your condition – if you understand more about MF then you will have a better chance of sharing it in simple terms with other people.

2 Change perspectives

Think about what your family and friends might like to know about your condition. Perhaps have a read of the section for carers and loved ones at www.mpnvoice.org.uk to find out the kinds of questions they may be wanting to ask.

3 Print information on your condition

It might help to print out basic information about your condition, and share this with family and friends. Not only can they take the print-out away with them to read in their own time, but it also takes the pressure off you having to explain all the details yourself.

4 Explain your needs

Family and friends will be keen to help – think about what people can help you with and make some suggestions, whether it's doing your weekly shop, coming round to cook dinner or driving you to an appointment.

5 Be open about how you feel

"If you find you're feeling you don't want to burden your loved ones and friends with your problems, remember people who care about you will want to help so don't be afraid to be open about how you're feeling," says Health Psychology Specialist Dr Rachel Davis. "Saying that, you don't need to be open about your feelings at all times – it may help to explain to family and friends in advance that you are likely to have days when you feel like talking and other days when you don't. People can then take their cue from you."

Coping with a rare condition

"Discovering you have a rare and potentially life-threatening condition can be difficult. It is common to feel lost, confused and isolated because you are unlikely to know anyone else who's going through the same experience, and there's so much new and complex information – and jargon – to take in," says Dr Rachel Davis. "But help and support does exist – and seeking it out can help you to get your head around the facts and help you feel you are part of a community of people all coping with MF, too."

"An MPN patient may need support in coming to terms with their diagnosis and any changes they experience in their condition"

There are a number of helpful sources including:

- Your haematologist and healthcare team
- Your family and friends
- A psychologist (ask your doctor for a referral)
- Specialist clinics in your area or nearby city that study MF where you can find out further information
- Patient support groups (see www.mpnvoice.org.uk)

See www.mpnvoice.org.uk for more ideas on how you can talk to your haematologist about MF, questions you may want to ask and how you can prepare yourself for each appointment.

Complementary therapies

Although there's no scientific evidence to support the use of complementary therapies alone in MF, many people do find that they may help them cope with their disease alongside standard medical treatments. Options include acupuncture, exercise, yoga, meditation and relaxation. Always discuss any complementary therapies with your haematologist.

Glossary

In this glossary some words and medical terms often associated with MF are accompanied by a brief explanation.

A

Abdomen The abdomen is an anatomical area that is bounded by the lower margin of the ribs and diaphragm above, the pelvic bone (pubic ramus) below, and the flanks on each side. Organs within the abdominal cavity include the stomach, small intestine, colon, liver, gallbladder, spleen, and pancreas.

Abdominal pain Pain that is felt in the abdomen [above].

Acute myeloid leukaemia (AML, acute myelogenous leukaemia or acute non-lymphocytic leukaemia) A quickly progressive malignant disease in which there are too many immature blood-forming cells in the blood and bone marrow, the cells being specifically those destined to give rise to types of white blood cells that fight infections.

Anaemia A medical condition in which the red blood cells count or haemoglobin is less than normal.

B

Blood cells See red blood cells, white blood cells, platelet.

Blood clot (or thrombus, or thrombosis) Blood that has been converted from a liquid to a solid state; also called a thrombus. A blood clot is stationary within a vessel or the heart. If it moves from that location through the bloodstream, it is referred to as an embolus.

Blood transfusion The transfer of blood or blood components from one person (the donor) into the bloodstream of another person (the recipient). Often a life saving technique to replace blood cells or blood products lost through bleeding, or when your body can't make blood properly because of an illness such as some MPNs.

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

Bone marrow transplant (BMT) A procedure in which bone marrow that is diseased or damaged is replaced with healthy bone marrow. The bone marrow to be replaced may be deliberately destroyed by high doses of chemotherapy and/or radiation therapy. The replacement marrow may be the patient's own marrow, treated, or it may come from another person (donor).

C

CALR Calreticulin is a protein involved in calcium flow within the cell which is also important in growth signals.

Chemotherapy Therapy for cancer using chemicals that stop the growth of cells.

Chronic A chronic condition is one that is long term in nature. It comes from the Greek 'chronos' and means lasting a long time. MPNs are considered chronic conditions, and many symptoms of MPNs are also chronic.

Complementary therapy
Complementary therapies such as meditation, yoga, acupuncture and aromatherapy are therapeutic disciplines that can be used alongside conventional medicine.

Complete Blood Cell count (CBC)
A set values of the cellular (formed elements) of blood. These measurements are determined by machines that analyse the different components of blood. It is possible to take blood counts for each individual element of the blood, e.g. white blood cells, red blood cells, platelets, etc.

D

Deep vein thrombosis (DVT)
A blood clot in a deep vein in the thigh or leg. The clot can break off as an embolus and make its way to the lung, where it can cause respiratory distress or respiratory failure.

E

Erythropoietin A hormone produced by the kidney that promotes the formation of red blood cells by the bone marrow.

Essential thrombocythaemia (ET)
A rare acquired myeloproliferative neoplasm (MPN) characterised by a sustained elevation of platelet number with a tendency for thrombosis and haemorrhage.

F

Fatigue A condition characterised by a lessened capacity for work, domestic and social activities, and reduced efficiency of accomplishment, usually accompanied by a feeling of weariness and tiredness. Fatigue can be acute and come on suddenly or be chronic and persist.

Fever Technically any body temperature above 98.6 degrees F / 37 degrees C. In practice a person is usually not considered to have a fever until the temperature is above 100.4 degrees F / 38 degrees C.

Full Blood Cell count (FBC) A set of values of the cellular (formed elements) of blood. These measurements are determined by machines that analyse the different components of blood. It is possible to take blood counts for each individual element of the blood, e.g. white blood cells, red blood cells, platelets, etc.

H

Haematocrit HCT or Packed Cell Volume. The proportion (or percentage) of red blood cells in relation to the rest of the blood cells.

Haematologist A doctor who is specially trained in haematology [below].

Haematology The diagnosis, treatment, and prevention of diseases of the blood and bone marrow as well as of the immunologic, haemostatic (blood clotting) and vascular systems.

Haemorrhage Bleeding or the abnormal flow of blood. Bleeding into the spleen or liver is an internal haemorrhage. Bleeding from a cut on the face is an external haemorrhage.

Heart attack The death of heart muscle due to the loss of blood supply. The loss of blood supply is usually caused by a complete blockage of a coronary artery, one of the arteries that supplies blood to the heart muscle. A heart attack can cause chest pain and electrical instability of the heart muscle tissue.

Hepatomegaly An enlarged liver.

J

JAK2 JAK2 is a molecule (called an enzyme) that exists in all people. It forms a communications pathway for messages travelling inside the cell.

JAK2 V617F A mutation (known as JAK2 V617F) in the JAK2 molecule in people with MPNs. This affects the signalling performance of the JAK2 molecule.

L

Liver An organ in the upper abdomen that aids in digestion and removes waste products and worn-out cells from the blood.

M

Myelofibrosis (MF) Fibrosis or spontaneous scarring of the bone marrow. It is characterised by significant anaemia and an enlarged spleen.

Myeloproliferative neoplasms (MPNs) Myeloproliferative neoplasms are diseases of the blood and bone marrow, and sometimes referred to as blood cancers. Four main types of MPNs make up around 95% of cases: myelofibrosis, essential thrombocythaemia, polycythaemia vera and chronic myeloid leukaemia (CML).

N

Neutropenia A condition in which the number of neutrophils (a type of white blood cell) in the bloodstream is decreased.

Next generation sequencing (NGS) DNA sequencing technology which has revolutionised genomic research.

Night sweats Severe hot flushes that occur at night and result in a drenching sweat.

P

Packed cell volume (PCV)
See haematocrit.

Phlebotomy (or venesection)
The removal of a volume of blood (usually 450mls) for medical reasons.

Platelet An irregular, disc-shaped element in the blood that assists in blood clotting. During normal blood clotting, the platelets clump together (aggregate). Although platelets are often classed as blood cells, they are actually fragments of large bone marrow cells.

Platelet count The calculated number of platelets [see above] in a volume of blood, usually expressed as platelets per cubic millimetre of whole blood. Normal platelet counts are in the range of 150,000 to 450,000 per microlitre (or 150 - 450 x 10^9 per litre).

Plateletpheresis Procedure in which thrombocytes (blood platelets) are removed.

Polycythaemia vera (PV). PV is a myeloproliferative neoplasm that results from an overproduction of red blood cells.

Pruritus Another word for itching. Pruritus can result from drug reaction, food allergy, kidney or liver disease, cancers, parasites, aging or dry skin, contact skin reaction, such as poison ivy, and for unknown reasons.

Pulmonary embolism The obstruction of the pulmonary artery or a branch of it leading to the lungs by a blood clot, usually from the leg, or foreign material causing sudden blockage of the vessel.

R

Radiotherapy (or radiation) The treatment of disease with ionising radiation. High-energy rays are used to damage cancer cells and stop them from growing and dividing.

Red blood cell (or erythrocyte) The blood cell that carries oxygen. Red cells contain haemoglobin, which permits them to transport oxygen (and carbon dioxide).

S

Spleen An organ located in the upper left part of the abdomen near the stomach. The spleen produces lymphocytes (a type of white blood cell), filters blood, serves as a reservoir for blood, and destroys old blood cells. The spleen can also supplement the bone marrow in the production of red blood cells in certain situations (as with MPNs). This can sometimes lead to an enlarged spleen (splenomegaly). An operation to remove the spleen is called a splenectomy.

Stem cells Stem cells are cells that have the potential to develop into many different or specialised cell types.

Stroke (Cerebrovascular Accident or CVA) The sudden death of some brain cells due to a lack of oxygen when the blood flow to the brain is impaired by blockage or rupture of an artery to the brain.

T

Thrombocythaemia An abnormally high number of platelets in the blood, typically above 450,000 per microlitre.

Thrombocytopenia A lower than normal number of platelets in the blood, typically below 150,000 per microlitre.

Thrombosis The formation or presence of a blood clot in a blood vessel. The vessel may be any vein or artery. The clot itself is termed a thrombus [see blood clot].

V

Venesection See phlebotomy.

W

White blood cell (or leukocyte) One of the cells the body makes to help fight infections. There are several types of white blood cells. The two most common types are the lymphocytes and neutrophils.

Y

Yoga A way of life that includes dietary prescriptions and physical exercise. Yogic meditation can help control such things as blood pressure, heart rate, respiratory function, and many other bodily functions.

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