
Myelofibrosis (MF)

A Guide for
Patients

Leukaemia Care
YOUR Blood Cancer Charity

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 booklet covers the basics for you.

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

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In this booklet you will see some quotations. These are the real experiences and words of blood cancer patients, so will hopefully help you to understand your disease and situation a little better.

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

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About Leukaemia Care

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

Our services

Helpline

Our helpline is available 9.00am - 10.00pm on weekdays and 9.30am - 12.30pm on Saturdays. If you need someone to talk to, call **08088 010 444**

Nurse service

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

Patient Information Booklets

We have a number of patient information booklets like this available to anyone who has been affected by a blood cancer. A full list of titles – both disease specific and general information titles – can be

found on our website at **www.leukaemicare.org.uk/support-and-information/help-and-resources/information-booklets/**

Support Groups

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

Buddy Support

We offer one-to-one phone support with volunteers who have had blood cancer themselves or been affected by it in some way. You can speak to someone who knows what you are going through. For more information on how to get a buddy call

08088 010 444 or email
support@leukaemicare.org.uk

Online Forum

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

Patient and carer conferences

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

Website

You can access up-to-date information on our website, **www.leukaemicare.org.uk**, as well as speak to one of our care advisers on our online support

service, LiveChat (9am-5pm weekdays).

Campaigning and Advocacy

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

Patient magazine

Our quarterly magazine includes inspirational patient and carer stories as well as informative articles by medical professionals. To subscribe go to **www.leukaemicare.org.uk/communication-preferences/**

What is Myelofibrosis?

Myelofibrosis (MF) is a disorder of the bone marrow. MF occurs when the marrow – the soft, fatty tissue inside your bones that produces blood cells – is replaced by fibrous (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.

There are two main types of MF:

- 1. Primary myelofibrosis:** If you have MF that occurs spontaneously, you have what is known as primary myelofibrosis. This is most commonly seen in people over the age of 50 years.
- 2. Secondary myelofibrosis:** 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 secondary myelofibrosis (either 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 a rare disease as it affects around one to two people in every 100,000 each year. It affects women and men in relatively equal numbers. The rarity of MF might explain why you probably have not 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 cancer?

Cancer is a disease that occurs when normal cells grow in an uncontrolled way. There has been some debate within medical circles about whether or not MPNs are types of cancer. This is because the word neoplasm (new growth) is a term used both for cancers (malignant neoplasms) and non-cancerous tumours

(benign neoplasms). Because MF is characterised by uncontrolled cell growth, most haematologists and cancer organisations do classify it as a blood cancer. Whatever it is called, remember that the symptoms and prognosis can vary widely. Your haematologist will advise you depending on your individual circumstances.

What causes MF?

As time goes on, we are learning more about MF. However, what causes MF is not fully understood. We do know that one of the features of the disease is dysregulated signalling in haematopoietic stem cells (cells in the bone marrow that make your blood cells). One of the signaling pathways that has been found to be frequently involved is called JAK-STAT. About 50 to 60% of MF patients have a change (mutation) in JAK2, a protein that regulates blood cell production (this mutation is known as JAK2 V617F). However, even MF patients who do not have this JAK2 mutation may still have dysregulated JAK-STAT signalling that underlies their disease.

It has recently been discovered that a further 30% of MF patients have a mutation in a gene called calreticulin-R (CALR) and about 5 to 10% of patients have mutation of a gene called MPL. New genetic mutations that are associated with MF continue to be discovered and it is likely that, in most patients, the disease is caused by a combination of these mutations.

All of the genetic mutations described previously are examples of acquired mutations; people are not born with them but develop them as they go through life. It is also important to note that MF is rarely inherited and is not passed on from parent to child, although some families do seem to develop the disease more readily than others.

Risk factors

Apart from the presence of the JAK2, CALR and MPL mutations, there are certain risk factors that increase the risk of you developing MF:

- **Age:** MF is more common in people aged over 50 years, although there are cases of MF in the young.

What is Myelofibrosis? (cont.)

- **Another bone marrow disorder:** A small proportion of people with MF develop it as a complication of other bone marrow disorders such as ET and PV.
- **Exposure to certain chemicals:** MF has been linked to exposure to industrial chemicals, such as toluene and benzene.
- **Exposure to radiation:** People exposed to high ionising radiation have an increased risk of MF.

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, muscle and brain cells, to replenish those lost or damaged. Blood stem cells (called haematopoietic stem cells) are found primarily in bone marrow. In the bone marrow, they have the potential to develop into mature blood cells, such as red cells, white cells and platelets.

Symptoms of Myelofibrosis

How does MF affect your body?

It is important to understand how MF affects the body compared to someone who does not have the condition.

In someone 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 chronic inflammation and fibrosis (scarring). The consequence is that the marrow is not able to make enough normal blood cells. The spleen and then the liver (and sometimes other organs) 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 (anaemia), white blood cells (sometimes called neutropenia) and/or platelets (thrombocytopenia). These changes may lead to some of the symptoms of MF.

What are the most common symptoms of MF?

The different manifestations and symptoms of MF vary tremendously between different people and no two patients will share exactly the same features. While most patients are diagnosed having presented with symptoms, others experience few or no symptoms at all in the early stages of MF. In fact, many patients are diagnosed after having tests for an unrelated condition.

Symptoms of MF include:

- Fatigue
- Sweats (which may be predominantly at night)

Symptoms of Myelofibrosis (cont.)

- Itching (pruritus) - this may be worse after baths or showers
- Bone pain (arthralgia)
- Muscle pain (myalgia)
- Weight loss
- Fever

Enlargement of the spleen is common in MF, and may cause abdominal pain, discomfort, loss of appetite and a feeling of filling up quickly during meals (sometimes referred to by doctors as early satiety).



Diagnosis of Myelofibrosis

The diagnosis of MF may not be confirmed all at once, but will usually be made following a number of tests which may be done over a period of several visits to the haematology clinic. These tests may include:

- **Full Blood Count (FBC)** - This is a routine blood test which measures the number of red cells, different types of white cells and platelets in the blood. In the laboratory a blood film may be made, allowing the haematologist to look at the blood cells under the microscope. In many patients with MF, this reveals the presence of immature cells in the blood that are normally only seen in the bone marrow (this is called a leucoerythroblastic blood picture).
- **Bone marrow biopsy** - In most cases, a bone marrow biopsy will need to be taken to confirm the diagnosis of MF. A small amount of liquid bone marrow (aspirate) and bone marrow tissue (trepine) are taken from the hip bone using special

biopsy needles, allowing the haematologists to look at the cells under the microscope. This procedure is performed under local anaesthetic.

- **Tests for gene mutations** - Blood tests may be performed to check for mutations in genes such as JAK2, CALR and MPL. The presence of these may help the diagnosis and also provide prognostic information. Although other gene mutations are being linked to MF, these are generally not yet part of routine testing.
- **Abdominal ultrasound scan** - Often it will be easy for doctors to feel your spleen, but an ultrasound might be done in patients with a spleen that is less enlarged. This will also help to look for liver enlargement or abnormalities of other organs.

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 different people very differently, and an individual patient's prognosis may vary depending on a number of factors, such as age, blood count and symptoms. Therefore, life expectancy varies from person to person.

Your haematologist will be able to provide you with an accurate picture for your individual situation. Establishing the likely prognosis is important when planning the best approach to treatment.

Prognostic scoring

In recent years, international research has more clearly identified different factors that influence the prognosis in MF and this has led to the establishment of 'prognostic scoring systems' that can be used by doctors either at the time that the diagnosis of MF is made (International Prognostic Scoring System [IPSS]) or at later points in the course of the disease (Dynamic International Prognostic Scoring

System [DIPSS] or DIPSS Plus).

Generally, these scores allow MF to be categorised as 'low risk', 'intermediate-1', 'intermediate-2' or 'high risk'. The following factors may all influence prognosis and will be used by your doctor to calculate the various prognostic scores:

- The degree of anaemia (lowering in the number of red blood cells) and whether blood transfusions are needed to treat this
- 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
- Whether the white blood cell count is raised
- The presence of 'constitutional symptoms' such as fever, night sweats, or weight loss

The DIPSS Plus score also considers lowering of the platelet count, need for transfusion and whether 'cytogenetic changes'

Diagnosis of Myelofibrosis (cont.)

(changes in the chromosomes that contain the genetic material) are seen in the bone marrow.

The presence of certain genetic mutations is increasingly being considered when working out prognosis. 'Triple negativity' refers to the situation where JAK2, CALR and MPL mutations are all absent and this is associated with a less favourable prognosis. New mutations that may influence the prognosis of MF are being discovered by researchers all the time, and eventually are likely to be added routinely to prognostic scoring systems but this is not yet part of standard practice.

About 10 to 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.

It is important when you are reading up about MF to remember that in many patients the disease may remain stable or only progress very gradually over

time. This means that many of the symptoms and outcomes you read about may not happen to you. If you have any concerns, ask your haematologist.

If you would like more information about acute myeloid leukaemia (AML), you can order one of our booklets by calling Patient Advocacy on **08088 010 444** or emailing **support@leukaemiacare.org.uk**

Treating Myelofibrosis

The majority of treatments for MF are aimed at managing symptoms and reducing complications, so that your quality of life is better.

Watch and Wait

If there are no symptoms when you are first diagnosed, a 'watch and wait' approach is often recommended. This usually involves regular check-ups and blood tests, as well as your haematologist advising on ways to live a healthy lifestyle.

Our booklet on Watch and Wait tells you all you need to know. Get your copy by calling **08088 010 444**, emailing support@leukaemiacare.org.uk or downloading at www.leukaemiacare.org.uk/support-and-information/help-and-resources/information-booklets/

Treatment options for MF

Bone marrow or stem cell transplant

A bone marrow or stem cell transplant offers the only curative treatment for MF. You will be 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 (called graft-versus-host disease). Transplants are usually only considered an option for fit patients with advanced disease who have a matched donor. This may or may not be a relative. The first step (known as conditioning) involves very high levels of chemotherapy or radiation therapy.

Some people may have less intense doses of chemotherapy to weaken their immune system enough to allow the donor stem cells to grow in their bone marrow. This is called reduced intensity conditioning (RIC).

Treating Myelofibrosis (cont.)

Apart from stem cell transplantation, all other treatments used in MF are aimed at improving quality of life by controlling symptoms, reducing the size of the spleen or improving the blood count. In general terms, these can best be divided into two main groups of treatments:

1. Treatments that are used to reduce the size of the spleen and to improve constitutional symptoms, such as fever, night sweats, or weight loss.
2. Treatments that are used primarily to treat anaemia and to improve low blood counts.

In reality though, the manifestations of MF in individual patients may result in a mixture of these two scenarios and your doctor may need to combine treatments or work out with you which to prioritise.

Treatments used principally to manage splenomegaly and control constitutional symptoms of MF

An enlarged spleen

(splenomegaly) is a common symptom of MF, often leading to pain, discomfort and a feeling of fullness or a loss of appetite. Treatment options include the following:

Chemotherapy Hydroxycarbamide

Chemotherapy is the use of anti-cancer (cytotoxic) drugs to destroy cancer cells. Hydroxycarbamide is the most commonly used chemotherapy drug to treat MF and is taken daily as a tablet. It can be used to reduce spleen size and constitutional symptoms and to manage myeloproliferative features, such as raised white cell or platelet counts. Hydroxycarbamide can cause side effects, but generally these are mild. Side effects can include lowered resistance to infection, mouth and leg ulcers, reduced red blood cell numbers (anaemia), reduced white cell numbers (neutropenia), diarrhoea or constipation.

Hydroxycarbamide may also affect fertility so if you are taking it you will be advised not to get

pregnant or father a child, as there may be a risk of harming the developing baby. It is advisable to use effective contraception while taking the drug and also for a few months afterwards. If hydroxycarbamide is used either alone or in combination with other chemotherapy drugs over a long period of time, it could potentially increase the chance of the MF developing into a leukaemia.

JAK Inhibitors

Most patients with MF have over-activity of signalling through JAK proteins. In 2011, the first JAK inhibitor, known as ruxolitinib (Jakavi), was licensed for treating patients with MF. Ruxolitinib is given twice per day as a tablet and large clinical trials have found it to be effective at reducing the size of the spleen and reducing constitutional symptoms of MF compared to standard therapies. It has been approved by NICE to be used within the NHS to treat splenomegaly and constitutional symptoms in MF patients with intermediate-2 and high-risk disease. Since ruxolitinib has been approved, its use as a drug of first choice has greatly

increased.

Due to its mechanism of action many patients develop anaemia with ruxolitinib as the haemoglobin often falls by about 10 to 20% over the first few months of therapy and the platelet count is also reduced. This can limit the dose that is used for some patients, especially those who already have anaemia. Ruxolitinib can work quickly but when tablets are stopped or missed the symptoms and spleen (including an enlarged spleen) can come back quickly so it is important not to run out of tablets or stop taking them suddenly unless directed by your doctor.

Recently, trial data is starting to suggest that using ruxolitinib may prolong life for some patients with MF; this is the first time that a drug treatment has shown these effects in this disease.

Other important things to be aware of with ruxolitinib are that it can interact with other medicines and foodstuffs such as grapefruit and Seville oranges. Also, its use is associated with increased risk of infections;

Treating Myelofibrosis (cont.)

usually these are minor, but they can be more serious, such as shingles.

Several other JAK inhibitory drugs, such as pacritinib, fedratinib and momelotinib, are currently under development in clinical trials and may become more widely available over the next few years.

Other chemotherapy drugs (melphalan, busulphan, cladribine and radioactive phosphorous (P32))

These treatments are used relatively rarely in MF nowadays, but may sometimes help when other medications are not working or are causing side effects. Your haematologist will explain the potential benefits and risks of these medications.

Busulphan, melphalan and cladribine affect the bone marrow directly and can lead to a fall in blood counts. They are usually given in short courses with regular monitoring.

Radioactive phosphorous is

given as a single injection. It is a weak radioactive drug and is usually given in nuclear medicine /medical physics departments. Injections are needed only very infrequently.

Splenectomy (surgical removal of the spleen)

This may be considered if your enlarged spleen is painful, causing complications and not responding to other more standard treatments. However, as a major surgical procedure, it carries significant risks of complications including infections, bleeding and thrombosis (blood clots).

Radiotherapy

Radiotherapy or irradiation 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 beams such as x-rays. It usually provides temporary relief that lasts between three and six months, but can lead to prolonged spells of

anaemia and lowering of the white blood cell and platelet counts.

In some MF patients, the body tries to make blood in parts of the body other than the bone marrow and spleen. This is called extramedullary haematopoiesis and may be painful. Radiotherapy can be effective in these circumstances.

Treatments used for patients with anaemia

Anaemia is common in people with MF. Anaemia means having too few red blood cells. 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 involves the transfer of red blood cells from a compatible donor into your body. If you have symptoms caused by a low red blood cell count (anaemia), you can be given blood transfusions. This can increase red blood cell count and quickly reduce symptoms of anaemia, often within 24-hours. Blood transfusion is a relatively safe

procedure and does not 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. It is usually possible to have a blood transfusion as a day patient, but it may sometimes involve an overnight stay in hospital. Blood transfusions can be repeated as often as necessary, until your haematologist says otherwise.

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.

Erythropoiesis-stimulating agents (ESAs)

Erythropoietin (EPO) is a special type of protein called a growth factor that is normally made in the kidney and stimulates the bone marrow to make red blood cells. Erythropoietin can also be made outside the body and used as a treatment for anaemia in a range of diseases, including MF. There are four different types of EPO currently available – epoetin alpha, epoetin beta, epoetin zeta and epoetin theta, all sold under

Treating Myelofibrosis (cont.)

various brand names. Darbepoetin alpha is a longer acting form of EPO that can be given less frequently. Before starting EPO therapy, your doctor may first take a blood test to determine your baseline erythropoietin level as this influences the likelihood of responding to erythropoietin therapy; if, as is frequently the case, your own EPO level is high, this makes it less likely that EPO therapy will work.

Erythropoietin is usually given as an injection under the skin (subcutaneously), most often in the thigh or abdomen. You, or a person caring for you, can be taught how to give the injections so that you can continue the treatment at home. Alternatively, you may be given the injections by a district nurse or GP practice nurse.

Each person's reaction to treatment is different. Some people have very few side effects while others may experience more. Some of the most common side effects include headaches, high blood pressure and flu-like symptoms. If you notice any effects that are not listed here,

discuss them with your doctor or specialist nurse.

Thalidomide

Thalidomide may be used to treat MF, usually in combination with steroid tablets (prednisolone). It is generally used to treat anaemia, sometimes when this is occurring in combination with enlargement of the spleen. It is a type of biological therapy that affects the way the immune system works. It can be given on its own or in combination with steroid tablets. Thalidomide is taken as a tablet. The side effects can include feeling tired or drowsy, constipation and numbness or tingling of the hands and feet. If you feel drowsy, it is important not to drive.

Thalidomide can cause birth defects, so it should not be given to pregnant women. People taking thalidomide who are sexually active should use a barrier form of contraception.

Danazol

Danazol is a synthetic form of male hormones (androgens), taken as a tablet, that may be

used to help improve anaemia. Danazol therapy is usually used for a period of up to six months and only continued beyond this if it is helping to improve anaemia or reducing the number of blood transfusions. It can cause side effects such as fluid retention and male pattern hair growth. While taking it, it is important that liver tests are monitored.

Interferon Alpha

Interferons are proteins that can be used to treat MF. They occur naturally in our bodies and help us fight infection. They are proteins that can also be given as medications and are used to treat many types of disorders. In MF, interferon may be used to treat anaemia and also to treat some of the proliferative features of the disease, such as raised white cell/platelet counts and constitutional symptoms.

Interferons are made by a variety of drug companies and are known by several brand names. They are given as an injection just under the skin and suppress the production of blood cells and reduce spleen size. They also have the potential to reduce bone marrow fibrosis and itching.

Some people may not be able to tolerate the side effects of interferon, which include flu-like feelings, nausea, headaches, depression, liver and thyroid inflammation and diarrhoea. You should inform your haematologist or clinical nurse specialist if you are experiencing any of these side effects, as there are often ways of controlling them. Conventionally, interferon is injected three or more times per week. Pegylated interferon, which is a slow release weekly formulation, is now becoming more widely available and is often better tolerated with a lower rate of side effects.

New treatments and treatments on the horizon

Many new drugs - such as alternative JAK inhibitors used alone or in combination, and other agents, for example inhibitors of a pathway known as the Hedgehog pathway, telomerase inhibitors and histone deacetylase inhibitors - are being actively investigated in MF. There are likely to be many different treatment options available to patients in the years to come.



Living with Myelofibrosis

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

Emotional impact of MF

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

"There was a total overwhelming feeling of helplessness and being out of control of my normal everyday life. But I had to carry on regardless for everyone else."

Looking after you

You may want to make changes to your lifestyle to try to stay as well

as possible after your diagnosis and during treatment. Do not try to change too much at once. Adopting a healthy way of living is about making small, manageable changes to your lifestyle.

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

You can find more information about living well with leukaemia, (including diet, exercise, complimentary therapies and appearance) on our website: www.leukaemiacare.org.uk/support-and-information/information-about-blood-cancer/living-well-with-leukaemia/

Living with Myelofibrosis (cont.)

One of the most commonly reported side effects of the treatment of MF is fatigue. This is not normal tiredness and does not improve with sleep.

Some general tips on how to deal with fatigue include:

- Have a regular lifestyle – try going to bed and waking up approximately the same time every day and try to avoid lying in.
- Take part in regular, gentle exercise to maintain your fitness levels as much as possible.
- Reserve your energy for what you find important and build rest periods around those times.
- Before going to bed avoid stimulants such as alcohol, coffee, tea or chocolate, or using laptops, tablets or mobile phones.
- Keep your bedroom quiet and at a comfortable temperature.
- Talk about your worries with family, friends, your doctor or nurse, or patient support

groups.

- Discuss your fatigue with your doctor or nurse.

"Don't get me wrong, it's hard living with a chronic condition. I get tired a lot and have to be careful of picking up infections, as my immune system is lower than others. But all in all, I will not let it take over my life. I don't intend on fighting it; it will have to fight me."

Practical support Work and finances

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

Your consultant or your GP can arrange letters to your employer to confirm your diagnosis and the effects it may have on your work



Living with Myelofibrosis (cont.)

life. It is often worth taking time to explain MF to your employer, as it is likely they will never have heard of the disease.

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

Macmillan has published a booklet about financial support following a diagnosis of cancer. They can also give you personal advice over the phone via their helpline and you can discuss which benefits you are eligible for. Some Macmillan centres can arrange face-to-face meetings with a benefits advisor. They can also provide financial assistance in the form of grants – ask your nurse in the hospital how to apply.

As MF is regarded as a cancer, you will also be entitled to apply for a medical exemption certificate which means that you are entitled to free NHS prescriptions. Your GP

or specialist nurse at the hospital can provide you with the details on how to apply for this.

If you would like more information about some of the things you may be entitled to, you can speak to our Advocacy Officer by emailing advocacy@leukaemiacare.org.uk or by calling 08088 010 444.

Talking about Myelofibrosis

Talking to your haematologist

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

The following gives advice on working well with your haematologist:

- If it is an initial consultation, take along a list of your current medications and doses, and a list of any allergies you may have.
- If you have a complicated medical history, take a list of diagnoses, previous procedures and/or complications.
- Make a list of questions to take to your appointment. This will help the discussion with your haematologist.
- It can be useful to repeat back what you have heard so that you can be sure that you fully understood.
- Note information down to help

you remember what was said.

- Be open when you discuss your symptoms and how you are coping. Good patient-doctor communication tends to improve outcomes for patients.

Other tips:

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

You need to tell your haematologist if...

You are having any medical treatment or taking any products such as prescribed medicines, over the counter treatments or vitamins. It is important to understand that treatments, including complementary therapies, which are perfectly safe for most people, may not be safe if you are being treated for MF. Remember, if you choose to start any form of complementary therapy outside

Talking about Myelofibrosis (cont.)

of your medical treatment, consult your haematology consultant or clinical nurse specialist, prior to beginning it. It is important to understand the difference between complementary therapies, used alongside standard treatment, and alternative therapies, used instead of standard treatment. There is no evidence that any form of alternative therapy can treat MF.

For help with talking to your haematologist, you can access more information about MF at www.leukaemiacare.org.uk/support-and-information/information-about-blood-cancer/blood-cancer-information/about-myeloproliferative-neoplasms-mpn/myelofibrosis-mf/ which includes a list of questions which you may want to ask your medical team.

Talking to other people

Telling people you have a rare condition like MF can be hard to explain. You might find it useful to let your close family and friends, as well as your employer, know about your health condition. It

might be easier to provide people with basic information and give them information about MF if they want to know more in-depth details.

"I made a conscious decision to be very open about my illness. Telling family was tough. But I encouraged people to ask questions."

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

These points may help you:

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

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

- **Find out more** - Try to find out as much as you can about your condition, from reliable internet sources, charitable organisations or your consultant haematologist. The more you know, the more you can share.
- **Have a print-out to hand** - It may help to have some information to hand to share with family and friends. This will take the pressure off you having to remember everything they

may want to know.

- **Explain your needs** - Try and be clear about what your needs may be. Perhaps you need help with the weekly food shop, help with cooking dinner, or someone to drive you to and from appointments. You may find that friends and family are pleased that they can do something to help you.
- **Be open about how you feel** - Do not be afraid of opening up about how you feel, as people who care will want to help you as best they can. Talk as and when you feel comfortable, so those around you will know when you need them most.

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

Glossary

Acute myeloid leukaemia

A rapidly progressive malignant disease in which there are too many immature blood-forming cells in the blood and bone marrow.

Blood clot

Also referred to as thrombosis. These form when there is damage to the lining of a blood vessel, either an artery or a vein. Also, blood will begin to clot if it stops moving and becomes stagnant.

Bone marrow

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

Calreticulin

Calreticulin is a multifunctional protein in humans and is encoded by the CALR gene. The role of calreticulin is to organise where proteins are inside cells. Mutations of the gene encoding calreticulin have recently been discovered in approximately 20

to 25% of patients with MF and essential thrombocythaemia (ET).

Chemotherapy

The treatment of cancer using specific chemical agents or drugs that are selectively destructive to malignant/cancer cells and tissues.

Chronic

A human health condition or disease that is persistent or otherwise long-lasting in its effects. The term chronic is usually applied when the course of the disease lasts for more than three months.

Cytoreductive therapy

Therapy or treatment used to reduce the size of a cancerous tumour (sometimes also called debulking). Surgery, chemotherapy and radiation therapy are common cytoreductive treatments used to reduce/debulk tumours.

Deep Vein Thrombosis (DVT)

This is the formation of a blood clot in a deep vein, predominantly in the thigh or lower leg; a clot

inside a blood vessel is called a thrombosis. Non-specific signs may include pain, swelling, redness, and warmth.

Essential Thrombocythaemia (ET)

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

Fatigue

Extreme tiredness, typically resulting from mental or physical exertion or illness. 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 Count (FBC)

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.

Haematologist

A doctor who is specially trained in diseases of the blood and blood forming organs.

Haematology

This is the study of blood, the blood-forming organs, and blood diseases.

Hepatomegaly

An enlargement of the liver.

JAK2 (and JAK2V617F)

JAK2 is a molecule (called an enzyme) that exists in all people. It forms a communications pathway for messages travelling inside the cell which is particularly important in the coordination of blood cell production in the bone marrow.

JAK2V617F

Researchers in 2005 found a mutation (known as JAK2V617F) in the JAK2 molecule in people with MPNs. The mutation causes an increase in signalling through the JAK2 pathway.

Glossary (cont.)

MPL

This is a type of gene that provides instructions for making a protein which helps promote the growth and division of cells.

Myelofibrosis (MF)

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

Myeloproliferative Neoplasms (MPNs)

A group of diseases of the blood and bone marrow. Three main types of MPNs make up around 95%: myelofibrosis (MF), essential thrombocythaemia (ET) and polycythaemia vera (PV).

Myelosuppression

A condition in which bone marrow activity is decreased, resulting in fewer red blood cells, white blood cells, and platelets. Myelosuppression is a side effect of some cancer treatments.

Night sweats

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

Phlebotomy

Phlebotomy (or venesection). Obtaining blood from a vein, usually large amounts. This is sometimes used in the treatment of patients with a raised red cell count, such as is seen in polycythaemia vera.

Platelet pheresis

Procedure in which platelets are removed from the blood and the remaining components are returned to the body.

Platelet

A 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

A normal platelet count in a healthy individual is between 150,000 and 450,000 per microlitre of blood. In general, low platelet counts increase bleeding risks. High counts may lead to

thrombosis, although this is mainly when the elevated count is due to myeloproliferative disorder.

Polycythaemia Vera (PV)

PV is a myeloproliferative neoplasm (MPNs) that results from an overproduction of red blood cells, sometimes platelets and white cells can be increased too.

Pruritus

Another word for itching. Pruritus is a common symptom of myeloproliferative neoplasms and can also 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 closure of the vessel.

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 from the lungs to the body, and carbon dioxide from the body back to the lungs.

Spleen

Similar in structure to a large lymph node, the spleen acts primarily as a blood filter. As such, it is a non-vital organ, with life possible after removal. The spleen plays important roles in regard to red blood cells (also referred to as erythrocytes) and the immune system. It is 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.

Splenomegaly

Medical term for an enlargement of the spleen. This can be seen in all of the myeloproliferative neoplasms but is particularly

Glossary (cont.)

associated with MF.

Stem cells

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

Thrombocythaemia

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

Thrombocytopenia

Medical term for an abnormally low platelet count, below 150,000 per microliter.

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].

Venesection

See phlebotomy.

White blood cell (or leukocytes)

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 neutrophils and lymphocytes.

Tell us what you think!

If you would like to give us some feedback about this patient information booklet, please hover over the code to the right using your phone or tablet's camera. Click the link as it appears and this will take you to a short web form to fill in.

Suitable for Android, iPhone 7 and above.



Useful contacts and further support

There are a number of helpful sources to support you during your diagnosis, treatment and beyond, including:

- Your haematologist and healthcare team
- Your family and friends
- Your psychologist (ask your haematologist or CNS for a referral)
- Reliable online sources, such as Leukaemia Care
- Charitable organisations

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

Leukaemia Care

We are a charity dedicated to supporting anyone affected by the diagnosis of any blood cancer.

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

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

Bloodwise

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

020 7504 2200
www.bloodwise.org.uk

Cancer Research UK

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

0808 800 4040
www.cancerresearchuk.org

Macmillan

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

0808 808 0000
www.macmillan.org.uk

Maggie's Centres

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

0300 123 1801
www.maggiescentres.org

Citizens Advice Bureau (CAB)

Offers advice on benefits and financial assistance.

08444 111 444
www.adviceguide.org.uk

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

Around 34,000 new cases of blood cancer are diagnosed in the UK each year. We are here to support you, whether you're a patient, carer or family member.

Want to talk?

Helpline: **08088 010 444**

(free from landlines and all major mobile networks)

Office Line: **01905 755977**

www.leukaemiacare.org.uk

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Leukaemia Care
YOUR Blood Cancer Charity

