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# Essential Thrombocythaemia (ET)

**A Guide for  
Patients**

**Leukaemia Care**  
YOUR Blood Cancer Charity

# Introduction

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

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

This booklet was originally written and subsequently revised by Ken Campbell MSc (Clinical Oncology) and the review was conducted by Lisa Lovelidge. It was then peer

reviewed by Manos Nikolousis, Consultant Haematologist at Heart of England NHS Trust. The booklet has then been updated by our Patient Information Writer, Isabelle Leach, and reviewed by Dr. Mallika Sekhar, UCLH. We are also grateful to Linda Phyll, Sabine Lurcook, Rachel Worzenkraft, Sue Law, Dr Dick Morris, Paula Davis, Jeanette Bennett, Fiona Bridge, Claire Todd, Cath Owens and Lucy Geering for their valuable contributions as patient reviewers.

If you would like any information on the sources used for this booklet, please email [communications@leukaemiacare.org.uk](mailto: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 8:30am – 5:00pm Monday - Friday and 7:00pm – 10:00pm on Thursdays and Fridays. If you need someone to talk to, call **08088 010 444**.

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

### Nurse service

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

### Patient Information Booklets

We have a number of patient information booklets like this available to anyone who

has been affected by a blood cancer. A full list of titles – both disease specific and general information titles – can be found on our website at **www.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@leukaemiacare.org.uk**

### Online Forum

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

### Patient and carer conferences

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

### Website

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

### Campaigning and Advocacy

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

### Patient magazine

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

# What is essential thrombocythaemia (ET)?

ET is a chronic condition characterised by too many platelets in the blood. It belongs to a group of conditions called myeloproliferative neoplasms (MPNs), which also includes polycythaemia vera (PV) and myelofibrosis (MF).

If you would like more information about PV and MF, you can download our booklets from our website at [www.leukaemiacare.org.uk](http://www.leukaemiacare.org.uk) or request a copy of the booklets by emailing [support@leukaemiacare.org.uk](mailto:support@leukaemiacare.org.uk) or calling the helpline on 08088 010 444.

There has been some debate about whether or not MPNs are types of cancer. This is because the word neoplasm (which means new growth) is a term used for cancers (malignant neoplasms) and noncancerous tumours (benign neoplasms). In ET there is an uncontrolled increase in cells; therefore, many haematologists and cancer organisations do consider MPNs as blood cancers. This includes the World Health

Organisation who updated their classification in 2016 to include MPNs such as ET.

Blood cells which are produced from stem cells in the bone marrow include:

1. Red blood cells (to carry oxygen to the tissues of your body)
2. White blood cells (to fight infection and disease)
3. Platelets (to help blood to clot)

Production of new blood cells is very closely controlled in the bone marrow so that it is balanced with the loss of worn-out cells or cells lost by bleeding or damage. In people with ET, there is an excess of platelets. Although it is platelets that are primarily affected, white blood cell levels may also be elevated.

In patients with ET, the blood becomes thicker than normal and the excess number of platelets may cause blood clots to form more easily. Clots can block blood flow through veins and arteries, potentially leading to heart attacks or strokes.

ET is also associated with an increased risk of bleeding

complications.

In some cases, ET can transform to MF or acute myeloid leukaemia (AML).

## Who is affected by ET?

ET is considered to be a rare disease. The number of people diagnosed with ET in Europe is 0.6 per 100,000 patients per year. This may explain why you might not have heard of ET or met anyone with the condition before.

ET is most common in those aged over 60 years.

It is more common in women than men, with two women being diagnosed for every man, particularly in patients who are less than 60 years of age. However, it must be noted that 20% of patients are older than 40 years of age at diagnosis.

## What causes ET?

While the exact cause of ET is not known, research has found that approximately 55% of people who have ET have a mutation in a protein that regulates blood cell production. This protein is known as Janus Kinase 2 (JAK2) and the mutation itself is JAK2 V617F. This mutation arises during

your lifetime and is not present at birth.

In addition, a mutation in a gene known as CALR is present in 15% to 30% of patients with ET, and the MPL mutation is seen in 4% to 8% of patients.

Some 10% to 20% of patients with ET do not express any of these three mutations. This is referred to as triple-negative.

It is also important to be aware that, although some families seem to develop the disease more readily than others, ET is usually not inherited nor passed on from parent to child. However, and this is less common, an increased likelihood of the gene mutating (which can then lead to developing ET) can be inherited and is called familial ET. It resembles non-familial ET both in terms of clinical symptoms and mutations.

Finally, some researchers believe MPNs may also be triggered by past exposure to ionising radiation (a type of radiation that has very high energy, like medical x-rays or nuclear fallout) or to some chemical substances such as benzene and toluene.

# Symptoms of ET

Many patients with ET do not have any symptoms when they are diagnosed. Typically, these patients are identified following abnormal results during a routine full blood test for something else. If symptoms do develop, they tend to do so over time.

If you have persistent symptoms of ET, make an appointment to see your GP. Common symptoms of ET include:

- Fatigue
- Night sweats
- Headaches (this can also include migraines with visual disturbances)
- Itching
- Fever
- Swollen spleen (located under the ribs on the left of the abdomen, this organ helps to rid the body of toxins, waste and other unwanted materials)
- Bone pain
- Weight loss
- Dizziness or light headedness

- Reddish or purple skin
- Bleeding or clotting
- Unexplained bruising

This is not an exhaustive list of symptoms. There is a wide variety of symptoms that can be attributed to a diagnosis ET, so it is important to make your doctor aware of these. Not everyone will have the same symptoms or to the same degree of severity. It is also important to understand that the presence of these symptoms does not always indicate ET and they could be due to other conditions and illnesses.

In patients with ET, the increased numbers of platelets and white blood cells can result in blood clots (thrombosis) in a vein or artery, or they may cause bleeding (haemorrhage).

Blood clots or bleeding are a major complication of ET. There is a 1% to 3% risk of arterial or venous blood clots occurring in a patient per year; however, if the patient has a JAK2 mutation, this risk increases to 7.7%.

The blood clots prevent the

blood from flowing and the most common complications caused by these blood clots are:

- Heart attacks, strokes or damage to the gastro-intestinal tract due to blood clots in the arteries.
- Venous thrombosis such as deep vein thrombosis in the calf.
- Pulmonary embolisms, where a clot in the vein travels through the blood stream and causes a blockage in one of the arteries in the lungs.

The risks of bleeding are less clearly defined, mostly because the definition of bleeding in various studies is different. Risks of bleeding are known to be increased if the platelet count is very high (between  $1000 \times 10^9/L$  and  $1500 \times 10^9/L$ ). It could also be related to acquired von Willebrand syndrome with extreme levels of platelets.

If you have experienced some bleeding, you may be screened for acquired von Willebrand syndrome. If the test is negative,

you will then have the functioning of your platelets tested. Your haematologist will closely monitor any antiplatelet drugs or anticoagulant drugs you may be receiving. Any significant bleeding episodes will be managed with tranexamic acid, which stops bleeding in the short term, or a transfusion of platelets.

General risk factors that can increase your likelihood of blood clots or bleeding include:

- Being older than 60 years
- Previous blood clots or bleeding
- A high platelet count
- JAK2 mutations
- Having a cardiovascular risk factor, such as high blood pressure, diabetes, smoking or high cholesterol

Enlargement of the spleen is present in 10% to 20% of patients with ET at diagnosis.

# How is ET diagnosed?

ET is often suspected if a routine blood test shows that a patient has a high platelet count, also called thrombocytosis.

To make a diagnosis of ET, the following criteria must be present:

- A platelet count of  $450 \times 10^9/L$  or higher
- A bone marrow biopsy showing an increase in the numbers of enlarged, mature megakaryocytes (cells in the bone marrow that produce platelets)
- Not meeting the diagnosis for another MPN, such as PV or MF
- The presence of a JAK2, CALR or MPL mutation

If none of the mutations are present, other causes of primary or secondary thrombocytosis as well as diagnoses of PV and MF must be excluded. Primary refers to a condition that is the root cause of the illness, whereas secondary refers to a condition that has developed as a result of another one.

ET is diagnosed using laboratory

tests including:

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

# What is the prognosis of ET?

Treatment is focussed mainly on preventing any complications from blood clots or bleeding which is associated with ET. Low-risk patients are best managed with low-dose aspirin or watch and wait alone, and patients with high-risk ET will benefit mainly with cytoreductive therapy and anticoagulation.

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

For the majority of patients whose ET does not progress to MF or AML, a normal or very slightly reduced life expectancy can be anticipated.

Having the mutations JAK2, CALR

or MPL does not affect the survival of patients with ET. However, older age, a high white blood cell count and a previous history of blood clots and anaemia are associated with a poor survival.

In some patients, ET can develop into MF or AML. However, this is rare. Transformation to AML risk factors include:

- Advanced age
- Leucocytosis (a high white blood cell count)
- Exposure to previous treatments which suppress the bone marrow, such as chemotherapy

The prognosis of patients with ET can vary widely. Your haematologist is the best person to advise you based on your individual circumstances.

# Treating ET

## Overview of treatment

Most treatments for ET are intended to manage your symptoms and prevent any associated problems from blood clots or bleeding in order to maintain your quality of life.

Treatments for ET are aimed at lowering the production of platelets and maintaining normal blood counts. This is called cytoreductive therapy. There are a number of medications which can achieve this.

## Treatment according to risk factors

Treatment will be based on an assessment of your risk factors regarding blood clots and bleeding. While cytoreductive therapy is fundamental in treating ET, managing any potential blood clots or bleeding is equally important. Your medical team will give you all the information about the treatment which is best for you.

Low-dose aspirin and anticoagulants may be used to prevent any blood clots, especially if you have a history of developing

blood clots.

## Medications

### Cytoreductive therapy

Cytoreductive therapy is important in enabling ET and its complications to be controlled. It is the only treatment significantly associated with reducing the occurrence of blood clots. In patients with ET, first-line treatment is hydroxycarbamide, with second-line treatment being anagrelide.

### Hydroxycarbamide (also known as hydroxyurea)

This is the most commonly used chemotherapy drug to treat ET and is available as a tablet or capsule.

An early study of patients with high-risk ET confirmed that the percentage of patients receiving hydroxycarbamide had significantly less blood clots or bleeding (3.6%) compared with patients not receiving any treatment (24%). In recent controlled studies of patients with high-risk ET, neither anagrelide nor interferon alpha could be shown to be superior to

hydroxycarbamide for reducing blood clots or bleeding.

Hydroxycarbamide can cause mild side effects such as:

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

Hydroxycarbamide may also affect fertility. If you are taking it, you will be advised not to get pregnant or father a child, as there may be a risk of harming the developing baby. It is advisable to use effective barrier 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 may slightly increase the chance of the ET

developing into AML.

### Anagrelide

Taken as a capsule, anagrelide is a drug that prevents the maturation of platelets. It is used to counter the overproduction of platelets. It lowers the platelet count and has some effect on the red blood cells.

Anagrelide is approved for reducing elevated platelet counts in at-risk patients with ET who are intolerant to their current therapy or whose elevated platelet counts are not reduced to an acceptable level by their current therapy.

Side effects of anagrelide include:

- Headaches
- Diarrhoea
- Palpitations
- Fluid retention

### Interferon alpha

Interferon alpha is a substance which occurs naturally in the body and reduces the rate at which blood cells, including platelets, are made in the bone marrow. It can be made into a medicine to be given as an injection under the skin to treat a wide range of

# Treating ET (cont.)

conditions, including ET.

Side effects of interferon alpha include:

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

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

## Busulfan

Busulfan is usually given to patients who have side effects when taking hydroxycarbamide. It can be given as a tablet. Like hydroxycarbamide, busulfan affects the bone marrow directly and can lead to a fall in the blood cell counts. Its main side effects are nausea and low platelet counts.

Your haematologist can advise you if busulfan is suitable, and will monitor your progress carefully during your treatment. There is evidence that busulfan can increase the risk of secondary leukaemia and can cause lung

scarring.

## New treatments

- **JAK2 inhibitors** – JAK2 inhibitors block the function of the JAK2 mutation which slows down blood cell production, reduces spleen size and improves symptoms. Up to 55% of patients with ET have been found to have a JAK2 mutation. JAK2 inhibitors are being looked into as a treatment for ET.

## Prevention of blood clots and bleeding

### Aspirin

As well as being used for reducing pain and lowering temperature, aspirin is known to prevent platelets sticking together and may reduce your risk of developing a blood clot.

In patients with very low-risk ET, aspirin might be required. In patients with low-risk ET, treatment with low-dose aspirin is advised, especially if the patient has cardiovascular risk factors, as it has been shown to reduce both venous and arterial blood clots.

Research shows that aspirin is very effective at reducing risks of

heart attacks and strokes in many people with different levels of risk. Patients with intermediate-risk ET and high-risk ET will be given low-dose aspirin whether they have cardiovascular risk factors or not.

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

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

### Anticoagulation treatment

Long-term anticoagulation treatment uses medication, also referred to as blood-thinners, to help prevent blood clots. This can be started if you have had

a venous blood clot that has occurred with no identifiable risk factor. This will be done after your bleeding risk has been assessed.

Patients with ET and other MPNs more commonly have unusual sites of venous clots, meaning that the use of anticoagulation treatment can be particularly beneficial for them. These unusual sites of venous clots include:

- **The splanchnic vein** – this drains the blood from the stomach, pancreas, spleen and intestines.
- **The cerebral vein** – this drains blood from the brain.

# Living with ET

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

## Emotional impact of ET

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

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

### Looking after you

You can live a long and normal life with ET, but you may want to

make changes to your lifestyle to try to stay as well as possible after your diagnosis and during treatment. Don't try to change too much at once. Adopting a healthy way of living is about making small, manageable changes to your lifestyle.

A healthy lifestyle includes having a well-balanced diet and being physically active. With some of the side effects you may be experiencing, the idea of getting out and being active may be the last thing you want to do, 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.

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

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

Some general tips on how to deal with fatigue include:

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

- Discuss your fatigue with your doctor or nurse.

## Practical support

### Work and finances

Being diagnosed with ET 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 for your employer to confirm your diagnosis and the effects it may have on your work life. It is often worth taking time to explain ET to your employer, as it is likely they will never have heard of the disease before.

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

# Living with ET (cont.)

arrangements and adjustments relating to your disease.

If you would like advice about some of the financial help available to you, then you can speak to our Patient Advocacy team on **08088 010 444**.

Alternatively, Macmillan has published a booklet about financial support following a diagnosis of cancer that might be useful to you. They can also give you personal advice over the phone via their helpline at **0808 808 000** 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 ET 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 in England and Wales. Your GP or specialist nurse at the hospital can provide you with the details of how to apply for this.



# Talking about ET

## Talking to your haematologist

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

Here are some tips for 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 have fully understood.
  - Note down information to help you remember what was said.
- Be open and honest when you discuss your symptoms and how you are coping. Good patient-doctor communication tends to improve outcomes for patients.
  - Bring someone along to your appointment. They can provide support, ask questions and take notes.
  - Don't be afraid to ask for a second opinion – most haematologists are happy for you to ask.

## You need to tell your haematologist if...

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

## Talking to other people

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

It is probably best to focus conversations on the symptoms that you are experiencing, how the condition affects you and how you feel about it. Often people misunderstand and, unfortunately, it will mostly fall to you to educate them as best as you can. Where possible, it's advisable to let people know what you find helpful and unhelpful, in

terms of what others say and do. Often people make assumptions and do what they think helps. For example, saying you look well, recounting stories of others they know with a similar diagnosis or encouraging you to look ahead and stay positive, isn't always what people really want to hear.

In many ways, the more you communicate with them the better. However, it is perfectly natural if you feel at some points that you would like to keep some things to yourself.

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).

# Talking about ET (cont.)

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

- **Find out more** – Try to find out as much as you can about your condition from reliable internet sources, charitable organisations or your consultant haematologist. The more you know, the more you can share.
- **Have a print out to hand** – It may help to have a factsheet, or a booklet like this one, 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** – Don't be afraid of opening up about how you feel, as people

who care will want to help you as best as 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 (AML)

A rapid and aggressive cancer of the myeloid cells in the bone marrow.

## Anaemia

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

## Anticoagulation

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

## Bone Marrow

A 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 (CALR)

A soluble protein which is known to be involved in many body cell

processes including regulation of calcium, cell adhesion and gene expression. Mutations in CALR are common in ET and MF.

## Chromosomes

Thread-like structures which carry the genes, and are located in the nuclei of every cell in the body. There are 46 chromosomes (23 pairs) in humans.

## Chronic Myeloid Leukaemia (CML)

A leukaemia in which the myeloid cells start multiplying in the bone marrow leading to large numbers of abnormal, immature myeloid cells called blasts, which prevent the bone marrow from producing enough healthy blood cells of all types.

## First-line Treatment

The first treatment given for a disease. It is generally the treatment accepted by the medical establishment for initial treatment of a given type and stage of cancer.

# Glossary (cont.)

## Genes

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

## Irradiation

Particles or rays falling on to a surface (radiation wave on the surface of the skin).

## Leukaemia

A group of cancers that usually begin in the bone marrow and result in high numbers of abnormal blood cells. These cells are not fully developed and are called blasts or leukaemia cells. Depending on the type of blood cell involved, there are different types of leukaemia with varying characteristics, such as being acute (develops quickly) or chronic (develops slowly).

## Myeloproliferative Neoplasms (MPNs)

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

## Myeloid

Relates to bone marrow.

## Myelofibrosis (MF)

A reactive and reversible process which occurs with many cancerous and non-cancerous diseases of the bone marrow.

## Platelets

One of the types of blood cells which help to stop bleeding.

## Polycythaemia Vera (PV)

A chronic condition belonging to the myeloproliferative neoplasms group of diseases. It is characterised by too many red blood cells, and sometimes too many platelets and white cells, in the blood.

## Prognosis

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

## Pulmonary Embolism

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

## Red Blood Cells

Small blood cells that contain haemoglobin and carry oxygen and other substances to all tissues of the body.

## Second-line Treatment

Treatment other than the type used the first time (first-line treatment).

## Spleen

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

## Stem Cell

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

## Thrombocytosis

A condition in which there is an

excessive number of platelets in the blood (a platelet count greater than  $450 \times 10^9/L$ ). Platelets are blood cells in plasma that stop bleeding by sticking together to form a clot. Too many platelets can lead to certain conditions, including stroke, heart attack, or a clot in the blood vessels.

## Thrombosis

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

## von Willebrand syndrome

A common hereditary blood-clotting disorder which causes varying degrees of bleeding. It can be hereditary or acquired. In acquired von Willebrand syndrome, the patient has auto antibodies to the von Willebrand clotting factor and this often occurs in conjunction with another autoimmune disease such as rheumatoid arthritis.

## White blood cells

White blood cells are one of the types of cells found in the blood and bone marrow, along with red

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## Glossary (cont.)

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

# Useful contacts and further support

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

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

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

## Leukaemia Care

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

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

Helpline: **08088 010 444**  
**[www.leukaemicare.org.uk](http://www.leukaemicare.org.uk)**  
**[support@leukaemicare.org.uk](mailto:support@leukaemicare.org.uk)**

## Blood Cancer UK

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

**0808 2080 888**  
**[www.bloodcancer.org.uk](http://www.bloodcancer.org.uk)**

## Cancer Research UK

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

**0808 800 4040**  
**[www.cancerresearchuk.org](http://www.cancerresearchuk.org)**

## Macmillan

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

**0808 808 0000**  
**[www.macmillan.org.uk](http://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](http://www.maggiescentres.org)**

## Citizens Advice Bureau (CAB)

Offers advice on benefits and financial assistance.

**08444 111 444**  
**[www.adviceguide.org.uk](http://www.adviceguide.org.uk)**

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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.leukaemicare.org.uk](http://www.leukaemicare.org.uk)**

**[support@leukaemicare.org.uk](mailto:support@leukaemicare.org.uk)**

Leukaemia Care,  
One Birch Court,  
Blackpole East,  
Worcester,  
WR3 8SG

Leukaemia Care is registered as a charity in England and Wales (no.1183890) and Scotland (no. SC049802).  
Company number: 11911752 (England and Wales).  
Registered office address: One Birch Court, Blackpole East, Worcester, WR3 8SG

**Leukaemia Care**  
YOUR Blood Cancer Charity