Myeloproliferative Neoplasms (MPN)

A guide for patients, families & whānau

leukaemia & blood cancer new zealand

our mission is to care, our vision is to cure

Myeloproliferative Neoplasms

Formerly called Myeloproliferative Disorders (MPD)

A guide for patients, families & whānau

The term 'Myeloproliferative Neoplasms' is the general name given to a group of blood conditions. The following are discussed in this booklet:

- Polycythaemia Vera (PV)
- Essential Thrombocytosis (ET)
- Primary Myelofibrosis (PMF)
- Chronic Eosinophilic Leukaemia (CEL)
- Chronic Neutrophilic Leukaemia (CNL)
- Mastocytosis

There is a separate Leukaemia & Blood Cancer New Zealand booklet entitled 'Chronic Myeloid Leukaemia – a guide for patients, families & whānau' available from Leukaemia & Blood Cancer New Zealand.

Contents

Introduction

This booklet has been written to help you and your family or whānau understand more about myeloproliferative neoplasms (MPN).

If you or someone you care for has been diagnosed with a myeloproliferative neoplasm, you may be feeling anxious, or a little overwhelmed and this is normal. Perhaps you have already started treatment, or you are discussing different treatment options with your doctor and your family. Whatever point you are at, we hope that the information contained in this booklet is useful in answering some of your questions. It may raise other questions, which you should discuss with your doctor or specialist nurse.

You may not feel like reading this booklet from cover to cover. It might be more useful to look at the list of contents and read the parts that you think will be of most use at a particular point in time.

We have used some medical words and terms that you may not be familiar with. Their meaning is either explained in the text, in the glossary of terms at the back of this booklet, or in the 'Dictionary of Terms' booklet available from Leukaemia & Blood Cancer New Zealand.

Some people may require more information than is contained in this booklet. We have included some internet addresses that you might find useful. In addition, many of you will receive written information from the doctors and nurses at your treatment centre.

It is not the intention of this booklet to recommend any particular form of treatment to you. You need to discuss your circumstances at all times with your doctor and treatment team.

We hope that you find this booklet useful. There is a feedback form at the back of this booklet, please feel free to fill this in and return it to us to assist in the production of future editions.

Acknowledgements

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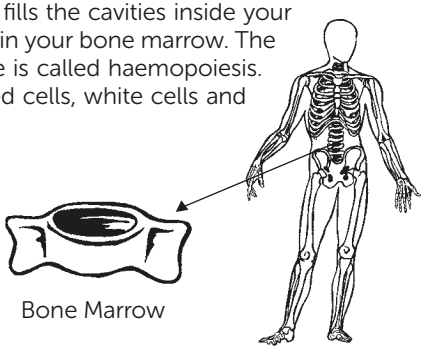
Bone marrow, stem cells & blood cell formation

Bone marrow

Bone marrow is the spongy tissue that fills the cavities inside your bones. All of your blood cells are made in your bone marrow. The process by which blood cells are made is called haemopoiesis. There are three types of blood cells; red cells, white cells and platelets.

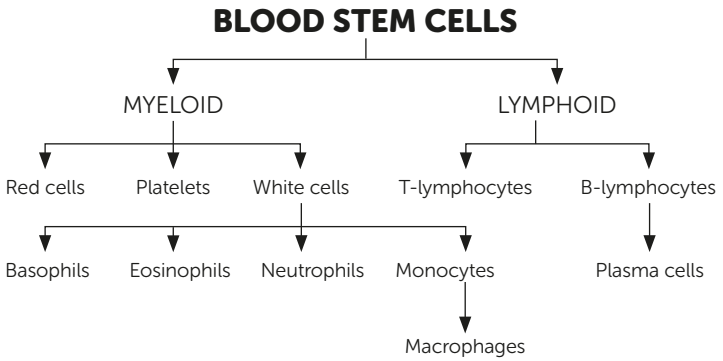


As an infant, haemopoiesis takes place in the centre of all bones. As an adult, fewer new blood cells are needed - the marrow space in the arms and legs is replaced by fat, and active bone marrow is limited to the hips, pelvis, ribs, breastbone (sternum) and back bones (spine or vertebrae).

You might like to think of the bone marrow as the blood cell factory. The main workers at the factory are the blood stem cells. They are relatively few in number, but are able not only to replicate themselves, but also to grow and divide into slightly more mature stem cells called myeloid or lymphoid stem cells. In turn, these can multiply and mature further to produce all the circulating blood cells.

Myeloid ('my-loid") stem cells develop into red cells, white cells (neutropils, eosinophils, basophils and monocytes) and platelets.

Lymphoid ('lim-foid') stem cells develop into two other types of white cells called T-lymphocytes and B-lymphocytes.



Blood

Blood consists of blood cells and plasma. Plasma is the straw-coloured fluid part of the blood which contains important proteins, such as clotting factors. Plasma carries these essential nutrients, including blood cells, around the body.

Blood cells

Red cells and haemoglobin

Red cells contain haemoglobin (Hb) which transports oxygen from the lungs to all parts of the body. Haemoglobin also carries carbon dioxide to the lungs where it can be breathed out. Haemoglobin is what makes red blood cells red.

The normal haemoglobin range for a man is between 130 - 170 g/L

The normal haemoglobin range for a woman is between 120 - 160 g/L

Red cells are by far the most numerous blood cells and the proportion of the blood that is occupied by these is called the haematocrit. A low haematocrit suggests that the number of red cells in the blood is lower than normal.

The normal haematocrit range for a man is between 0.40 – 0.52 (40 – 52%)

The normal haematocrit range for a woman is between 0.36 – 0.46 (36 – 46%)

Anaemia is a condition caused by a reduction in the haemoglobin level of the red blood cells. The severity of the anaemia relates to the level of measured haemoglobin. The lower the level of haemoglobin, the greater the severity of anaemia. If you are anaemic you may feel run down and weak. You may be pale and short of breath, or you may tire easily because your body is not getting enough oxygen. In this situation a red blood cell transfusion may be given to increase the red cell numbers and therefore the haemoglobin level.

White cells

White cells, also known as leucocytes, fight infection. There are different types of white cells which fight infection together and in different ways.

Neutrophils	kill bacteria and fungi
Eosinophils	kill parasites
Basophils	work with neutrophils to fight infection
T-lymphocytes	kill viruses, parasites and cancer cells; produce cytokines
B-lymphocytes	make antibodies which target microorganisms
Monocytes	work with neutrophils and lymphocytes to fight infection; they also help with antibody production and act as scavengers to remove dead tissue. These cells are known as monocytes when they are found in the blood and macrophages when they migrate into body tissues to help fight infection

If your white cell count drops below normal you are at risk of infection.

The normal adult white cell range is between 4.0 – 11.0 x 10^9/L

Neutropenia is the term given to describe a lower than normal neutrophil count. If you have a neutrophil count of less than 1.0 (1.0 x10^9/L) you are considered to be neutropenic and at risk of developing frequent and sometimes severe infections.

The normal adult neutrophil range is between 2.0 – 7.5 x 10^9/L

Platelets

Platelets are disc-shaped cellular fragments that circulate in the blood and play an important role in clot formation. They help to prevent bleeding. If part of your body is injured or cut or a blood vessel is damaged, the platelets gather at the site of the injury, stick together and form a plug to help stop the bleeding.

The normal adult platelet range is between 150 - 400 x 10^9/L

Thrombocytopenia is the term used to describe a low platelet count. If your platelet count is low, you are at higher risk of bleeding and will tend to bruise more easily. Platelet transfusions are sometimes given to bring the platelet count back to a higher level. In certain situations, especially when patients are receiving treatment, platelets may be transfused if the blood level falls below 10 x 10^9/L.

The normal blood counts provided here may differ slightly from the ones used at your treatment centre. You can ask for a copy of your blood results, which should include the normal range for each blood cell type.

Children

In children, some normal blood cell counts vary with age. If your child is being treated for a myeloproliferative neoplasm you can ask your doctor or nurse for a copy of their blood results, which should include the normal values for each blood type for a male or female child of the same age.

Growth factors and cytokines

All normal blood cells have a limited survival in the circulation and need to be replaced on a continual basis. This means that the bone marrow remains active tissue throughout your life. Natural chemicals in your blood called growth factors or cytokines control the process of blood cell formation. Different growth factors stimulate the blood stem cells in the bone marrow to produce different types of blood cells.

Many growth factors can be made in the laboratory (synthesised) and are available for use in people with blood disorders. For example, granulocyte-colony stimulating factor (G-CSF) stimulates the production of white cells called neutrophils, while erythropoietin (EPO) stimulates the production of red cells. Unfortunately, drugs to stimulate platelet production has been less successful, but research is continuing in this area.

Which Doctor?

If your general practitioner (GP) suspects that you might have a myeloproliferative neoplasm you will be referred to a haematologist for further tests and treatment.

A haematologist is a doctor who specialises in the care of people with diseases of the blood, bone marrow and immune system.

What are myeloproliferative neoplasms?

'Myelo' is the Greek word for marrow and 'proliferative' is another word for growing or reproducing. Myeloproliferative Neoplasms are a group of disorders in which the bone marrow cells grow and reproduce abnormally. In myeloproliferative neoplasms abnormal bone marrow stem cells produce excess numbers of one or more types of blood cells (red cells, white cells and/or platelets). These abnormal cells cannot function properly and can cause serious health problems unless properly treated and controlled.

It is important to remember, as you read through this booklet, that myeloproliferative neoplasms are chronic diseases that in most cases, remain stable for many years or progress gradually over time. The symptoms and complications of myeloproliferative neoplasms described in this booklet do not occur in everyone, and may not occur for many years.

Types of myeloproliferative neoplasms

Myeloproliferative neoplasms are usually described according to the type of blood cell which is most affected. There are four main types of myeloproliferative neoplasms that together represent around 95 per cent of all cases:

1. **Chronic myeloid leukaemia** – too many white cells
2. **Polycythaemia vera (PV)** – too many red cells
3. **Essential thrombocythaemia (ET)** – too many platelets
4. **Idiopathic myelofibrosis** – bone marrow tissue is replaced by fibrous scar-like tissue. This disrupts normal blood cell production.

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Myeloproliferative neoplasms are closely related diseases, so it's not uncommon for people to have features of more than one myeloproliferative neoplasm when they are first diagnosed, or during the course of their illness. In some cases, one neoplasm may transform over time to another, or to a type of leukaemia called acute myeloid leukaemia (AML).

Less common types of myeloproliferative neoplasms include:

- **Chronic neutrophilic leukaemia (CNL)** – too many neutrophils (a type of white cell) in blood and bone marrow
- **Chronic eosinophilic leukaemia (CEL) / hypereosinophilic syndrome** – too many eosinophils (another type of white cell) in blood and bone marrow
- **Chronic myelomonocytic leukaemia (CMML)** – too many monocytes (a type of white cell) in blood and bone marrow. CMML also has features of myelodysplasia, another blood stem cell disorder characterised by abnormal blood maturation.
- **Systemic mastocytosis** – too many mast cells (a type of white cell) in blood, bone marrow, skin and other tissues
- **Myeloproliferative disease** – unclassifiable

What causes myeloproliferative neoplasms?

The exact cause of myeloproliferative neoplasms remains unknown but there are likely to be a number of factors involved. Myeloproliferative neoplasms are sometimes described as being clonal blood stem cell disorders. This means that they result from a change, or mutation, in the DNA (genetic code) of a single blood stem cell. This change (or changes) results in abnormal blood cell development and in this case the overproduction of blood cells. In myeloproliferative neoplasms the original mutation is preserved when the affected stem cell divides (proliferates) and produces a 'clone'; a group of identical stem cells all with the same defect. Mutations in dividing cells occur all the time and healthy cells have sophisticated mechanisms within them to stop these abnormalities persisting. But the longer we live, the more chance we have of acquiring mutations that manage to escape these safe-guards. That's why myeloproliferative neoplasms, like most leukaemias and other cancers, become more common as we get older.

A mutation of a particular gene (a segment of DNA that makes proteins) known as Janus kinase 2 (JAK 2) is found in a large proportion of people with myeloproliferative neoplasms. The exact effect of this mutation remains unclear but it appears to play a role in the overproduction of blood cells seen in these disorders. A better understanding of how JAK2 gene influences blood formation is important because it may have a significant impact on the way myeloproliferative neoplasms are treated in the future.

Finally, myeloproliferative neoplasms are not contagious; you cannot 'catch' the disorders by being in contact with someone who has one. Most people with a myeloproliferative neoplasm have no family history of the disease.

Polycythaemia (rubra) vera

Polycythaemia (rubra) vera, also known as primary polycythaemia vera, is a disorder in which too many red cells are produced in the bone marrow, without any identifiable cause. These cells accumulate in the bone marrow and in the blood stream where they increase the blood volume and cause the blood to become thicker, or more 'viscous' than normal. In many people with polycythaemia vera, too many platelets and white cells are also produced.

Polycythaemia vera is a rare chronic disease diagnosed in an estimated 2 to 3 per 100,000 population. Although it can occur at any age, polycythaemia vera usually affects older people, with most patients diagnosed over the age of 55 years. Polycythaemia vera is rare in children and young adults. It occurs more commonly in males than in females.

What is secondary or reactive polycythaemia?

In secondary or reactive polycythaemia, red cell production is increased in response to excess amounts of erythropoietin (a red cell growth factor) circulating in the bloodstream. High levels of erythropoietin can be a response to lower than normal levels of oxygen in the blood (for example at high altitudes, in heavy smokers and in people with heart or lung disease). This is a useful compensatory mechanism that helps the body to produce more red cells and haemoglobin to transport more oxygen around the body. Erythropoietin levels, and therefore red cell production may also be increased abnormally in some types of kidney disease and in some types of cancer.

In a condition known as relative, apparent or spurious polycythaemia, the volume of plasma (the liquid portion of the blood) is reduced, usually as a result of dehydration, vomiting or diuretic (fluid loss) therapy. This increases the concentration of red cells in the blood, but the actual red cell mass (the total number of red cells) remains normal.

Symptoms of polycythaemia vera

Many people have no symptoms when they are first diagnosed with polycythaemia vera and the disease is picked up incidentally during a routine blood test or physical examination. When symptoms do occur, they usually develop gradually over time. They are mainly due to the increased thickness (hyperviscosity) and abnormally high numbers of blood cells in the circulating blood. Common symptoms include:

- Headaches
- Blurred vision
- Fatigue
- Weakness
- Dizziness
- Itchiness (pruritus), especially after a hot bath
- Night sweats

Complications of polycythaemia vera

Enlargement of the spleen (splenomegaly) is also common and occurs in around 75 per cent of cases. Symptoms include feelings of discomfort, pain, or fullness in the upper left-side of the abdomen. An enlarged spleen may also cause pressure on the stomach causing a feeling of fullness, indigestion and a loss of appetite.

In some cases the liver may also be enlarged. This is called hepatomegaly.

Some people experience gout, which usually presents as a painful inflammation of the big toe or foot. This can result from a buildup of uric acid, a byproduct of the increased production and breakdown of blood cells.

Some individuals may develop erythromelalgia, a rare condition that primarily affects the feet and, less commonly, the hands. It is characterised by intense, burning pain of affected extremities, and increased skin temperature that may be episodic or almost continuous in nature.

In many cases, people with polycythaemia vera have a ruddy (red) complexion, and a reddening of the palms of the hand and soles of the feet, ear lobes, mucous membranes and the eyes. This is due to the high numbers of red cell in the circulation.

A raised blood pressure (hypertension) is also common.

As the blood is thicker than normal it cannot flow as easily, especially through the smaller blood vessels. If left untreated, this increases the risk of thrombosis, the formation of a blood clot within a blood vessel. Blood clots can form in various parts of the body including the deep and superficial veins of the legs, in the heart (causing a myocardial infarction or heart attack) and in the brain (causing a stroke). Blood clots are a common complication of polycythaemia vera and occur in around 30 per cent of people, even before they are diagnosed. Older people and those with a history of a previous blood clot are at increased risk. A major aim of treatment in polycythaemia vera is to maintain a normal blood count and reduce your risk of thrombosis.

Bleeding and easy bruising can also occur. This is usually minor and occurs in around one quarter of all patients. Occasionally bleeding into the gut can be prolonged or severe.

How is polycythaemia vera diagnosed?

Polycythaemia vera is diagnosed using a combination of laboratory tests and a physical examination.

Laboratory tests

Full blood count

People with polycythaemia vera have a high red cell count, haemoglobin level and haematocrit (more than 52 per cent in men, and more than 48 per cent in females) due to the excessive production of red cells. The haematocrit is the proportion of the whole blood that is made up of red cells. A raised white cell count (especially a raised neutrophil count) and a raised platelet count are also common findings.

JAK2 mutation testing

JAK2 mutations (particularly the V617F mutation) can be found in more than 95 per cent of people with polycythaemia vera. This test can be performed on a blood sample and will help to confirm the diagnosis of a myeloproliferative neoplasm. It doesn't help distinguish polycythaemia vera from essential thrombocythaemia or primary myelofibrosis.

Bone marrow aspirate and biopsy

A procedure that involves removing a sample of the liquid bone marrow and a small core of bone marrow for examination in the laboratory. The biopsy (or trephine) is taken under local or general anaesthetic, from the back of the pelvis (hip girdle).

In polycythaemia vera the bone marrow is often very active with abnormally high numbers of normal cells. Iron stores may be depleted since iron is being used to make more and more red cells

Other possible blood tests

- Serum vitamin B-12 levels
- Uric acid levels
- Erythropoietin levels
- Coagulation studies (to see if your blood is clotting normally)
- Blood oxygen levels

Physical examination

Other possible tests

- Chest X-ray – to rule out lung disease
- Abdominal ultrasound and/or CT scan – to rule out kidney disease and measure spleen/liver size

How is polycythaemia vera treated?

The goal of treatment for polycythaemia vera is to reduce the number of cells in your blood and help you to maintain a normal blood count. This helps control any symptoms of your disease and reduces the risk of complications due to blood clotting or bleeding. The treatment, or combination of treatments, chosen for you will depend on several factors including the duration and severity of your disorder, whether or not you have a history of blood clots, your age and your general health.

Venesection

Venesection (or phlebotomy) is a procedure in which a controlled amount of blood is removed from your bloodstream. This procedure is commonly used when people are first diagnosed with polycythaemia vera because it can help to rapidly reduce a high red cell count. In a process similar to a blood donation, 450mls to 500mls of your blood is removed, usually from a large vein in the arm, inside the elbow bend. This is usually done in the outpatient's department of the hospital. It takes about 30 minutes to complete. You will need to have a blood test before to check your blood count, and you must make sure you drink plenty of water before and after the procedure.

This procedure may need to be repeated frequently at first, usually every few days, until your haematocrit is reduced to the desired level. After this, you may need to have the procedure repeated periodically, for example at monthly intervals, to help maintain a normal blood count. For many people, particularly younger patients and those with mild disease, regular venesection (every few months) may be all that is needed to control their disease for many years.

Many people with polycythaemia vera also need other treatments in addition to, or instead of venesection, to help control their blood count.

Myelosuppressive Drugs

Myelosuppressive (bone marrow suppressing) drugs or chemotherapy are commonly used to reduce blood cell production in the bone marrow. These drugs are commonly used for people with an extremely high platelet count, complications due to blood clotting or bleeding, or symptoms of an enlarged spleen. They are also used for some people who are unable to tolerate venesection, or whose disease is no longer responding to venesection. The most commonly used myelosuppressive agent is a chemotherapy drug called hydroxycarbamide which is also known as hydroxyurea. Hydroxyurea is particularly useful in controlling a high platelet count (thrombocytosis) and therefore reducing the risk of thrombosis. Hydroxyurea is taken in the form of capsules at home every day. As hydroxyurea is a chemotherapy drug, it may effect fertility and should be avoided during pregnancy because it can cause harm, or death to the foetus. If this could be an issue for you, you should ask your haematologist about your options.

Another less commonly used chemotherapy drug is busulphan. This drug is also given in tablet form.

Chemotherapy taken in capsule form is tolerated well by most people and side effects tend to be few and mild. As these drugs work by suppressing blood formation, periodic blood tests should be performed when taking these drugs to monitor the blood count and to guard against severe reductions in the white cell or platelet counts. There is a very small risk of developing leukaemia later on, in people who receive some chemotherapy for prolonged periods of time. It is still unclear whether there is a very small increase in the risk of leukaemia in people receiving hydroxyurea and this must be weighed against the potentially serious complications of uncontrolled disease (thrombosis). Discuss with your doctor if this is a concern to you.

Interferon

Interferon is a substance produced naturally by the body's immune system. It plays an important role in fighting disease. In polycythaemia vera, interferon is sometimes prescribed for younger patients to help control the production of blood cells. Interferon is usually given three times a week as an injection under the skin (subcutaneous injection) using a very small needle. You or a family member (or friend) will be taught how to do this at home. A weekly injection is now available and is becoming more widely used.

Side effects of interferon can be unpleasant, but they can be minimised by starting with a small dose, and building up to the full dose over several weeks. The main side-effects are flu-like symptoms such as chills, fevers, aches and pains and weakness. Your doctor or nurse will explain any side effects you might experience while you are having these treatments and how they can be managed.

Other Treatments

Aspirin

Many people are prescribed small daily doses of aspirin, which have been shown to significantly reduce the risk of thrombosis in people with polycythaemia vera.

Aspirin works by preventing your platelets from clumping together to form harmful blood clots in different parts of your body. Aspirin can irritate the lining of the stomach, which can result in pain or discomfort in the stomach, causing nausea, heartburn or loss of appetite. Taking your aspirin with food or milk may help prevent this. In addition, many people are prescribed specially coated aspirin that allows the drug to pass through the stomach and into the intestine before being dissolved. This helps to reduce the risk of stomach irritation. You should see your doctor if you are experiencing stomach irritation while on aspirin.

Aspirin is taken at home in tablet form. Drug interactions can occur, so it is important to avoid taking other medications while you are on aspirin, unless you are advised to do so by your doctor.

Anagrelide hydrochloride

Anagrelide hydrochloride (Agrylin®) is a drug used to reduce high platelet counts in people with polycythaemia vera and essential thrombocythaemia. Anagrelide affects platelet-producing cells in the bone marrow called megakaryocytes, slowing down platelet production and therefore reducing the number of platelets in the circulating blood. This can help to reduce symptoms and the risk of clotting complications in the future. Although anagrelide lowers platelet counts to more normal levels, it does not affect the body's natural process to form a clot when needed. Anagrelide is taken in capsule form by mouth. It can be taken with or without food. The capsule strength and the number of times a day you need to take anagrelide will depend on your platelet count, your response to treatment and how well you tolerate the drug.

Your doctor will keep track of your response to anagrelide and adjust your dose as needed to maintain your platelet count at the desired level. Side effects are generally mild to moderate and may decrease with continued therapy. The most commonly reported side effects include headaches, fast or forceful heart beat (palpitations), diarrhoea, weakness, fluid retention, nausea, dizziness, abdominal pain and shortness of breath. You should report any side effects you are experiencing to your doctor as many of them can be treated to reduce any discomfort to you. You need to contact your doctor immediately if you experience the following symptoms: shortness of breath or difficulty breathing, swollen ankles, fast or irregular heartbeat, and/or chest pain.

You should not stop taking this, or any other medication for polycythaemia vera unless instructed by your doctor. Stopping these medications suddenly can be harmful.

Radioactive phosphorus (32P)

Radioactive phosphorus (32P) is a radioisotope which may be used for long-lasting control of blood counts in older people. One or two doses of 32P are usually given, by injection into a vein in the hand or arm, in the nuclear medicine department of the hospital. This substance is taken up and concentrated in bone marrow where it suppresses the overactive bone marrow and helps to control blood counts.

In addition to the treatments described above, your doctor will advise you on ways to stay healthy and reduce any 'life-style' factors that might increase your risk of thrombosis. For example you may be advised to stop smoking, and / or take a series of steps to maintain a healthy weight range and blood pressure.

Prognosis

A prognosis is an estimate of the likely course of a disease. It provides some guide regarding the chances of curing the disease or controlling it for a given time.

The natural course of polycythaemia vera can vary considerably between individuals. In many patients, with treatment, the disease remains stable for long periods of time, often many years. In around 10 per cent of all cases, polycythaemia vera transforms over time into another type of myeloproliferative neoplasm called myelofibrosis, and less commonly, in up to 3 per cent of cases into acute myeloid leukaemia.

In some people, polycythaemia progresses over time despite treatment. The spleen may become increasingly enlarged. Anaemia and thrombocytopenia (low numbers of circulating platelets) is common as the bone marrow is no longer able to produce adequate numbers of red cells or platelets. In addition, abnormal immature blood cells, known as blast cells, may start to appear in the blood.

Treatment during this time is supportive and involves making every effort to improve the patient's quality of life, by relieving any symptoms they might have and by preventing and treating any complications that arise from their disease, or its treatment. This may involve blood transfusions if required, pain relief and careful myelosuppression. In selected cases, surgical removal of the spleen, or low dose radiation to the spleen may be required to relieve symptoms.

Your doctor is the best person to give you an accurate prognosis regarding your disease as they will have all the necessary information to make this assessment.

Essential thrombocythaemia (ET)

Essential thrombocythaemia (ET) is a myeloproliferative neoplasm in which too many platelets are produced in the bone marrow. Platelets are normally needed in the body to control bleeding. However, excess numbers of platelets can lead to abnormal blood clotting, which can block the flow of blood in the blood vessels. In essential thrombocythaemia, the blood platelet count is persistently elevated as a result of increased bone marrow production of platelets, in the absence of any identifiable cause.

Apart from essential thrombocythaemia, there are a number of conditions that can cause a rise in the number of platelets in the circulating blood (secondary thrombocytosis). These include bleeding, infection and some types of cancer. Secondary thrombocytosis is not essential thrombocythaemia and is not discussed further in this booklet.

Essential thrombocythaemia is a rare chronic disease diagnosed in an estimated 3 per 100,000 population. Although it can occur at any age, essential thrombocythaemia usually affects older people, with most patients diagnosed between the ages of 50 and 70 years.

Symptoms of essential thrombocythaemia

Many people have no symptoms when they are first diagnosed with essential thrombocythaemia and their disease is identified accidentally during a routine blood test. Symptoms can result from excessive numbers of platelets causing blockages in small or large blood vessels in different parts of the body and can include:

- Tingling in the hands and feet
- Headaches
- Visual problems
- Weakness
- Dizziness

Complications of essential thrombocythaemia

An enlarged spleen is much less commonly found at presentation. Symptoms include feelings of discomfort, pain or fullness in the upper left-side of the abdomen. An enlarged spleen may also cause pressure on the stomach causing a feeling of fullness, indigestion and a loss of appetite. In some cases the liver may also be enlarged (hepatomegaly). Other symptoms include weight loss and generalised itching.

Thrombosis is a major complication of essential thrombocythaemia. Older patients and those with a high platelet count, or a prior history of thrombosis, may be at increased risk. A major aim of treatment in essential thrombocythaemia is to reduce your risk of thrombosis. If you are at high risk of thrombosis this is achieved by making the blood count as normal as possible using hydroxyurea.

Blood clots can occur in large or small arteries interfering with the blood and therefore oxygen supply to various organs or tissues. Blockages in the smaller blood vessels in the toes and fingers can cause redness of the skin and burning and throbbing pains. These pains are often made worse by heat or exercise and relieved by cooling and elevating the affected area. These symptoms are often dramatically improved using small daily doses of aspirin, and/or reducing the patient's platelet count.

Blockages in the arteries supplying the heart (causing a myocardial infarction or heart attack), kidneys or brain (causing a stroke) can be serious and can lead to significant tissue damage, or ischaemia (tissue death). Blood clots can also develop in the veins of the legs (causing deep vein thrombosis), and less commonly, the spleen and liver occluding the blood flow and causing pain in these areas. A blood clot that breaks off the wall of the vein and travels in the blood stream is known as an embolism. When a blood clot travels to the lungs it is known as a pulmonary embolism and can cause breathing problems.

People experience bleeding including bruising for no apparent reason, or exaggerated or prolonged bleeding following minor cuts or injury. Some people notice frequent, or severe nose bleeds or bleeding gums and some women may have unusually heavy menstrual periods. Bleeding complications are particularly seen when the platelet count is very high.

In pregnancy, uncontrolled essential thrombocythaemia can reduce the blood supply to the placenta or foetus. This can cause problems with foetal growth and may in some cases lead to miscarriage.

How is essential thrombocythaemia diagnosed?

The diagnosis of essential thrombocythaemia is only made when other causes of a raised platelet count have been excluded.

Full blood count

A persistently raised platelet count is the most common sign of essential thrombocythaemia. The platelet count can range from slightly higher than normal to many times higher than normal. Under the microscope the platelets may be abnormally large and pale blue-stained. Fragments of megakaryocytes, the cells from which platelets are released, may also be seen in the blood film. Around a third of people with essential thrombocythaemia will also have a mildly raised red cell and/or white cell count.

If the results of your blood test suggest that you may have essential thrombocythaemia, further investigation and tests including a bone marrow examination may be required to help confirm the diagnosis and rule out other secondary, or 'reactive' causes of a raised platelet count.

Bone marrow examination

In essential thrombocythaemia the bone marrow is usually found to be overactive. An excess number of abnormal megakaryocytes are a common finding. Cytogenetic and molecular analysis of blood and bone marrow cells may be carried out to help confirm the diagnosis. A mutation in the JAK2 gene is found in a significant proportion (50-60 per cent) of people with essential thrombocythaemia. Mutations in the c-MPL gene (which produces a protein that responds to a growth factor that stimulates platelet production) account for approximately 10 per cent of cases.

Other blood tests may be done to check your general health and how well your kidneys, liver and other vital organs are functioning.

How is essential thrombocythaemia treated?

The goal of treatment for people with essential thrombocythaemia is to prevent complications like abnormal bleeding and bruising, and in some cases reducing the number of platelets in the blood. Patients are at higher risk of these complications if they are older and if they have had any complications before. Other blood vessel diseases such as high blood pressure and diabetes can also add to the risk of complications. If you are at lower risk of complications your doctor may recommend a 'watch and wait' strategy which involves regular check-ups and blood counts to carefully monitor your health. In addition they will advise you on the steps you can take to stay healthy and reduce any 'lifestyle-related' risk factors you may have that increase your chances of developing a blood clot. You may be advised for example on ways to help you stop smoking, and / or maintain a healthy weight range and blood pressure.

For the majority of people, essential thrombocythaemia will require some form of treatment to reduce their platelet count and therefore their risk of thrombosis. The treatment chosen for you will depend on a number of factors that influence your particular risk of complications due to thrombosis or bleeding. These include your age, platelet count and whether or not you have had any previous episodes of blood clots or bleeding. A history of smoking or high blood pressure can influence your risk of thrombosis. These factors and others are taken into account when planning the most appropriate treatment for your disease.

For people at high-risk of thrombosis, a chemotherapy drug called hydroxyurea, with or without low-dose aspirin, is often used as first-line treatment. Hydroxyurea works by suppressing the function of your bone marrow and thereby controlling platelet production, while aspirin prevents your platelets from aggregating and forming harmful clots in your body.

Anagrelide hydrochloride (Agrylin®) and interferon may also be used. Anagrelide slows down platelet production in your bone marrow, thereby helping to reduce symptoms and your risk of thrombosis. Interferon works by suppressing the abnormal megakaryocyte clone in your bone marrow thereby reducing the overproduction of platelets.

Those at low-risk may be simply treated using low-dose aspirin, or an equivalent drug alone. They usually have a very good outlook with no difference to the general population. Your doctor will be able to discuss with you all of the treatment options suitable for you.

Plateletpheresis

If your platelet count is very high and you have symptoms of clotting or bleeding, your platelet count will need to be reduced quickly to prevent further complications. In these emergency situations, excess platelets can be removed from your bloodstream using a procedure known as plateletpheresis. During this procedure all your blood is passed through a special machine called a cell separator. The blood is drawn from a cannula (plastic needle) placed in a vein in one arm. The machine spins the blood very quickly and removes the excess platelets. This is a continuous process. While platelets are being removed the rest of your blood is being returned to you via another cannula, placed in your other arm. If your veins are not suitable for this procedure, a special catheter device can be inserted into a large vein and might be used instead. This line allows blood to be drawn from one of the bigger veins in your body when your smaller veins are hard to access.

Plateletpheresis is usually carried out in hospital and usually takes about two hours to complete.

Prognosis

Essential thrombocythaemia is regarded as an incurable disease but in many people with treatment, the disease remains stable for long periods of time, often 10-20 years or more. In the longer term, a small number of people with essential thrombocythaemia may develop myelofibrosis. The risk of transforming to acute myeloid leukaemia is relatively low (less than 1 per cent).

Your doctor is the best person to give you an accurate prognosis regarding your disease as they will have all the necessary information to make this assessment.

Primary myelofibrosis

Primary myelofibrosis (also called chronic idiopathic myelofibrosis, agnogenic myeloid metaplasia) is a disorder in which normal bone marrow tissue is gradually replaced with a fibrous scar-like material. Over time, this leads to progressive bone marrow failure.

Under normal conditions, the bone marrow provides a fine network of fibres on which the stem cells can divide and grow. Specialised cells in the bone marrow known as fibroblasts make these fibres. In primary myelofibrosis, chemicals released by high numbers of platelets and abnormal megakaryocytes (platelet forming cells) over-stimulate the fibroblasts. This results in the overgrowth of thick coarse fibres in the bone marrow, which gradually replace normal bone marrow tissue. Over time this destroys the normal bone marrow environment, preventing the production of adequate numbers of red cells, white cells and platelets. This results in anaemia, low platelet counts and the production of blood cells in areas outside the bone marrow, for example in the spleen and liver, which become enlarged as a result.

Primary myelofibrosis is a rare chronic disorder diagnosed in an estimated 1 per 100,000 population. It can occur at any age, but is usually diagnosed later in life, between the ages of 60 and 70 years. The cause of primary myelofibrosis remains largely unknown. It can be classified as either JAK2 mutation positive (having the JAK2 mutation) or negative (not having the JAK2 mutation).

Long-term exposure to high levels of benzene or very high doses of ionising radiation may increase the risk of primary myelofibrosis in a small number of cases. Around one third of people with myelofibrosis have been previously diagnosed with polycythaemia (postpolycythaemic myelofibrosis), or essential thrombocythaemia (post-ET myelofibrosis).

Symptoms of primary myelofibrosis

Around 20 per cent of people have no symptoms of primary myelofibrosis when they are first diagnosed and the disorder is picked up incidentally as a result of a routine blood test. For others, symptoms develop gradually over time. Symptoms of anaemia are common and include:

- Unexplained tiredness
- Weakness
- Shortness of breath and palpitations
- Fever
- Unintended weight loss
- Pruritus (generalised itching)
- Excess sweating, especially at night

Other less common symptoms include bone and joint pain, and bleeding problems.

Complications of primary myelofibrosis

Virtually all patients with primary myelofibrosis have an enlarged spleen (splenomegaly) when they are first diagnosed. In around a third of cases the spleen is very enlarged. Common symptoms include feelings of discomfort, pain or fullness in the upper left-side of the abdomen. An enlarged spleen may also cause pressure on the stomach causing a feeling of fullness, indigestion and a loss of appetite. Abdominal discomfort can also result from an enlarged liver (hepatomegaly), which occurs in around two-thirds of cases.

How is myelofibrosis diagnosed?

Primary myelofibrosis is diagnosed using a combination of a physical examination which may identify an enlarged spleen, blood tests and a bone marrow examination. Primary myelofibrosis is only diagnosed when other causes of marrow fibrosis (including leukaemia, lymphoma, and other types of cancer that have spread to the bone marrow) have been ruled out.

Full blood count

People with primary myelofibrosis commonly present with varying degrees of anaemia. When examined under the microscope the red cells are often described as being 'teardrop-shaped'. Higher than normal numbers of white cells and platelets may be found in the early stages of this disorder, but low white cell and platelet counts are common in a more advanced disease.

Bone marrow examination

It is frequently impossible to obtain any samples of bone marrow fluid using a needle and syringe (bone marrow aspiration) due to marrow fibrosis. This is known as a 'dry tap'. The bone marrow trephine biopsy typically shows abnormal fibrosis of the marrow cavity.

Cytogenetic and molecular analysis of blood and bone marrow cells is also carried out to help confirm the diagnosis and may help with prognosis. A mutation in JAK2 is found in about 50 per cent of people with primary myelofibrosis. It is unclear at present why some patients with mutations in JAK2 develop myelofibrosis and others don't.

How is myelofibrosis treated?

Some people have no symptoms when they are first diagnosed with primary myelofibrosis and do not require treatment straight away, apart from regular check-ups with their doctor to carefully monitor their disease.

For others, treatment is largely supportive and is aimed at preventing complications due to low blood counts and an enlarged spleen (splenomegaly). This involves making every effort to improve your quality of life, by relieving any symptoms of anaemia or an enlarged spleen, and preventing and treating any complications that might arise from your disease or its treatment.

This may include periodic blood transfusions and taking antibiotics to prevent and treat any infections.

A chemotherapy drug such as hydroxyurea (see polycythaemia vera), or low-doses of a drug called thalidomide may be used to reduce an enlarged spleen. In some cases, the surgical removal of the spleen (splenectomy) may be considered, especially when the spleen has enlarged so much that it is causing severe symptoms. A splenectomy may also be considered if there is an increased need for blood transfusions. This sometimes happens because the spleen is destroying blood cells, particularly platelets, at a very fast rate. Small doses of radiation to the spleen can also be given to reduce its size. This usually provides temporary relief for about 3 to 6 months.

Some younger patients who have a suitably matched donor may be offered an allogeneic (donor) stem cell transplant. This is a medical procedure that offers the only chance of cure for patients with myelofibrosis. It involves the use of very high doses of chemotherapy, with or without radiotherapy, followed by infusion of blood stem cells, which have been donated by a suitably matched donor. Stem cell transplants carry significant risks.

There are separate booklets about stem cell transplants available from Leukaemia & Blood Cancer New Zealand.

Patients with myelofibrosis, whether or not they have the JAK2 mutation, may respond to drugs which inhibit JAK2, with improvement of symptoms and reduction in the size of the spleen. While no JAK2 inhibitors are currently licenced for use in New Zealand, a number may be available in clinical trials, or may become available soon. Side effects of JAK2 inhibitors may include worsening anaemia, or a low platelet count.

Blood and platelet transfusions

If symptoms of anaemia are interfering with your normal daily activities, your doctor may recommend that you have a red blood cell transfusion.

Platelet transfusions are sometimes given to prevent or treat bleeding (for example a persistent nose bleed) when the platelet count is below a critical level.

You do not need to be admitted to hospital for a red blood cell or platelet transfusion. They are usually given in the outpatient department. Transfusions these days are relatively safe and they don't usually cause any serious complications. Nevertheless you will be carefully monitored throughout the transfusion.

In the meantime, remember to call the nurse if you are feeling hot, cold and shivery or in any way unwell during the transfusion, as this might indicate that you are having a reaction. Steps can be taken to minimise these symptoms and ensure that they don't happen again.

Prognosis

Primary myelofibrosis is generally regarded as an incurable disease, but with treatment many people can remain comfortable and symptom-free.

The natural course of the disease can vary considerably between individuals. In some people their disease remains stable for long periods and they are free to live a normal life with minimal interruptions from their disease, or its treatment. For others, myelofibrosis progresses more quickly and people require treatment to help relieve symptoms of their disease. Transformation to acute myeloid leukaemia occurs in between 10 and 20 per cent of cases.

Your doctor is the best person to give you an accurate prognosis regarding your disease as they will have all the necessary information to make this assessment.

Chronic eosinophilic leukaemia (CEL) / hypereosinophilic syndrome

Chronic eosinophilic leukaemia (also known as hypereosinophilic syndrome) is a rare myeloproliferative neoplasm in which too many eosinophils (a type of white blood cell) are made in the bone marrow. These cells spill out of the bone marrow and accumulate in the blood and other tissues around the body.

Symptoms of chronic eosinophilic leukaemia

Some people with chronic eosinophilic leukaemia don't have any symptoms and the disease is picked up incidentally during a routine blood test. Others may go to their doctor because they have one or more of a range of symptoms, including:

- Fever
- Fatigue
- Cough
- Muscle pains
- Pruritis (generalized itching)
- Diarrhoea

Complications of chronic eosinophilic leukaemia (CEL) / hypereosinophilic syndrome

Sometimes there can be chronic changes to the lungs with increased lung fibrosis, which can make you short of breath on exertion. Sometimes the heart muscle can be affected by fibrosis. This makes the heart muscle stiff and less able to pump blood effectively. The heart valves can develop fibrin deposits which can cause the heart valves to leak and this can be heard as a heart murmur.

How is chronic eosinophilic leukaemia diagnosed?

This is often a difficult diagnosis to make. While a simple blood test will demonstrate that high numbers of eosinophils are present, it is often difficult to find out what the cause of this is. Eosinophils are increased in the blood in allergic reactions, in response to some infections (such as intestinal worms), related to abnormal lymphocytes, or due to a myeloproliferative neoplasm.

In some people mutations that affect the growth of eosinophils can be detected by a blood test.

These tests may help in deciding the best treatment to give.

Mutations in CEL

There has been increasing knowledge of how CEL develops in some people. Three types of mutations have been described:

- those that affect the platelet derived growth factor receptor (PDGFR). These can involve either the alpha (PDGFRα) or beta (PDGFRβ) chain that makes up the complete receptor.
- Mutations of the fibroblast growth factor receptor (FGFR1).

Finding mutations can be useful for diagnosis as they are not found in reactive eosinophilia states. The presence of mutations can also predict the response to treatment.

Treatment and prognosis of chronic eosinophilic leukaemia

The natural course of chronic eosinophilic leukaemia can vary considerably between individuals. The disease may remain stable for many years, even decades, or it may quickly progress and transform to an acute leukaemia. Because of this, the most appropriate treatment for each patient is decided upon on an individual basis.

Treatment may include corticosteroids, chemotherapy drugs such as hydroxyurea, and interferon therapy. Some patients may respond to a newer drug called imatinib mesylate, most often used in the treatment of chronic myeloid leukaemia. A stem cell transplant may be considered in selected cases.

Chronic neutrophilic leukaemia

Chronic neutrophilic leukaemia is a rare myeloproliferative neoplasm in which too many neutrophils are made in the bone marrow. These cells spill out into the circulating blood and tend to accumulate in the liver and spleen, which become enlarged as a result. Chronic neutrophilic leukaemia is usually a slowly progressing disease, closely related to another type of leukaemia called chronic myeloid leukaemia, but the cause and treatment are different. Its natural course can vary considerably between individuals with survival times ranging from 6 months to over 20 years. Treatment options may include the use of chemotherapy drugs such as busulphan or hydroxyurea, which are given in tablet or capsule form. These drugs are used to control the high white cell count.

Mastocytosis

Mastocytosis is a disorder that results from the overproduction of mast cells (a type of white blood cell), in the bone marrow. These cells accumulate in the blood, spleen, skin and other body tissues. Excess numbers of mast cells release large amounts of histamine and other substances which can cause allergic-type reactions in affected tissues. For example, when these substances collect in the skin they tend to cause an itchy rash. Other allergic-type symptoms may include abdominal pain and difficulty breathing. Medications known as antihistamines are used to prevent and reduce allergic reactions.

There are two main types of mastocytosis:

- Cutaneous mastocytosis. This only affects the skin and is most commonly seen in children. It has a good prognosis, often resolving at puberty.
- Systemic mastocytosis. More common in adults, this affects the bone marrow and other tissues, including the skin.

Symptoms and complications of mastocytosis

One feature of mast cell collections in the skin is that they release histamine when stroked. This causes a raised weal to come up on the skin, called urticaria. This can be more pronounced in some people and can results in blisters forming, especially in children under the age of 3 years. In some people you can draw on the skin and raised lines appear, a symptom called dermatographism.

Symptoms from the mast cells can be grouped into 4 categories:

1. Constitutional symptoms, such as fatigue, weight loss, sweating and fever
2. Skin symptoms, including itch, urticaria and dermatographism
3. Systemic symptoms related to the release of mast cell granules, such as abdominal pain, flushing, fainting, headache, rapid pulse, cough and shortness of breath
4. Musculoskeletal problems including bone pain, osteoporosis, fractures, joint and muscle aches

Some people with mastocytosis may have an enlarged spleen, less commonly the lymph nodes are enlarged. Spleen and lymph node enlargement are more common if the disease is faster growing. Some people have abnormal mast cells circulating in the blood.

Diagnosis and treatment of mastocytosis

The diagnosis of mastocytosis can be made on skin or bone marrow biopsies, and by measuring the blood level of tryptase, an enzyme released by the mast cells. Treatment decisions tend to be made on an individual basis and may include chemotherapy in tablet form and/or interferon therapy to help control the overproduction of mast cells in the bone marrow. Many cases of mastocytosis in adults have been shown to have a mutation of a protein called KIT, which is a growth factor for the cells that causes them to keep growing. New drugs that block this mutant protein are being developed and may prove useful for treating mastocytosis.

Making treatment decisions

Many people feel overwhelmed when they are diagnosed with a myeloproliferative neoplasm. In addition to this, waiting for test results and then having to make decisions about proceeding with the recommended treatment can be very stressful. Some people do not feel that they have enough information to make such decisions while others feel overwhelmed by the amount of information they are given, or that they are being rushed into making a decision. It is important that you feel you have enough information about your illness and all of the treatment options available, so that you can make your own decisions about which treatment to have.

Anxiety, shock, denial or grief can make it difficult, at times, to absorb or remember discussions you have had with your doctor, and it is common for people not to remember much of the information given to them at diagnosis. Before going to see your specialist doctor, make a list of the questions you want to ask. It may be useful to keep a notebook with you and write questions down as you think of them, as often questions are forgotten between appointments.

Sometimes it is hard to remember everything the doctor has said. It may help to bring a family member or a friend along who can write down the answers to your questions or prompt you to ask others, be an extra set of ears or simply be there to support you.

Your doctor will spend time with you and your family discussing what he or she feels is the best option for you. Feel free to ask as many questions as you need to, at any stage. You are involved in making important decisions regarding your wellbeing.

You should feel that you have enough information to do this and that the decisions made are in your best interests. Remember, you can always request a second opinion if you feel this is necessary.

The Haematology Patient Diary, available from Leukaemia & Blood Cancer New Zealand, may be useful for recording details of treatment and making notes from clinic appointments.

Informed consent

Giving your informed consent means that you understand and accept the risks and benefits of a proposed procedure or treatment. It means that you are happy that you have adequate information to make such a decision.

Your informed consent is also required if you agree to take part in a clinical trial, or if information is being collected about you or some aspect of your care (data collection).

If you have any doubts or questions regarding any proposed procedure or treatment do not hesitate to talk to the doctor or nurse again.

Standard therapy

Standard therapy refers to a type of treatment which is commonly used in particular types and stages of disease. It has been tried and tested and has proven to be effective in a given situation.

Clinical trials

Your specialist doctor may ask you to consider taking part in a clinical trial (also called a research study). Clinical trials test new treatments, or existing treatments given in new ways to see if they work better. Clinical trials are important because they provide vital information about how to improve treatment by achieving better results with fewer side effects.

Participation in a trial may also involve giving blood or bone marrow samples in order to contribute to a better understanding of the disease. Clinical trials often give people access to new therapies not yet funded by governments.

Taking part in a clinical trial is entirely voluntary and you are under no obligation to participate. If you are considering taking part in a clinical trial, make sure that you understand the reasons for the trial and what it involves for you. You should always take time to consider all the implications of a trial and discuss this thoroughly with your specialist doctor and other support people before giving your informed consent. Your specialist doctor can guide you in making the best decision for you.

There is a separate booklet called 'Clinical Trials' available from Leukaemia & Blood Cancer New Zealand.

Interpreting services

New Zealand's Health and Disability Code states that everyone has the right to have an interpreter present during a medical consultation. Family or friends may assist if you and your doctor do not speak the same language, but you can also ask your doctor to provide a trained interpreter if using a family member is not appropriate.

Reproductive health

Fertility

Fertility is the ability to produce a child. In males, fertility means having enough healthy sperm to get a female pregnant. In females, fertility is the ability to become pregnant.

Some types of chemotherapy and radiotherapy may cause a temporary or permanent reduction in your fertility. It is very important that you discuss any questions or concerns you might have regarding your future fertility with your doctor, if possible, before you commence treatment.

In women, some types of chemotherapy and radiotherapy can cause varying degrees of damage to the normal functioning of the ovaries. In some cases this leads to menopause (change of life) earlier than expected. In men sperm production can be impaired for a while but the production of new sperm may become normal again in the future.

There are some options for preserving your fertility, if necessary, while you are having treatment. These are described below.

Protecting your fertility - Men

Sperm banking is a relatively simple procedure whereby the man donates semen, which is then stored at a very low temperature (cryopreserved), with the intention of using it to achieve a pregnancy in the future. You should discuss sperm banking with your doctor before starting any treatment that might impact on your fertility. In some cases, however, people are not suitable for sperm banking when they are first diagnosed because they are too unwell and therefore unable to produce the sperm in sufficient quantity or quality.

If possible, semen should be donated on more than one occasion. It is important to realise that there are many factors that can affect the quality and quantity of sperm collected in a semen donation and its viability after it is thawed out. There is no guarantee that you and your partner will be able to achieve a pregnancy and healthy newborn in the future. You should raise any concerns you have with your doctor who can best advise you on your fertility options.

The use of donor sperm might be another option for you and your partner. The sperm is donated from another male to achieve a pregnancy.

Protecting your fertility - Women

There are several approaches that may be used to protect a woman's fertility. These are outlined below.

Embryo storage - this involves collecting your eggs, usually after taking drugs to stimulate your ovaries to produce a number of eggs, so that more than one egg can be collected. This process takes at least several weeks and this can be a problem if your treatment needs to start immediately. Once the eggs are collected they are then fertilised with your partner's sperm and stored to be used at a later date. Your unfertilised eggs can also be collected and stored in a similar manner (egg storage).

Ovarian tissue storage - this is still a fairly new approach to protecting your fertility and to date there is very little experience with this technique in New Zealand. It involves the removal and storage at a very low temperature of some ovarian tissue (cryopreservation). It is hoped that at a later date the eggs contained in this tissue can be matured, fertilised and used to achieve a pregnancy.

To date, these first two approaches have unfortunately shown little success in cancer patients.

The use of donor eggs might be another option for you and your partner. These eggs could be fertilised using your partner's sperm and used in an attempt to achieve a pregnancy in the future.

It is important to understand that these methods are still quite experimental and for many reasons achieving a pregnancy and subsequently a baby is not guaranteed by using any of them. In addition, some are time consuming and costly, while others may simply not be acceptable to you or your partner.

Early menopause

Some cancer treatments can affect the normal functioning of the ovaries. This can sometimes lead to infertility and an earlier than expected onset of menopause, even at a young age. The onset of menopause in these circumstances can be sudden and, understandably, very distressing.

Hormone changes can lead to many of the classic symptoms of menopause including menstrual changes, hot flushes, sweating, dry skin, vaginal dryness and itchiness, headache and other aches and pains. Some women experience decreased sexual drive, anxiety and even depressive symptoms during this time.

It is important that you discuss any changes to your periods with your doctor or nurse. He or she may be able to advise you or refer you to a specialist doctor (a gynaecologist) or clinic that can suggest appropriate steps to take to reduce your symptoms.

Menstruation

Cancer treatment can also affect your periods; you may find your periods stop, become irregular or sometimes much heavier and longer in length. You may be prescribed a birth control pill to stop your periods. This prevents heavy bleeding and blood loss when your platelets are low. If you are having chemotherapy, it is best to use pads instead of tampons if you are menstruating as this will reduce the risk of infections. Always let your doctor know if you are having your period.

Body image, sexuality and sexual activity

It is likely that the diagnosis and treatment of your blood cancer or condition will have had some impact on how you feel about yourself as a man or a woman and as a 'sexual being'. Hair loss, skin changes and fatigue can all interfere with feeling attractive.

During treatment you may experience a decrease in libido, which is your body's sexual urge or desire, sometimes without there being any obvious reason. It may take some time for things to return to 'normal'. It is perfectly reasonable and safe to have sex after your transplant, but there are some precautions you need to take. Men may sometimes experience some degree of erectile dysfunction during treatment and in the stages of recovery. Please discuss this with your doctor so any help can be initiated.

It is usually recommended that you or your partner do not attempt to become pregnant while undergoing treatment, as some of the treatments given might harm the developing baby. It is important not to assume infertility following intensive treatment. You need to ensure that you and your partner use a suitable form of contraception. Condoms (with a spermicidal gel) offer good contraceptive protection as well as protection against infection or irritation. Your partner may be worried that sex might in some way harm you. This is not likely as long as your partner is free from any infections and the sex is relatively gentle. It is important to be extra careful when your white cell count or platelet levels are low as you may be more prone to infection and bleeding/spotting. Finally, if you are experiencing vaginal dryness, a lubricant can be helpful. This will help prevent irritation. Using a condom is also important to protect your partner from chemotherapy drugs that can be excreted in body fluids in the first few days after they are administered.

If you have any questions or concerns regarding sexual activity and contraception don't hesitate to discuss these with your doctor or nurse, or ask for a referral to a doctor or health professional who specialises in sexual issues.

Supportive care

Supportive care plays an important role in treatment. This involves making every effort to improve your quality of life, by relieving any symptoms you might have and by preventing and treating any complications that arise from your disease or treatment. Supportive care begins prior to transplant and continues until you have recovered.

Blood transfusions, antibiotics and intravenous fluids can all be important elements of medical supportive care. Non-medical supportive care may involve complementary therapies, nutrition support, exercise, counselling and similar services. Many aspects of supportive care are things that you can do yourself.

Inform your haematologist if any surgery or treatment is planned by another practitioner, as advice may be required from your haematologist as to the best supportive treatment, such as transfusions, blood tests or other monitoring, to ensure that your treatment happens successfully without problems due to your disease, current or previous treatment.

Complementary therapies

Complementary therapies are therapies which are not considered standard medical therapies. Many people find that they are helpful in coping with their treatment and recovery from disease. There are many different types of complementary therapies. These include yoga, exercise, meditation, prayer, acupuncture, relaxation and herbal and vitamin supplements.

If you have not already done so you might consider learning some relaxation techniques such as meditation, yoga or breathing exercises that you can use while you are in hospital, and while you are recovering from your transplant.

Complementary therapies should 'complement' or assist with recommended medical treatment. They are not recommended as an alternative to medical treatment. It is important to realise that no complementary or alternative treatment alone has proven to be effective against cancer.

It is also important to let your doctor or nurse know if you are using any complementary or alternative therapies in case they interfere with the effectiveness of chemotherapy or other treatments you may be having.

Nutrition

A healthy and nutritious diet is important in helping your body to cope with treatment and recovery. Talk to your doctor or nurse if you have any questions about your diet or if you are considering making any radical changes to the way you eat. It is likely that during your hospital stay you will see a hospital dietician who will advise you on planning a balanced and nutritious diet.

If you are thinking about using herbs or vitamins it is very important to talk this over with your doctor first. Some of these substances can interfere with the effectiveness of chemotherapy or other treatment you are having.

Social and emotional effects

People cope with a diagnosis of a myeloproliferative neoplasm in different ways, and there is no right or wrong or standard reaction. For some people, the diagnosis can trigger any number of emotional responses ranging from denial to devastation. It is not uncommon to feel angry, helpless and confused. Naturally people fear for their own lives or that of a loved one.

It is worth remembering that information can often help to take away the fear of the unknown. It is a good idea for you and your family to speak directly to your doctor regarding any questions you might have about your disease or treatment. It can also be helpful to talk to other health professionals including social workers or nurses who have been specially educated to take care of people with haematological diseases. Some people find it useful to talk with other patients and family members who understand the complexity of feelings and the kinds of issues that come up for people living with blood cancers and conditions.

Leukaemia & Blood Cancer New Zealand facilitate patient support groups around the country.

Many people are concerned about the social and financial impact of the diagnosis and treatment on their families. Normal family routines are often disrupted and other members of the family may suddenly have to fulfil roles they are not familiar with, for example, cooking, cleaning, and taking care of children. The social worker at your treatment centre is available for you to talk through and discuss these issues with you and your family.

If you have a psychological or psychiatric condition, please inform your doctor and don't hesitate to request additional support from a mental health professional.

There is a variety of assistance available to help ease the emotional and financial strain created by a diagnosis of a blood cancer or condition. In some cases a travel allowance may be available. Support Services staff at Leukaemia & Blood Cancer New Zealand are available to provide you and your family with information and support to help you cope during this time.

Contact details for Leukaemia & Blood New Zealand are provided on the back of this booklet.

Looking after yourself

Focusing on the things you can do to help yourself recover both physically and emotionally is important. Enjoying simple pleasures every day, looking to better times in the future, making plans and having hope are all important in maintaining a sense of control in a time of uncertainty.

Maintain a healthy lifestyle by:

Avoiding smoking

Eating a healthy diet

Taking regular exercise

Drinking alcohol sensibly

Maintaining a healthy weight

Wearing appropriate sun protection

Useful internet addresses

The value of the internet is widely recognised; however, not all the information available may be accurate and up to date. For this reason, we have selected some of the key sites that people with MDS might find useful.

With the exception of our own websites, Leukaemia & Blood Cancer New Zealand do not maintain these listed sites. We have only suggested sites we believe may offer credible and responsible information, but we cannot guarantee the information on them is correct, up to date or evidence based medical information.

Leukaemia & Blood Cancer New Zealand
www.leukaemia.org.nz

Cancer Society of New Zealand
www.cancernz.org.nz

Leukaemia Foundation of Australia
www.leukaemia.com.au

MPD Foundation (USA)
www.mpnresearchfoundation.org

National Cancer Institute (USA)
www.cancer.gov

CancerBACKUP (A UK cancer information site)
www.macmillan.org.uk

Leukemia & Lymphoma Society USA (note different spelling)
www.lls.org

Leukaemia Research Fund (UK)
leukaemialymphomaresearch.org.uk

Information on Essential thrombocythaemia
www.nhlbi.nih.gov

Information on Secondary or reactive polycythaemia
www.nhs.uk

Leukaemia & Blood Cancer New Zealand

Leukaemia & Blood Cancer New Zealand (LBC) is the only organisation in New Zealand dedicated to supporting patients and their families living with leukaemia, lymphoma, myeloma and related blood conditions.

Since 1977, our work has been made possible through our fundraising events and the generous support we receive from individuals, companies, trusts and grants. We do not receive government funding.

LBC manages the New Zealand Bone Marrow Donor Registry (NZBMDR), which works towards finding matched volunteer donors from New Zealand or overseas for New Zealand patients who need a bone marrow or stem cell transplant and who do not have a family donor. The registry maintains information on New Zealand donors and has access to a worldwide database of over 20 million donors.

Patient Support

Leukaemia & Blood Cancer New Zealand's Support Services provide personalised support programmes for patients and their families. This can include regular visits, phone or email contact, as well as face to face education and support programmes, and an online information forum. We also provide a toll free number for advice and support which is 0800 15 10 15.

Research

Research plays a critical role in building a greater understanding of blood cancers and conditions. LBC supports and funds investigation into these conditions. Improved treatments for patients can lead to less side effects and increased survival rates.

Information

We provide vital information to patients, families, health professionals and the community to improve understanding about blood cancers and conditions.

Awareness

We work to increase public knowledge of blood cancers and conditions. This is achieved through specifically focused campaigns for the public, health professionals and health agencies.

Advocacy

We represent the needs of patients and their families to the government, related agencies and other relevant organisations.

Contacting us

Leukaemia & Blood Cancer New Zealand provides services and support throughout New Zealand. Each person's experience of living with a blood cancer or condition is different. Living with leukaemia, lymphoma, myeloma or a related blood condition is not easy, but you do not have to do it alone.

Call **0800 15 10 15** to speak to a local Support Services Coordinator or to find out more about the services offered by Leukaemia & Blood Cancer New Zealand. Alternatively, contact us via email by sending a message to info@leukaemia.org.nz or by visiting www.leukaemia.org.nz.

We welcome visitors to our offices in Auckland, Hamilton, Wellington and Christchurch. Please phone for an appointment.

Glossary of terms

Acute leukaemia
Rapidly progressing cancers of the blood and bone marrow, usually of sudden onset and characterised by uncontrolled growth of immature blood cells which crowd the bone marrow and spill out into the bloodstream. These include acute myeloid leukaemia (AML) and acute lymphoblastic leukaemia (ALL).

Allogeneic stem cell transplant
The transplant of blood stem cells from one person to another. The donor is usually a sister or brother or an unrelated volunteer donor.

Alopecia
Hair loss. This is a side effect of some kinds of chemotherapy and radiotherapy. It is usually temporary.

Anaemia
A reduction in the haemoglobin level in the blood. Haemoglobin normally carries oxygen to all the body's tissues. Anaemia causes tiredness, paleness and sometimes light-headedness, and shortness of breath.

Antiemetic
A drug used to prevent or reduce feelings of sickness (nausea) and vomiting.

Autologous stem cell transplant
A type of stem cell transplant using blood stem cells collected from the patient's own bone marrow. These cells are collected and stored in advance, at an early disease stage. They are returned to the patient at a later stage, to rescue the function of their bone marrow, after the patient has received high doses of chemotherapy to destroy their disease.

Blasts
Abnormal immature blood cells that multiply in an uncontrolled manner, crowding out the bone marrow and preventing it from producing normal blood cells. These abnormal cells can also spill out into the bloodstream and accumulate in other organs. High numbers of blasts are present in acute leukaemia.

B-lymphocyte
A type of white cell normally involved in the production of antibodies to combat infection.

Bone marrow
The tissue found at the centre of many flat or big bones of the body. Active or red bone marrow contains stem cells from which all blood cells are made and in the adult this is found mainly in the bones making up the axial skeleton – hips, ribs, spine, skull and sternum.

Bone marrow aspirate
A procedure that involves removing (or aspirating) a small sample of bone marrow fluid for examination in the laboratory. The fluid is aspirated, under local or general anaesthetic, usually from the back of the pelvic bone, or occasionally from the breastbone (sternum).

Bone marrow biopsy
A procedure that involves removing a small core of bone and bone marrow for examination in the laboratory. The biopsy (or trephine) is taken under local or general anaesthetic, from the back of the pelvic bone.

Cancer
A malignant disease characterised by uncontrolled growth, division, accumulation and invasion into other tissues of abnormal cells from the original site where the cancer started. Cancer cells can grow and multiply to the extent that they eventually form a lump or swelling. This is a mass of cancer cells known as a tumour. Not all tumours are due to cancer; in which case they are referred to as non-malignant or benign tumours.

Cannula
A small plastic tube which can be inserted into a vein to allow fluid and drugs to enter the bloodstream.

Central venous catheter (CVC)
A line or tube passed through the large veins of the arm, neck, chest or groin and into the central blood circulation. It can be used for taking samples of blood, giving intravenous fluids, blood products, chemotherapy and other drugs without the need for repeated needles. Also known as a central venous access device (CVAD). There are several different types of central venous catheters, these include Hickman lines, PICC lines, Groshong lines and portacaths.

Chemotherapy
Single drugs or combinations of drugs which may be used to kill and prevent the growth and division of cancer cells. Although aimed at cancer cells, chemotherapy can also affect rapidly dividing normal cells and this is responsible for some common side effects including hair loss and a sore mouth (mucositis). Nausea and vomiting are also common, but nowadays largely preventable with modern anti-nausea medication. Most side effects are temporary and reversible.

Chromosomes
Chromosomes are made up of coils of DNA (deoxyribonucleic acid). DNA carries all the genetic information for the body in sequences known as genes. There are approximately 40,000 genes on 23 different chromosomes. The chromosomes are contained within the nucleus of a cell.

Chronic leukaemia
A group of cancers that affect the blood and bone marrow. Chronic leukaemias usually develop gradually and slowly progress, particularly in the early stages of disease. The leukaemia is called chronic because the leukaemia cells divide and increase in number more slowly than in acute leukaemia. Typically, chronic leukaemic cells are more mature than those found in acute leukaemia. Chronic leukaemias are sometimes diagnosed by chance, during a routine blood test.

Chronic myeloid leukaemia (CML)
A type of leukaemia which is an initially slow growing (indolent) disease where the bone marrow produces too many white cells. Overtime, CML usually transforms into acute leukaemia, a more aggressive type of disease where the bone marrow produces large numbers of abnormal immature granulocytes, known as blast cells or leukaemic blasts. CML is also called chronic myelogenous or chronic granulocytic leukaemia (CGL).

Clinical trial
A controlled and carefully monitored assessment of new forms of treatment. Trials can vary in design and size from small-scale trials of experimental treatments to large national trials that compare subtle variations in current therapies. The patient will be informed and will always be given the option not to join, or not without detriment to their treatment when their treatment is part of a trial. Likewise, patients can opt out of a clinical trial at any time.

Clone
A population of genetically identical cells arising from a single parent cell. Leukaemia is believed to be a clonal disease, that is, all the leukaemia cells may originate from one abnormal cell.

Computerised axial tomography (CT scan or CAT scan)
A specialised x-ray or imaging technique that produces a series of detailed three dimensional (3D) images of cross sections of the body.

Cure
This means that there is no evidence of disease and no sign of the disease reappearing, even many years later.

Cytogenetic tests
The study of the genetic make-up of the cells, in other words, the structure and number of chromosomes present. Cytogenetic tests are commonly carried out on samples of blood and bone marrow to detect chromosomal abnormalities associated with disease. This information helps in the diagnosis and selection of the most appropriate treatment.

DNA (Deoxyribonucleic acid)
Molecules found in the centre of the cell that carry all the genetic information for the body. There are four different chemical compounds of DNA (bases) arranged in coded sequences called genes, which determine an individual's inherited characteristics.

Echocardiogram
A special ultrasound scan of the heart.

Electrocardiogram (ECG)
A recording of the electrical activity of the heart.

Full Blood count
Also called a Complete Blood Count (CBC). A routine blood test that measures the number and type of cells circulating in the blood.

Genes
Collections of DNA. Genes direct the activity of cells. They are responsible for the inherited characteristics that distinguish one individual from another. Each person has an estimated 100,000 separate genes.

Granulocytes
A family of white blood cells that contains granules in their cytoplasm (neutrophils, eosinophils and basophils). They protect the body by seeking out and destroying micro-organisms.

Growth factors and cytokines
A complex family of proteins produced by the body to control the growth, division and maturation of blood cells by the bone marrow. Some are now available as drugs as a result of genetic engineering and may be used to stimulate normal blood cell production following chemotherapy or bone marrow or peripheral blood stem cell transplantation.

Haemoglobin
The iron containing pigment in red blood cells, which carries oxygen to all the body's tissues.

Haemopoiesis (or Haematopoiesis)
The processes involved in blood cell formation. In adults this occurs in the bone marrow.

Haematologist
A doctor who specialises in the diagnosis and treatment of diseases of the blood, bone marrow and immune system.

High dose therapy
The use of higher than normal doses of chemotherapy to kill off resistant and / or residual (left over) cancer cells that have survived standard-dose therapy.

Imatinib mesylate (Glivec®)
A drug used to treat chronic myeloid leukaemia and other Philadelphia chromosome positive (Ph+) leukaemias. Imatinib is classified as a tyrosine kinase inhibitor. It works by targeting the abnormal BCR-ABL gene thereby blocking the leukaemia-causing effects of the enzyme tyrosine kinase. Also known as imatinib (Glivec®).

Immune system
The body's defence system against infection and disease.

Immunocompromised
When someone has decreased immune function.

Leukaemia
A cancer of the blood and bone marrow characterised by the widespread, uncontrolled production of large numbers of abnormal blood cells. These cells take over the bone marrow often causing a fall in blood counts. If they spill out into the bloodstream however they can cause very high abnormal white cell counts.

Leucopheresis (leukopheresis)
A procedure that uses a special machine called a 'cell separator' to separate and remove white blood cells from the circulation before returning the remainder of the blood to the patient (see apheresis). Leucopheresis is the technique used to collect stem cells from the blood for use in a stem cell transplant. It is also sometimes used to reduce a dangerously high white cell count.

Lymph nodes or glands
Structures found throughout the body, for example in the neck, groin, armpit and abdomen, which contain both mature and immature lymphocytes. There are millions of very small lymph glands in all organs of the body. Their role is to filter the lymph fluid, which "washes" the tissues, and to detect infection.

Lymphocytes
Specialised white blood cells involved in defending the body against disease and infection. There are two types of lymphocytes: B-lymphocytes and T-lymphocytes. They are also called B-cells and T-cells.

Lymphoid
Term used to describe a pathway of maturation of blood cells in the bone marrow. White blood cells (B-lymphocytes and T-lymphocytes) are derived from the lymphoid stem cell line.

Malignancy
A term applied to tumours characterised by uncontrolled growth and division of cells (see cancer).

Mucositis
Inflammation of the lining of the mouth and throat, which also can extend to the lining of the whole of the gastro-intestinal tract (stomach and intestines).

Mutation
A change in the DNA code of a cell, caused for example by exposure to hazardous chemicals or copying errors during cell division. If mutations affect normal cell function this can lead to the development of disease due to the loss of normal function, or the development of abnormal functions of that cell.

Myeloid
Term used to describe a pathway of maturation of blood cells in the bone marrow. Red cells, white cells (neutrophils, eosinophils, basophils and monocytes) and platelets are derived from the myeloid stem cell line.

Myeloproliferative neoplasms
A group of disorders characterised by the over-production of blood cells by the bone marrow. One or more of the cell families - red, white, platelets or support tissue, may be involved and treatment varies depending on the type and severity of the disease. Includes chronic myeloid leukaemia (CML), polycythaemia vera (PV), essential thrombocythaemia (ET) and primary myelofibrosis.

Neutropenia
A reduction in the number of circulating neutrophils, an important type of white blood cell. Neutropenia is associated with an increased risk of infection.

Neutrophils
Neutrophils are the most common type of white blood cell. They are needed to mount an effective fight against infection, especially bacteria and fungi.

Oncologist
General term used for a specialist doctor who diagnoses and treats cancer by different means, e.g. medical, radiation, surgical oncologist that have not originated in the bone marrow.

Pathologist
A doctor who specialises in the laboratory diagnosis of disease, and how disease is affecting the organs of the body.

Petechiae
Red or purple flat pinhead sized spots on the skin, especially on the legs. They are caused by tiny bleeds under the skin, usually as a result of a severe shortage of platelets.

Philadelphia chromosome
The abnormal chromosome present in nearly all cases of chronic myeloid leukaemia and some cases of acute lymphoblastic leukaemia. It is formed when part of chromosome 9 (the abl gene) breaks off and attaches itself to part of chromosome 22 (the bcr gene) in a process known as translocation.

Platelets
Tiny disc-like fragments that circulate in the blood and play an important role in clot formation.

Prognosis
An estimate of the likely course of a disease. It provides some guide regarding the chances of curing the disease or controlling it for a given time.

Purpura
Purple spots on the skin, often accompanied by bleeding from the gums. It is caused by a shortage of platelets as well as fragile skin.

Radiotherapy (radiation therapy)
The use of high energy x-rays to kill cancer cells and shrink tumours.

Resistant or Refractory Disease
This means that the disease is not responding to treatment.

Remission
Otherwise known as complete remission. When there is no evidence of disease detectable in the body. Note this is not equivalent to a cure as relapse may still occur.

Spleen
An organ that accumulates lymphocytes, acts as a reservoir for red cells for emergencies, and destroys blood cells at the end of their lifespan. The spleen is found high in the abdomen on the left-hand side. It cannot normally be felt on examination unless it is enlarged. It is often enlarged in diseases of the blood – this is known as splenomegaly.

Splenomegaly
Another term used to describe an enlarged spleen.

Standard therapy
The most effective and safest therapy currently being used.

Stem cells
Stem cells are primitive blood cells that can give rise to more than one cell type. There are many different types of stem cells in the body. Bone marrow stem cells have the ability to grow and produce all the different blood cells including red cells, white cells and platelets.

Stem cell transplant
(peripheral blood stem cell or bone marrow transplant)
These treatments are used to support the use of high-dose chemotherapy and/or radiotherapy in the treatment of a wide range of blood cancers including leukaemias, lymphomas, myeloma and other serious diseases.

Translocation
A chromosomal abnormality in which part of the one chromosome is transferred to another.

T-lymphocyte
A type of white cell involved in controlling immune reactions.

Tumour
An abnormal mass of cells which may be non-malignant (benign) or malignant (cancerous).

Ultrasound
Pictures of the body's internal organs built up from the interpretation of reflected sound waves.

White blood cells (white cells)
Specialised cells of the immune system that protect the body against infection. There are five main types of white blood cells: neutrophils, eosinophils, basophils, monocytes and lymphocytes.

X-ray
A form of electronically produced radiation used in diagnosis and treatment.

Please refer to the 'Dictionary of Terms' booklet for further definitions.

Please send me a copy of the following patient information booklets:

- ☐ Dictionary of Terms
- ☐ Haematology Patient Diary
- ☐ Clinical Trials
- ☐ Autologous Stem Cell Transplants
- ☐ Allogeneic Stem Cell Transplants
- ☐ Myloproliferative Neoplasms
- ☐ Myelodysplastic Syndromes
- ☐ Myeloma
- ☐ Acute Lymphoblastic Leukaemia in Adults
- ☐ Acute Lymphoblastic Leukaemia in Children
- ☐ Acute Myeloid Leukaemia
- ☐ Chronic Lymphocytic Leukaemia
- ☐ Chronic Myeloid Leukaemia
- ☐ Hodgkin Lymphoma
- ☐ Non-Hodgkin Lymphoma
- ☐ Haemochromatosis
- ☐ My Guide to Blood Cancer - for adolescents and young adults

Or information on:

- ☐ Leukaemia & Blood Cancer New Zealand's Support Services
- ☐ How to make a bequest to Leukaemia & Blood Cancer New Zealand

Newsletters:

- ☐ LifeBlood
- ☐ Lymphoma Today
- ☐ Leukaemia Today
- ☐ Myeloma Today

Name: _____

Address: _____

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Email: _____

Send to: Leukaemia & Blood Cancer New Zealand
PO Box 99182, Newmarket, Auckland 1149
Phone: 09 638 3556 or 0800 15 10 15
Email: info@leukaemia.org.nz

Leukaemia & Blood Cancer New Zealand will record your details to facilitate services and keep you informed about blood cancers and conditions. We value your privacy and take all the necessary steps to protect it. You can access, change or delete this information by contacting us at info@leukaemia.org.nz

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Myeloproliferative Neoplasms (MPN)

We hope that you found this information booklet useful. We are interested in what you thought of the booklet, whether you found it helpful or not. If you would like to give us your feedback, please fill out this questionnaire and send it to Leukaemia & Blood Cancer New Zealand, at the address at the bottom of the following page.

1. Did you find this booklet helpful?

☐ Yes ☐ No

Comments _____

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2. Did you find this booklet easy to understand?

☐ Yes ☐ No

Comments _____

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3. Where did you get this booklet from?

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4. Did you have any questions that were not answered in the booklet?

☐ Yes ☐ No

If yes, what were they?

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5. What did you like the most about this booklet?

6. What did you like least about this booklet?

7. Any other comments?

Thank you for helping us review this booklet. We will record your feedback and consider it when this booklet is reviewed for the next edition.

Please return to: Leukaemia & Blood Cancer New Zealand
PO Box 99182 Newmarket, Auckland 1149

Important information for haematology patients

It is important that you contact your doctor or the hospital for advice immediately (at any time of the day or night) if you are feeling very unwell, or if you experience any of the following:

- a temperature of 38°C or over and/or an episode of shivering
- bleeding or bruising, for example blood in your urine, faeces, sputum, bleeding gums or a persistent nose bleed
- nausea or vomiting that prevents you from eating or drinking or taking your normal medications
- diarrhoea, stomach cramps or constipation
- coughing or shortness of breath
- the presence of a new rash, reddening of the skin, itching
- a persistent headache
- a new pain or soreness anywhere
- if you cut or otherwise injure yourself
- if you notice pain, swelling, redness or pus anywhere on your body

leukaemia & blood cancer new zealand

our mission is to care, our vision is to cure

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