Understanding Leukaemias Lymphomas Myeloma

and related blood disorders

A guide for patients and families

Leukaemia Foundation®

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NOTES

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INTRODUCTION

This booklet has been written to provide you and your families with general information about *leukaemias, lymphomas, myeloma* and the related blood disorders.

Some of you may be feeling anxious or overwhelmed if you or someone you care for has been diagnosed with a leukaemia, lymphoma, myeloma or a related blood disorder. This is understandable. 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 also raise others, which you should bring up 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. Because this is a general information booklet, not everything written here will necessarily apply to you and your experience of a disease or its treatment. You need to discuss your particular circumstances at all times with your treating doctor.

Many of you may require more information than is contained in this booklet. There are separate Leukaemia Foundation booklets that provide more detailed information about different types of diseases. These can be obtained by contacting the Leukaemia Foundation. A list of available booklets and contact details for the Leukaemia Foundation are provided in the back of this booklet. We have also included some internet addresses at the back of the booklet that you might find useful. In addition, many of you will receive written information from the doctors and nurses at your treating hospital.

We have used some medical words and terms which you may not be familiar with. These are highlighted in *italics*. Their meaning is explained in the booklet and / or in the glossary of terms at the back of the booklet.

Finally, we hope that you find this information useful and we would appreciate any feedback from you so that we can continue to serve you and your families better in the future.

THE LEUKAEMIA FOUNDATION

The Leukaemia Foundation is the only national not-for-profit organisation dedicated to the care and cure of patients and families living leukaemias, lymphomas, myeloma and related blood disorders. Since 1975, the Foundation has been committed to improving survival for patients and providing much needed support. It does not receive direct ongoing government funding, relying instead on the continued and generous support of individuals and corporations to develop and expand its services.

The Foundation provides a range of **free** support services to patients and their carers, family and friends. This support may be offered over the telephone, face to face at home, hospital or at the Foundation's accommodation centres, depending on the geographical and individual needs. Support may include providing information, patient education seminars and programs that provide a forum for peer support and consumer representation, practical assistance, accommodation, transport and emotional support/ counselling.

The Leukaemia Foundation funds leading research into better treatments and cures for leukaemias, lymphomas, myeloma and related blood disorders. Through its National Research Program, the Foundation has established the Leukaemia and Lymphoma Tissue Bank and the Leukaemia Foundation Research Unit at the Queensland Institute for Medical Research. In addition, the Foundation funds research grants, scholarships and fellowships for talented researchers and rural health professionals.

Support Services

"Foundation staff provide patients and their families with information and support across Australia"

The Leukaemia Foundation has a team of highly trained and caring Support Services staff with qualifications and/or experience in nursing or allied health that work across the country. They can offer individual support and care to you and your family when it is needed.

Support Services may include:

Information

The Leukaemia Foundation has a range of booklets, fact sheets and resources such as this one that are available free of charge. These can be ordered via the form at the back of this booklet or downloaded from the website. Translated versions (in languages other than English) of some booklets and fact sheets are also available from our website.

Education & Support programs

The Leukaemia Foundation offers you and your family disease-specific and general education and support programs throughout Australia. These programs are designed to empower you with information about various aspects of diagnosis and treatment and how to support your general health and well being.

Emotional support/counselling

A diagnosis of a blood cancer/disorder can have a dramatic impact on a person's life. At times it can be difficult to cope with the emotional stress involved. The Leukaemia Foundation's Support Services staff can provide you and your family with much needed support during this time. They may refer you or a loved one to a specialist health professional eg Psychologist if required.

The Foundation has established an on-line information and support group for people living with leukaemia, lymphoma, multiple myeloma, or a related blood disorder. Registration is free and participants can remain anonymous, see www.talkbloodcancer.com

Accommodation

Some patients and carers need to relocate for treatment and may need help with accommodation. The Leukaemia Foundation staff can help you to find suitable accommodation close to your hospital or treatment centre. In many areas, the Foundation's fully furnished self-contained units and houses can provide a 'home away from home' for you and your family.

Transport

The Foundation also assists with transporting patients and carers to and from hospital for treatment. Courtesy cars and other services are available in many areas throughout the country.

Practical Assistance

The urgency and lengthy duration of medical treatment can affect you and your family's normal way of life and there may be practical things the Foundation can do to help. In special circumstances, the Leukaemia Foundation provides financial support for patients who are experiencing financial difficulties or hardships as a result of their illness or its treatment. This assistance is assessed on an individual basis.

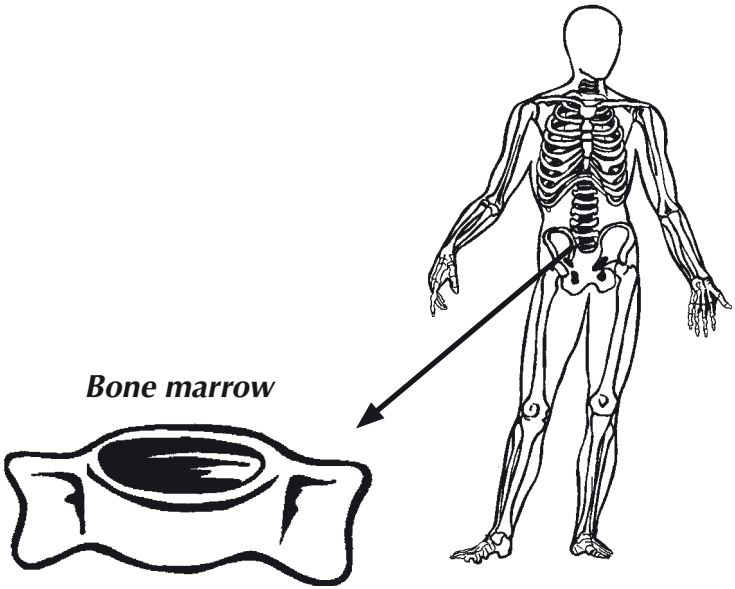
Contacting us

The Leukaemia Foundation provides services and support in every Australian state and territory. Every person's experience of living with these blood cancers and disorders is different. Living with leukaemias, lymphomas or myeloma is not easy, but you don't have to do it alone. Please call 1800 620 420 (Freecall) to speak to a local support service staff member or to find out more about the services offered by the Foundation. Alternatively, contact us via email by sending a message to info@leukaemia.org.au or visit www.leukaemia.org.au

NORMAL BLOOD CELL DEVELOPMENT

There are many different types and sub-types of blood cancers and related blood disorders. They all affect, to a greater or lesser extent, the normal production of blood cells in the bone marrow and the normal function of blood cells circulating in the blood stream.

In this section of the booklet we provide a brief overview of the production and function of blood cells, which we hope will help you to understand your disease better.



Bone marrow

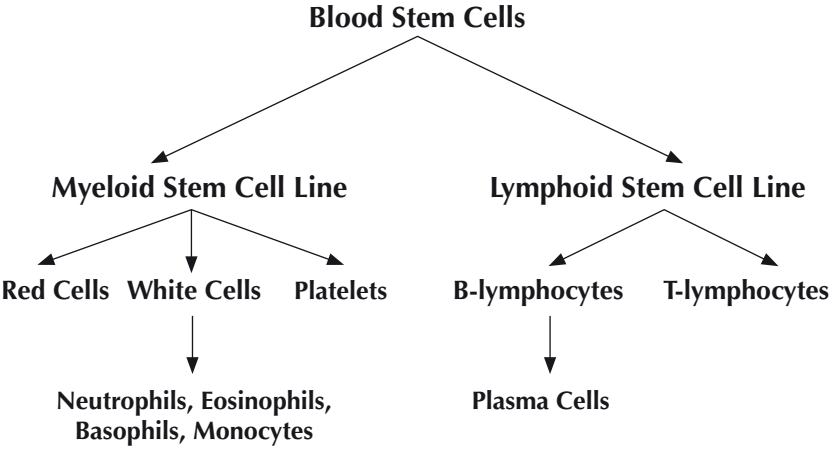
Bone marrow is the spongy tissue that fills the cavities inside your bones. Most of your blood cells are made in your bone marrow. The process by which blood cells are made is called *haemopoiesis*. In infants, haemopoiesis takes place at the center of all bones. In adults, it is limited to the hips, ribs, spine, skull and breastbone (sternum).

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 small in number but are able, when stimulated, not only to divide to replicate themselves, but to grow and divide into slightly more mature stem cells called myeloid stem cells and lymphoid stem cells. These cells multiply and mature further to produce all the circulating blood cells. There are three main types of blood cells; red cells, *white cells* and platelets.

Myeloid ('my-loid') stem cells develop into red cells, white cells (neutrophils, 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.



Growth factors and cytokines

All normal blood cells have a limited survival in circulation and need replaced on a continual basis. This means that the bone marrow remains a very 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.

These days some growth factors can be made in the laboratory (synthesized) 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 erythropoeitin (EPO) stimulates the production of red cells.

Blood

Blood consists of blood cells and plasma. Plasma is the straw coloured fluid part of the blood that blood cells use to travel around your body.

Red cells and haemoglobin

Red cells contain haemoglobin (Hb), which gives the blood its red colour and 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.

The normal haemoglobin range for a man is between 130 and 170 (130 - 170 g/L)
The normal haemoglobin range for a woman is between 120 and 160 (120 - 160 g/L)

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

The normal range of the haematocrit for a man is between 40% and 52%
The normal range of the haematocrit for a woman is between 36% and 46%

Anaemia

Anaemia is a condition caused by a reduction in the number of red cells, which in turn results in a low haemoglobin level. Measuring either the haematocrit or the haemoglobin will provide information regarding the degree of anaemia.

If you are anaemic you will 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 some cases a red cell transfusion may be given to restore the red cell numbers and therefore the haemoglobin to more normal levels.

White cells

White cells fight infection. There are different types of white cells that fight infection together and in different ways.

Granulocytes:	
Neutrophils	*kill bacteria and fungi.*
Eosinophils	*kill parasites.*
Basophils	*work with neutrophils to fight infection.*
Monocyte-Phagocyte system (Lymphocytes)	
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.*

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

The normal adult white cell count varies between 3.7 and 11 (3.7 - 11 x 109/L)

Neutropaenia

Neutropaenia is the term given to describe a lower than normal neutrophil count. If you have a neutrophil count of less than 1 (1 x 109/L), you are considered to be neutropenic and at risk of developing frequent and sometimes severe infections.

The normal adult neutrophil count varies between 2.0 and 7.5 (2.0 - 7.5 x 109/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 a blood vessel is damaged (e.g. by a cut) the platelets gather at the site of injury, stick together and form a plug to help stop the bleeding.

The normal adult platelet count varies between 150 and 400 (150 - 400 x 109/L)

Thrombocytopenia

Thrombocytopenia is the term used to describe a reduction in the platelet count to below normal. If your platelet count drops below 20 (20 x 109/L) you are at increased risk of bleeding, and tend to bruise easily. Platelet transfusions are sometimes given to bring the platelet count back to a safer level.

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

In children, some normal blood cell counts vary with age (see table next page).

Normal range of blood values for children

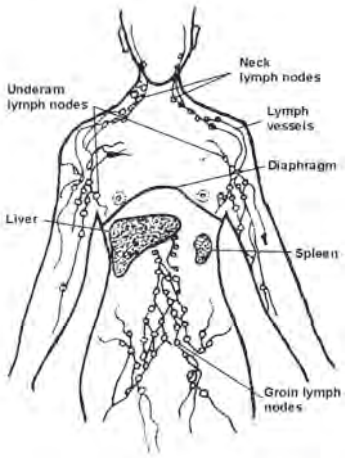
	1 month	1 year	3 years	5 years	9 years	16 years
Haemoglobin (g/L)	102-130	104-132	107-136	110-139	113-143	115-165 F 130-180 M
White cell count (x 109/L)	6.4-12.1	5.4-13.6	4.9-12.8	4.7-12.3	4.7-12.2	3.5-11
Platelets (x 109/L)	270-645	205-553	214-483	205-457	187-415	150-450
Neutrophils (x 109/L)	0.8-4.9	1.1-6.0	1.7-6.7	1.8-7.7	1.8-7.6	1.7-7.0

If your child is being treated for a blood cancer or related disorder 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 range.

THE LYMPHATIC SYSTEM

The *lymphatic* system is made up of a vast network of vessels, similar to blood vessels that branch out into all the tissues of the body. These vessels contain lymph, a colourless watery fluid that carries lymphocytes, specialised white blood cells that fight infection. There are two types of lymphocytes, *B-lymphocytes* and *T-lymphocytes* (are called B-cells and T-cells). These cells protect us by making antibodies and destroying harmful microorganisms like bacteria and viruses. As such, the lymphatic system forms part of the immune system, which protects our bodies against disease and infection.

Clusters of small bean-shaped organs called *lymph nodes* (also known as lymph glands) are found at various points throughout the lymphatic system. The lymph nodes, which are filled with lymphocytes, act as important filtering stations, cleaning the lymph fluid as it passes through them. Here bacteria, viruses and other harmful substances are removed and destroyed. When you have an infection, for example a sore throat, you may notice that the lymph nodes under your jaw bone become swollen and tender. This is because the lymphocytes which live there become activated and multiply in response to the virus or bacteria causing the infection.



The spleen (an organ on the left side of the abdomen), thymus (a gland found behind the breast bone), tonsils and adenoids (glands in the throat) and bone marrow (spongy material inside bones) all contain lymphatic tissue and are therefore considered to be part of the lymphatic system. Lymphatic tissue is also found in other parts of the body.

BLOOD CANCERS

In this section of our booklet we provide a brief overview of blood cancers and related blood disorders. It is important to point out that the information provided here is of a general nature and may not necessarily apply to the specific type or severity of disease you or your loved one has.

Leukaemias

Leukaemias are a group of cancers that affect the blood and bone marrow. All leukaemias start in the bone marrow where developing blood cells, usually developing white cells, undergo a *malignant* (cancerous) change. This means that they multiply in an uncontrolled way, crowding the marrow and interfering with normal blood cell production. Increasing numbers of abnormal cells, called blast cells or *leukaemic blasts* eventually spill out of the bone marrow and travel around the body in the bloodstream. In some cases these abnormal cells accumulate in various organs including the lymph nodes, spleen, liver and central nervous system (brain and spinal cord).

Types of leukaemias

Leukaemias are broadly classified by how quickly the disease develops, and by the type of blood cell involved.

- *Acute leukaemias* develop quickly and need to be treated urgently.
- *Chronic leukaemias* develop more slowly and may not need to be treated for some time after they are diagnosed.
- *Myeloid leukaemias* arise from myeloid stem cells and are characterised by the accumulation of cancerous cells called *myeloblasts*.
- *Lymphoid leukaemias* arise from lymphoid stem cells and are characterised by the accumulation of cancerous cells called *lymphoblasts*.

Therefore there are four main types of leukaemia:

1. Acute myeloid leukaemia (AML)
2. Acute lymphoblastic leukaemia (ALL)
3. Chronic myeloid leukaemia (CML)
4. Chronic lymphocytic leukaemia (CLL)

Incidence

Each year in Australia, about 3,000 adults and around 300 children (0 – 14 years) are diagnosed with leukaemia. The most common types of leukaemia in adults are CLL and AML. ALL is the most common type of leukaemia in children, and the most common type of childhood cancer.

Causes

In most cases, the cause of leukaemias remains unknown, but there are likely to be a number of factors involved. Like all cancers, leukaemias may result from a change in one or more of the genes that normally control the growth and development of blood cells. In a small number of cases, exposure to high levels of radiation and particular chemicals, especially benzene and some chemotherapy drugs used to treat another cancer, may be involved. Some people with pre-existing blood disorders or particular genetic disorders may have a higher chance of developing some types of leukaemia.

Symptoms

The main symptoms of leukaemia are caused by a lack of normal blood cells.

Without enough red cells, normal white cells and platelets people with leukaemia may become fatigued, more susceptible to infections and to bleeding and bruising more easily.

In some cases, people with chronic leukaemias don't have any troublesome symptoms and the disease is picked up during a routine blood test.

Treatment

Treatment varies depending on the exact type of leukaemia involved, the person's age, and their general health.

The main treatment is chemotherapy. This is given to destroy the leukaemic cells and allow the bone marrow to function normally again. Other types of treatment are also used. For example the sub-type of leukaemia called acute promyelocytic leukaemia is treated with chemotherapy and a non-chemotherapy drug called all-trans retinoic acid (ATRA), a derivative of vitamin A which helps make the leukaemic cells either mature properly or die. Chronic myeloid leukaemia is commonly treated with imatinib mesylate, a relatively new drug that works by blocking the leukaemia-causing effects of an abnormal protein found in of CML.

Occasionally a stem cell transplant is used because it gives some people a better chance of cure or long-term control of their disease than chemotherapy alone.

Lymphomas

Lymphomas are cancers of the lymphatic system. Lymphomas arise when developing *lymphocytes* (a type of white blood cell) undergo a malignant (cancerous) change and multiply in an uncontrolled way. Increasing numbers of abnormal lymphocytes, called *lymphoma cells* accumulate and form collections of cancer cells called malignant tumours in lymph nodes and other parts of the body.

Types of lymphomas

There are many different types of lymphoma which are broadly divided into two main groups:

- Hodgkin lymphoma (also known as Hodgkin's disease)
- Non-Hodgkin lymphomas (NHL) – can be divided into B-cell and T-cell types

Incidence

Each year in Australia, over 4,000 people are diagnosed with lymphoma making it the sixth most common type of cancer in men and the fifth most common type of cancer in women. Of these about 3,600 will have a type of B-cell or T-cell lymphoma and, around 400 will have a type of Hodgkin lymphoma.

Lymphomas can develop at any age but the majority of NHL occurs in people over the age of 50 years. The peak age for diagnosis of Hodgkin lymphoma is between 15 and 30 years.

Causes

The incidence of lymphoma is increasing every year. In most cases we don't know why but there are likely to be a number of factors involved. Like all cancers, lymphomas may result from a change in one or more of the genes that normally control the growth and development of blood cells. We know that people with a weakened immune system (either due to an immunodeficiency disease or drugs that suppress the function of the immune system) are at an increased risk of developing lymphomas. Certain types of viral infections may also play a role, especially in people with a weakened immune system.

Symptoms

Lymphomas commonly present as a firm painless swelling of a lymph node (swollen glands), usually in the neck, under the arms or in the groin. Other symptoms may include:

- Recurrent fevers
- Excessive sweating at night
- Unintentional weight loss
- Persistent lack of energy
- Generalised itching

Lymphoma may develop in the lymph nodes in deeper parts of the body like those found in the abdomen (causing bloating), or in the chest (causing coughing, discomfort in the chest and difficulty breathing).

In some cases people don't have any troubling symptoms and the disease is picked up during a routine examination or chest x-ray.

Treatment

Treatment will vary depending on the exact type of lymphoma involved, and how fast it is likely to grow and cause problems in the body. It may also depend on the extent of the disease in a person's body at diagnosis, their age and general health.

Some lymphomas grow slowly and cause few troubling symptoms, and may not need to be treated urgently. Others grow more quickly and need to be treated as soon as they are diagnosed. Treatment can involve chemotherapy, radiotherapy and immunotherapy. Occasionally, a stem cell transplant is used to treat disease which has relapsed (come back), or where there is a high likelihood that the disease will relapse in the future.

Myeloma

Myeloma, or *multiple myeloma*, is a cancer of plasma cells. *Plasma cells* are mature B-lymphocytes that live predominantly in the bone marrow and normally produce antibodies to help fight infection. In myeloma, plasma cells undergo a malignant (cancerous) change and multiply in an uncontrolled way causing problems in different parts of the body. Large numbers of abnormal plasma cells, called myeloma cells, collect in the bone marrow and may interfere with blood cell production and damage the adjacent bones causing pain. Myeloma cells produce an abnormal type of antibody called *paraprotein* that can usually be detected in blood and/or urine.

Incidence

Each year in Australia over 1,100 people are diagnosed with myeloma. The majority of those diagnosed (almost 80 per cent) are over the age of 60.

Causes

In most cases, the cause of myeloma remains unknown, but there are likely to be a number of factors involved. Like all cancers, myeloma may result from a change in one or more of the genes that normally control the growth and development of blood cells. In a small number of cases, exposure to high doses of radiation and ongoing exposure to certain industrial or environmental chemicals may be involved.

Symptoms

The most common of myeloma symptom is bone pain. Other common symptoms include persistent tiredness and fatigue due to anaemia and kidney failure or frequent infections.

The symptoms of multiple myeloma depend on how advanced the disease is. In the earliest stages, there may be no symptoms and myeloma is picked up during a routine blood test.

Treatment

The main form of treatment is chemotherapy, usually in combination with cortico-steroids. High dose chemotherapy followed by stem cell transplantation is also used for younger patients (under 75 years) who are fit enough and would benefit by this type of treatment. Thalidomide and other newer drugs may also be used to help keep myeloma under control or in some cases be used as part of initial treatment. Other types of treatment are also used depending on the kinds of problems myeloma is causing in the body. Drugs called *bisphosphonates* are a standard part of therapy used to strengthen bones. Radiotherapy and surgery may also used to prevent and treat problems caused by bone damage.

RELATED BLOOD DISORDERS

Myelodysplastic syndromes

Myelodysplastic syndromes (MDS) are a group of disorders that affect normal blood cell production in the bone marrow. In MDS, the bone marrow produces *too* few red cells, white cells and / or platelets, and an excess of immature blood cells known as blast cells.

There are several different types of MDS and the disease can vary in its severity, and the extent to which blood cell production is disrupted. In mild cases, people may be anaemic, or have few symptoms of their disease. In more severe cases, a shortage of circulating blood cells can cause severe anaemia, increased susceptibility to infection, and bruising and bleeding more easily.

Furthermore, in up to 30 per cent of cases, MDS can progress to a type of leukaemia. Because of this it is sometimes called a pre-leukaemic disorder.

While MDS can occur at any age, the majority of cases (over 90 per cent) develop over the age of 60 years.

Causes

MDS occurs as a result of a change (or mutation) in one or more of the genes that normally control the growth and development of blood cells. The precise reasons for this change (or changes) remain unclear but there are likely to be a number of factors involved. Increasing age remains the greatest risk factor for developing primary MDS, thus the longer we live the greater the chance we have of acquiring the sorts of mutations likely to cause MDS. Exposure to high levels of benzene, petroleum products and cigarette smoking have also been linked to the development of MDS.

People who have been previously treated for cancer or other conditions with cytotoxic chemotherapy are at an increased risk of developing what is called secondary or treatment-related MDS.

Symptoms

In general the types of symptoms that people experience depends on the severity of their disease, and the type of blood cell which is most affected. In many cases MDS develops slowly, people do not have any symptoms and the disease is picked up during a routine blood test.

The most common symptoms are those caused by anaemia, and include: persistent tiredness, dizziness, paleness or shortness of breath when physically active. Other symptoms may include frequent or repeated infections and slow healing, and increased or unexplained bleeding or bruising.

Treatment

Treatment will vary depending on several factors including the severity of disease, an estimation of its likely course and the chances of curing or controlling it for a given time. Other important factors considered include the person's age and general health.

Many people, particularly in the early stages of MDS don't have any symptoms and don't need to be treated. In these cases the doctor may simply recommend regular checkups to carefully monitor their health.

In more severe or progressive disease, chemotherapy may be used to control a rising blast cell count, and allow the bone marrow to resume normal blood cell production. This may involve low dose chemotherapy given in tablet form, or more intensive treatment using a combination of drugs given intravenously (into a vein), similar to those used to treat acute myeloid leukaemia. More recently newer drugs including azacitidine, decitabine, lenalidomide and thalidomide have been used in the treatment of MDS.

The main treatment for the majority of people with MDS is supportive care. This involves the use of antibiotics to treat infection and where necessary blood transfusions to replenish vital numbers of red cells and platelets. In some cases growth factors are used to promote normal blood cell production in the bone marrow.

A stem cell transplant is used in selected younger cases. This type of treatment may increase the chance of cure for some people with MDS.

Myeloproliferative disorders

Myeloproliferative disorders are a group of disorders that affect normal blood cell production in the bone marrow. In myeloproliferative disorders, the bone marrow produces too many of one or more types of blood cells (red cells, white cells or platelets). When present in large numbers, these cells cannot function properly and cause various problems in the body.

There are several different types of myeloproliferative disorders. They are generally distinguished from each other by the type of cell which is most affected.

- Essential thrombocythemia involves an overproduction of platelets.
- Polycythemia rubra vera involves an overproduction of red cells and frequently platelets and/or white blood cells.
- Myelofibrosis causes excessive blood cell production which damages bone marrow tissue and is gradually replaced with abnormal fibrous tissue.
- Other less common types of myeloproliferative disorders also exist.

In most cases these blood disorders develop slowly and get worse gradually over many years. In rare cases myeloproliferative disorders can progress to leukaemia.

While myeloproliferative disorders can occur at any age, the majority of cases occur between the ages of 40 and 60 years. They are uncommon under the age of 20 years and rarely occur in children.

Causes

The precise cause of myeloproliferative disorders remains unknown but there are likely to be a number of factors involved including a mutation or change in one or more of the genes that normally control the growth and development of blood cells.

Symptoms

Symptoms vary depending on the particular type of myeloproliferative disorder involved.

Symptoms of an enlarged spleen (splenomegaly) are common and include feelings of discomfort, pain or fullness in the upper left-side of the abdomen. Excess circulating blood cells can cause easy bruising and bleeding, or blood clotting problems.

Treatment

Treatment will vary depending on the type of myeloproliferative disorder and its severity, the person's age and their general health. Treatment is generally aimed at reducing excess numbers of blood cells in circulation, and preventing and treating any symptoms and complications of the disease. It may include the use of oral chemotherapy drugs or other agents such as interferon, aspirin or anagrelide. Sometimes patients may also need to have the regular removal of small quantities of blood via a procedure known as venesection (this is a very similar procedure to donating blood at the Red Cross but is done at the treating hospital).

Aplastic anaemia

Aplastic anaemia is a rare disorder in which the bone marrow fails to produce enough blood cells (red cells, white cells and platelets).

Causes

In the majority of cases aplastic anaemia is an acquired disorder that develops at some stage in the person's life. This means that it is usually not inherited (passed down from parent to child), and it is not present at birth. Several potential triggers for the development of aplastic anaemia have been identified and these include viruses, radiation exposure and exposure to certain chemicals and drugs. In many cases however the cause remains unknown. Fanconi Anaemia is a rare inherited form of aplastic anaemia.

Although aplastic anaemia is not a malignant disease (not a cancer) it can be very serious, especially if the bone marrow is severely affected and there are very few blood cells being produced.

Symptoms

Without adequate numbers of circulating blood cells people with aplastic anaemia can become anaemic, more susceptible to infections and to bleeding and bruising more easily.

Treatment

The treatment for aplastic anaemia depends on several factors including the cause of the disease (if this can be identified), its severity and the age and general health of the patient. Immunosuppressants (drugs which affect the function of the immune system) are commonly used.

In some cases an allogeneic stem cell transplant may be recommended as a curative option for younger people. This involves replacing abnormal blood stem cells with healthy ones from a suitable donor. Supportive therapies are also important and include where necessary, blood transfusions to replace circulating blood cells and antibiotics to treat infections.

Amyloidosis

Amyloidosis is the general name given to a group of disorders in which an abnormal protein 'amyloid' builds up in the blood and is deposited in organs and tissues around the body. These deposits progressively accumulate and disrupt the normal function of the tissues, eventually leading to organ failure. The organs most commonly affected include heart, liver, kidneys, nervous system and the gut.

While amyloidosis is not a type of cancer, it is a very serious and life threatening disorder.

There are three main types of amyloidosis. In Systemic AL Amyloidosis (also known as primary amyloidosis) amyloid deposits are derived from abnormal plasma cells in the bone marrow. AA Amyloidosis (also known as secondary amyloidosis) results from a chronic inflammatory disease (for example chronic arthritis) or infection (for example tuberculosis, osteomylitis, familial Mediterranean fever) in the body. ATTR Amyloidosis is an inherited disease (passed down from one generation to the next). In this case amyloid is derived from an abnormal transthyretin protein which made in the liver. Other types of amyloidosis also exist.

Symptoms

Symptoms are often vague and they can vary considerably from person to person. Fatigue, unexplained weight loss and oedema (fluid retention commonly in the lower limbs and ankles) are common. Other symptoms vary depending on the organ or

tissues most affected and may include shortness of breath, loss of appetite, enlarged tongue, unexplained bruising around the eyes and numbness or tingling in the hands and feet. Because amyloidosis is relatively rare and many of the symptoms mimic other diseases, amyloidosis often goes undiagnosed until the patient is very sick.

Treatment

The choice of treatment will depend on the type of amyloidosis involved and the extent and severity of organ involvement. Although there is currently no cure for amyloidosis there are effective treatments that can help to slow down the progress of the disease and support and preserve organ function.

Clinical trials play an increasingly important role in the treatment of all types of amyloidosis. The development of new and improved treatments means that the outlook for people with amyloidosis is gradually improving.

The main aims of treatment for amyloidosis are twofold; to reduce the production of the abnormal amyloid-forming protein by treating any underlying disease, and to support and preserve normal organ function. In systemic AL amyloidosis, chemotherapy and in some cases an autologous stem cell transplant may be used to destroy abnormal plasma cells that are making the abnormal amyloid proteins. Cortico-steroids and a drug called thalidomide may also be used.

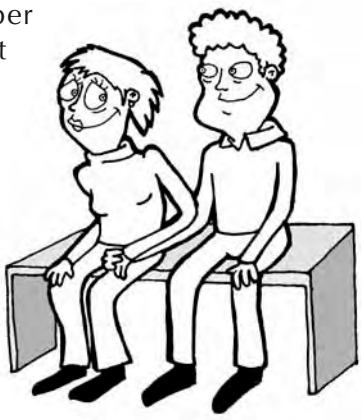
In AA amyloidosis, treatment is used to control an underlying disease or infection. In inherited forms of ATTR amyloidosis, treatments are aimed at supporting existing organ function, sometimes in association with organ transplantation.

MAKING TREATMENT DECISIONS

Many people feel overwhelmed when they are diagnosed with a blood cancer or related disorder. 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.

Before going to see your doctor make a list of the questions you want to ask. It is handy to keep a notebook or some paper and a pen handy as many questions are thought of in the early hours of the morning.

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



Your treating doctor (haematologist) will spend time discussing with you and your family 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 well-being. 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.

INFORMATION AND SUPPORT*

People cope with a diagnosis of a blood cancer or related disorder in different ways, and there is no right or wrong or standard reaction. It is not uncommon to feel concerned, afraid, angry or confused.

It is worth remembering that information can often help to take away the fear of the unknown. It is best for patients and families to speak directly to their doctor regarding any questions they might have about their disease or treatment. It can also be helpful to talk to other health professionals like social workers or nurses who have been specially educated to take care of people with diseases like yours. Some people find it useful to talk with other patients and family members who understand the different feelings and issues that come up for people living with an illness of this nature.

If you have a psychological or psychiatric condition, please inform your doctor and request additional support from a mental health professional.

Many people are concerned about the social and financial impact of the diagnosis and treatment on themselves and their loved ones. Medical appointments and traveling to and from the hospital can be inconvenient, costly and they can interfere with your everyday life. In some cases normal family routines are disrupted and other members of the family may suddenly have to fulfill roles they are not familiar with, for example cooking, cleaning, doing the banking and taking care of children.

Many hospitals have psychologists, social workers and pastoral care workers who can assist you and your loved ones in coping better with any psychological, emotional or financial difficulties you may be experiencing.

There are a variety of programs designed to help ease the emotional and financial strain created by cancer. The Leukaemia Foundation is there to provide you and your family with information and support to help you cope during this time. The Leukaemia Foundation's support service coordinators are at hand to help and are just a phone call away. Contact details for your state office of the Leukaemia Foundation are provided on the back of this booklet.

**There is a separate Leukaemia Foundation booklet called 'Living with Leukaemias, Lymphomas, Myelomas and Related Disorders'. This booklet addresses the impact of the diagnosis, family matters, support, survivorship, and other general issues around treatment.*

USEFUL INTERNET ADDRESSES

- American Cancer Society
  www.cancer.org
- Amyloidosis Australia
  www.amyloidosisaustralia.org/
- Amyloidosis Support Network
  www.amyloidosis.org
- Aplastic Anaemia & MDS International Foundation
  www.aamds.org
- Arrow Foundation
  www.arrow.org.au
- Australian Bone Marrow Donor Registry
  www.abmdr.org.au
- Australian Clinical Trial's Registry
  www.actr.org.au/about.aspx
- Australian Cord Blood Bank
  www.sch.edu.au/departments/acbb
- Australian Organization for Young People Living with Cancer (CANTEEN)
  www.canteen.org.au
- Bone & Marrow Transplant Information Network
  www.bmtinfonet.org
- Bone Marrow Transplant Network NSW
  www.bmtnsw.com.au
- Camp Quality
  www.campquality.org.au
- CancerBACUP (A UK cancer information site)
  www.cancerbacup.org.uk
- Cancer Council of Australia
  www.cancercouncil.org.au
- Cancer Voices New South Wales (Consumer organisation)
  www.cancervoices.org.au
- Centre for Grief and Loss
  www.grief.org.au

- CLL Global Research Foundation
  www.cllglobal.org
- International Myeloma Foundation (IMF)
  www.myeloma.org
- International Waldenstrom's Macroglobinemia Foundation
  www.iwmf.com
- Leukaemia Foundation
  www.leukaemia.org.au
- Leukaemia Foundation in Australia – on line forum
  www.talkbloodcancer.com
- Leukaemia Foundation's Network for Young Adults
  www.teamrevive.com
- Leukemia & Lymphoma Society of America
  www.leukemia-lymphoma.org
- Leukaemia Research Fund (UK)
  www.lrf.org.uk
- Look Good … Feel Better program
  www.lgfb.org.au
- Make-a-Wish Foundation of Australia®
  www.makeawish.org.au
- Myelodysplastic Syndromes Foundation
  www.mds-foundation.org
- Myeloproliferative disorders Australia
  www.mpd-oz.org
- Myeloproliferative disorders (online resource for MPD information)
  www.mpdinfo.org
- National Cancer Institute
  www.cancer.gov/cancerinfo/
- NSW Health – NSW IPTAAS
  www.health.nsw.gov.au (and follow the links)
- Redkite (previously the Malcolm Sargent Cancer Fund for Children)
  www.redkite.org.au
- Starlight Children's Foundation of Australia
  www.starlight.org.au

GLOSSARY OF TERMS

Alopecia

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

Anaemia

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

Antibodies

Naturally produced substances in the blood, made by white blood cells called B-lymphocytes or B-cells. Antibodies target antigens on other substances such as bacteria, viruses and some cancer cells and cause their destruction.

Bisphosphonates

A group of drugs commonly used to treat and prevent osteoporosis. These drugs work by protecting the bone surfaces from the action of osteoclasts, cells normally involved in bone breakdown.

B-lymphocyte (B-cell)

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

Bone marrow

The tissue found at the center of many flat or big bones of the body. The bone marrow contains stem cells from which all blood cells are made.

Blood count

A routine blood test that measures the number and type cells circulating in the blood.

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 plastic tube which can be inserted into a vein to allow fluid to enter the blood stream.

Central venous catheter (CVC)

Also known as a central venous access device (CVAD). A line tube passed through the large veins of the neck, chest or groin and into the central blood circulation. It can be used for taking samples of blood, giving intravenous fluids, blood, chemotherapy and other drugs without the need for repeated needles.

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. Most side-effects of are temporary and reversible.

Complete remission

Anti-cancer treatment has been successful and so much of the disease has been destroyed that it can no longer be detected using current technology.

Cure

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

Disease progression

Where the disease is getting worse on or off treatment.

Growth factors

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 cell transplantation. For example G-CSF (granulocyte colony stimulating factor).

Haemopoiesis

The formation of blood cells.

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.

Hyperviscosity

Increased viscosity (thickness) of the blood, usually caused by a build up of paraprotein in the blood. Blood flow becomes more sluggish ands the blood supply to various parts of the body including the brain and eyes may be affected.

Immune system

The body's defense system against infection and disease.

Immunoglobulins

Proteins produced by plasma cells which function as antibodies and play an important role in protecting the body against infection and disease.

Leukaemia

A cancer of the blood and bone marrow characterised by the widespread, uncontrolled production of large numbers of abnormal and / or immature 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.

Leukaemic blasts

Abnormal blast cells which multiple in an uncontrolled manner, crowding out the bone marrow and preventing it from producing normal blood cells. These abnormal cells also spill out into the blood stream and can accumulate in other organs.

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.

Lymphatic system

A vast network of vessels, similar to blood vessels, that branch out into all the tissues of the body. These vessels carry lymph, a colourless watery fluid that carries lymphocytes, specialised white cells that protect us against disease and infection. The lymphatic system is part of the body's immune system.

Lymphocytes

Specialised white cells which are 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.

Lymphoma

Cancer that arises in the lymphatic system.

Malignancy

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

Myeloma

Also called multiple myeloma or myelomatosis. Myeloma is a cancer that usually arises in the bone marrow when mature B-lymphocytes known as plasma cells, undergo a malignant change.

Neutropaenia

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

Neutrophils

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

Paraprotein

Also called monoclonal immunoglobulin, myeloma protein, or M protein. Paraprotein is the abnormal protein produced by myeloma cells.

Pathologist

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

Plasma cells

Mature B-lymphocytes that have become activated in response to bacteria, viruses and other substances in the body. Plasma cells secrete antibodies that help protect the body from infection and disease.

Plasmapheresis

A procedure that uses a special machine called a 'cell separator' to remove the straw-coloured fluid part of the blood (plasma) while returning the rest of the blood and a suitable plasma substitute to the patient.

Prognosis

An estimate of the likely course of a disease.

Radiotherapy (radiation therapy)

The use of high energy x-rays to kill cancer cells and shrink tumours.

Relapse

The return of the original disease.

Resistant or refractory disease

The disease is not responding to treatment.

Remission

When there is no evidence of disease detectable in the body. This is not the same as 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 hypersplenism.

Splenomegaly

Another term used to describe an enlarged spleen.

Stable disease

When the disease is stable it is not getting any better or worse with treatment.

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 (blood) 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, certain solid tumours, and other serious diseases.

T-lymphocyte

A type of white cell involved in controlling immune reactions.

White cells

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

X-ray

A form of radiation used in diagnosis and treatment.

Leukaemia Foundation
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MISSION TO CARE

A BEQUEST
Your planned gift to the Leukaemia Foundation

A wonderful way to make a significant gift is through a bequest in your will. After making due allowance for loved ones, a bequest of a specific amount or a proportion of the residue of your estate, is a way of leaving a real and lasting legacy to the future.

Your bequest to the Leukaemia Foundation will be used to support our mission to care for patients, carers and families and help us achieve our vision to find a cure for leukaemias, lymphomas, myeloma and related blood disorders.

Wording your bequest to the Leukaemia Foundation

You may choose to make a general bequest and allow the Leukaemia Foundation to decide how your bequest will be used, or you may prefer to make that decision yourself e.g. direct your bequest to patient support or research. Your legal adviser can provide further information on the different types of bequests, and on the appropriate wording for a bequest.

As a guide, the following wording may be useful:

'I give and bequeath free of all duties (here state the amount/percentage or share/residue or assets to be gifted) to the Leukaemia Foundation of (here insert the address) absolutely -

- for the general charitable purposes of the said Foundation (this is the Leukaemia Foundation's preferred option); or
- for the purpose of patient and family support; or
- for the purpose of research into the cause, cure or treatment of leukaemia, lymphoma, myeloma and related blood disorders

and I direct that a receipt of the proper officer for the time being of the Leukaemia Foundation shall be a good and sufficient discharge to my trustee/s'.

Please see the next page for the response form.

Response Form

- ❑ I have already made a bequest to the Leukaemia Foundation in my will
- ❑ I am considering/it is my intention to make (please circle) a bequest to the Leukaemia Foundation
- ❑ I would like more information about making a bequest and/or where to direct my bequest
- ❑ I would like to speak to the Planned Giving Manager about appropriate recognition for my bequest
- ❑ I would like to receive invitations to functions

Dr/Mr/Mrs/Ms/Miss: .....

Address: .....

..... Postcode.....

Telephone: (h).....

(w) .....

Email: .....

Please return this form to the:

Planned Giving Manager,
The Leukaemia Foundation,
GPO Box 9954,
in your Capital City
(marked Private & Confidential)

If you are interested in leaving a bequest to the Leukaemia Foundation in your will and you would like further information, without any obligation, in strictest confidence, please contact the Planned Giving Manager in your state on Freecall 1800 620 420 .

Leukaemia Foundation
VISION TO CURE
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The Leukaemia Foundation is the only national not-for-profit organisation dedicated to the care and cure of patients and families living with leukaemias, lymphomas, myeloma and related blood disorders.

You can help by making a donation. Please fill out the form below or visit www.leukaemia.org.au to make your gift online.

Dr/Mr/Mrs/Ms/Miss: .....

Address: .....

..... Postcode.....

Telephone: (h).....

(w) .....

Email: .....

Please accept my tax deductible donation for $ .....

My cheque, made payable to the Leukaemia Foundation, is enclosed, or please charge $..... to my credit card:

❑ Bankcard ❑ Visa ❑ Mastercard ❑ Amex ❑ Diners

__ __ __ __ / __ __ __ __ / __ __ __ __ / __ __ __ __

Cardholder's name: .....

Cardholder's signature: .....

Expiry date: ...../.....

Contact Telephone number: .....

Please send to:
The Leukaemia Foundation
GPO Box 9954
in your Capital City.

Please send me a copy of the following information booklets:

- ❑ Living with Leukaemias, Lymphomas, Myeloma & Related Blood Disorders: Information & Support
- ❑ Understanding Leukaemias, Lymphomas, Myeloma and Related Blood Disorders
- ❑ Understanding Acute Myeloid Leukaemia
- ❑ Understanding Acute Lymphoblastic Leukaemia in Adults
- ❑ Understanding Acute Lymphoblastic Leukaemia in Children
- ❑ Understanding Chronic Lymphocytic Leukaemia
- ❑ Understanding Chronic Myeloid Leukaemia
- ❑ Understanding Lymphomas (Non-Hodgkin's Lymphomas or B-cell and T-cell lymphomas)
- ❑ Understanding Hodgkin Lymphoma
- ❑ Understanding Allogeneic Transplants
- ❑ Understanding Autologous Transplants
- ❑ Understanding Myelodysplastic Syndromes
- ❑ Understanding Myeloma
- ❑ Young Adults with a Blood Cancer
- ❑ Eating Well: A practical guide for people living with leukaemias, lymphomas and myeloma

Or information about:

- ❑ The Leukaemia Foundation's Support Services
- ❑ Workplace giving
- ❑ Regular deduction scheme
- ❑ National fundraising campaigns
- ❑ Community fundraising opportunities
- ❑ Volunteering
- ❑ Receiving the Foundation's newsletters

Name: .....

Street or Postal Address:.....

Suburb.....

State/Postcode .....

Email:..... Tel: (....).....

Please send to:
Leukaemia Foundation, GPO Box 9954, In Your Capital City

or Freecall 1800 620 420

or email: info@leukaemia.org.au

Further information is available on the Leukaemia Foundation's website **www.leukaemia.org.au**

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Leukaemia Foundation®

VISION TO CURE
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This information booklet is produced by the Leukaemia Foundation and is one in a series on leukaemias, lymphomas, myeloma and related blood disorders.

Some booklets are also available in other languages. Copies of this booklet and the other booklets can be obtained from the Leukaemia Foundation in your state by contacting us on

Freecall: 1800 620 420
Email: info@leukaemia.org.au
Website: www.leukaemia.org.au

The Leukaemia Foundation is a non-profit organisation that depends on donations and support from the community.

Please support our work by calling 1800 620 420
or by mailing your donation to:
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in your capital city

October 2008