Understanding Myeloma

A guide for patients and families

Leukaemia Foundation
VISION TO CURE
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NOTES

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Acknowledgements

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INTRODUCTION

This booklet has been written to help you and your family understand more about myeloma. Myeloma, also known as multiple myeloma, is a plasma cell (type of blood cell) cancer that usually arises in the bone marrow.

Some of you may be feeling anxious or a little overwhelmed if you or someone you care about has been diagnosed with myeloma. This is understandable. 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 which you may not be familiar with. These are highlighted in *italics*. Their meaning is explained in the booklet or in the glossary of terms at the back of the booklet.

In some parts of the booklet we have provided additional information you may wish to read on selected topics. This information is presented in the shaded boxes. Some of you 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 treating hospital.

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

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 with 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 giving 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 PwC Foundation Leukaemia and Lymphoma Tissue Bank and the Leukaemia Foundation Research Laboratory at the Queensland Institute for Medical Research.

The Foundation also 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 at the Foundation's accommodation centres across Australia.

The Leukaemia Foundation has an extensive team of cancer nurses and allied health professionals working across the country. They offer personalised support and care to you and your family right throughout the journey.

The support services provided include:

Information, Education and Support

Support is provided by telephone and through visits to hospitals, office or home, referral to specialist services and practical assistance.

The Foundation also offers you and your family free disease-specific or general education programs and workshops on subjects such as Taking Control, Caring for the Carer, survivorship issues and grief. We also coordinate patient and family support groups, and have established an on-line support group at www.talkbloodcancer.com.

The Leukaemia Foundation has a range of booklets 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 are also available from our website.

Accommodation

Accommodation and other housing arrangements close to hospitals are available if you have to relocate for treatment. The self contained modern units and houses are fully furnished, providing a 'home away from home' for you and your family.

Transport

Courtesy cars and other forms of transport support are available in many areas to transport patients and carers to and from hospital for treatment.

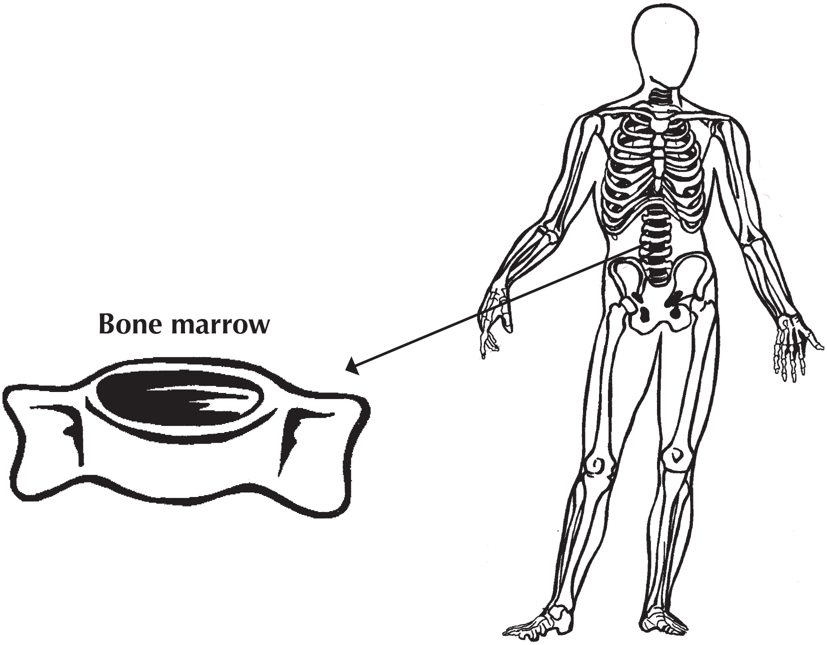
Practical Assistance

Leukaemias, lymphomas and myeloma can affect you and your family's normal way of life and there may be practical things the Foundation can do to help. By working with you, your doctors and social workers, the Foundation can find out the best way to care for you and your family. In special circumstances, the Leukaemia Foundation provides financial support for patients who are experiencing financial difficulties.

Contacting us

The Leukaemia Foundation provides services and support in every Australian state and territory. Every person's experience of living with these 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

BONE MARROW, STEM CELLS AND BLOOD CELL FORMATION



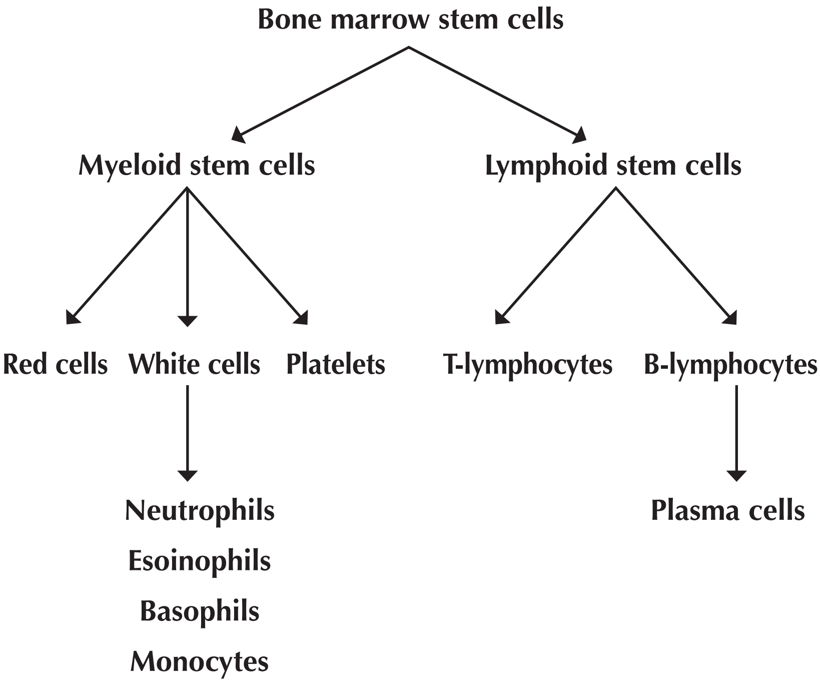
Bone marrow

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 centre of all bones. In adults, it is limited to the hips, ribs, spine, skull and breastbone *(sternum)*. Some of you may have had a *bone marrow* biopsy taken from the bone at the back of your hip (the *iliac crest*) or the breastbone.

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 *stem cells develop into red cells, white cells (neutrophils, eosinophils, basophils and monocytes) and platelets.*

Lymphoid *stem cells develop into two other types of white cells called T-lymphocytes and B-lymphocytes. Some B-lymphocytes develop further into* plasma cells, *which are the antibody-producing cells. Antibodies are substances that target bacteria, viruses and other harmful substances and help remove them from the body.*



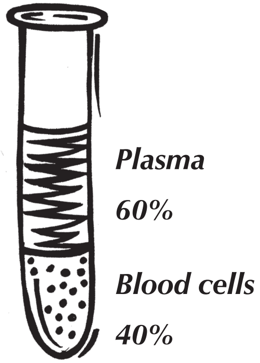
Growth factors and cytokines

All normal blood cells have a limited survival in circulation and need to be 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 (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 *erythropoeitin* (EPO) stimulates the production of red cells. Drugs to stimulate platelet production have been less successful but research is continuing in this area.

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 in a man is between 40% and 52%

The normal range of the haematocrit in 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. 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 this situation 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.

Neutrophils	*kill bacteria and fungi*
Eosinophils	*kill parasites*
Basophils	*work with neutrophils to fight infection*
Monocytes	*work with neutrophils and lymphocytes to fight infection; they also help with antibody production and act as scavengers to remove dead tissue*
T-lymphocytes	*kill viruses, parasites and cancer cells; produce cytokines*
B-lymphocytes	*make antibodies which target microorganisms*

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 10^9/L)

Neutropenia

Neutropenia 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 seriously 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 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 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 10^9/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 risk of bleeding, and tend to bruise easily. Platelet transfusions are sometimes given to bring the platelet count back to a safe 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 blood type.

WHAT IS MYELOMA?

Myeloma, also known as *multiple myeloma*, is a cancer of *plasma cells* (mature B-lymphocytes) that usually arises in the bone marrow. Myeloma develops when plasma cells undergo a cancerous, or malignant change and become *myeloma cells*. These myeloma cells multiply without any proper order, forming collections known as tumours that accumulate in different parts of the body, especially in the bone marrow and on the surfaces of different bones in the body. These tumours secrete chemicals that stimulate other bone marrow cells (osteoclasts) to remove calcium from the bone. As a result bones can become weaker, more brittle and break more easily.

Under normal conditions, plasma cells produce *immunoglobulins* or antibodies that help protect the body from infection and disease. Myeloma cells produce an abnormal type of immunoglobulin called *paraprotein*, (also known as *monoclonal immunoglobulin*, *myeloma protein*, or simply *M protein*). This can be detected in the blood. Sometimes excessive amounts of fragments of immunoglobulin known as *light chains* are produced. These light chains can be detected in the blood and they also appear in the urine. Light chains detected in the urine are called Bence-Jones protein. Some light chains can cause kidney damage.

As myeloma cells multiply, they crowd the bone marrow and prevent it from making normal numbers of red cells, white cells and platelets. Myeloma cells can also interfere with the production of normal antibodies. This can make people with myeloma anaemic, more susceptible to infections and to bleeding and bruising more easily.

What are plasma cells and immunoglobulins? (Imm-you-no-glob-you-lins)

Plasma cells are a type of blood cell that develops from mature B-lymphocytes in the bone marrow. They play an important role in protecting the body against infection and disease by producing proteins called immunoglobulins (Ig), also known as antibodies.

Immunogblobulins (Ig) *are produced by plasma cells in response to bacteria, viruses and other harmful substances found in the* *body. Once released into the blood stream, immunoglobulin circulates about and attaches itself to the target for which it was originally made (the target antigen). This makes it easier for other white blood cells to destroy harmful organisms and other unwanted substances and remove them from the body.*

There are different types of plasma cells that develop in response to different foreign substances (for example bacteria and viruses) within the body. They produce different types of immunoglobulins specific to the substance recognised as foreign and potentially harmful to the body. Each type of immunoglobulin is designated a letter (IgA, IgD, IgE, IgG and IgM) and has a slightly different function.

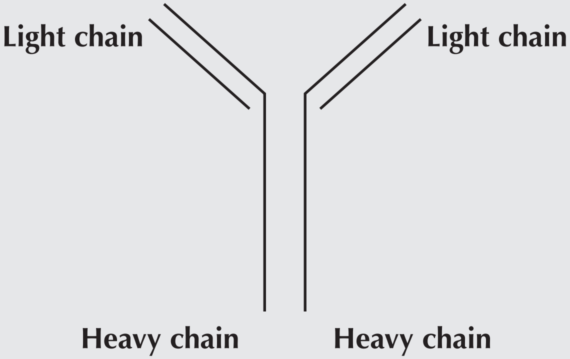
Immature B-lymphocytes
↓ *develop into*

Mature B-lymphocytes
↓ *which develop into*

Plasma cells
↓ *which produce*

Immunoglobulin (antibody)
(IgA, IgG, IgD, IgM and IgE)

Immunoglobulins are Y-shaped molecules made up of two heavy chains and two light chains (see below). There are five main families of immunoglobulins which are named after the heavy chains that form an important part of their structure. These are: IgA, IgG, IgD, IgM and IgE. There are two classes of light chains: kappa (κ) and lambda (λ).



WHO GETS IT?

The majority of people diagnosed with myeloma (93%) are over the age of 50. Myeloma is rare under the age of 40 and it has been reported in children or adolescents. Myeloma is more common in men than women.

HOW COMMON IS IT?

Myeloma is a relatively rare disease. It accounts for approximately 1 per cent of all cancers and 10 per cent of all blood and bone marrow cancers. In 2001, 1115 people were diagnosed with myeloma in Australia.

WHAT CAUSES MYELOMA?

Many people who are diagnosed with myeloma ask the question “why me? “ Naturally, they want to know what has happened or what they might have done to cause their disease. The truth is that no one knows exactly what causes myeloma. We do know that it is not contagious. You cannot ‘catch’ myeloma by being in contact with someone who has it. In most cases people who are diagnosed with myeloma have no family history of the disease.

There are certain factors that *may* put some people at a higher risk of developing this disease. These include exposure to high doses of radiation and ongoing exposure to certain industrial or environmental chemicals.

WHAT ARE THE SYMPTOMS OF MYELOMA?

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

Bone pain

The most common symptom of myeloma is bone pain. This is usually felt in the back or ribs and may be made worse by movement. Bone pain is usually the result of the gradual erosion of bone caused by substances secreted by myeloma cells. Over time bones can become weakened and thinned *(osteoporosis)* and holes *(lytic lesions)* may develop, increasing the risk of fracture (breaking of bone).

When bone tissue is damaged, calcium is released from the bone into the blood stream. An excess of calcium in the blood is called *hypercalcaemia*. If you have a higher than normal calcium level in your blood you may feel nauseated, constipated, tired, thirsty or even confused. There are several ways to prevent and reduce bone damage and its related problems and these are discussed later in this booklet.

Other symptoms of myeloma arise when these cancer cells crowd the bone marrow and prevent it from making normal red cells, white cells and platelets. This can lead to anaemia, frequent or repeated infections and to bleeding or bruising more easily.

Anaemia

A low haemoglobin level in the blood can cause symptoms of anaemia. These include a lack of energy, persistent tiredness and fatigue, weakness, dizziness or feeling unusually short of breath when physically active. In addition, people with anaemia often have a pale complexion.

Frequent or repeated infections

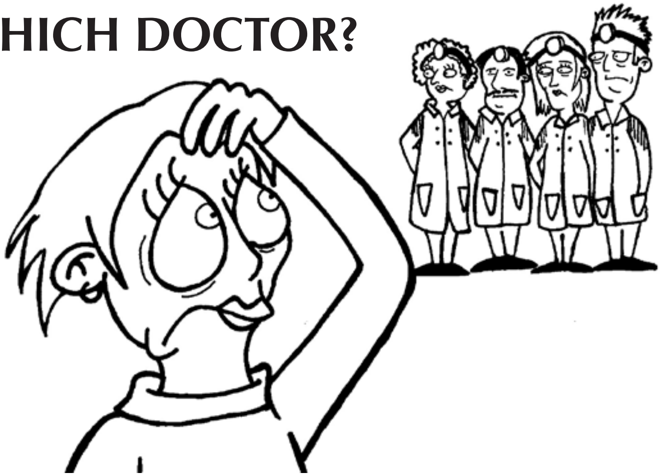
People with myeloma are at risk of developing frequent or repeated infections. This is because their bone marrow may be unable to produce adequate numbers of white cells and because they may be unable to produce adequate amounts of normal antibody. Common infections include chest infections, urinary tract infections and skin infections.

Increased bleeding or bruising

A very low platelet count can cause bruising for no apparent reason, or excessive or prolonged bleeding following minor cuts or injury. Some people notice bruises on their skin for no apparent reason, or more frequent or severe nose bleeds, bleeding gums or excessive or prolonged bleeding following minor cuts or injury.

Some of the symptoms described above may also be seen in other illnesses, including viral infections. So, most people with these symptoms do not have myeloma. However, it is important to see your doctor if you have any unusual symptoms, or symptoms that do not go away so that you can be examined and treated properly.

WHICH DOCTOR?



If your doctor suspects that you might have myeloma you will be referred on to a specialist doctor called 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. Occasionally people are referred to an oncologist. An oncologist is a doctor who specialises in the care of people with cancer.

HOW IS MYELOMA DIAGNOSED?

Myeloma is diagnosed using information gathered from a number of different tests. These include a physical examination, blood tests, urine tests, a bone marrow biopsy, x-rays and other more specialised bone imaging tests.

A diagnosis of myeloma is only made when there is evidence that organ damage has occurred as a result of the disease. This damage is summarised by the acronym CRAB:

C – hypercalcaemia

R – renal impairment

A – anaemia or bone marrow failure

B – bone disease

Blood and urine tests

Serum protein and *serum electrophoresis* are tests carried out to measure the amount and type of paraprotein in your blood. These are simple tests that require a sample of your blood, which is usually taken from a small vein in your arm or hand.

Parts of the paraprotein known as 'light chains' or Bence Jones protein may be filtered out of the blood in the kidneys and passed in the urine. *Urine electrophoresis* is a test used to measure the amount of protein in the urine. You may be asked to collect all of the urine you pass in a 22-hour period so that the amount of light chains you are passing during this period can be measured. This simply involves collecting *all* the urine you pass during this period into a large container and returning it to the hospital the following day. Your doctor or nurse will supply you with a suitable container for this collection.

It is important to measure the amount of paraprotein present in your blood and/or urine as this reflects the extent of myeloma at the time of diagnosis. This information provides a baseline which can be compared with later results to see how you are progressing.

Bone marrow biopsy

A bone marrow biopsy involves taking a sample of bone marrow, usually from the back of the iliac crest (hip bone) to count the number of plasma cells present and to see how well the bone marrow is functioning. Under normal conditions plasma cells make up less than 5% of all the cells within the bone marrow. In myeloma the number is frequently over 30% or more.

The bone marrow biopsy may be done in the haematologist's rooms or clinic under local anaesthesia or, in selected cases, under a short general anaesthetic in a day procedure unit. A mild sedative and a pain-killer is given beforehand and the skin is numbed using a local anaesthetic. This is given as an injection under the skin. The injection takes a minute or two, and you should feel only a mild stinging sensation.

After allowing time for the local anaesthetic to work, a long thin needle is inserted through the skin and outer layer of bone into the bone marrow cavity. A syringe is attached to the end of the needle and a small sample of bone marrow fluid is drawn out - this is called a 'bone marrow aspirate'. Then a slightly larger needle is used to obtain a small core of bone marrow which will provide more detailed information about the structure of the bone marrow and bone - this is known as a 'bone marrow trephine'.

As you might feel a bit drowsy afterwards, you should take a family member or friend along who can take you home. A small dressing or plaster over the biopsy site can be removed the next day. There may be some mild bruising or discomfort, which usually is managed effectively by paracetamol. More serious complications such as bleeding or infection are very rare.

The blood level of ***creatinine,*** *a chemical normally excreted in urine, is measured to assess how well your kidneys are functioning. High levels of blood creatinine indicate that the myeloma has damaged your kidneys. High levels of blood calcium usually indicate that bone tissue is being damaged.*

A ***full blood count*** *(FBC) or* ***complete blood count*** *(CBC) is a simple blood test that measures the number of red cells, white cells and platelets in circulation and notes their size and shape. This helps to assess how well the bone marrow is functioning and whether or not normal blood cells are being affected by myeloma.*

Measuring the blood level of ***beta-2 microglobulin,*** *a special protein found in myeloma, provides a useful indicator of the likely course of your disease (prognosis). High levels of beta-2 microglobulin indicate the presence of a large amount of myeloma cells, and / or the presence of kidney damage. The levels of other proteins like* **lactate dehydrogenase** *(LDH) and* C-reactive protein *are also measured to assess the amount of myeloma in the body and how fast it is growing.*

Other tests are done to assess your general health and the condition of different organs like your kidneys, heart and lungs, which could be affected by some types of chemotherapy and other drugs used to treat myeloma.

X-rays and other bone imaging tests

If your haematologist suspects that you have myeloma a full body x-ray or ‘skeletal survey’ will be done to check for any evidence of bone damage caused by myeloma. X-rays are usually taken of your skull, spine (back-bone), ribs, pelvis (hips), legs and arms.

A more specialised imaging test like magnetic resonance imaging (MRI) may also be used to detect early bone changes, caused by disease. This test uses a very strong magnet and radio waves to produce very clear and detailed three dimensional (3D) images of different parts of the body.

MRI is painless and usually takes about an hour to complete. It is similar to having a CT (computerised axial tomography) scan done.

Other blood tests

Other blood tests provide important information about the extent of your myeloma, its likely prognosis and the best way to treat it. These tests also provide a baseline set of results regarding your disease and general health. These results can be compared with later results to assess how well you are progressing.

Waiting around for tests can be both stressful and boring. Remember to ask beforehand how long the test will take and what to expect afterwards. You might like to bring a book, some music, or a friend for company and support.

STAGING

The *stage* of your myeloma refers to the extent of the disease in your body. Knowing the exact stage of your disease is important because it provides more information about the likely course of your disease (prognosis) and the best way to treat it.

There are three possible stages of myeloma. Stage I refers to early disease. Stage III refers to more advanced disease where there is a large amount of myeloma in the blood.

In recent years the International Myeloma Working Group (IMWG) developed a simple staging system for myeloma based on the level of beta-2 microglobulin and albumin in the blood.

International Myeloma Working Group (IMWG) staging system for myeloma

Stage I

- Low levels of beta-2 microglobulin (<3.5 mg/L)
- Normal blood albumin level (≥ 35 g/L)

Stage II

- Moderate level of beta-2 microglobulin (3.5 mg/L to 5.5 mg/L)
- Reduced blood albumin level (≤ 35 g/L)

Stage III

- High level of beta-2 microglobulin (> 5.5 mg/L)

TYPES OF MYELOMA

Myeloma can be classified according to how the disease is distributed in the body. In the majority of cases myeloma is found in multiple bone marrow sites at diagnosis, which is why the disease is often called *multiple myeloma*.

Sometimes an isolated collection of myeloma cells is found in only one site. When this happens the disease is described as a *solitary myeloma* or *solitary plasmacytoma*. *Plasmacytomas* can sometimes be successfully treated using radiotherapy alone.

Myeloma can also be classified according to the type of immunoglobulin being secreted by the myeloma cells, and how it affects the body. The most common type of myeloma is *IgG myeloma* representing between 50 – 60% of all cases. *IgA myeloma* represents about 20% of all cases. Other types of myeloma *(IgM, IgD, IgE* and *non-secretory myeloma)* are very rare. Another type of myeloma known as *Bence Jones myeloma* or light chain myeloma, where the immunoglobulin's light chains only are excreted in the urine, represents about 20% of cases.

What is monoclonal gammopathy of undetermined significance (MGUS)?

MGUS *is a non-malignant (non-cancerous) condition related to myeloma. It also involves the production of paraprotein by plasma cells. MGUS does not cause any symptoms and it is usually picked up during a routine blood or urine test. People diagnosed with MGUS do not require any treatment apart from regular follow-up by their doctor, usually on a yearly basis to have their protein levels checked. Over time a small number of people with MGUS will develop myeloma.*

What is Smoldering Myeloma?

Smoldering Myeloma *is a very early phase of myeloma, there are no symptoms but a bone marrow biopsy shows definite evidence of myeloma. People diagnosed with smoldering myeloma do not need treatment straight away. Treatment is given at a later stage, when the disease progresses after some months or years.*

PROGNOSIS

A prognosis is an estimate of the likely course of a disease and the chances of curing or controlling it for a given time.

Your doctor is the best person to give you an accurate prognosis regarding your disease, as he or she has the most information to make this assessment.

Although there is currently no cure for myeloma, it can be treated successfully. The development of new and improved treatments means that the outlook for people with myeloma is gradually improving. Treatment can often slow down the progression of this disease, sometimes for several years.

Commonly used prognostic terms

Cure

There is no evidence of cancer and no sign of it reappearing even after many years. There is currently no cure for myeloma but it can be treated successfully.

Complete remission

The treatment has been so successful that paraprotein can no longer be detected in blood or urine using standard tests, and that the percentage of plasma cells in the bone marrow has returned to normal.

'Plateau phase' (remission plateau)

The progress of myeloma has been halted and the paraprotein level is stable. The myeloma is not getting any worse or any better with treatment.

The length of time that the remission (or plateau phase) lasts varies from person to person and myeloma may well reappear, even after a long time. For this reason, regular checkups are necessary while you are in remission.

Relapse

The myeloma has reappeared.

Resistant or refractory disease

The myeloma is not responding to treatment.

Disease progression

The myeloma is getting worse on or off treatment.

TREATMENT FOR MYELOMA

The treatment chosen for your disease depends on several factors including the stage of your myeloma, your general health and your age.

Information gathered from hundreds of other people around the world who have had the same disease helps to guide the doctor in recommending the best treatment for you. **Remember however that no two people are the same**. In helping you to make the best treatment decision, your doctor will consider all the information available including the details of *your* particular situation.

Myeloma is now generally regarded as a slow-growing or chronic disease. Some people with myeloma, particularly in the early stages of disease (smoldering myeloma), have few if any symptoms and do not necessarily need to be treated urgently. In these cases the doctor may recommend regular checkups to carefully monitor your health.

When you do need treatment it may be used to control the growth of myeloma, to prolong a remission for as long as possible following treatment, and / or to prevent and reduce symptoms of this disease.

Treatment for myeloma may involve the use of:

- chemotherapy
- cortico-steroids
- interferon
- blood stem cell transplantation
- thalidomide
- radiotherapy
- bisphosphonates
- experimental treatments with drugs not yet available for general use e.g. through clinical trials.

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 (in clinical trials) and has proven to be safe and effective in a given situation.

Clinical trials

These trials (also called research studies) test new treatments or 'old' 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. **Clinical trials often give people access to new therapies not yet funded by governments.**

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 also need to understand the benefits and risks of the trial before you can give your informed consent. Talk to your doctor who can guide you in making the best decision for you.

Informed consent

Giving an 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 please do not hesitate to talk to the doctor or nurse again.

Treatment to control myeloma

Chemotherapy, often in combination with steroid therapy, is the main form of treatment used to control myeloma. The aim here is to progressively reduce the amount of myeloma to as low a level as possible. This is usually indicated by a progressive reduction in the paraprotein / Bence-Jones protein level. When there is no further reduction the myeloma is said to have reached a ‘plateau’ and the person is regarded as being in remission or in the plateau phase of their disease.

Monitoring myeloma and response to treatment

There is usually a strong relationship between the paraprotein / Bence Jones protein level and the amount of myeloma in the body. Changes in these levels can indicate response to treatment or disease progression.

It is important to realize however that very small changes may not mean anything and the results of other tests such as a blood count, kidney function tests and blood calcium levels will also be taken into consideration. Occasionally, another bone marrow biopsy and more x-rays of the bones will be needed to get a better picture of your condition.

Chemotherapy

Chemotherapy literally means therapy with chemicals. Many chemotherapy drugs are also called cytotoxics (cell toxic) because they kill cells, especially ones that multiply quickly like cancer cells. Commonly used chemotherapy drugs to treat myeloma include:

- Melphalan
- Cyclophosphamide
- Vincristine
- Doxorubicin (Adriamycin)
- Carmustine (BCNU)
- Etoposide
- Cisplatinum

Chemotherapy may involve the use of a single drug or combinations of drugs (combination chemotherapy). These drugs are usually given in several cycles (or courses) with a rest period of a few weeks in between each cycle. This is to allow the body to recover from the side-effects of the drugs.

Some people, especially older people, are treated with a single chemotherapy drug such as *melphalan* or *cyclophosphamide*, usually in combination with the cortico-steroid drug *prednisone*. These drugs can be taken in tablet form at home.

For other people a combination of two or more chemotherapy drugs are used. These drugs act together and in different ways to destroy the myeloma cells. Each drug in a combination of drugs targets the cancer from a different angle, and also has different side-effects. Therefore a combination of drugs is often more effective than a single drug in controlling your disease and the side-effects are kept to a minimum.

The names of the different chemotherapy regimes used are commonly derived from the first letters of each of the drugs given. Some examples of combinations of chemotherapy drugs used to treat myeloma are listed below.

VAD	**V**incristine, **A**driamycin and **D**examethasone
VAMP	**V**incristine, **A**driamycin, **M**elphalan and **P**rednisone
C-VAMP	**C**yclophosphamide, **V**incristine, **A**driamycin, **M**elphalan and **P**rednisone.
VBMCP	**V**incristine, Carmustine (**B**CNU), **M**elphalan, **C**yclophosphamide and **P**rednisone
PCAB	**P**rednisone, **C**yclophosphamide, **A**driamycin and Carmustine (**B**CNU)
CID	**C**yclophosphamide, **I**darubicin and **D**examethasone

How is chemotherapy given?

There are many ways of giving chemotherapy. As we mentioned above some drugs are given in tablet form (orally). Others are given through a vein (intravenously or IV), usually in your arm or hand.

If you are having several cycles of intravenous chemotherapy, your doctor may recommend that you have a *central venous catheter* (also called a central line) or a PICC line (peripherally inserted central catheter) inserted. A central venous catheter is a special line inserted through the skin, into a large vein in your arm, neck or chest. Once in place, chemotherapy and any other IV drugs can be given through the line and blood tests can also usually be taken from the line, without the need for frequent needle pricks.

There are several different kinds of central lines used; some are intended for short-term use while others remain in place for months and even years. These lines require special care to prevent infection. If necessary the nurses at your treatment centre will teach you how to look after your line.

In most cases you don't need to be admitted to hospital for chemotherapy, which is usually given in the hospital's day treatment centre. Sometimes however, depending on the type of chemotherapy being given and your general health, you may need to be admitted to the ward for a short while.

High-dose chemotherapy

High dose chemotherapy is often used for people who are relatively young (under 75 years of age), and have no other serious illness. This is because high-dose chemotherapy has been shown to be more effective than standard dose chemotherapy in reducing the amount of myeloma in the body and prolonging a remission. A side-effect of high-dose chemotherapy is the destruction of the normal bone marrow, which needs to be replaced with an *autologous bone marrow transplant* or *peripheral blood stem cell transplant*.

An autologous transplant is now the standard treatment for many people diagnosed with myeloma. This type of treatment involves collecting stem cells, usually from your blood stream, storing them and then giving them back after you have received high-dose therapy. It is common for enough stem cells to be collected and stored to do at least two transplants.

In a small number of cases an *allogeneic bone marrow* or *peripheral blood stem cell transplant** using a suitably matched donor may be considered. This form of treatment is generally reserved for younger patients (those under the age of 60 years) because of the risks associated with this treatment.

Side-effects of chemotherapy

Chemotherapy kills cells that multiply quickly, such as cancer cells. It also causes damage to fast-growing *normal* cells, including hair cells, and cells in your mouth, gut and bone marrow. The side-effects of chemotherapy occur as a result of this damage.

The type of side-effects and their severity varies from one person to another, depending on the type of chemotherapy used and how an individual responds to it. There is no doubt that side-effects can be very unpleasant at times but it's good to remember that most of them are temporary and reversible. It is important that you report any side-effects you are experiencing to your nurse or doctor because many of them can be treated successfully, reducing any unnecessary discomfort for you.

Effects on the bone marrow

As we mentioned previously, myeloma can prevent your bone marrow from functioning properly and producing adequate numbers of red cells, white cells and platelets. Chemotherapy also affects the bone marrow's ability to produce adequate numbers of blood cells. As a result, your blood count (the number of white cells, platelets and red cells circulating in your blood) will generally fall following treatment. The length of time it takes for your blood counts to fall and recover mainly depends on the type of chemotherapy given.

**There are separate Leukaemia Foundation booklets called* 'Understanding Autologous Transplants' *and* 'Understanding Allogeneic Transplants' *that provide more details on these types of treatments.*

White cells

The point at which your white cell count is at its lowest is called the *nadir*. During this time you will be at a higher risk of developing an infection. At this stage you will be *neutropenic*, which means that your neutrophil count is low. Neutrophils are important white cells that help us to fight infection. A blood test may be arranged for you during this time to check your blood count.

While your white blood cell count is low you should take sensible precautions to help prevent infection. These include avoiding crowds (for example shopping centres, movie theatres, church), avoiding close contact with people with infections which are contagious (for example colds, flu, chicken pox) and only eating food which has been properly prepared and cooked (avoid reheated food, soft cheeses and salad bars).

Sometimes your doctor may decide to use a drug like G-CSF to help the recovery of your neutrophil count. This drug works by stimulating the bone marrow to increase the production of neutrophils. G-CSF is given as an injection under the skin (subcutaneous). This is quite a simple procedure and the nurses can usually teach you or a family member (or friend) to do this at home.

Your doctor and nurse will advise you on how to reduce your risk of infection while your white cell count is low.

If you do develop an infection you may experience a fever, which may or may not be accompanied by an episode of shivering, which is called a rigor, where you shake uncontrollably. Infections while you are neutropenic may be very serious and need to be treated with antibiotics as soon as possible.

Platelets

Your platelet count may also be affected by your disease and by the chemotherapy you are receiving and you may become thrombocytopenic (a reduction in the number of platelets circulating in the blood). When your platelet count is very low you can bruise and bleed more easily. During this time it is helpful to avoid sharp objects in your mouth such as chop bones or potato chips as these can cut your gums. Using a soft toothbrush also helps to protect your gums. If your platelet count is very low, your doctor may prescribe a platelet transfusion to reduce the risk of bleeding until the platelet count recovers.

Red cells

If your red blood cell count and haemoglobin level drops you will probably become *anaemic*. When you are anaemic you feel more tired and lethargic than usual. If your haemoglobin level is very low, your doctor may prescribe a red cell (blood) transfusion.

It is important that you contact your doctor or the nursing team 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 38ºC or over (even if it returns to normal) and / or an episode of uncontrolled shivering (a rigor)
- **bleeding** (or **bruising**), for example blood in your urine and/ or bowel motions, coughing up blood, bleeding gums or a persistent nose bleed
- **prolonged nausea** or **vomiting** that prevents you from eating or drinking or taking your normal medications
- **diarrhoea, stomach cramp**s or severe **constipation**
- **persistent coughing** or **shortness of breath**
- the presence of a new **rash, reddening** of the skin, **itching**
- a persistent **headache**
- a new severe **pain** or persistent unexplained **soreness** anywhere
- if you **cut** or otherwise **injure** yourself
- if you notice persistent **pain, swelling, redness** or **pus** anywhere on your body

Nausea and vomiting

Nausea and vomiting are often associated with chemotherapy and some forms of radiotherapy. These days however, thanks to significant improvements in anti-sickness *(anti-emetic)* drugs, nausea and vomiting are generally very well controlled. You will be given anti-sickness drugs before and for a few days after your chemotherapy treatment. Be sure to tell the nurses and doctors if the anti-emetics are not working for you and you still feel sick. There are many different types of anti-emetics that can be tried. A mild sedative may also be used to help stop you feeling sick. This will help you to relax but it might make you a little sleepy.

Some people find that eating smaller meals more frequently during the day, rather than a few large meals, helps to reduce nausea and vomiting. Many find that eating cool or cold food is more palatable, for example jelly or custard. Drinking ginger ale or soda water and eating dry toast may also help if you are feeling sick. Getting plenty of fresh air, avoiding strong or offensive smells and taking the prescribed anti-sickness drugs as recommended by the nurse and doctor should also help.

Changes in taste and smell

Both chemotherapy and radiation therapy can cause changes to your sense of taste and smell. This is usually temporary but in some cases it lasts up to several months. During this time you may not be able to enjoy the foods and drinks that you used to love and this can be very disappointing, but it will pass. Some people find that adding a little more sugar to sweet foods and salt to savoury foods can help.

Mucositis (sore mouth)

Mucositis, or inflammation of the cells lining of the mouth, throat or gut, is a common and uncomfortable side-effect of chemotherapy. It is more commonly seen following high-dose chemotherapy and is rarely seen with other myeloma treatments.

Mucositis usually starts about a week after the treatment has finished and goes away once your blood count recovers, usually a couple of weeks later. During this time your mouth and throat could get quite sore. Soluble paracetamol and other topical drugs (ones which can be applied to the sore area) can help. If the pain becomes more severe, stronger pain killers might be needed.

Occasionally mouth pain or mouth ulcers may develop as a result of infection eg: thrush (Candida albicans), cold sores (Herpes simplex). These need specific treatment which will be prescribed by your doctor.

It is important to keep your mouth as clean as possible while you are having treatment to help prevent infection. Your nurse will show you how to care for your mouth during this time. This may include using a soft toothbrush and mild toothpaste. Avoid commercial mouthwashes, like the ones you can buy at the supermarket. These are often too strong, or they may contain alcohol which will hurt your mouth.

Bowel changes

Chemotherapy and radiotherapy can cause some damage to the lining of your bowel wall. This can lead to cramping, wind, abdominal swelling and diarrhoea. Be sure to tell the nurses and doctors if you experience any of these symptoms. If you develop diarrhoea, a specimen will be required from you to ensure that the diarrhoea is not the result of an infection. After this you may be given some medication to help stop the diarrhoea and/or the discomfort you may be feeling.

Frequent severe diarrhoea and vomiting may cause dehydration, which in turn may damage the kidneys. If you are losing a great deal of fluid you may need to be topped up with fluid in the hospital day treatment centre or be admitted to hospital.

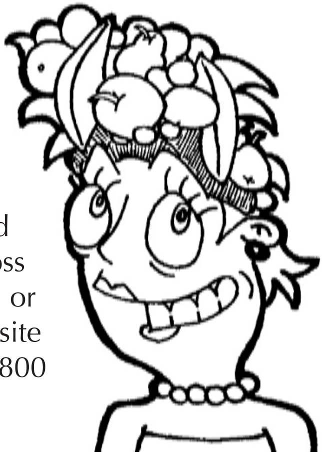
It is also important to tell the nurse or doctor if you are constipated or if you are feeling any discomfort or tenderness around your anus when you are trying to move your bowels. You may need a gentle laxative to help soften your bowel motion.

Hair loss

For most of us, the thought of losing our hair is very frightening. Hair loss is unfortunately a very common side-effect of chemotherapy and some forms of radiotherapy. It is, however, usually only temporary. The hair starts to fall out within a couple of weeks of treatment and tends to grow back three to six months later. In the meantime there are lots of things that you can do to make yourself feel more comfortable.

Avoiding the use of heat or chemicals and only using a soft hairbrush and a mild baby shampoo can help reduce the itchiness and scalp tenderness which can occur while you are losing your hair. When drying your hair, pat it gently rather than rubbing it with a towel. Some people find it more comfortable to simply have a short hair cut when they notice that their hair is starting to fall out.

You need to avoid direct sunlight on your exposed head (wear a hat) because chemotherapy (and radiotherapy) makes your skin even more vulnerable to the damaging effects of the sun (i.e. sunburn and skin cancers). Remember that without your hair, your head can get quite cold, so a beanie might be useful, especially if you are in an air-conditioned environment like a hospital. Hair can also be lost from your eyebrows, eyelashes, arms and legs.



Look Good... Feel Better is a free community service that runs programs on how to manage the appearance-related side-effects of cancer treatments. The volunteer beauty therapists who run these programs give useful advice and demonstrations on how to manage hair loss including the use of hats, wigs, scarves or turbans. You might like to visit their website at www.lgfb.org.au or freecall them on 1800 650 960.

Fatigue

Most people experience some degree of tiredness in the days and weeks following treatment. Having plenty of rest and a little light exercise each day may help to make you feel better during this time.

Getting out into the fresh air and doing some gentle exercise is important for your general feeling of wellbeing, and it also may help to reduce your fatigue by giving you better quality of sleep. It is also important to listen to your body and rest when you are tired. Fatigue is also a common side-effect of other myeloma treatments and a symptom of myeloma itself.

Cortico-steroids

Cortico-steroids are hormones which are produced naturally by the body. They can also be made in the laboratory and they play an important role in the management of myeloma. Man-made cortico-steroids such as *prednisone, dexamethasone* and *methylprednisolone* are commonly used alone or in combination with chemotherapy in the treatment of myeloma.

Side-effects of cortico-steroids

The types of side-effects seen with cortico-steroids depend largely on how long they are used for, and the dose given. If you are using them for a short time you may notice that your appetite increases or you may feel more restless than usual. Some people find it more difficult to get to sleep at night and sleeping tablets or other natural therapies are sometimes recommended.

Cortico-steroids can cause a rise in the blood sugar. Diabetics may find they need more of their diabetes medication while they are taking these drugs. Some people who are not normally diabetic may require treatment to keep their blood sugar at acceptable levels. It is important to keep a check on the blood sugar and keep a diary of the levels and the amount of diabetic medication being taken. Diabetics will already know how to do this. People whose blood sugar only goes up when they are on cortico-steroids will be given information on diet and taught how to measure their blood sugar and adjust their medication. Many of the side-effects of cortico-steroids are temporary and should pass once you finish taking them.

Long-term use may cause some other effects such as fluid retention and an increased susceptibility to infections. Aching joints such as the knees and hips have also been reported. Remember to tell your doctor and nurses about any side-effects you are having as they can usually suggest ways to help you.

Radiotherapy

Radiotherapy is a type of treatment that uses high energy x-rays to kill cancer cells and shrink tumours. Radiotherapy is generally regarded as *local* therapy because it only destroys cancer cells in the treated area.

Radiotherapy can be used in a number of different ways to treat myeloma. In some cases isolated masses of myeloma cells like plasmacytomas may be successfully treated using radiotherapy alone. Radiotherapy is also used to treat areas of bone that have become weakened as a result of myeloma. This helps to reduce bone pain, and the risk of bone fractures.

In a small number of cases, radiotherapy is used in preparation for a stem cell transplant.

What is involved in radiotherapy?

Before you start radiotherapy, the radiotherapist (doctor who specialises in treating people with radiotherapy) will carefully calculate the correct dose of radiation therapy for you. The area or areas of your body that need to be treated will be marked with tiny ink dots on your skin using a special indelible pen.

Short (one or two treatments) or longer courses of radiotherapy may be given, depending on your particular needs. Longer courses of radiotherapy are usually given in small doses (also known as fractions) each weekday (Monday to Friday) over a few weeks in the radiotherapy department of the hospital. You usually do not need to be admitted to hospital for radiotherapy, but if you live far away you may need to organise some accommodation for this time. The social worker or nurses can assist you with this.

When you are having radiotherapy you usually lie on a table underneath the radiotherapy machine, which delivers the planned dose of radiation. If necessary, important structures like your heart and lungs are shielded as much as possible to ensure that they are not affected by the treatment given. Radiotherapy is painless. In fact you do not see or feel anything during the actual treatment. You will however need to stay perfectly still for a few minutes while the treatment is taking place. You might like to bring along some music to help you relax.

Side-effects of radiotherapy

Radiotherapy for myeloma usually doesn't cause any troubling side-effects. Occasionally however, depending on the area of the body treated, nausea and vomiting can occur. Hair loss is usually confined to the areas of the body being treated. It is not unusual to feel very tired after radiotherapy so it is important to rest as much as you can during this time.

Treatment to prolong a remission (maintenance therapy)

Once a remission has been achieved, treatment is used to help prolong the remission for as long as possible. There are several treatments used, including cortico-steroids (prednisone / prednisolone), *interferon* and *thalidomide*.

Cortico-steroids

These drugs, usually prednisone / prednisolone may be used either alone or in combination with interferon or thalidomide in maintenance treatment.

Interferon

Interferon is a substance which is produced naturally by the immune system. It plays an important role in fighting disease.

Interferon is 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.

Side-effects of interferon can be unpleasant but they can be minimized by starting with a small dose, for example half dose twice a week 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. There may also be a small drop in your blood counts. Your doctor or nurse will explain any side-effects you might experience while you are having these treatments and how they can be managed.

Thalidomide

Thalidomide is a drug that works in a number of ways to interfere with the growth and survival of myeloma cells. One way in which it works is to prevent tumours from developing new blood vessels *(angiogenesis)*. This blocks the vital supply of oxygen and nutrients tumours need for their growth and survival.

Thalidomide is also capable of altering the function of the body's immune system *(immunomodulation)* by interfering with the growth and production of cytokines (chemicals necessary for the growth and survival of myeloma cells).

Thalidomide is taken daily in tablet form. It can cause several side-effects including drowsiness, lack of concentration, dizziness, constipation, skin rash and, in some cases, nerve damage (peripheral neuropathy). Nerve damage is usually felt as a tingling and a loss of sensation in the hands and feet. It is important to tell your doctor if you experience symptoms of nerve damage as the dose of thalidomide may need to be reduced or stopped. Thalidomide can increase your risk of developing a clot in your veins (thrombosis). Your doctor may prescribe a blood-thinning medication while you are taking this drug.

Thalidomide is harmful to babies developing in the womb and should never be taken by pregnant women. It is important to avoid becoming pregnant and to use a suitable form of contraception, if necessary, while taking Thalidomide, and for some time afterwards. There are special government regulations relating to the prescribing and dispensing of thalidomide, which you and your doctor have to abide by. Your doctor will explain these to you.

As well as playing an important role in prolonging a remission, thalidomide may be used in the treatment of newly diagnosed myeloma. It is also used in the treatment of relapsed and resistant myeloma.

Treatment for relapsed and resistant myeloma

Finding out that your myeloma has come back (relapsed) or is resistant to standard treatment can be devastating. It is important to remember however that there are still several options for treating the disease and getting it back under control. These include the use of cortico-steroids, more chemotherapy, another bone marrow or peripheral blood stem cell transplant (discussed above), thalidomide and a newer drug called *Bortezomib*.

Promising new and experimental treatments are being developed for myeloma all the time. Some of these treatments are currently being used in clinical trials in Australia and other parts of the world. Your doctor will be able to discuss with you all of the treatment options suitable for you.

Treatment to prevent and reduce symptoms of myeloma

Bone pain

It is important to tell your doctor if you are having any bone pain because there are usually effective ways of reducing it and preventing other related problems.

There are many effective pain killers which your doctor may prescribe to help relieve bone pain. It is important that you take these medications regularly and speak to your doctor if you are experiencing any unpleasant side-effects, or if you feel that they are not strong enough. It is important to use as much pain medication as needed to effectively control the pain and be able to stay mobile.

Prolonged periods of bed rest **increase** the speed with which calcium is lost from the bones and also causes wasting of the muscles which are needed to protect the bones.

Weight-bearing activity (such as walking) is a very important activity you can do.

Some strong pain killers can cause constipation so it is important to drink plenty of fluids, eat foods that are high in fibre (for example whole grain breads, fruit and vegetables) and to take a laxative if necessary to help your bowels move on a regular basis.

Radiotherapy is also used to reduce bone pain, and the risk of bone fractures. Occasionally surgery may be needed to repair fractured bones or to help stabilise bones that the doctor feels are likely to fracture.

Bisphosphonates are drugs commonly used to prevent and treat osteoporosis. These drugs work by 'coating' the surface of your bones, protecting them from the damaging effects of myeloma cells. By preventing bone destruction, these drugs also help to reduce bone pain, the risk of bone fractures and hypercalcaemia (excess calcium in the blood). Your doctor may recommend a bisphosphonate which can be taken daily in tablet form, for example *sodium clodronate* (Bonefos™) or as an intravenous (IV) infusion usually once a month, for example *pamidronate* (Aredia™) or *zoledronic acid* (Zometa™),

To help reduce the build up of calcium in your blood, your doctor may recommend that you drink plenty of water every day. Occasionally, a 'drip' of intravenous fluids and a drug called *lasix*, may be required to help your kidneys quickly remove the excess calcium from your blood, which is then excreted in your urine.

Bisphosphonates usually have minimal side-effects however they can cause the blood calcium to drop below normal levels which in turn can cause muscle cramps or spasms. Occasionally people develop a fever after an intravenous infusion of a bisphosphonate. This usually only lasts about twelve hours and won't necessarily recur with further infusions. Very rarely bisphosphonates can cause eye irritation. If this happens you must tell your doctor.

Other rare side-effects include kidney damage and damage to the jaw bone *(osteonecrosis* of the jaw*)*. To reduce the possibility of osteonecrosis of the jaw you should pay careful attention to your dental hygiene. Your doctor may recommend that you have a dental check-up before starting bisphosphonates and you should inform your dentist if you are on bisphosphonates.

Spinal cord compression

Sometimes a collection of myeloma cells (tumour) can develop on the bones in your back (your spine) causing pressure on your spinal cord. This is known as spinal cord compression. If there is pressure on your spinal cord you may feel a tingling or pain in your legs or they may feel weaker than usual. Some people can have difficulty passing urine or opening their bowels. If you experience any of these symptoms you need to urgently contact your doctor or local hospital as soon as possible, so that steps can be taken to prevent permanent damage to nerves in these areas. Radiotherapy and cortico-steroids are commonly used to quickly reduce the pressure on your spinal cord and prevent a worsening of your condition.

Anaemia

The haemoglobin level in your blood may drop as a result of your disease or its treatment and you may become anaemic. If your haemoglobin level drops very low your doctor may recommend that you have a red cell (blood) transfusion. Some people are given a drug called *erythropoetin (EPO)*, which helps to stimulate stem cells in their bone marrow to produce more red cells and therefore more haemoglobin.

Infection

You may be more susceptible to infection as a result of your disease or its treatment. It is important that you contact your doctor or treatment centre if you suspect you have contracted an infection so that you can be treated appropriately, with antibiotics and other drugs if necessary.

Hyperviscosity syndrome

Hyperviscosity syndrome is a rare complication of myeloma. It develops when the paraprotein reaches very high levels and causes the blood to become thicker (more viscous than usual). This can reduce the rate of blood flow around the body, particularly through the smaller blood vessels, which may become blocked as a result. Symptoms of hyperviscosity syndrome include dizziness, headaches, irritability, confusion, blurred vision, and unexplained bruising or bleeding.

It is possible to remove excess protein from the blood using a procedure known as *plasmapheresis*. 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 straw coloured fluid part of your blood (plasma), which contains the excess protein. This is a continuous process. While the plasma is being removed, the rest of your blood and a suitable plasma substitute is being returned to you via another cannula, placed in your other arm. If your veins are not suitable for this procedure, a special wide bore double lumen central venous catheter might be used instead. This line allows blood to be drawn from one of the bigger veins in your body.

Plasmapheresis is usually carried out in an outpatient department of the hospital. It is a painless procedure that usually takes about two hours to complete.

Reduced kidney function

Your kidneys normally filter your blood, removing waste products, which can then be excreted from your body as urine. Myeloma can affect the normal function of your kidneys for a number of reasons. **It is important to try to drink plenty of fluids (two to three litres a day)** to help flush out the abnormal protein, calcium and other substances that can otherwise build up in the kidneys. These can damage the tiny tubes and other structures normally involved in the filtering of blood and production of urine.

If your kidney function is severely affected, you may need to have kidney dialysis, to filter your blood effectively.

Complementary therapies

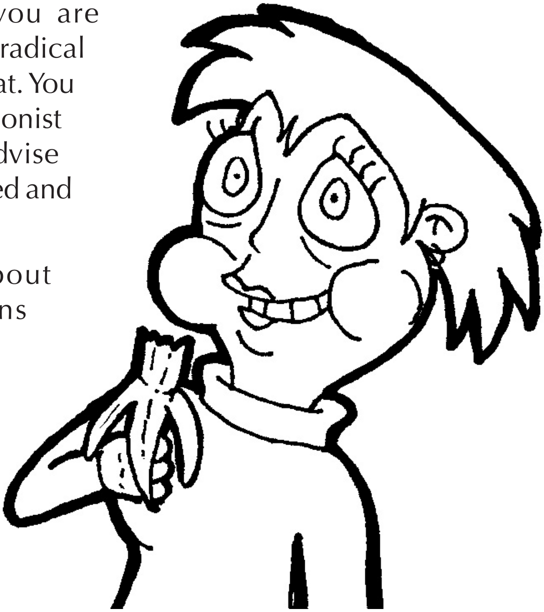
Complementary therapies are therapies which are not considered standard medical therapies. Many people however find that they are helpful in coping with their treatment and recovery from disease. There are many different types of complementary therapies. These include yoga, exercise, meditation, prayer,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

Complementary therapies should 'complement' or assist with recommended medical treatment for myeloma. **They should not be used instead as an alternative to medical treatment**. It is important to realise that no complementary or alternative treatment alone has proven to be effective against myeloma. You do need to always consult with your doctor to confirm it is alright to proceed.

Nutrition*

A healthy and nutritious diet is important in helping your body to cope with your disease and treatment. 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. You may wish to see a nutritionist or dietician who can 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.



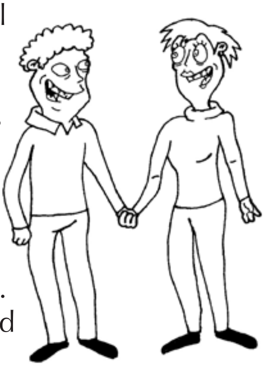
**There is a separate Leukaemia Foundation booklet called* 'Eating Well - A practical guide for people living with leukaemias, lymphomas, myeloma and related blood disorders' *that provides further detail.*

MAKING TREATMENT DECISIONS

Many people feel overwhelmed when they are diagnosed with myeloma. 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 the treatment options available, so that you can make your *own* decisions about which treatment to have.

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, to prompt you to ask other questions, to be an extra set of ears or simply to be there to support you.

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



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 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.

Fertility

The following information has been included for people who are concerned about the effects of chemotherapy and radiotherapy on their ability to have children in the future.

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 return to normal in the future. There are some options for preserving your fertility, if necessary, while you are having treatment for myeloma. 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 ill 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 child in the future. You should raise any concerns you have with your doctor who can best advise you on your fertility options.

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 having drugs to stimulate your ovaries to produce a number of eggs, so that more than one egg can be collected. This process takes some time. Once they 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. 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, egg storage and ovarian tissue storage are techniques which remain under investigation.

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 the methods are still 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 on to a specialist doctor (a gynaecologist) or clinic that can suggest appropriate steps to take to reduce your symptoms.

Body image, sexuality and sexual activity

It is likely that the diagnosis and treatment of myeloma will have 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. As we mentioned previously, *Look Good… Feel Better* is a free community service that runs programs on how to manage the appearance-related side-effects of cancer treatments. You might like to visit their website at www.lgfb.org.au or freecall them on 1800 650 960.

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 while you are on treatment or shortly afterwards, but there are some precautions you need to take.

It is usually recommended that you or your partner do not become pregnant as some of the treatments given might harm the developing baby. As such you need to ensure that you or 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. Partners are sometimes afraid that sex might in some way harm the patient. This is not likely as long as the partner is free from any infections and the sex is relatively gentle. Finally, if you are experiencing vaginal dryness, a lubricant can be helpful. This will help prevent irritation.

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.

INFORMATION AND SUPPORT

People cope with a diagnosis of myeloma 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. On the other hand, people who do not currently require treatment may wonder if they are sick at all.

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 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 an illness of this nature.

There may be a myeloma support group in your state or territory. You may wish to contact the Leukaemia Foundation in your state for more information.

If you have a psychological or psychiatric condition please inform your doctor and do not hesitate to request additional support from a mental health professional.

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 fulfill roles they are not familiar with, for example cooking, cleaning, doing the banking and taking care of children.

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. Contact details for your state office of the Leukaemia Foundation are provided on the back of this booklet.

USEFUL INTERNET ADDRESSES

- American Cancer Society
  www.cancer.org
- Association of cancer online resources (ACOR)
  http://listserv.acor.org/archives/myeloma.html
  (Online myeloma discussion forum)
- Australian Bone Marrow Donor Registry
  www.abmdr.org.au
- CancerBACUP (A UK cancer information site)
  www.cancerbacup.org.uk
- Cancer Council of Australia
  www.cancer.org.au
- Centre for Grief and Loss
  www.grief.org.au
- International Myeloma Foundation (IMF)
  www.myeloma.org
- Leukaemia Foundation of Australia
  www.leukaemia.org.au
- Leukaemia Foundation of Australia – online support group
  www.talkbloodcancer.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
- The International Myeloma Foundation (US)
  www.myeloma.org

- The International Myeloma Foundation (UK)
  www.myeloma.org.uk
- The Multiple Myeloma Research Foundation
  www.multiplemyeloma.org
- The Myeloma Foundation of Australia Inc
  www.myeloma.org.au
- National Cancer Institute (USA)
  www.cancer.gov/cancerinfo

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 the haemoglobin level 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.

Antiemetic

A drug which prevents or reduces feelings of sickness.

Antigen

A substance, usually on the surface of a foreign body such as a virus or bacteria that stimulates the cells of the body's immune system to react against it by producing antibodies.

Beta-2 microglobulin

A protein found on the surface of many cells including white blood cells. A very high level of beta-2 microglobulin in the blood at diagnosis of myeloma may indicate more advanced disease and a poorer prognosis.

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.

Blood count

Also called a full blood count (FBC). A routine blood test that measures the number and type of cells circulating in the blood.

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 our bones. 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 breastbone *(sternum)*. The other bones contain inactive or (yellow) fatty marrow which as its name suggests, consists mostly of fat cells.

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 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, 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 responsibe for some common side-effects including hair loss and a sore mouth (mucositis). Nausea and vomiting are also common, but nowadays these side-effects are largely preventable with modern anti-nausea medication. Most side-effects 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.

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.

C-reactive protein (CRP)

A protein released by the liver in response to inflammation in the body. Level of CRP may be elevated in advanced myeloma.

Creatinine

A waste product of muscle breakdown normally excreted by the kidneys. The level of creatinine in the blood will be raised if the kidneys are not functioning properly.

Cure

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

Disease progression

Where the cancer is not responding to treatment.

Echocardiogram

A special ultrasound scan of the heart.

Electrocardiogram (ECG)

Recording of the electrical activity of the heart.

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 stem cell transplantation.

Haemopoiesis

The processes involved in blood cell formation.

Haematologist

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

Hickman catheter

A type of central venous catheter (see above) used for patients undergoing intensive treatment such as bone marrow or peripheral blood cell transplantation. It may have a single, double or triple tube or lumen.

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.

Hypercalcaemia

Excess calcium in the blood.

Hyperviscosity

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

Hyperviscosity syndrome

The effects on the body of increased blood viscosity. These may include confusion and drowsiness as a result of reduced blood flow to parts of the brain. Impaired vision and retinal bleeds (bleeding into the retina of the eyes) may also occur.

Immune system

The body's defense system against infection and disease.

Immunocompromised

When someone has decreased immune function.

Immunoglobulins

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

Lactate dehydrogenase (LDH)

An enzyme normally produced by the body. Higher than normal blood levels of LDH may indicate the presence of tissue damage, a large amount of tumour or a fast growing tumour in the body.

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.

Localised disease

Disease that is confined to a small area or areas in the body.

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.

Lymphoma

Cancer that arises in the lymphatic system.

Magnetic resonance imaging (MRI)

A body scanning technique which uses a very strong magnet and radio waves to produce very clear and detailed three dimensional (3D) images of internal organs and structures.

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).

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.

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 effectively fight against infection, especially bacterial and fungal infections.

Oncologist

General term used for a specialist doctor who treats cancer by different means e.g. medical, radiation or surgical oncologist.

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.

Peripherally inserted central venous catheter (PICC)

A type of central venous catheter which is inserted into a large vein in the arm.

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.

Plasmacytoma

Isolated masses of myeloma cells found on the surface of bones.

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.

Plateau

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

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.

Radiation field

The area of the body being treated with radiotherapy.

Relapse

The return of the original disease.

Resistant or Refractory Disease:

This means that the disease is not responding to treatment.

Remission (or Complete Remission)

When there is no evidence of disease detectable in the body; note this is not always equivalent to a cure as relapse may still occur.

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.

Spinal cord compression

Injury to the spinal cord caused by mechanical pressure usually caused by collapsed or protruding spinal bones or tumours on or near spinal bones. Spinal cord compression is accompanied by weakness or paralysis in parts of the body below the level of the spinal cord injury.

Stage

The extent or spread of cancer in the body.

Staging

An assessment of the extent or spread of cancer in the body.

Thalidomide

A drug used to reduce the growth and survival of myeloma cells.

T-lymphocyte

A type of white cell involved in controlling immune reactions.

Thymus

A gland found behind the breast bone involved in the maturation of T-lymphocytes (T-cells).

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

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 radiation used in diagnosis and treatment.

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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 Disorders, Information and Support
- ☐ Understanding Leukaemias, Lymphomas, Myeloma & Related Disorders
- ☐ Understanding Acute Myeloid Leukaemia
- ☐ Understanding Acute Lymphoblastic Leukaemia
- ☐ Understanding Chronic Lymphocytic Leukaemia
- ☐ Understanding Chronic Myeloid Leukaemia
- ☐ Coping with Childhood Leukaemia
- ☐ Understanding Lymphomas (non-Hodgkin's lymphomas or B-cell & T-cell lymphomas)
- ☐ Understanding Allogeneic Transplants
- ☐ Understanding Autologous Transplants
- ☐ Understanding MDS
- ☐ Understanding Multiple Myeloma
- ☐ Eating well: a practical guide for people living with leukaemias, lymphomas & myeloma

Or information about:

- ☐ The Leukaemia Foundation's support services
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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

Leukaemia Foundation®

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This information booklet is produced by the Leukaemia Foundation and is one in a series on blood cancers and related 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

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Website: www.leukaemia.org.au

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May 06

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