

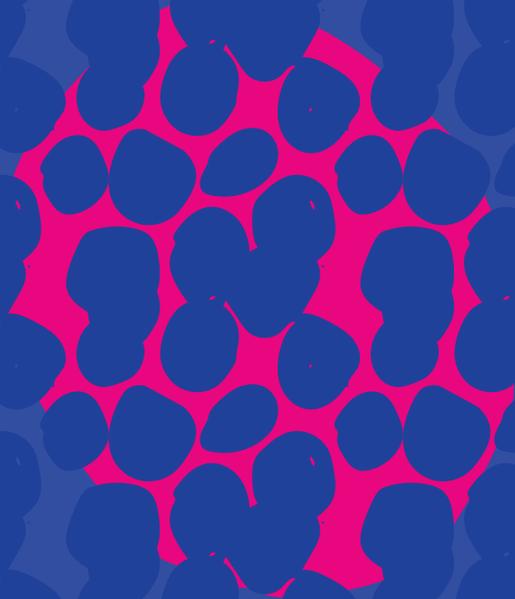


Leukaemia  
Foundation

VISION TO CURE  
MISSION TO CARE

# MYELOMA

A guide for patients and families



1800 620 420



[leukaemia.org.au](http://leukaemia.org.au)



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People who have experienced myeloma as a patient or carer, Leukaemia Foundation blood cancer support staff, haematology nursing staff and clinical haematologists.

The Leukaemia Foundation values feedback from people affected by myeloma and the health care professionals working with them. If you would like to make suggestions, or tell us about your experience of using this booklet, please contact [info@leukaemia.org.au](mailto:info@leukaemia.org.au)

**March 2018**

## INTRODUCTION

This booklet has been written to help you and your family understand more about myeloma.

Some of you may be feeling anxious or a little overwhelmed if you or someone you care for has been diagnosed with myeloma. This is normal.

Perhaps you have already started treatment or you are discussing different treatment options with your doctor and your family.

Whatever point you are at, we hope 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 you think will be of most use at a particular point in time.

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

Some of you may require more information than is contained in this booklet; we have included some internet addresses 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 and team.

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

# THE LEUKAEMIA FOUNDATION

The Leukaemia Foundation is the only national charity dedicated to helping those with leukaemia, lymphoma, myeloma and related blood disorders survive and then live a better quality of life.

Since 1975, the Leukaemia Foundation has been committed to improving survival for patients and providing much needed support. The Leukaemia Foundation does not receive direct ongoing government funding, relying instead on the continued and generous support of individuals and corporate supporters to develop and expand its services.

The Leukaemia Foundation provides a range of support services to patients at no cost. This support may be offered over the telephone, face to face or online 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 also funds leading research into better treatments and cures for myeloma and related blood disorders. The Leukaemia Foundation funds research grants, scholarships and fellowships for talented researchers and health professionals.



Leukaemia Foundation staff with health professional qualifications provide patients and their families with information and support

## **Support Services**

The Leukaemia Foundation has a team of highly trained and caring Support Services staff with qualifications and experience in nursing or allied health that work across the country.

We 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, DVDs, fact sheets and other resources that are available at no cost. These can be ordered via the form at the back of this booklet or downloaded from **[leukaemia.org.au](http://leukaemia.org.au)**.

### ***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 wellbeing.

### ***Emotional support***

A diagnosis of myeloma can have a dramatic impact of a person's life. At times it can be difficult to cope with the emotional stress involved. The Leukaemia Foundation's Blood Cancer Support staff can provide you and your family with much needed emotional support during this time.



### ***Blood Buddies***

This is a program for people newly diagnosed with myeloma or their carer, introducing them to a trained ‘buddy’ who has been living with myeloma for at least two years, to share their experience, their learning and to provide some support.

### ***Telephone discussion forums***

This support service enables anyone throughout Australia who has myeloma to share their experiences, provide tips, and receive education and support in a relaxed forum. Each discussion is facilitated by a member of the Leukaemia Foundation Blood Cancer Support team who is a trained health professional.

### ***Accommodation***

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

### ***Transport***

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

With the cost of hospital car parking and how difficult it can be to find a car park, the Foundation’s transport service has made my hospital visits so much easier.

The health system can feel so big and overwhelming. Sometimes I don't even know what questions to ask to get what I need. The Leukaemia Foundation's staff help by pointing me in the right direction.

### ***Practical assistance***

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

### ***Advocacy***

The Leukaemia Foundation is a source of support for you as you navigate the health system. While we do not provide treatment recommendations, we can support you while you weigh up your options. We may also provide information on other options such as special drug access programs and available clinical trials.

### ***Contacting us***

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

## BLOOD CANCERS

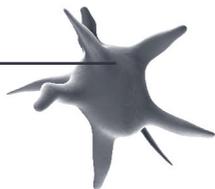
In this section of our booklet we provide a brief overview of blood cancers and related blood disorders. It is important to point out that the information provided here is of a general nature and may not necessarily apply to the specific type or severity of disease you or your loved one has.

Blood cancers occur in cells that originate in the bone marrow and are defined by the uncontrolled growth of faulty cells.

To best understand these cancers we first need to understand the bone marrow and blood.

## Platelets

Support blood clotting to stop bleeding



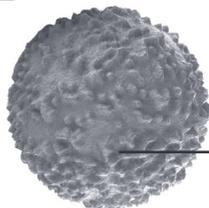
## Red blood cells

Carry oxygen for the body to produce energy



## White blood cells

Form part of the immune system



# GETTING TO KNOW YOUR BONE MARROW, STEM CELLS AND BLOOD

## 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 *haematopoiesis*. There are three main types of blood cells: red cells, white cells and platelets.

As an infant, haematopoiesis takes place at the centre of all bones. In later life, it is limited mainly to the hips, ribs and breast bone (sternum). Some of you may have had a bone marrow biopsy taken from the bone at the back of your hip (the iliac crest).

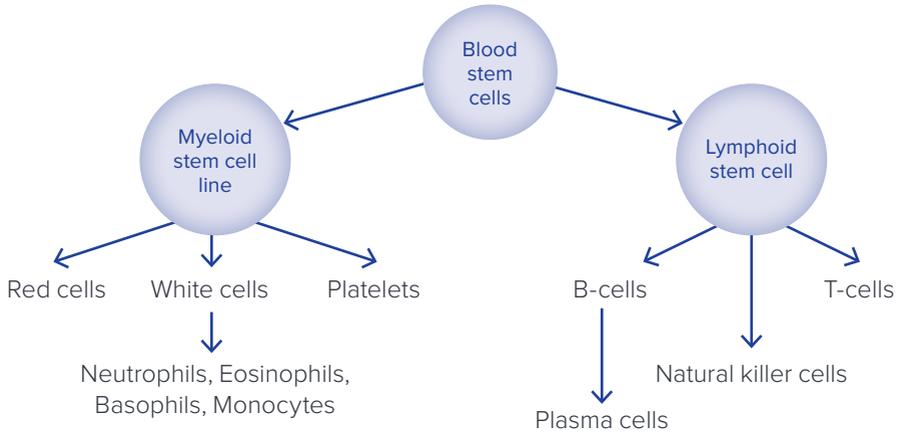
You might like to think of the bone marrow as the blood cell factory. The main workers at the factory are the *stem cells*. They are relatively small in number but are able, when stimulated, to reproduce vital numbers of red cells, white cells and platelets. All blood cells need to be replaced because they have limited life spans.

There are two main families of stem cells, which develop into the various types of blood cells.

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

**Lymphoid** ('lim-foid') stem cells develop into other types of white cells including T-cells, B-cells and Natural Killer Cells.

## Blood cell formation



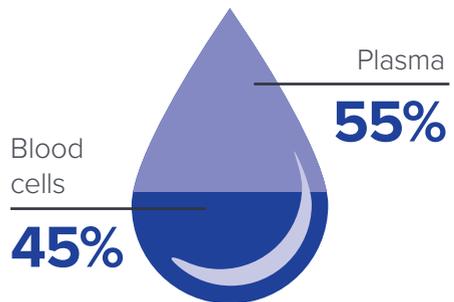
## Growth factors and cytokines

All normal blood cells have a limited lifespan in the circulation and need to be replaced on a continual basis. This means the bone marrow remains very active throughout life. Natural chemicals circulating in your blood called growth factors, or cytokines, control this process of blood cell formation. Each of the different blood cells is produced from stem cells under the guidance of a different growth factor.

Some of the growth factors can now 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 certain white cells, including *neutrophils*, while erythropoietin (EPO) stimulates the production of red cells.

## Blood

Blood consists of blood cells and plasma. Plasma is a straw-coloured fluid that blood cells use to travel around your body and also contains many important proteins and chemicals.



## Blood cells

### *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. The body uses this oxygen to create energy.

### *Haematocrit*

About 99% of all blood cells in circulation are red blood cells. The percentage of the blood that is occupied by red blood cells is called the haematocrit. A low haematocrit suggests the number of red cells in the blood is lower than normal.

### *Anaemia*

Anaemia is a reduction in the number of red cells or low haemoglobin. Measuring either the haematocrit or the haemoglobin will provide information regarding the degree of anaemia.

If you are anaemic you may feel rundown 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 blood transfusion may be given to restore the red blood cell numbers and therefore the haemoglobin to more normal levels.

## Normal ranges for adults

	 <b>Men</b>	 <b>Women</b>
<i>Haemoglobin (Hb)</i>	130 – 170 g/L	120 – 160 g/L
<i>Haematocrit (Hct)</i>	40 – 52%	36 – 46%
<i>White cell count (WBC)</i>	3.7 – 11.0 x 10 <sup>9</sup> /L	
<i>Neutrophils (neut)</i>	2.0 – 7.5 x 10 <sup>9</sup> /L	
<i>Platelets (Plt)</i>	150 – 400 x 10 <sup>9</sup> /L	

## **White cells**

White cells, also known as leukocytes, fight infection. The following is a list of some of the different types of white cells:

**Neutrophils:** Mainly kill bacteria and remove damaged tissue. Neutrophils are often called the first line of defence when infections occur. They are often the first white blood cell at the site of infection and attempt to destroy the foreign pathogen before it becomes a problem to the body.

**Eosinophils:** Mainly kill parasites

**Basophils:** Mainly work with neutrophils to fight infection

**Monocytes:** Mainly work with neutrophils and lymphocytes to fight infection; they also act as scavengers to remove dead tissue. These cells are known as monocytes when found in the blood, and called macrophages when they migrate into body tissue to help fight infection.

**B-cells:** Mainly make antibodies that target micro-organisms, particularly bacteria.

**T-cells:** Mainly kill viruses, parasites and cancer cells and produce cytokines which can recruit other cells to make antibodies which target micro-organisms.

These white cells work together to fight infection as well as having unique individual roles in the fight against infection.



### **Neutropenia**

Neutropenia is the term given to describe a lower than normal neutrophil count. If you have a neutrophil count of less than  $1 (1 \times 10^9/L)$ , you are at an increased risk of developing more frequent and sometimes severe infections.

### **Platelets**

Platelets are 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 (for example by a cut) the platelets gather at the site of the injury, stick together and form a plug to help stop the bleeding. They also release chemicals, called clotting factors that are required for the formation of blood clots.

### **Thrombocytopenia**

Thrombocytopenia is the term used to describe a reduction in the platelet count to below normal. If your platelet count drops too low, you are at an increased risk of bleeding and tend to bruise easily. Each treatment centre will have their own guidelines on the specific platelet count level when interventions may need to be taken. Platelet transfusions are sometimes given to return the platelet count to a safer level.

Regular washing of my hands has become part of my new normal

## WHAT IS MYELOMA?

Myeloma, also known as multiple myeloma, is a cancer of plasma cells that arises in the bone marrow.

Myeloma develops when plasma cells undergo a cancerous (malignant) change and become myeloma cells. These myeloma cells multiply at an increased rate and lack the controls that normally tell them to die when they are no longer needed. Myeloma cells also make chemicals that stimulate other bone marrow cells (osteoclasts) to remove calcium from the bone. As a result bones can become weaker and break more easily.

Under normal conditions, plasma cells produce immunoglobulins (antibodies) that help protect the body from infection and disease. Myeloma cells produce an abnormal type of immunoglobulin called a 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 fragments known as light chains are produced. These light chains can be detected in the blood and they may also appear in the urine. Light chains detected in the urine are called Bence Jones protein.

Most of the problems associated with myeloma are caused by the accumulation of myeloma cells in the bone marrow or bone and/or the effects of paraproteins in the blood or urine.

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.

Abnormal accumulation of myeloma cells or paraprotein in the kidney can cause kidney failure, and in the bones can cause osteoporosis, fractures or bone pain.



## What are plasma cells and immunoglobulins?

*(Imm-you-no-glob-you-lins)*

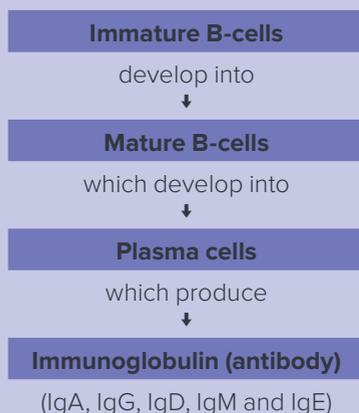
Plasma cells are a type of white blood cell that develops from mature B-cells in the bone marrow. They play an important role in protecting the body against infection and disease by producing proteins called immunoglobulins (Igs), also known as antibodies.

Immunoglobulins are produced by plasma cells in response to bacteria, viruses and other harmful organisms found in the body.

Once released into the blood stream, immunoglobulins circulate and attach to the target for which they were 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 five main classes of immunoglobulin (IgA, IgD, IgE, IgG and IgM) and each class has slightly different functions. In a normal immune response several different immunoglobulins are made and this mixture of immunoglobulins is described as 'polyclonal'. In myeloma, all the myeloma cells

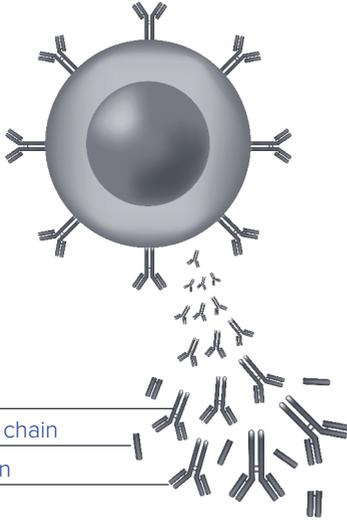
make identical immunoglobulin molecules and this is referred to as 'monoclonal'.



Immunoglobulins are Y-shaped molecules made up of two identical long pieces of protein (the heavy chains) and two identical short pieces of protein (the light chains).

There are five main families of immunoglobulins which are named after the heavy chains. (A, G, D, M and E). They can be joined to one of two classes of light chains: kappa (K) or lambda (Λ) to make, for example, IgA/ K or IgG/ Λ etc.

# PLASMA CELLS AND ANTIBODIES

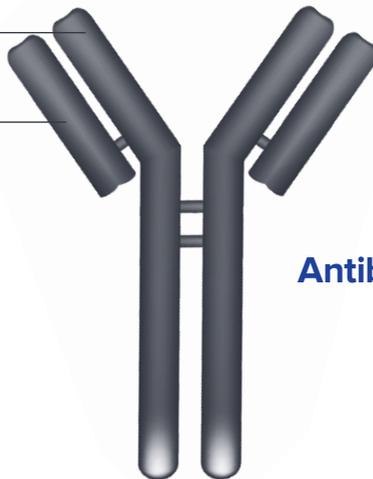


*Plasma cells are a type of white blood cell that develop from mature B-cells in the bone marrow.*

*Plasma cells in the bone marrow produce whole antibodies (=immunoglobulins) and immunoglobulin free light chains, which enter the blood stream, circulate around the body, and may be found in the urine where they are known as Bence Jones protein.*

Heavy chains

Light chains



**Antibodies**

## WHO GETS MYELOMA?

Myeloma mainly affects older people, with an average age of 70 years at the time of diagnosis, but, rarely, can still affect people in their 20s and 30s.

Myeloma is more common in men than women.

### How common is it?

Myeloma is a relatively rare disease. It accounts for approximately 1% of all cancers and 15% of all blood and bone marrow cancers. Each year in Australia, approximately 1,800 people are diagnosed with myeloma, equivalent to five people every day.

### What causes myeloma?

Many people who are diagnosed with myeloma ask the question 'Why me?'. Naturally, they want to know what has

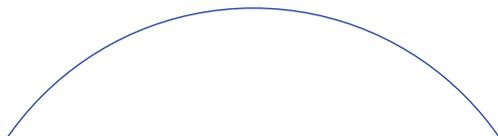
happened or what they might have done to cause their disease. The truth is that no one knows exactly what causes myeloma. We do know that it is not contagious.

You cannot 'catch' myeloma by being in contact with someone who has it.

Although there are rare cases where myeloma occurs in families, in the vast majority of 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.

It is widely accepted that all people with myeloma originally had a disorder called Monoclonal Gammopathy of Uncertain Significance (MGUS). However only a small number of people with MGUS are actually diagnosed with it as it is a condition where people have an abnormal paraprotein but no other noticeable symptoms.





## WHAT ARE THE SYMPTOMS OF MYELOMA?

The symptoms of myeloma depend on which parts of the body are most severely affected and how advanced the disease is. A person with myeloma may not necessarily experience all of the symptoms described below. In the earliest stages, there may be no symptoms and myeloma may be incidentally 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 that stimulate other bone marrow cells (osteoclasts) to remove calcium from the bone. Over time bones can become weakened and thinned (osteoporosis) and areas that look like holes on X-rays (lytic lesions) may develop, increasing the risk of fracture (breaking of bone).

### Elevated blood calcium level (hypercalcaemia)

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.

### Anaemia

Anaemia is a reduction in the number of red blood cells circulating in the blood, or a low haemoglobin level.

Haemoglobin is the red pigment in the red blood cells that makes blood appear red in colour. The level of anaemia is most commonly measured by the level of haemoglobin in the blood. The symptoms of anaemia include a lack of energy, persistent tiredness and fatigue, weakness, dizziness and feeling unusually short of breath when physically active.

People with anaemia often have a pale complexion.

## Kidney damage

There are many causes of kidney damage in myeloma. The paraprotein found in the urine (known as Bence Jones protein) is produced by the myeloma cells and can damage the kidneys. Other factors such as dehydration and hypercalcaemia can also cause kidney damage. The symptoms of kidney damage may include fatigue, mental confusion, swollen ankles and altered urinary output.

## Frequent infections

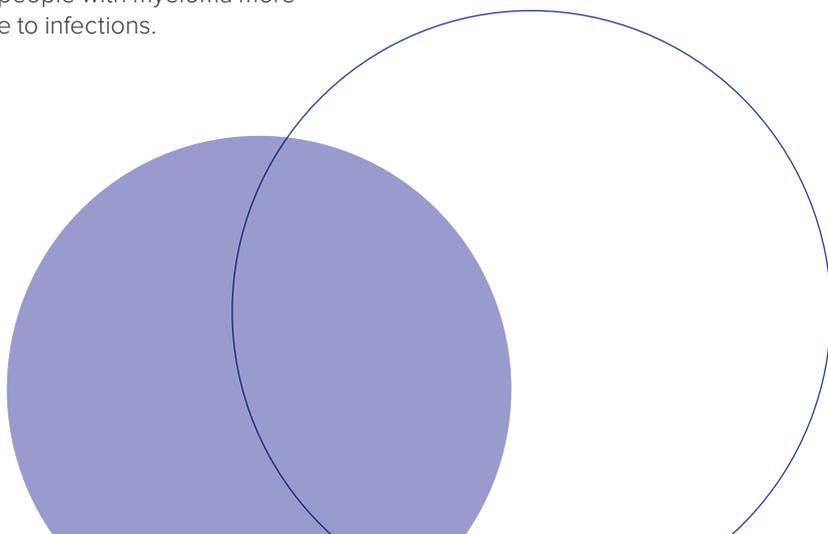
People with myeloma are at risk of developing frequent infections. As myeloma cells multiply, they crowd the bone marrow and prevent it from making normal numbers of white cells which can also interfere with the production of normal antibodies. This can make people with myeloma more susceptible to infections.

## Increased bleeding and bruising

A low platelet count can cause bruising for no apparent reason, or excessive or prolonged bleeding following injury.

Some people notice bruises on their skin for no apparent reason, more frequent or severe nose bleeds, bleeding gums or excessive or prolonged bleeding following minor cuts or injury.

Excessive bleeding and bruising in myeloma can also be due to interference of the paraprotein or light chains with the ability for blood clots to form, or merely to very high levels of paraprotein causing hyperviscosity (increased thickness) of the blood.





## 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, bone marrow biopsy, X-rays and other more specialised bone imaging tests.

### Blood and urine tests

Serum (blood) and urine electrophoresis and immunofixation (IFE) are tests carried out to detect paraprotein and Bence Jones protein and measure the amount and type of protein in the blood and urine.

For accurate measurement of the Bence Jones protein, you may be required to collect a 24-hour urine specimen (every bit of urine you pass during a whole day). You will be supplied with a suitable container for this collection.

*The amount of paraprotein present in your blood and the amount of Bence Jones protein in your urine is usually a good indicator of the amount of myeloma in your body at the time of diagnosis or after treatment. Generally rises or falls in the paraprotein parallel the progress of myeloma. This information provides a baseline which can be compared with later results to see how you are progressing.*

### ***Serum biochemistry (measurement of the various chemicals in the blood)***

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 the kidneys are not working properly.

A high blood calcium level usually indicates calcium is being removed from the bones faster than the kidneys can remove it from the blood. To try to get rid of the excess calcium the kidneys produce increased amounts of urine and this can cause dehydration and kidney failure.

Reduced levels of albumin (a normal blood protein) may be found in myeloma as chemicals (cytokines)

produced by myeloma cells may suppress albumin production.

Beta-2-microglobulin is a small molecule that sits on the surface of lymphocytes and plasma cells. The level of beta-2-microglobulin is helpful in staging myeloma and in determining the likely course of the myeloma (prognosis). High levels of beta-2-microglobulin indicate the presence of a large number of myeloma cells, and/or the presence of kidney damage. The beta-2-microglobulin level and the level of albumin together form the basis of the prognostic system called the International Staging System (ISS) in myeloma. The levels of other proteins, the lactate dehydrogenase (LDH) and C-reactive protein (CRP) also may indicate the way the myeloma is likely to behave.

### ***Serum free light chains***

The measurement of the level of the light chains in the blood is a test which can be used as an alternative to other blood or urine tests in some cases. The level of light chains (either kappa or lambda) can reflect the amount of myeloma present. This test is especially useful for following the progress of the disease in the small percentage of patients who previously were said to be 'non-secretory', meaning there was no detectable paraprotein in their blood or Bence Jones protein in their urine.

### ***Blood count***

A full blood count (FBC) or complete blood count (CBC) measures the number of red cells, white cells (including the different kinds of white cells) and platelets in circulation. This helps to assess how well the bone marrow is functioning and whether or not normal blood cells are being affected by myeloma.

## Bone marrow biopsy

The presence of more-than-normal numbers of plasma cells in the bone marrow is the most important part of the diagnosis of multiple myeloma. Under normal conditions plasma cells make up less than 5% of all the cells in the bone marrow.

In myeloma the number is usually over 10% and can be as high as 90%. As well as the assessment of the number of plasma cells, the bone marrow is also examined for changes in the chromosomes and genes of the myeloma cells.

Cytogenetic tests or analysis of chromosomes may be predictive factors of response to treatment and the ultimate outcome i.e. the prognosis, and guide treatment choices.

A bone marrow biopsy involves taking a sample of bone marrow, usually from the back of the iliac crest (hip bone) to see if there are any myeloma cells present in the bone marrow, and to see how well it is functioning.

A sedative, short-acting anaesthetic or a pain killer is usually given before the biopsy 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 will probably feel an uncomfortable stinging sensation.

A long thin needle is then inserted through the skin and into the bone and bone marrow. 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).

In addition, a small sample of bone with the intact bone marrow inside is obtained for further tests (this is called a bone marrow biopsy). You might have some soreness and bruising at the biopsy site afterwards and you may need to use paracetamol or a stronger painkillers for a few days.

Because you might feel a bit drowsy afterwards you should be accompanied by a family member or friend who can take you home after the procedure.

## 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 (backbone), ribs, pelvis (hips), legs and arms. Areas of bone damage, or 'lytic lesions', may show up on standard X-rays. Whole body low dose CT are also increasingly used in place of skeletal survey as this is more sensitive for detecting myeloma bony lesions. It uses a much lower dose of radiation compared to usual CT and is generally well tolerated.

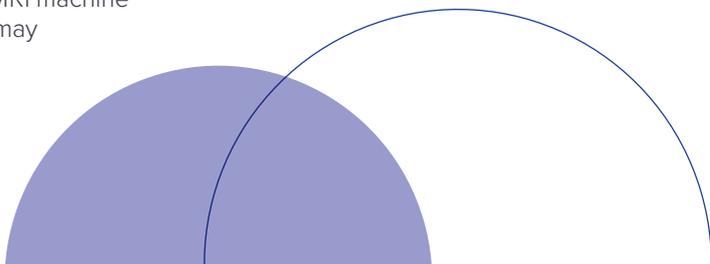
Magnetic resonance imaging (MRI) may also be used to detect early bone changes as well as soft tissue myeloma tumours (e.g. involving the spinal canal, liver etc) caused by disease. This test uses a very strong magnet to produce very clear and detailed three dimensional (3D) images of the part of the body being examined. Some MRI machines are capable of imaging the whole body in one examination.

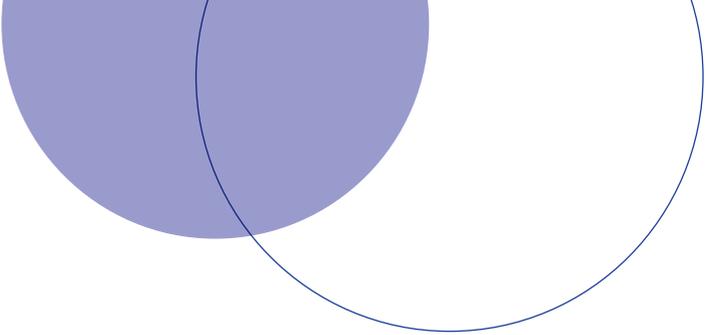
MRI is painless and usually takes about an hour to complete. However, some people find being in the MRI machine very claustrophobic and may

experience some anxiety during the examination. It is also noisy. If you feel this may be an issue for you, discuss it with the staff prior to your test and they may have some suggestions to help alleviate your anxiety.

PET (positron emission tomography) relies on the different way cancer tissue uses glucose, compared with normal tissues. Myeloma cells come up brighter in the pictures because they are more active and take up more of the sugar radiotracer that is placed in your body. This test is being used increasingly in myeloma, as it is capable of differentiating active cancer from 'burnt out' cancer tissue.

Waiting around for tests can be stressful, time consuming 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.





## PROGNOSIS

**A prognosis is an estimate of the likely course of a disease and the chance of it being cured or controlled.**

Your doctor is the best person to tell you your prognosis. You should be aware prognostic predictions are only estimates based on the available information about your disease and currently available treatments. You are unique and these estimates may not represent the actual course of your disease.

Although there is currently no cure for myeloma, in most cases the progression of the disease can be halted for a period of time, and some people with myeloma can have a long duration of stable disease, which can also be termed remission.

In this period of remission you may to some extent be able to return to your normal activities and lifestyle. The development of new and improved treatments in recent years has meant the outlook for people with myeloma is improving and there is promise of further improvements in the future.



## STAGING

The stage of your myeloma refers to the extent of the disease in your body and is one of the factors used to predict the course of your disease and potential treatments.

Currently, the International Staging System (ISS) is the most commonly adopted prognostic model for myeloma, based on two blood tests, which can be performed in most pathology laboratories: the level of beta-2 microglobulin and albumin in the blood.

There are three stages of myeloma. Stage I is early disease. Stage III is advanced disease where there is a large amount of myeloma in the body. Stage II is in between.

Besides the staging system described above, the outcome of the disease and its treatment is also determined by the presence or absence of other test abnormalities including chromosome abnormalities. A bone marrow biopsy is performed to allow a test called cytogenetics (where the genetic material from the myeloma cell can be looked at under a microscope). The presence or absence of certain chromosome abnormalities, influences the prognosis, such as these chromosome abnormalities, t(4;14),

t(14;16) and del(17p), which are known as higher risk abnormalities.

This has recently led to a proposed revised staging known as (R)-ISS risk stratification system which incorporates ISS stage, LDH and high risk cytogenetic features. The R-ISS risk stratification system was recently shown to clearly identify three different prognostic groups in patients who were treated in the era of the newer myeloma treatment agents (IMiDs and proteasome inhibitors). The R-ISS staging system is being increasingly used by doctors treating patients with myeloma and may replace the ISS method.

The CRAB criteria is often used to identify whether a person has active myeloma that may require treatment. The acronym CRAB consists of (C) increased calcium level, (R) renal (kidney) problems, (A) anaemia and (B) bone changes (lytic lesions or bone loss). One or more of these CRAB symptoms indicates symptomatic myeloma which requires treatment.

Additionally, there can be indications for treatment outside of these CRAB criteria where there is a heavy bone marrow burden of myeloma (>60% of plasma cells); heavy burden of serum free light chains (ratio >100); and bone lesions seen on a MRI scan.



## TYPES OF MYELOMA

### **International Staging System (ISS) 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)

Myeloma can be classified according to its distribution in the body. In the majority of cases myeloma can be found throughout all the bone marrow, which is why the disease is also called multiple myeloma.

Occasionally there is an isolated myeloma lesion found only in one site. This is called a solitary plasmacytoma. Such plasmacytomas can sometimes be cured with radiotherapy alone.

Myeloma can be classified according to the type of immunoglobulin being secreted. The most common type of myeloma is IgG myeloma representing between 50 – 60% of all cases. IgA myeloma represents about 20% of all cases. Light chain myeloma, in which the myeloma cells make only kappa or lambda light chains, represents about 20% of cases. IgM, IgD, IgE and non-secretory (no paraprotein or excess light chains) myelomas are rare.

Some people have an increased number of plasma cells in their bone marrow, but do not fit the criteria for a diagnosis of multiple myeloma. Their disease may be classified as 'MGUS' or smouldering myeloma.

### ***Monoclonal gammopathy of undetermined significance (MGUS)***

MGUS is a non-malignant (non-cancerous) condition related to myeloma. It also involves the production of paraprotein and/or excess light chains, but the number of plasma cells in the bone marrow is normal or only slightly increased. None of the symptoms associated with myeloma are present.

MGUS is quite common in elderly people. 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 as over time only some people with MGUS will develop myeloma. A higher level of the paraprotein in the blood can be indicative of a greater likelihood of myeloma developing and can be used to decide the frequency of follow-up.

Currently there are no treatments available that have been shown to be beneficial in preventing the progression of MGUS.

### ***Smouldering or Asymptomatic Myeloma***

Smouldering or asymptomatic myeloma is a very early phase of myeloma. There are no symptoms associated with myeloma, but a bone marrow biopsy shows definite evidence of myeloma.

People diagnosed with smouldering myeloma do not need treatment straight away. They should be carefully followed up so treatment can be given when the disease progresses and before significant complications develop. This may happen after a few months or after several years. As with MGUS, the level of the light chains in the blood can be indicative of the likelihood of progression and can be used to decide the frequency of follow-up.

Currently there are no treatments available that have been shown to be beneficial in preventing the progression of smouldering myeloma.

# TREATMENTS

The treatment for myeloma consists of both therapeutic and supportive therapy. Therapeutic therapy targets the cancerous myeloma cells with the aim of reducing the amount of these cells.

Supportive therapy is designed to prevent and manage the symptoms of myeloma, and is appropriate for all patients.

The treatment chosen for your disease depends on several factors including the stage of your myeloma, your general health, your age, the treatment you have had in the past, the response you have had to previous treatment and any complications or side-effects you have experienced with previous treatments.

Information gathered from thousands 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.

If you have symptoms and require treatment, it will be aimed at reducing the amount of myeloma to achieve remission as long as possible, or at a minimum to stabilise the disease to stop its progression. Over time you may be treated with several different types of therapy depending on the effectiveness and side-effects experienced.

Additionally, treatments may be needed to control symptoms, such as pain or nausea and to prevent or treat complications of the disease, such as hypercalcaemia, bone fractures and kidney failure.

For me fatigue is my worst side effect. It is more than just feeling tired and it is hard to explain to others how it feels

Treatment for myeloma may involve the use of:

- » chemotherapy (intravenous or tablet form)
- » corticosteroids
- » immunomodulatory drugs (also known as IMiDs), such as thalidomide, lenalidomide or pomalidamide
- » targeted treatment such as proteasome inhibitors, including bortezomib or carfilzomib
- » high-dose therapy and stem cell transplantation
- » antibodies, such as daratumumab and elotuzumab
- » radiotherapy
- » bisphosphonates
- » new treatments on clinical trials not yet available for general use.

### **Standard therapy**

Standard therapy refers to types of treatment that are commonly used and have been tried and tested both in clinical trials and through long-term clinical use. These have been proven to be effective and have acceptable side-effects.

### **Clinical trials**

These trials (also called research studies) test new treatments and compare them to the current standard treatment to see if the new treatment works better or has more or less side-effects. Some clinical trials are randomised, so some patients will be receiving the new treatment and others the standard treatment. Clinical trials are important because they provide vital information about how treatment could be improved. Clinical trials can also be very beneficial as they give people with myeloma access to new therapies not otherwise available in Australia.

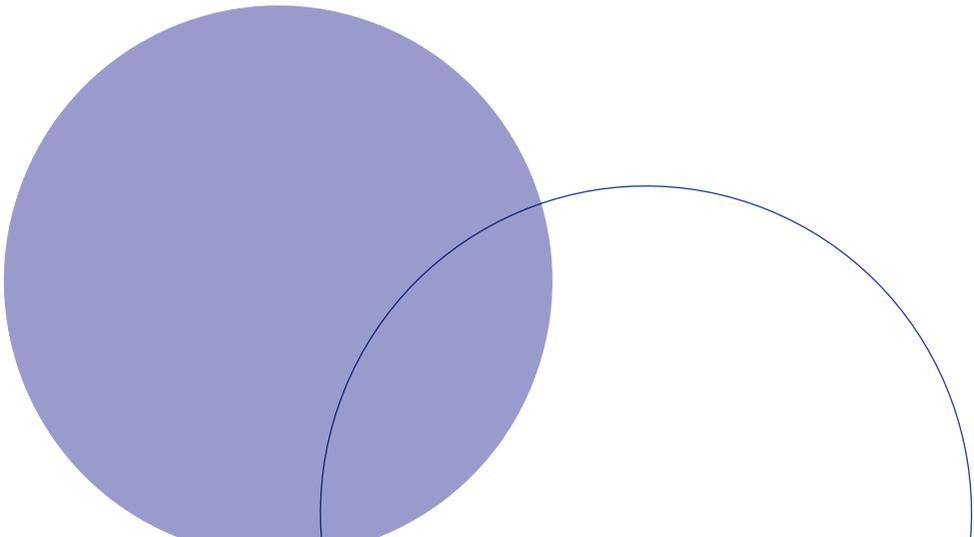
Previous myeloma clinical trial participants have helped double the average expected survival time in myeloma patients over the last 10 years.

If you are considering taking part in a clinical trial make sure 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 consent. Talk to your doctor who can guide you in making the best decision for you.

### ***Informed consent***

Giving an informed consent means the potential risks and potential benefits of a proposed procedure or treatment have been fully explained to you. You will then be asked if you are willing to comply with the requirements of the therapy, including treatments, tests and the collection of relevant information about you and your disease. If you feel happy you have adequate information to make such a decision, you may then give your informed consent.

You should never feel compelled to be in a clinical trial. The decision to participate must always be voluntary, after you have received all the necessary information. Even after you have agreed to be in a clinical trial and signed the consent form, you can change your mind, whether or not trial treatment has already started.



# TREATMENTS TO CONTROL MYELOMA

There has been a great deal of research in myeloma over the last 10 years and new forms of treatments have become available. These therapies work very differently to standard chemotherapy. The addition of these therapies has improved the outcomes for people being treated with myeloma and they are now part of standard treatment in myeloma therapy.

## Initial therapy

Initial treatment regimes will usually consist of a combination of medications, usually a newer agent, such as an immunomodulatory agent (IMiDs) or proteasome inhibitor, together with corticosteroids and often with chemotherapy.

The choice of initial treatment will often depend on whether your doctor thinks high dose chemotherapy and autologous stem cell transplantation\* (using your own cells) will be of benefit to you. This treatment has been shown to keep the myeloma under control

effectively in many people for a period of time. It achieves this better than standard chemotherapy alone and better than IMiDs and proteasome inhibitors as well.

The drug combination you initially receive will depend on what the ultimate treatment plan for you is. This will be discussed with you before you start treatment.

*\* There is a separate Leukaemia Foundation booklet called 'Understanding Autologous Stem Cell Transplants' which provide more details on these types of treatments.*

Treatment regimens consist of a combination of medications given in cycles. The length of the cycle is most commonly between three to six weeks.

The total number of cycles received is generally referred to as a course of treatment. The cycles are repeated at regular intervals. However the interval can be extended if the side-effects, most often blood counts, have not recovered sufficiently.

## Immunomodulatory drugs (IMiDs)

Immunomodulatory drugs are derived from thalidomide and act by impacting the immune system, have anti-inflammatory properties, reduce the body's ability to produce new blood vessels, and reduce the ability for the myeloma cells to multiply.

### *Thalidomide [capsules]*

Thalidomide is a drug developed in the 1950s. It was originally used as a sedative and to treat nausea and vomiting associated with early pregnancy. It caused severe birth defects in the babies of mothers who were taking it. As a result, the use of thalidomide was restricted to very severe medical conditions like leprosy.

In the late 1990s thalidomide was found to be an effective anti-myeloma drug and over the last 10 years has become part of routine treatment.

Thalidomide is more effective when it is combined with prednisolone or dexamethasone and/or chemotherapy.

Its use is now governed by very strict regulations to ensure it can never again be taken by pregnant women.

Thalidomide is a drug that works in a number of ways to interfere with the growth and survival of myeloma cells. In general thalidomide works by preventing the growth of new blood vessels (angiogenesis) which myeloma cells need to survive. It also stimulates the body's own immune system to attack myeloma cells, blocking chemicals that myeloma cells need to grow and survive and directly kills myeloma cells.

Thalidomide has side-effects which have to be carefully monitored.

These include:

- » Sleepiness
- » Constipation
- » Rashes
- » Increased tendency to form blood clots
- » Peripheral neuropathy (damage to the nerves) leading to numbness and tingling, particularly in the fingers and toes.

It is important to tell your doctor if you experience symptoms as the dose of thalidomide may need to be reduced or stopped.

### ***Lenalidomide [capsules]***

Lenalidomide was derived by modifying the chemical structure of thalidomide. It is effective as part of induction therapy and in people whose myeloma has relapsed or is refractory to other treatment. Lenalidomide is often given in combination with dexamethasone and is generally given as a tablet for 21 days out of every 28, with treatment continuing until the myeloma stops responding or side-effects make continuing treatment undesirable.

The most common side-effects of lenalidomide include:

- » Reduction in white cell and platelet counts
- » Fatigue
- » Blood clots.

Lenalidomide is also thought to cause birth defects so the same precautions as for thalidomide are required to prevent pregnancy.

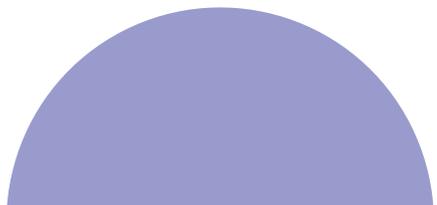
### ***Pomalidomide [tablet]***

Pomalidomide is a third generation immunomodulatory (IMid) which has a similar chemical structure to thalidomide and lenalidomide. Pomalidomide is given orally usually for 21 days followed by a seven-day rest. This makes up one 28-day cycle and treatment is normally continued until disease progression i.e. it is given as an ongoing treatment rather than in a fixed number of treatment cycles. It is usually given in combination with a steroid, such as dexamethasone or prednisolone.

The most common side effects of pomalidomide include:

- » Lowered blood cell counts
- » Fatigue
- » Nausea, vomiting, diarrhoea and constipation
- » Blood clots.

Pomalidomide is also thought to cause birth defects so the same precautions as for thalidomide are required to prevent pregnancy.



## Proteasome inhibitors

Proteasome inhibitors are targeted therapies that work by blocking the action of proteasomes, which are found in all cells throughout the body.

Proteasomes act as the 'waste disposal unit' for cells, breaking down and recycling proteins from old cells.

Proteasome inhibitors block the waste disposal unit in myeloma cells, affecting their growth and survival.

### ***Bortezomib [injection]***

Bortezomib is effective as initial (induction) treatment, generally used in combination with corticosteroids (dexamethasone or prednisolone) and sometimes with chemotherapy (such as cyclophosphamide). It is also effective in people whose myeloma has relapsed or is refractory (resistant) to other treatments. Bortezomib is given by injection either under the skin (subcutaneously) or into the vein (intravenously) once or twice a week in a three to four week cycle.

Bortezomib has a number of side-effects including:

- » Nausea
- » Vomiting
- » Diarrhoea
- » Low numbers of white cells and platelets
- » Low blood pressure
- » Peripheral neuropathy.

### ***Carfilzomib [injection]***

Carfilzomib is a newer proteasome inhibitor, working in a similar way to bortezomib but has been developed to target a different part of the proteasome.

It is given as an infusion into the vein (intravenously) on two consecutive days per week for three weeks followed by a one week break. Although it can be given on its own it has been shown to be more effective when used in combination with other myeloma therapies, and is usually given with steroids. Carfilzomib received PBS reimbursement in Australia in January 2018 for use in relapsed myeloma after one prior line of treatment.

Carfilzomib has a number of potential side-effects, and special monitoring is required for those who have pre-existing heart or blood pressure problems as in a small number of cases it has been found to worsen these conditions. The most common side effects include:

- » Reduction in red cell and platelet blood counts
- » Fatigue
- » Nausea
- » Difficulty breathing
- » Nausea, diarrhoea
- » Increased blood pressure.

## Chemotherapy

As myeloma is usually present throughout the bone marrow, chemotherapy is one of the forms of treatment used to control it.

Chemotherapy literally means therapy with chemicals. Chemotherapy drugs (also called cytotoxics) damage and kill cells. Most cancer cells are more vulnerable to the effects of chemotherapy than normal cells, but some normal cells are also damaged. This is why most patients experience some side-effects while they are having chemotherapy.

Myeloma treatment often combines chemotherapy drugs with corticosteroids, most commonly dexamethasone or prednisolone, and an immunomodulatory drug or a proteasome inhibitor.

## Corticosteroids

Corticosteroids are hormones produced naturally by the body in the adrenal glands. Man-made corticosteroids such as prednisolone, dexamethasone and methylprednisolone are very important in the treatment of myeloma and are commonly used alone or in combination with chemotherapy or targeted therapies in the treatment of myeloma.

Corticosteroids are not chemotherapy, however they can make myeloma cells less likely to survive by affecting lymphocytes and the immune system. When used with certain other therapies they can also enhance the effects of these drugs in killing the myeloma cells.

The types of side-effects seen with corticosteroids depend on the dose and the length of therapy. If you are taking a high dose for a short time (also called pulsed steroid therapy) you may notice that your appetite increases.

Some people find it more difficult to get to sleep at night and it is generally recommended that steroids are taken early in the morning and medications,

such as sleeping tablets or natural therapies, are used to help with sleep if needed. Depression, mania and other changes in mood (e.g. anxiety, increased aggression) can also occur. When you are on high dose corticosteroids, it is important to remember to avoid making major life decisions or spending large amounts of money without taking into consideration that your choices may be influenced by the medication.

Corticosteroids can also cause higher blood sugar levels. People with diabetes may find they need more of their diabetes medication when taking corticosteroids.

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 corticosteroids will be given information on diet and taught how to measure their blood sugar levels and adjust their medication. Advice from a dietitian may also be useful. Many of the side-effects of corticosteroids are temporary and should pass once you finish taking them.

Corticosteroids may also cause stomach irritation or reflux (heartburn) and medications are often used to prevent or treat this symptom during courses of chemotherapy or during transplant. It is also helpful to take oral corticosteroids with food which may reduce these symptoms.

Long-term use may cause other effects such as fluid retention, weight gain, cataracts, muscle weakness and an increased susceptibility to infections. Aching joints (knees and hips) have also been reported. Remember to tell your doctor, nurse or pharmacist about any side-effects you are having as they can usually suggest ways to help you.

## Autologous stem cell transplants

In autologous stem cell transplantation, you are given some (induction) chemotherapy, injected with a growth factor (G-CSF) that causes your bone marrow to produce many stem cells. These stem cells are then collected, frozen and stored for when you need them. Frequently, enough stem cells are collected for two autologous stem cell transplants.

When you are to undergo your transplant, you will then be given a high dose of chemotherapy to kill off the remaining cells in your bone marrow.

When this chemotherapy has been excreted from your body, your collected stem cells are then thawed, and re-infused into your veins, where they make their way back into your bone marrow to start the process of restoring, or 'rescuing', it.

Some of the chemotherapy drugs commonly used to treat myeloma can make it difficult to collect stem cells. The

initial choice of treatment drugs will depend on whether or not your doctor thinks that in your case a stem cell transplant will be beneficial. This will mostly depend on your general health.

People who have a significant other illness, or are very elderly, may not be well enough to undergo a stem cell transplant and will be treated with conventional chemotherapy only. In some people, other types of therapy will be used at the early stage of treatment, often in combination with chemotherapy.

If transplantation is planned, melphalan is usually avoided during induction treatment because it can make it difficult to collect the stem cells required. Instead treatment regimens that have little effect on the ability to successfully collect stem cells may be used. The types of combinations may include tablets/capsules, intravenous drugs (given into a vein), or subcutaneous drugs (given as an injection under the skin).

## Allogeneic stem cell transplants

Transplantation from a donor is also occasionally used in treating myeloma. At present allogeneic transplantation is the only form of treatment that offers hope of curing the disease.

Unfortunately, for people with myeloma there remains some significant risks associated with this form of transplant. A person undergoing this procedure needs to be in optimal health and therefore it is often reserved for younger patients.

Research is currently being done to find the safest and most effective way of using this treatment for people with myeloma.



## New and emerging treatments

As the understanding of myeloma and the immune system grows, research into new agents has advanced and there are promising new classes of drugs showing efficacy in myeloma treatment. The complexity of the immune system means there are many different ways the immune system can be triggered to kill myeloma cells.

In 2015 five new drugs were approved by the US food drug administration (FDA) for the treatment of relapsed and refractory myeloma: second generation proteasome inhibitors, carfilzomib (PBS funded in Jan 2018) and ixazomib; monoclonal antibodies (mAb), daratumumab and elotuzumab; and the histone deacetylase inhibitor (HDACi), panobinostat.

Although not all reimbursed by the Australian Pharmaceutical benefits scheme (PBS) or approved for use in myeloma in Australia, they are in use in clinical studies and show promise.

Other promising novel agents in early development include novel immune therapies of chimeric antigen receptors (CAR-T) and bispecific T-cell engagers (BiTEs) as well as small molecules such as the BH3-mimetic, venetoclax.

It is hopeful that the combination of these new agents may enhance the effect of established treatment combinations but more research into their use is still needed.

## How is treatment given?

Some treatment is given in tablet/capsule form (orally). Others are given by injection either under the skin (subcutaneously) or into a vein (intravenously or IV).

If you are having several cycles of intravenous chemotherapy, or if the chemotherapy has to be given continuously for several days your doctor may recommend that you have a PICC (peripherally inserted central catheter) inserted.

A PICC is a long intravenous tube threaded up one of the arm veins to the large vein leading from your neck to your heart (superior vena cava). This is usually done by a nurse or a doctor with ultrasound guidance so they can see the tube is going to the correct place.

PICCs can often stay in place for several months and can be used throughout your whole treatment for all intravenous drugs, blood transfusions and for blood sampling.

PICC lines require special care to prevent infection. The nurses at your treatment centre will teach you how to look after your line. There are also other less commonly used intravenous lines, such as central venous catheters, infusaports and portacaths.

In many cases you don't need to be admitted to hospital for treatment. Treatment is usually able to be given in the hospital day unit or at home.

At times, you may need to be admitted to hospital for investigation and treatment of complications of myeloma or side effects of treatment.

## 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.

Occasionally radiotherapy to the whole body (total body radiotherapy) is used in preparation for an allogeneic stem cell transplant.

### ***What is involved in radiotherapy?***

Before you start radiotherapy, the radiation oncologist (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 semi-permanent 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.

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 nurse 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.

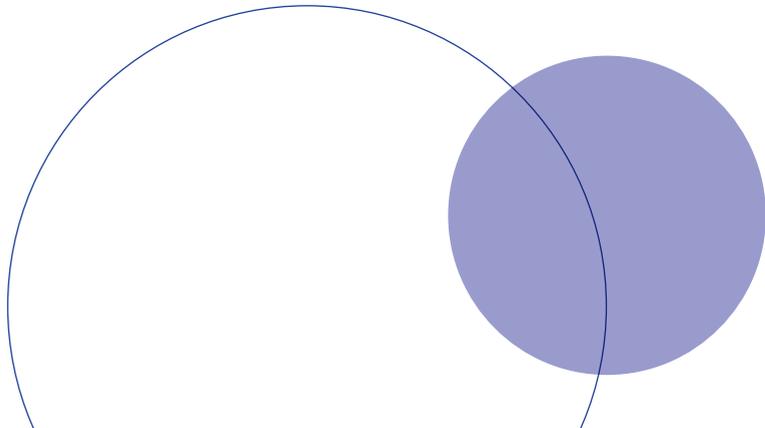
Radiotherapy is painless. In fact you do not see or feel anything during the actual treatment. You will however need to stay perfectly still for a few minutes while the treatment is taking place. You might like to bring along some music to help you relax.

### ***Monitoring myeloma and response to treatment***

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

It is important to realise 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 should 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.

See the next page for more about treatment responses.



## Commonly used treatment response terms

In myeloma, different levels of response to treatment are recognised and the quality of response has been shown to indicate future prognosis.

### ***Complete response (CR)***

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

### ***Very good partial response (VGPR) and partial response (PR)***

This indicates a good, but lesser response to treatment than CR, with persistent signs of myeloma in the various laboratory tests. VGPR refers to >90% decrease in paraprotein and PR >50% decrease in paraprotein.

### ***Minimal response***

More than 25% but less than 50% decrease in paraprotein.

### ***Stable disease***

Less than 25% decrease in paraprotein but the paraprotein is not increasing.

### ***'Plateau phase' or remission***

The progression of myeloma has been halted and the paraprotein level is stable. The myeloma is not

getting any worse or any better with further treatment.

The length of time that the remission lasts varies from person to person and eventually the myeloma will reappear. Regular checkups are necessary while you are in remission so reappearance of the disease can be detected and further treatment started before any serious complications develop.

### ***Resistant/refractory disease***

The myeloma is not improving with treatment.

### ***Disease progression/relapse***

The myeloma is getting worse on or off treatment.

### ***Minimal Residual Disease (MRD)***

Minimal residual disease is a term used to describe the level of myeloma cells that remain following treatment that are not routinely visible through conventional tests or measurements. Using a bone marrow sample very sensitive techniques are used to determine if there are even extremely low levels of myeloma remaining (residual) in the bone marrow. These techniques are being used in the clinical trial setting to help determine whether one treatment is better than another. It is not yet used in clinic setting as there are still a lot of questions to answer as to what technique should be used and how to use this information in guiding treatment.

## Side-effects of treatment

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

The side-effects you experience and the severity of the side-effects vary depending on the treatment regimen being given, the stage of disease and other individual factors.

There is no doubt side-effects can be very unpleasant but fortunately most of them are temporary and reversible. It is important you report any side-effects you are experiencing to your doctor, nurse or pharmacist because many of them can be treated successfully, reducing any unnecessary discomfort for you.

### ***Effects on the bone marrow***

Chemotherapy and some of the newer therapies can affect the bone marrow. As a result, your blood counts (the number of white cells, platelets and red cells circulating in your blood) may fall following treatment. The length of time it takes for your blood counts to fall and recover depends partly on the drugs used and on how severely the bone marrow is affected by the myeloma.

### ***White cells***

There are several different kinds of white cells normally circulating in the blood. The white cells that are the first line of defence against infection are called neutrophils. When the neutrophil count drops to a very low level (neutropenia) you are much more likely to develop an infection and it is much more difficult for your body to fight off an infection once you have one.

Most treatments cause a temporary neutropenia, but the level to which the count drops and how long it stays down varies between different individuals according to the treatment combination given and the doses of medication used.

While you are having treatment that can decrease the number of white cells it is important to take sensible precautions to help prevent infection. These include:

- » Avoiding crowds (for example shopping centres and movie theatres)
- » Avoiding close contact with people with infections that are contagious (for example colds, flu, chicken pox)
- » Eating food only if it has been properly prepared and cooked. Avoid reheated food, soft cheeses and salad bars.

Although precautions against infections are important, many infections arise from 'harmless' organisms that you carry in your body all the time, but which are capable of causing infection when your immunity is down.

### **Infection**

If you do develop an infection you may experience a fever, which may or may not be accompanied by episodes of shivering, called rigors. **Infections while you are neutropenic can be very serious, even life threatening, and need to be treated with antibiotics as soon as possible.**

### **Granulocyte-colony stimulating factors (G-CSF)**

Sometimes your doctor may decide to use injections of a medication that reduces the length of time you are neutropenic, thus reducing your chance of getting an infection. These medications, which are a manmade form of a naturally occurring cytokine in the body called granulocyte-colony stimulating factor, work by stimulating the bone marrow to increase the production of neutrophils.

These are given by an injection under the skin (subcutaneous). This is a simple procedure and the nurses can usually teach you or a family member (or friend) to do this at home.

### **Platelets**

Your platelet count may be reduced by your disease and by treatment.

A low platelet count is called thrombocytopenia. When your platelet count is low you can bruise and bleed more easily. During this time you should avoid sharp or harsh foods such as chop bones, potato crisps and crisp toast as these can damage your gums and cause bleeding and infection. Use only a soft toothbrush. 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 be anaemic. When you are anaemic you feel more tired and lethargic than usual. If your haemoglobin level is very low, your doctor may give you a red cell (blood) transfusion.

## ***Nausea and vomiting***

Nausea and vomiting may be associated with chemotherapy, bortezomib and some forms of radiotherapy. These days however, thanks to significant improvements in anti-sickness (antiemetic) medications, nausea and vomiting are generally very well controlled. You will be given antiemetic medications before and sometimes also for a few days after your treatment.

Be sure to tell your doctor, nurse or pharmacist if the antiemetics are not working for you and you still feel sick. There are many different types of antiemetic medications that can be tried.

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 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 antiemetic medications as recommended by your doctor, nurse and pharmacist should also help.

## ***Bowel changes***

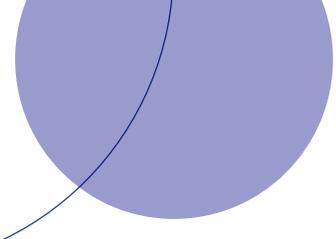
Chemotherapy and certain newer therapies, such as the IMiDs and proteasome inhibitors, can affect the functioning of your bowel.

Chemotherapy agents can also affect the lining of your bowel wall. This can lead to cramping, wind, abdominal swelling, constipation and diarrhoea. Be sure to tell the nurses and doctors if you experience any of these symptoms.

If you develop diarrhoea, a specimen may be required from you to ensure 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.

Severe diarrhoea and vomiting may cause dehydration, which in turn may damage the kidneys. If you have severe diarrhoea you may need intravenous fluid in the hospital day unit or you may need to be admitted to hospital.

It is also important to tell the doctor or nurse 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. It is common for haemorrhoids to flare up after chemotherapy treatment, and constipation will further inflame this.



## **Peripheral neuropathy**

The term peripheral neuropathy refers to damage to the nerves that branch off from the spinal cord. It changes the way they work resulting in altered sensation, tingling, numbness, pain or weakness, especially in the extremities, e.g. hands and feet. Peripheral neuropathy may result from the effects of the myeloma, paraprotein, from viral infection (shingles), or, more commonly, as a complication of the medications used to treat myeloma, including bortezomib, thalidomide and lenalidomide.

There is no simple way to prevent or treat peripheral neuropathy and for this reason it is important to report any symptoms you have to your treating doctor as soon as you become aware of them. The sooner the symptoms are reported, the better they can be managed.

Management often requires decreasing or stopping the drug thought to be causing the nerve damage, the use of pain relieving medications (including amitriptyline, pregabalin, gabapentin and opioid drugs), massage, relaxation techniques and steps to reduce the risk of further injury (such as care with footwear).

If the dose of the drug causing the peripheral neuropathy is reduced or stopped when the symptoms first appear the effects may improve or resolve completely. However in some cases the damage is irreversible and it is a case of managing the symptoms.

## **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.

Maintenance of muscle strength is important in myeloma because strong muscles help to support and protect the damaged bones. It is better to have periods of activity followed by short rests than to avoid exercise.

Unless there is some critical bone damage that may be aggravated by activity, it is better to be active with the help of pain relieving medicines than to be inactive and 'put up' with the pain. Continuous rest makes the muscles waste, increases the rate at which calcium is removed from the bones and reduces cardiovascular fitness. You should discuss with your doctor what form of exercise is right for you.

### ***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 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 not as commonly seen with other myeloma treatments.

Mucositis usually starts about a week after treatment has finished and goes away once your blood count recovers, usually a couple of weeks later. During this time your mouth and throat can 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 e.g. thrush (*Candida albicans*) and 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.

Seek advice from your treating team about 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.

### ***Alopecia (hair loss)***

For most of us the thought of losing our hair is very frightening. Hair loss can be a 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 you can do to make yourself feel more comfortable.

Avoiding the use of heat or chemicals in your hair and only using a soft hair-brush and a mild baby shampoo can help reduce the itchiness and scalp tenderness that 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 haircut when they notice their hair is starting to fall out.

You need to avoid direct sunlight on your exposed head (wear a hat) because chemotherapy and radiotherapy make your skin more vulnerable to the damaging effects of the sun (i.e. sunburn and skin cancers).

Remember 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. With some treatment regimens, hair can also be lost from your eyebrows, eyelashes, arms, legs and other parts of the body.



## IMPORTANT!

**It is important 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 38°C or above (even if it returns to normal) and/or an episode of uncontrolled shivering (rigors). Many hospitals would expect you to go straight into the Emergency Department of your treating hospital without first contacting your doctor. Be sure you know the policy from your treating centre
- » Bleeding (or significant bruising), for example 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 cramps or severe constipation
- » Persistent coughing or shortness of breath
- » Burning or stinging when passing urine
- » The presence of a new rash, reddening of the skin or 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
- » Any numbness, change in sensation or pain in the hands, feet, arms or legs.

## Treatment to prolong a remission (maintenance therapy)

Once a response has been achieved, lower dose treatment may be used if it is considered by your doctor as beneficial in maintaining a good initial treatment result to help prolong the remission. Whether maintenance therapy is used or not always involves a balance between the benefits of continuing treatment and the potential side-effects of treatment.

## Treatment for relapsed and resistant (refractory) myeloma

Finding out your myeloma has come back (relapsed) or is resistant to standard treatment can be devastating. It is important to remember there are usually several options for treating the disease and getting it back under control.

The choice of treatment will depend on your previous treatment; your response to that treatment; the side-effects you have experienced with treatment; your general health; and your personal preferences. Selected patients may also undergo a second autologous stem cell transplant or, less often, proceed to an allogeneic transplant.

Promising new and experimental treatments are being developed for myeloma all the time. Your doctor will be able to discuss with you all of the treatment options including suitable clinical trials.

# SUPPORTIVE THERAPIES

## *Pain relief*

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 medications which your doctor may prescribe to help relieve bone pain. It is important you take these medications regularly and not wait until you have pain before you take them.

If you are experiencing any unpleasant side-effects, such as drowsiness, constipation or nausea, or if you feel that the medication is not strong enough, talk to your doctor or pharmacist. 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 inactivity increases the loss of calcium from the bones as well as causing muscle wasting. Weight-bearing activity (such as walking) is a very important activity you can do.

Some strong pain medication 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 to help your bowels move on a regular basis.

## *Radiotherapy*

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 the doctor feels are likely to fracture.

Sometimes a collection of myeloma cells (tumour) can develop in the bones of the spine (vertebrae) and grow into the spinal canal causing pressure on the spinal cord. This is known as spinal cord compression.

If there is pressure on the 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 contact your doctor or local hospital urgently, so steps can be taken to prevent permanent damage and possible paraplegia. Radiotherapy and corticosteroids are usually used to quickly reduce the pressure on the spinal cord. Occasionally decompression surgery is required.

## ***Bisphosphonates***

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.

Your doctor may recommend a bisphosphonate which can be taken daily in tablet form, like sodium clodronate or as an intravenous (IV) infusion usually once a month, like pamidronate or zoledronic acid.

Bisphosphonates usually have minimal side-effects, however they can cause the blood calcium to drop below normal levels which may cause muscle cramps or spasms. Occasionally people develop a fever or flu-like symptoms after an intravenous infusion of a bisphosphonate. This usually only lasts about 12 hours and won't necessarily recur with further infusions. Very rarely bisphosphonates can cause eye irritation. If this happens you must tell your doctor.

Rare but potentially serious side-effects of bisphosphonate therapy (particularly intravenous bisphosphonates) include kidney damage and damage to the jaw bone (osteonecrosis of the jaw).

To reduce the risk of kidney problems with bisphosphonates you should drink at least 2-3 litres of water each day and avoid medications that may increase the risk of kidney damage, particularly non-steroidal anti-inflammatory drugs.

## ***Osteonecrosis of the jaw (ONJ)***

ONJ generally presents either with continuous jaw pain or as a persistent wound in the mouth that may be painful and slow to heal. To reduce the possibility of osteonecrosis of the jaw, you should pay careful attention to your dental hygiene.

Your doctor may recommend you have a dental check-up before starting bisphosphonates and you should inform your dentist if you are on bisphosphonates.

Osteonecrosis of the jaw is also known to be associated with invasive dental work including removal of teeth and is more common in people with diabetes or with poor dental hygiene and those who are on bisphosphonates for prolonged periods of time (over 12-18 months). For this reason it is very important you inform your doctor and dentist before having any dental work done.

## ***Plasmapheresis***

Hyperviscosity syndrome is a rare complication of myeloma. It develops when the paraprotein reaches very high levels and causes the blood to become thicker. 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. 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. This is a continuous process.

If your veins are not suitable for this procedure, a special 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.

## ***Dialysis***

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 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. It is also important to minimise medication that may increase the risk of kidney damage, such as anti-inflammatory drugs.

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

## **Lifestyle**

Fatigue is one of the most common and most difficult symptoms associated with myeloma. You should let your doctor know how you are feeling as there are many things you can do to help such as avoiding over-tiring yourself, ensuring that you get enough sleep, eating a healthy balanced diet and doing regular exercise.

A healthy and nutritious diet is important in helping your body to cope with your disease and treatment. Talk to your doctor, nurse or dietitian if you have any questions about your diet or if you are considering making any radical changes to the way you eat. A nutritionist or dietitian can also advise you on planning a balanced and nutritious diet.

## **Fertility treatments**

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 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.

### **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 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 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 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:

**Embryo storage** - this involves collecting your eggs, usually after having drugs to stimulate your ovaries to produce a number of eggs, so 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, at a later date, the eggs contained in this tissue can be matured, fertilised and used to achieve a pregnancy.

**Donor eggs** - 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.

While assisted reproductive technologies have significantly progressed in recent years and assist many women to have children, 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. For aggressive disease, the best treatment option may require immediate chemotherapy and fertility options may be limited.



## 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, headaches and other aches and pains. Some women experience decreased sexual drive, anxiety and even depressive symptoms during this time. It is important 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.

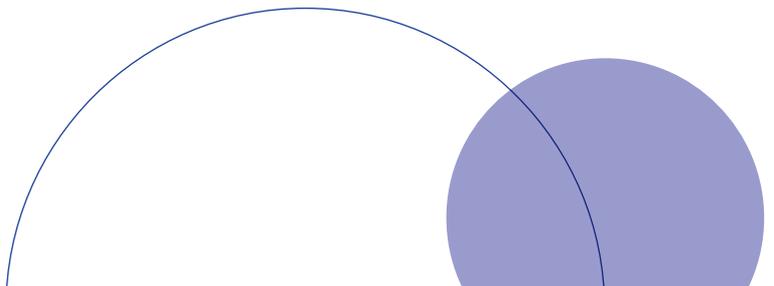
## Body image, sexuality and sexual activity

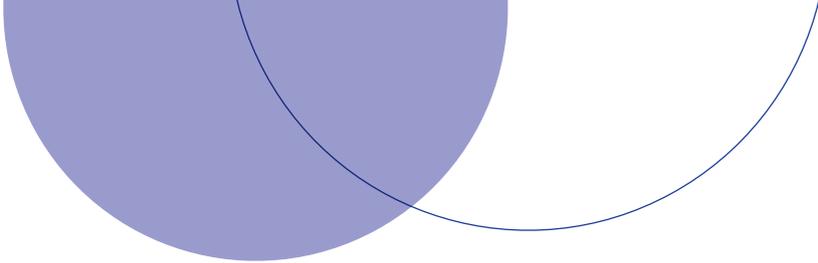
It is likely the diagnosis and treatment of myeloma will have some impact on how you feel about yourself as a man or a woman and as a 'sexual being'.

Hair loss, skin changes, and fatigue can all interfere with feeling attractive. *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](http://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 you or your partner do not become pregnant as some of the treatments might harm the developing baby. As such you need to ensure that you or your partner use a suitable form of contraception.

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.

It is important to realise no complementary or alternative treatments alone have proven to be effective against myeloma. It is also important to let your doctor, nurse and pharmacist know if you are contemplating or using any complementary or alternative treatments. Some of these therapies, such as green tea and high-dose vitamin C, may reduce the effectiveness or increase the toxicity of the chemotherapy or other treatments you may be having.

## Complementary therapies

Complementary therapies are not considered standard medical therapies. Some people however find they are helpful in coping with myeloma and its treatment. There are many different types of complementary therapies.

These include yoga, exercise, meditation, acupuncture, relaxation and vitamin supplements.

Should you choose to use these types of therapies they should be used to 'complement' or assist with recommended medical treatment for myeloma. They should not be used as an alternative to medical treatment for myeloma.

## MAKING TREATMENT DECISIONS

Many people feel overwhelmed when they are diagnosed with myeloma. In addition, 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 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 you feel you have enough information about your illness and all the treatment options available, so 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, to prompt you to ask other questions, to be an extra set of ears or simply to 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 wellbeing. You should feel you have enough information to do this and 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 myeloma in different ways, and there is no right or wrong 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 information can often help to take away the fear of the unknown. It is best for people 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 blood and bone marrow 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 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 take on fulfil roles they are not familiar with, for example cooking, cleaning, doing the banking and taking care of children. The social worker attached to your treating hospital can also provide some assistance.

There are a variety of programs designed to help ease the emotional and financial strain created by blood cancers and related disorders. 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 the Leukaemia Foundation are at the back of this booklet.

## USEFUL INTERNET ADDRESSES

There is an enormous amount of information about myeloma available on the internet. Many of these sites are based in the United States, United Kingdom or Europe and the quality of information provided on these sites varies.

For this reason, it is important to use reputable and up-to-date sites and to discuss information that you have gained from these sites with your doctor and other members of your health care team.

*Leukaemia Foundation of Australia*  
[www.leukaemia.org.au](http://www.leukaemia.org.au)

*International Myeloma Foundation (IMF)*  
[www.myeloma.org](http://www.myeloma.org)

*Myeloma UK*  
[www.myeloma.org.uk](http://www.myeloma.org.uk)

*The Multiple Myeloma Research Foundation (MMRF)*  
[www.themmr.org](http://www.themmr.org)

*Cancer Council of Australia*  
[www.cancer.org.au](http://www.cancer.org.au)

*Leukemia & Lymphoma Society (US)*  
[www.lls.org](http://www.lls.org)

*Look Good...Feel Better program*  
[www.lgfb.org.au](http://www.lgfb.org.au)

*Myeloma Australia*  
[www.myeloma.org.au](http://www.myeloma.org.au)

*National Cancer Institute (USA)*  
[www.cancer.gov/cancertopics](http://www.cancer.gov/cancertopics)

# GLOSSARY OF TERMS

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## **AL Amyloidosis**

A condition in which myeloma light chains are deposited in tissues and organs throughout the body.

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## **Alopecia**

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

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## **Anaemia**

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

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## **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.

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## **Antiemetic**

A drug which prevents or reduces feelings of sickness, nausea and vomiting.

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## **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.

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## **Bence Jones protein**

A myeloma protein present in the urine.

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## **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.

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## **Bisphosphonates**

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

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## **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.

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## **B-lymphocyte (B-cell)**

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

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## Bone marrow

The tissue found at the centre of many flat or big bones of the body. Active or red bone marrow contains stem cells from which all blood cells are made and in the adult this is found mainly in the bones making up the axial skeleton – hips, ribs, spine, skull and breastbone (sternum). The other bones contain inactive or (yellow) fatty marrow which as its name suggests, consists mostly of fat cells.

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## 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.

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## Cannula

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

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## Central venous catheter (CVC)

Also known as a central venous access device (CVAD). A line or 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.

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## Chemotherapy

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

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## 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.

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## 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.

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## Disease progression

Where the cancer is not responding to treatment.

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## 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.

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## Haematologist

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

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## Haemopoiesis/Haematopoiesis

The formation of blood cells.

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## High dose chemotherapy (HDT)

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.

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## Hypercalcaemia

Excess calcium in the blood.

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## 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.

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## 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.

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## Immune system

The body's defence system against infection and disease.

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## Immunoglobulins

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

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## 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.

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## 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.

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## Light Chain

One of the short protein chains that make up an immunoglobulin molecule. May be kappa or lambda type.

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## Localised disease

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

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## Lymphocytes

Specialised white blood cells involved in defending the body against disease and infection. There are three main types of lymphocytes: B- cells, T-cells, and natural killer cells.

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## Lymphoma

Cancer that arises in the lymphatic system.

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## Lytic lesion

A damaged area of bone that shows up as a dark spot on an x-ray.

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## 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.

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## 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).

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## Myeloma

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

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## 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.

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## Neutrophils

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

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## Paraprotein

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

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## Peripherally inserted central catheter (PICC)

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

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## 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.

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## Plasmacytoma

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

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## 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.

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## Plateau

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

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## Prognosis

An estimate of the likely course of a disease.

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### **Radiotherapy (radiation therapy)**

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

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### **Relapse**

The return of the original disease.

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### **Remission**

When there is no evidence of disease detectable in the body.

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### **Resistant or Refractory Disease**

This means that the disease is not responding to treatment.

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### **Standard therapy**

The most effective and safest therapy currently being used.

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### **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.

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### **Stage**

The extent or spread of cancer in the body.

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### **Staging**

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

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### **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.

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### **T-lymphocyte (T-cell)**

A type of white cell involved in controlling immune reactions.

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### **Tumour**

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

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### **Ultrasound**

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

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### **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.

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### **X-ray**

A form of radiation used in diagnosis and treatment.



# MAKING A DONATION

The Leukaemia Foundation is the only national charity dedicated to helping those with leukaemia, lymphoma, myeloma and related blood disorders survive and then live a better quality of life. It exists only because of the generous and ongoing support of the Australian community.

## How can I give?



**ONLINE** [leukaemia.org.au](http://leukaemia.org.au)



**PHONE** 1800 620 420



**POST** (complete this form or enclose cheque/money order and return)

The Leukaemia Foundation, Reply Paid 9954 BRISBANE QLD 4001

Name	
Address	
Postcode	
Phone	Mobile
Email	

I enclose my gift of (please tick box)

\$30  \$50  \$75  \$100  \$250 Other \$

I wish to make a regular monthly donation of \$

Commencing on / /

\*

\*You can cancel at any time by calling 1800 620 420.

My cheque/money order made payable to the Leukaemia Foundation is enclosed.

I wish to pay with my credit card and my details are included below:

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Card Number

Expiry Date /

CVV

Cardholder's Name

Signature

Your privacy is important to us. That is why we treat your personal information with confidence. To learn more about how and why we collect and use any personal or sensitive information about you, please view our Notification Statement at [www.leukaemia.org.au/privacy](http://www.leukaemia.org.au/privacy)



# PLEASE SEND ME A COPY OF THE FOLLOWING BOOKLETS:

- Leukaemia, Lymphoma, Myeloma, MDS, MPN and related blood disorders
- Acute Lymphoblastic Leukaemia in Adults (ALL)
- Acute Lymphoblastic Leukaemia in Children (ALL)
- Acute Myeloid Leukaemia (AML)
- Amyloidosis
- Chronic Lymphocytic Leukaemia (CLL)
- Chronic Myeloid Leukaemia (CML)
- Hodgkin Lymphoma
- Non-Hodgkin Lymphoma (NHL)
- Myelodysplastic Syndrome (MDS)
- Myeloma
- Myeloproliferative Neoplasms (MPN)
- Eating Well
- Living with Leukaemia, Lymphoma, Myeloma, MDS, MPN and related blood disorders
- Allogeneic Stem Cell Transplants (also called Bone Marrow Transplants)

- Autologous Stem Cell Transplants
- Young Adults with a Blood Cancer
- My Haematology Diary

## Books for children:

- Tom has Lymphoma
- Joe has Leukaemia
- Ben's Stem Cell Transplant
- Jess' Stem Cell Donation

## Or information about:

- The Leukaemia Foundation's Support Services
- Workplace Giving
- Monthly giving program
- National fundraising campaigns
- Volunteering
- Receiving our newsletters
- Leaving a gift in my will

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 **POST TO** The Leukaemia Foundation, Reply Paid 9954 BRISBANE QLD 4001

 **PHONE** 1800 620 420     **EMAIL** [info@leukaemia.org.au](mailto:info@leukaemia.org.au)

 **FURTHER INFORMATION ONLINE** [leukaemia.org.au](http://leukaemia.org.au)



This information booklet is produced by the Leukaemia Foundation and is one in a series on leukaemia, lymphoma, myeloma, MDS, MPN and related blood disorders.

Copies of this booklet can be obtained from the Leukaemia Foundation by contacting us.

The Leukaemia Foundation is a not-for-profit organisation that depends on donations and support from the community.

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## CONTACT US

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Leukaemia  
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