Amyloidosis

A guide for people with amyloidosis and their support people

Leukaemia Foundation®

This booklet has been written to help you and your support people understand more about amyloidosis.

We know you may be feeling anxious or overwhelmed if you or someone you care for has recently been diagnosed with amyloidosis. Maybe you are discussing different treatment options with your treatment team or maybe you have started treatment. Whatever point you are at, this booklet will answer some of your questions and it may raise others, which you can jot down and discuss with your haematologist or nurse.

If you don't feel like reading this booklet from cover to cover, take a look at the list of contents and choose which parts to read now. You can come back to read other parts later on. You may need more information, so towards the back of the booklet there is a list of useful resources. Your doctor or nurse might also give you some further reading. You can always call our Blood Cancer Support Coordinators to find out how we can help you.

You will meet many different types of healthcare professionals who work as a team to provide you with the best treatment available. The people you'll most often see will be haematologists and haematology nurses, and you'll need a regular GP, but you'll also meet pathology providers and allied health professionals, like dietitians. In this booklet, when we refer to 'your treatment team' we usually mean your haematologist and haematology nurses.

You will come across quite a few medical terms in this booklet. They are words that your treatment team will probably use and that you may not have heard before. They will be happy to explain any terms you don't understand, so never be afraid to ask. Many of these words are defined in the text or in the Glossary at the end of this booklet.

Although we provide some information about treatments, this booklet does not recommend any particular form of treatment and you must discuss your circumstances and best treatment options with your haematologist.

We hope you find this information useful. We'd love to hear any feedback so we can make sure we best meet your information needs.

The Leukaemia Foundation acknowledges the traditional owners of country throughout Australia and recognises their continuing connection to land, sea and community. We pay our respects to their Elders past, present and emerging.

Contents

Scan to learn more about how the Leukaemia Foundation can provide you with additional support and the latest information about your blood cancer.

Amyloidosis in brief

About amyloidosis

Amyloidosis is the name for a group of disorders where people have proteins that fold in an abnormal way to form fibrils. These protein fibrils are deposited as amyloid in the body's tissues and organs. They make it hard for the organs to function normally.

There are many different types of amyloidosis caused by about 30 different amyloid-forming proteins. Most types are systemic, affecting more than one organ or tissue.

Symptoms of amyloidosis vary depending on which organs and tissues are affected. Each type of amyloidosis has different signs and symptoms. People with the same type of amyloidosis may not share symptoms, so could need different treatments.

Amyloidosis is usually diagnosed by biopsy of either fatty tissue or the affected organ. Some types of amyloidosis can be diagnosed by a special scan.

We don't know what causes amyloidosis, although we do know some risk factors for certain types. There is no way to prevent it. Some types of amyloidosis are hereditary and can be passed on.

Who gets amyloidosis

Amyloidosis is quite rare. While research has shown that approximately 12 cases per million are diagnosed each year, it may be much more common than that. It's hard and slow to diagnose, which makes it difficult to work out how many Australians have it. We do know that ATTRwt amyloidosis is the most common type, and that amyloidosis usually diagnosed in people over age 50. In general, more men than women have amyloidosis.

What's the prognosis?

A prognosis is an estimate your haematologist will make of the likely course and outcome of your disease. Your haematologist will take into account many factors when considering your prognosis. Some of these are the type of amyloidosis you have, which organs are involved, your age, your overall health and how you respond to treatment. Outcomes for patients with amyloidosis have been continually improving over recent years due to earlier diagnosis and better treatments.

All about amyloidosis

What is amyloidosis?

Amyloidosis is a rare group of disorders where a protein folds abnormally to form amyloid which builds up in the tissues and organs in the body. It can be systemic (throughout the body) or localised (in one tissue or organ). Amyloid is formed when certain proteins fold in an abnormal way to form fibrils. Amyloid is said to be deposited in tissues and organs, so you may hear the term amyloid deposits.

Amyloid deposits can't be broken down easily. The body finds it difficult to remove them, which is why they build up in tissues and organs. When this happens, the organs can't function normally. Without treatment, amyloidosis can eventually lead to organ failure.

There are more than 30 types of proteins that cause all the types of amyloidosis. Each type has different signs and symptoms. The types develop at varied paces and can differ person to person. Even people with the same type of amyloidosis may not share symptoms, so could need different treatment. Amyloidosis is not contagious. You can't 'catch' it by being in contact with someone who has it.

Systemic amyloidosis

In systemic amyloidosis, the amyloid forming protein is made in one part of the body (like the bone marrow or liver) but the amyloid deposits are found in other tissues (like muscles) or organs (like your heart).

Local amyloidosis

In localised amyloidosis the amyloid forming protein is both made and forms amyloid deposits in one area, like the airways, bladder or bowel. Is amyloidosis cancer?

Amyloidosis is not a cancer. It is linked to some blood cancers, like multiple myeloma and some types of lymphoma.

Types of amyloidosis

There are more than 30 types of amyloidosis. Many types cause few problems. Each type of amyloidosis is different, so there are many treatments.

Amyloidosis is classified into types depending on what protein is causing the amyloid deposits.

Each type of amyloidosis is given a name where the first letter 'A' stands for amyloid and the following initial stands for the amyloid protein. For example, in AL amyloidosis the 'A' stands for amyloid and the 'L' stands for the type of protein, which is called a light chain.

The more common types of amyloidosis are:

- AL amyloidosis: a light chain amyloid caused by a bone marrow disorder.
- AA amyloidosis: where the serum amyloid A (SAA) protein increases a lot due to a long-term inflammatory disorder like rheumatoid arthritis.
- Hereditary amyloidosis: where a gene mutation is inherited, leading to lifelong production of an abnormal protein. The most common types of hereditary amyloidosis are ATTRv (transthyretin gene mutation) and AFib (fibrinogen alpha chain gene mutation).
- Wild-type transthyretin amyloidosis (ATTRwt) (previously known as *senile amyloidosis* or *age-related amyloidosis*): where amyloid appears in the heart and sometimes in the wrists, causing damage to the carpal tunnel. This is not an inherited disease.

Organ involvement

Amyloidosis is usually a *systemic* disease. This means that any tissues and organs of the body may be affected by the amyloid protein. There is no set pattern as to how. Kidneys and the heart are often involved. For many people with amyloidosis, more than one organ will be affected.

Amyloid	*Nature of amyloid*	*Other names*	*Major organs*
AL	Immunoglobulin light chain	Previously known as primary systemic amyloidosis Myeloma-associated amyloidosis	Kidney Heart Nervous system Liver Gastrointestinal Soft tissues
AA	Amyloid A protein	Previously known as secondary amyloidosis	Kidney Liver

ATTR	Mutated transthyretin	Familial amyloidotic polyneuropathy	Nervous system Heart
ATTR	'Wild-type' transthyretin	Senile amyloidosis	Heart
AFib	Mutated fibrinogen alpha chain		Kidney

Causes of amyloidosis

In most cases, there is no specific cause of amyloidosis. It can be acquired (not a condition you are born with but something that develops over time) or hereditary (due to a faulty gene passed down through the family).

Known risk factors

- **Ageing** because most types, particularly AL and ATTRwt amyloidosis, are more common in older people.
- **Family history** because some types of amyloidosis are hereditary (passed on via genes from parents to children).
- Having an **inflammatory disease** like rheumatoid arthritis.
- Having the blood cancer **myeloma**.

Most proteins that form amyloid are made in the bone marrow or the liver, circulate in the blood and then deposit as amyloid in the tissues. Amyloid forms:

- because too many amyloid-forming proteins are being made (e.g. AA amyloidosis), or
- because amyloid-forming proteins are abnormal (e.g. AL or hereditary amyloidosis), or
- because normal amounts of a normal blood protein very slowly forms amyloid over decades (e.g. ATTRwt amyloidosis).

Symptoms of amyloidosis

Which symptoms you have depends on which tissues and organs are affected and how much. Symptoms vary greatly from person to person and between the different types of amyloidosis.

The most common symptoms are:

- fatigue (extreme tiredness that sleep and rest don't help with)
- weight loss for no reason
- shortness of breath
- swollen ankles and legs (called *oedema*)

How is amyloidosis diagnosed?

Amyloidosis can be difficult to diagnose. There is no specific blood test and test results for one person can be very different to another person with amyloidosis. Your treatment team will decide which tests you need based on your symptoms. The tests will confirm if you have amyloidosis and will help work out which type you have.

Diagnostic testing may include:

- medical history and physical examination
- blood and urine tests
- tissue biopsy
- bone marrow biopsy
- scans, including DPD, PYP, MRI, CT scans
- X-rays
- ultrasound
- nerve conduction tests
- heart tests, including electrocardiogram and/or echocardiogram.

It's unlikely you will have all of these tests.

Medical history and physical exam

First, your treatment team will take a full medical history. They'll ask you to talk about past and present illnesses, health problems, infections and bleeding. They'll also need details of any old and new medicines you're taking including prescribed and any over the counter medications you take regularly.

Your doctor will also do a physical examination, to check your general health and your whole body for any signs of amyloidosis.

Tissue biopsy

The main test for anyone who may have amyloidosis is a biopsy. This involves taking a small piece of tissue from your body, most commonly from one of the organs or tissues affected by the amyloidosis. For people who may have systemic amyloidosis (amyloid deposits around the body), a biopsy may be taken of fat tissue from the belly (abdomen).

This tissue sample is sent to the laboratory, where a pathologist puts a dye called *Congo red* on the sample. The biopsy will appear red under normal light and green under special polarised light if there are amyloid deposits in the tissue.

Some types of amyloidosis, such as ATTRwt, can be diagnosed with a special scan (DPD, PYP) and don't need a tissue biopsy.

Blood and urine tests

Full blood count

You'll be asked to have a simple blood test called a full blood count (FBC). This test measures the number of red cells, white cells and platelets in circulation. Your treatment team will give you a referral and tell you where to go to have it done. They will also tell you if you need to fast (not eat or drink) for a certain amount of time before you have the blood test.

A nurse will take a sample of blood from a vein in your arm. It's sent to the lab, where a pathologist (a blood specialist) will look at the blood cells under a microscope.

Your blood might be tested for other things to rule out other conditions, so your nurse might take more than one tube of blood.

Blood chemistry tests

Blood chemistry tests measure the levels of different chemicals in your body. These blood tests are often taken at the same time as your FBC. Some substances that may be tested will be:

Substance tested	*What it indicates*
Creatinine	Kidney function
C reactive protein	Inflammation in the body
Brain natriuretic peptide	Heart function
Troponin	Heart function
Liver function tests	Liver function

Serum (blood) free light chains, electrophoresis and immunofixation

Immunoglobulins are made up of two identical long pieces of protein (called *heavy chains*) and two identical short pieces of protein (called *light chains*). It is these light chains that form AL amyloidosis. The level of light chains (kappa or lambda) in your blood will be measured for diagnosis and treatment response for AL amyloidosis. The free light chains test measures the free light chains and calculates the kappa/lambda ratio.

The whole immunoglobulin protein (also called a paraprotein) is measured by tests called serum protein electrophoresis and immunofixation. This also helps diagnose and monitor AL amyloidosis and can help rule out the blood cancer myeloma.

Urine tests: electrophoresis and immunofixation

Urine tests measure the type and the amount of protein in your urine. They help work out if amyloid proteins are affecting your kidneys. They also help rule out the blood cancer myeloma.

24-hour urine collection test

You may be asked to collect your urine over 24 hours at home. Your treatment team will give you the bottle with instructions and tell you where to take the bottle when you're done. Your urine be tested for the Bence Jones protein, which is a type of paraprotein seen in myeloma and may be associated with AL amyloidosis.

Genetic testing

Your treatment team may request genetic tests to check on gene abnormalities in hereditary amyloidosis. Sometimes, healthy people who may be at risk of inheriting a potential gene mutation that may cause amyloidosis can choose to have these tests.

Bone marrow biopsy

If your treatment team thinks you may have AL amyloidosis, the next step is a bone marrow test.

This test is more complex than a blood test, but it won't involve a hospital stay. It will be done in your haematologist's rooms or in a day procedure clinic or outpatient ward in a hospital. It's a good idea to bring a support person with you to keep you company while you wait and to help you home, as you may be able to drive.

What does a bone marrow biopsy involve?

A bone marrow biopsy involves using a needle to enter the bone marrow most commonly in the hip bone. This is an area where the bone is usually quite close to the skin and an area that can be easily accessed by the bone marrow needle. A small amount of liquid bone marrow is usually taken and placed onto slides that are examined in the laboratory. The liquid bone marrow is also sent for additional specialised tests. Usually a small piece of the bone marrow is also taken and examined in the laboratory.

Is a bone marrow biopsy painful?

Having a bone marrow biopsy is sometimes painful. With the use of local anaesthetic before the bone marrow needle is inserted this pain is usually well tolerated. Often you will be given a form of pain preventer that you breathe in, or a small dose of sedative is given (under appropriate hospital conditions) to assist in the management of the discomfort.

You should try to rest for the day. You may not have any pain but if you do, take a paracetamol tablet. You can take off the dressing and shower 24 hours after the test or as advised by your treatment team.

Imaging tests

Bone scans (DPD, PYP scans)

While originally used as bone scans, we now know that DPD and PYP scans are very useful to diagnose heart involvement in ATTR amyloidosis. You are given a special dye which is injected into a vein in your hand or arm before the scan. The scanner can then detect if the dye is showing amyloid in the heart.

CT scans

CT scans show three-dimensional (3D) images of parts of your body. While the scan is being done you lie flat and still on a cushioned table that moves slowly through the CT machine. Sometimes you may have a special dye (called a *contrast*), which you swallow or may be injected into a vein in your hand or arm before the scan. The CT scanner shows the contrast as it moves through your body.

MRI scans

Magnetic resonance imaging (MRI) uses a very strong magnet to make three dimensional (3D) images.

What happens next?

After diagnosis

When all your test results have been reviewed, you'll meet with your treatment team to discuss your amyloidosis type and treatment goals and options. It's natural to feel scared, confused or sad. You'll hear a lot of information and it can be overwhelming.

Don't be afraid to ask your treatment team to repeat things and for some written information. It's helpful to bring someone along to the appointment as second pair of ears and to take notes.

Treatment

Who makes up your treatment team?

Because amyloidosis can affect many organs, a few types of doctors work together to create your treatment plan. This is called a *multidisciplinary team*. They will be supported by specialist nurses too. Your treatment team includes your GP, pharmacist and any other allied health professionals you may see, like dietitians.

Your team may include:

- Haematologist: specialist in blood disorders
- Cardiologist: specialist in heart conditions
- Nephrologist: specialist in kidneys
- Respiratory physician: specialist in lungs
- Neurologist: specialist in brain and nervous system.

Often the lead will be your haematologist.

Goals of treatment

Your treatment team will recommend treatment based on your type of amyloidosis.

The goals of treatment in amyloidosis are:

- to stop or slow down production of the amyloid protein
- to support and preserve organs and tissues
- to improve your quality of life.

Because there is so much difference between the types of amyloidosis and how they affect the body, treatments vary. You can read more about the types of treatments (and their side effects) under each type of amyloidosis later in this booklet.

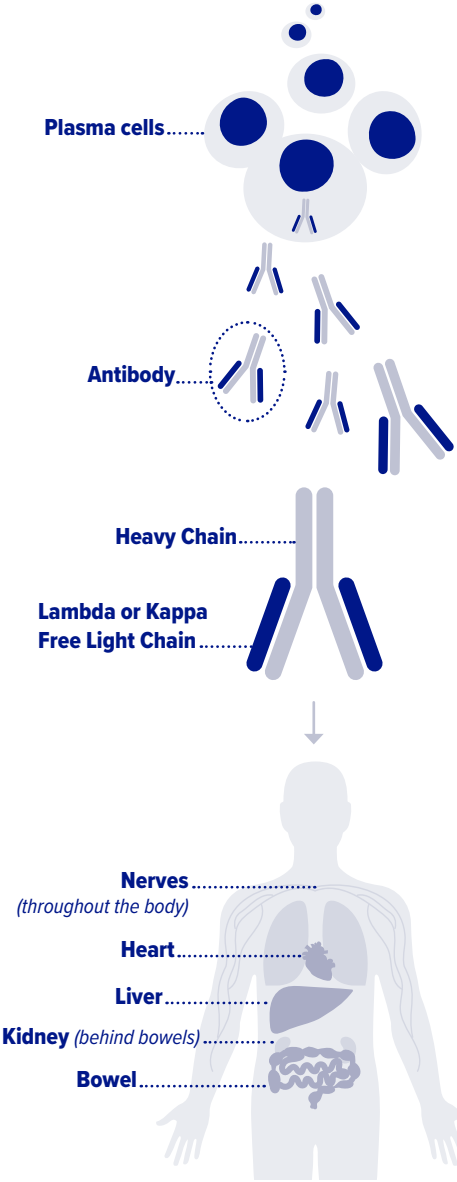
Treatment consent

Your treatment team will explain the treatments, their benefits and possible side effects. They will ask you to sign a consent form to agree to the treatment after you have thought about the options.

What is AL amyloidosis?

AL amyloidosis is one of the more common forms of amyloidosis. It used to be called primary amyloidosis. Most people diagnosed are over 50, but younger people can get it. It is not inherited or contagious.

AL amyloidosis is caused by an abnormal protein (the 'light chain' of the immunoglobulin or antibody protein) made by abnormal plasma cells found in the bone marrow. Plasma cells are a special type of white blood cells and are part of the body's immune system. They make antibodies, also called *immunoglobulins*, which are proteins that help protect us against and fight infection. Normally once these proteins have served their purpose they are broken down and recycled in the body.



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 (Kappa and Lambda type), which enter the blood stream and circulate around the body.

The free light chains misfold into amyloid fibrils, which deposit and build up in the tissues and organs in the body.

In AL amyloidosis a clone or single population of plasma cells grows and produces too many immunoglobulin light chains. These light chains circulate in the bloodstream and are deposited as amyloid fibrils in the body's tissues and organs. The amyloid fibrils can't be broken down easily. They stop the organs functioning normally. AL amyloidosis can lead to organ failure if it's not treated.

The way the amyloid protein is laid down organs and tissues varies. This means that there are many different symptoms. Often more than one organ is affected.

What are free light chains?

In AL amyloidosis the amyloid protein comes from the light chain of an antibody (also called an *immunoglobulin*). An antibody is made of two 'heavy' chains and two 'light' chains. Antibodies are made by plasma cells in the bone marrow.

Normally the body makes lots of different antibodies, which have different heavy chains and lots of different light chains. The plasma cells make more light chains than are needed to produce an antibody. Those excess light chains circulate around in the blood as *free light chains*.

There are two main types of light chains: kappa and lambda. Everyone has small amounts of normal kappa and lambda free light chains in their blood. For people with AL amyloidosis, abnormal plasma cells multiply and build up in the bone marrow. They make large amounts of a single type of free light chain, which forms amyloid deposits.

Which organs can be affected?

Any organ apart from the brain can be affected by AL amyloidosis. It most commonly affects the kidneys, heart, nerves and/or liver.

Kidney

The kidney is the most common organ affected by AL amyloidosis. Your kidneys are like sieves that filter your blood. When your kidneys are working well, waste fluid can filter from your blood. This waste fluid becomes your urine. As the waste fluid passes through the sieve/kidneys the sieve traps the normal products and keeps them in your bloodstream where they belong. When the sieve/kidneys are affected by the amyloid protein, the holes in the sieve are damaged and get bigger. Because of this, the normal 'good' blood proteins (including albumin) leak through the holes.

A urine test will show if you have too much normal protein (albumin) in your urine. This is called proteinuria. If you are losing protein:

- fluid leaks out of the blood vessels into tissues of the body. This fluid may build up to cause swelling (*oedema*), often in your feet and ankles
- over a long period of time, kidney function gets worse
- blood cholesterol can rise to very high levels.

Heart

The second most common organ involved in AL amyloidosis is the heart. The amyloid deposits make the heart muscle stiff so it can't pump blood properly. If your heart can't pump properly, you may have:

- extreme tiredness
- shortness of breath
- swollen ankles and legs
- chest pain
- light-headedness when you stand up due to low blood pressure.

This is different from normal heart failure where the heart muscle is weak, so many drugs used for heart failure may not work for amyloidosis patients. You may be given fluid tablets (called *diuretics*) to help with symptoms.

Cardiac amyloidosis can also affect the way electrical signals move through the heart (conduction system). This can lead to irregular heartbeat (*arrhythmia*) and a racing heartbeat (*palpitations*).

Nervous system

Your nervous system includes the central nervous system and the peripheral nervous system. The brain and the spinal cord make up the central nervous system. The central nervous system can send and receive signals to and from the rest of the body via the nerves (known as the *peripheral nervous system*) to ensure the body works normally. AL amyloidosis affects the peripheral nervous system.

Peripheral neuropathy

Amyloid deposits can affect the nerves in your arms, hands, legs and feet. These peripheral nerves act like electrical wiring, carrying signals caused by touch, pain, heat and cold, from the feet and hands to the brain. The nerves also send signals to our muscles telling them when to contract and relax.

When the amyloid protein affects the nerves it can cause a short circuit in this wiring. This leads to numbness, weakness, tingling and pain in your fingers, hands, toes and feet. It's called *peripheral neuropathy*.

Some people will have carpal tunnel syndrome, where the nerves are squashed by amyloid deposits in the wrist. It causes wrist and hand pain and tingling.

Autonomic neuropathy

Nerves that control heart rate, blood pressure, and movement of the gut that allows us to digest food can also be affected. They are *autonomic* nerves.

***Autonomic neuropathy* can cause:**

- nausea
- gut bloating or pain
- diarrhoea
- weight loss
- feeling full after eating only small amounts
- impotence
- dizziness on standing.

Digestive system and gastrointestinal tract (gut)

The main role of the gut is to break down the food you eat into small components so that you can absorb the nutrients into your body. Amyloid deposits can stop the regular movement of the gut, which helps break down the food particles, and can make it very difficult for the nutrients to pass from your gut into your body. This can cause diarrhoea or constipation, weight loss, and/or bleeding (bright red blood or black bowel movements).

Amyloid proteins can also deposit in your tongue. This makes it swell and become rubbery (called *macroglossia*). Macroglossia can cause problems with speech, eating, and even breathing.

Liver

Amyloid deposits in your liver can make it swell. Swollen liver is called *hepatomegaly*. Changes to your liver will be picked up in routine blood tests, which measure liver function. Sometimes liver involvement causes the skin to look yellow (this is called *jaundice*). You might also fell a fullness or some pain in your upper right belly.

Symptoms of AL amyloidosis

You may feel week and tired and lose weight and your appetite. Other symptoms depend on which organs are affected. You may have:

- swollen ankles (kidney or heart involvement)
- shortness of breath (heart involvement)
- diarrhoea (gut or nerve involvement)
- tingling or numbness in your fingers and toes (nerve involvement).

What tests will I need?

After you've been diagnosed with AL amyloidosis, you'll need more tests to work out:

- which organs are affected and how much, and
- any other medical problems that may affect treatment.

You'll also have these tests during and after treatment to check how treatment's working.

Ongoing Blood tests

Blood tests check:

- free light chains and serum protein electrophoresis: these measure the abnormal light chain protein that is causing the AL amyloidosis. This helps track your treatment progress too
- kidney, liver and heart function: important heart tests include serum troponin and NT-pro BNP (brain natriuretic peptide)
- normal protein (albumin) in the blood
- blood's clotting function
- the number of red and white blood cells and platelets.

Urine tests

Urine tests give information about kidney function. They check for free light chains in the urine (Bence Jones protein) and whether normal protein (albumin) is being lost into the urine.

Bone marrow biopsy

Bone marrow biopsy checks if you have abnormal plasma cells in your bone marrow. Samples taken may have further tests to check for a type of blood cancer called *myeloma*.

Other tests

Echocardiogram: assesses the structure and the function of the heart muscle and heart valves.

Electrocardiogram: measures electrical activity in the heart.

CT or MRI scan: which look at images of organs and tissues of the body.

Treatment for AL amyloidosis

Although AL amyloidosis can't be cured, treatment can help you live a long, active life.

To work out the best treatment for you, your treatment team will consider your age, general health and the extent to which your organs have been affected by the disease. They will also think about potential complications of therapy.

Treatment goals

The goal of treatment is to quickly lower the free light chains that cause the amyloid deposits. This is done by targeting plasma cells in the bone marrow, which make the free light chains.

Once production of the amyloid protein is slowed or stopped, the amyloid fibrils already deposited in organs may slowly move out of the affected organs. The function of the affected organs may then slowly improve.

Treatments and side effects

There are a few treatments for AL amyloidosis. You may have one or more of these:

- chemotherapy
- steroids
- targeted therapies
- autologous stem cell transplant
- supportive care

There may also be clinical trials which use newer therapies.

Standard drug therapies

Your treatment team might use the term ‘standard of care’ or ‘standard therapy’. This is a treatment that is commonly used by medical experts for a certain type of disease.

Combination therapy

Standard drug therapies for AL amyloidosis include targeted therapies (a type of immunotherapy), chemotherapy and steroids. Some therapies may be given via drip (IV) or subcutaneous injection (SC) but most are tablets you can take at home. You may have a combination of these types of therapies.

When you start treatment you’ll most likely have a combination of these types of therapies.

Your haematologist will recommend your drug therapies depending on:

- your overall health
- your age
- whether it is your first treatment, or your amyloidosis has come back after remission (relapsed), and
- whether you are preparing for a stem cell transplant. You can read more about stem cell transplants later in this booklet.

Targeted therapies

Targeted therapies target specific substances on the surface of abnormal cells but don't harm normal cells. They can act by switching genes on or off, or by sending substances straight to the abnormal cells to kill them or stop them growing. Like chemo drugs, targeted therapies can cause side effects. Not everyone will have every side effect.

A type of targeted therapy called a *proteasome inhibitor* blocks the waste disposal unit in light-chain producing plasma cells, which affects their growth and survival. Newer types of proteasome inhibitors are also sometimes used.

Immunotherapies

Immunotherapies are sometimes called *biologic therapies*. They use a part of your immune system to fight cancer. In this case, antibodies are created in a lab. In your body, antibodies fight infection by targeting parts of cancer cells to change how they grow. Immunotherapies are given via drip (IV) or in tablet form.

Monoclonal antibodies

Monoclonal antibodies are a type of *immunotherapy*. They work by attaching to specific cancer cells to tell the immune system to destroy those cells. They also make cancer cells grow more slowly.

Chemotherapy

Chemotherapy medications (also called *chemo*), sometimes also called *cytotoxic* (which means 'cell killing') medications, make cancer cells stop growing. They either kill the cells or stop them from dividing, replicating and reproducing. They also damage normal cells, but these cells can repair and recover.

Chemo can be given as tablets, injections, or infusions via a drip (IV). It's given in cycles of treatment days. This means that you will have treatment for a certain number of days, followed by a set number of rest days. Chemo cycle times depend on the drug.

Chemotherapy for AL amyloidosis

Although amyloidosis is not a cancer, chemotherapy drugs have been proven to work in the treatment of AL amyloidosis by destroying the light chain-producing plasma cells in the bone marrow.

The most common chemo drugs used to treat AL amyloidosis are both tablets and you will most likely be given the steroid dexamethasone to take too.

Treatment side effects

Everyone gets different side effects with treatment. You may have no side effects, or one or more of them, and they may change over time.

You can find more information on chemo side effects and how to manage them on our website.

Changes in blood counts

Some treatments affect your bone marrow's ability to produce enough blood cells.

Low circulating red blood cells cause anaemia. You may feel tired, short of breath, and look pale. Take it easy and contact your treatment team. You might need a blood transfusion to help your levels recover.

If your platelets are low, you can bruise and bleed more easily. If you notice these signs please let your treating team know. You may need a platelet transfusion.

When your white blood cell count (neutrophils) is too low, this puts you at a higher risk of developing an infection. You may be given G-CSF to help raise your neutrophils.

Feeling sick - nausea and vomiting

Nausea (feeling sick to your stomach) and vomiting are common side effects, but you will be given medicine to prevent or manage them. If you do feel nauseous, even with medicine to help, do not hesitate to contact your treating team to ensure it is managed and you can continue eating and drinking.

Keep an eye on your weight if you are eating less than usual. If you find it difficult to eat, talk to your treatment team. They can arrange for you to see a dietitian for some advice.

Sense of taste and smell

Changes to your sense of taste and smell can make you not enjoy food and drinks that you used to love. You might have a metallic taste in your mouth. These changes will pass when your treatment ends.

Mouth problems - mucositis

Your mouth or throat might become sore, or you might get ulcers. This is called *mucositis*. It is very important to keep your mouth clean by using an alcohol-free mouthwash, salty water or sodium bicarbonate in water.

Bowel changes

Chemo can damage the lining of your bowel. You might have cramping, wind, bloating, diarrhoea or constipation. You will be given medication to help. Tell your treating team if you are experiencing diarrhoea, are constipated or if it's painful or hard to pass faeces. If you have haemorrhoids, don't strain. Sometimes making a few changes to what you eat can help too. Your treatment team can give you tips on food choices. *You can read more about diet and nutrition later in this booklet and on our website.*

Feeling tired and weak (fatigue), even after resting

Most people feel tired following chemo. It can be frustrating if you're used to keeping busy. *You can find more information on how to manage fatigue later in this booklet and on our website.*

Bone, joint and muscle aches and pains (arthralgia/ myalgia)

You may find you feel achy or sore, particularly in the morning. The pain may be because of your amyloidosis, or it might be due to treatment. Talk to your treatment team if you have pain.

Chemo brain

You may find it difficult to concentrate ('foggy brain') or have trouble remembering things. It can take up to a year after treatment finishes to recover. There are no medicines to help with chemo brain and in some cases changes can be permanent. Set up some ways to remember things, like writing them down. Talk to your support network too, so they know what's going on.

Hair loss (alopecia) and thinning

The thought of losing our hair is scary. Hair thinning or loss is a very common side effect of chemo. You might lose your head hair, your eyebrows and your eyelashes, but it's only temporary. Hair starts to fall out a few weeks after you start treatment and tends to grow back three to six months after it finishes.

You might find your scalp is itchy as you lose your hair, but this will pass. There are some great wraps, turbans and beanies available. Your nurse can give you information on where to find them, and about where to get wigs.

Tingling or numbness in fingers and toes (peripheral neuropathy)

Some drugs can affect your nerves, usually in your hands or feet. This is called *peripheral neuropathy*. Symptoms can start any time during treatment. Tell your treatment team if you feel tingling, numbness, burning or pins and needles in your fingers or toes. It might be hard to do up buttons or to grip things.

Skin issues

There are a few different skin changes you might have as a result of treatment. You may have a rash, or you may have soreness, redness or skin changes where you are having regular injections.

Sun sensitivity

Some chemo drugs make you more prone to sunburn. You can go outside, but avoid direct sunlight.

Managing chemo side effects

Potential side effects	What might help
Low red blood cells (anaemia)	• you may be given a blood transfusion or recommended supplements
Low platelets	• avoid sharp objects in your mouth like chop bones or potato chips • be careful not to cut or injure yourself • use a soft toothbrush • use an electric razor • wear gloves and closed shoes in the garden
Low white blood cells (neutrophils) – risk of infection	• talk to your treatment team about vaccinations • avoid crowds • keep away from people who are sick and might be contagious (colds, flu, chicken pox) • eat food that has been properly prepared and freshly cooked • don't clean up pet faeces • wear gloves in the garden • don't swim in public pools, lakes or rivers

Feeling sick – nausea and vomiting	• eat smaller meals more often during the day • try cool or cold food, like jelly • let someone else cook for you • drink ginger ale or soda water • avoid strong smells • you'll be given medicine to help
Change to taste	• add a little more sugar to sweet foods • add a bit more salt to savoury foods • if you have a metallic taste, try rinsing your mouth
Mouth problems – mucositis	• use a soft toothbrush and mild toothpaste • brush every time after you eat • use salty water, sodium bicarbonate in water, or alcohol-free mouthwash • continue to floss but stop if your gums bleed
Bowel changes	• drink plenty of fluids • get some dietary advice from your treatment team • if you're constipated, don't strain • if you have haemorrhoids, do not push on them • tell your treatment team, you'll be given medicine to help
Skin problems (rashes, sensitivity)	• your treatment team will recommend body washes and cream that are safe to use • avoid soaps and perfumes on the affected area • take cooler showers or baths
Fatigue	• see later in this booklet for more info • rest or nap when needed • take regular gentle exercise
Bone, joint or muscle aches and pains	• talk to your treatment team • take some gentle exercise, like walking, may help • you may be given medicine to help

Chemo brain	• keep a notebook handy to write things down • ask your pharmacist to Webster-pak your medications • take regular gentle exercise • socialise – tell your loved ones what's going on
Hair loss and thinning	• prepare your family and friends • use a soft hairbrush and a mild baby shampoo • pat your hair dry gently with a towel • cut your hair shorter or have it shaved when you start chemo • use an electric shaver • avoid using heat or chemicals; don't dye or blow dry your hair • use sunscreen on your scalp
Sun sensitivity	• cover up with long sleeves and long pants • wear sunglasses and a hat or beanie to protect your scalp • talk to your nurse about which sunscreens are best to use • avoid sun exposure at high UV times of the day

Corticosteroids

During treatment you may be given drugs called corticosteroids or steroids for short. During treatment you may be given drugs called *corticosteroids* or *steroids* for short. Steroids are drugs that act like your body's own hormones. They can help treat many types of cancer, and they can keep you from having nausea and vomiting after a round of chemo. They can also prevent allergic reactions to other drugs and blood products.

Common steroids are prednisolone and dexamethasone. They can be given in tablet form and/or via drip (intravenously).

Steroid side effects and how to deal with them:

- difficulty sleeping – take in the morning after breakfast
- upset stomach – take with food or milk
- high blood sugar levels – diabetics should increase checks and talk to their treatment team about adjusting insulin
- mood changes – ask your treatment team to refer you to a counsellor
- increased appetite and weight gain – ask your treatment team to refer you to a dietitian
- swelling due to retaining fluid – keep an eye on swelling and let your treatment team know if it gets worse.

Relapsed AL amyloidosis

Sometimes amyloidosis comes back (relapses) after a period of remission. The treatment options will depend on your previous treatment, your age and overall health, and on how long the amyloidosis was in remission. The treatment goal is to achieve remission again. It may involve similar chemo or different drugs. An autologous stem cell transplant may be an option too.

If further treatment or a clinical trial is not an option, the treatment goal might change. You may receive supportive care, which treats any symptoms of amyloidosis. It does not try to cure it.

Stem cell transplantation

Stem cell transplants are also called *bone marrow transplant*, or a *hemopoietic cell transplant* or *HCT*. This treatment is risky because it has very serious side effects. Your haematologist will work out if you need and can have a stem cell transplant.

Stem cell transplant involves having very high doses of chemotherapy before the transplant. The aim is to destroy the cancer cells in your bone marrow. These cells are then replaced with healthy stem cells.

Autologous stem cell transplant

Your own stem cells are collected when your amyloidosis is stable. They're stored until after you've had chemo when they're returned to your bloodstream. This is the more common type of transplant for AL amyloidosis.

Stem cell transplant side effects include:

- low blood counts
- all the same side effects as chemo, but more severe.

You can find out more about stem cell transplants in our booklet *Autologous Stem Cell Transplants* and on our website.

Organ transplants

Slowing or stopping the production of amyloid may not be enough to repair a damaged organ. An organ transplant may be considered. Kidney, heart and liver transplants are sometimes offered in some treatment centres. To stop the amyloid protein depositing in the new organ, you'll need chemotherapy or a stem cell transplant.

Supportive care

Supportive care prevents and treats symptoms and side effects. It includes emotional and social support too.28 The goal is to improve symptoms of your Amyloidosis, but it doesn't treat the disease itself.

Blood transfusions

If you notice symptoms of anaemia, tell your treatment team. You may need a transfusion. Blood transfusions are slow injections of blood into a vein (intravenously, or IV). Transfusions are usually

given by a nurse in a clinic or in an outpatient ward of a hospital. The nurse will use your CVAD (a device that gives access through a central vein in your chest). or will insert a 'drip' (a cannula), a plastic tube, into a vein in your arm. The cannula connects to a bag of blood (called packed red blood cells). Each bag of blood will take 60-120 minutes to transfuse.

Platelet transfusions

If you have symptoms of low platelets (thrombocytopenia), you may need a platelet transfusion. This is like a red blood cell transfusion, but you will be given a bag of platelets instead of packed red blood cells.

Growth factors

Growth factors are chemicals in your blood that help the bone marrow produce different types of blood cells. Some growth factors can be made in the lab. They are used to boost low blood counts.

In Australia, white blood cell growth factors are given. Neutrophils are white blood cells that help fight infections. A growth factor called *granulocyte colony stimulating factor* (usually called G-CSF) makes the bone marrow produce more neutrophils. This is predominantly used when people are being treated with chemotherapy.

Growth factors don't usually cause any major side effects, but some people may have chills, headaches and bone pain after an injection.

Antibiotics

When your white blood cell count is low you have a higher risk of getting infections. If you do, it's important to be treated as soon as possible. Your treatment team will probably prescribe antibiotics, either in tablet form or given via 'drip' straight into your bloodstream (IV).

Antifungal and antiviral medicines

When you start chemo or if you are preparing for a stem cell transplant, you may be given antifungal and/or antiviral medicines. If you're low in certain cell types, you have a higher risk of fungal or viral infections. These types of medicines will be taken to prevent infection. This is called prophylaxis, or you might hear the drugs referred to as *prophylactics*. They will usually be in tablet form and your treatment team will tell you how often and for how long you need to take them.

Vaccines

Vaccines help prevent infections. Only some vaccines are safe, and these are called *inactivated* vaccines. You will need to check with your treatment team when to have them. You should not have any live vaccines.

Elastic stockings

If you have swollen ankles and/or legs (oedema), you may be given elastic stockings to wear. Ask your treatment nurse how to put them on properly. Putting your feet up when you're sitting can help with oedema too.

What is localised amyloidosis?

Localised amyloidosis is usually due to light chain amyloid deposits and so is sometimes also called localised AL amyloidosis.

Amyloidosis is most often systemic. This means that more than one tissue or organ of the body is affected by it. In localised amyloidosis, the amyloid-forming protein is produced *and* deposited in only one part of the body.

Which areas of the body can be affected?

Localised light chain amyloid deposits are most often found in the:

- skin
- upper airways and lungs
- eye, especially the conjunctiva
- bladder
- gastrointestinal tract

Symptoms of localised amyloidosis

Symptoms relate to the affected site. For example, localised amyloidosis of the skin may appear as flat or raised lumps. People with localised amyloidosis in the throat may have a hoarse voice and a sense that their throat is full.

What tests will I need?

Localised amyloidosis is diagnosed by biopsy of the affected site in the body. Your treatment team will do some other tests, like blood and urine testing, to rule out systemic amyloidosis.

Treatment for localised amyloidosis

Usually no treatment is needed. Some people may have surgery or laser therapy to treat the localised amyloidosis depending on where it is in the body.

What is AA amyloidosis?

AA amyloidosis used to be known as *reactive* or *secondary* amyloidosis because people who develop AA amyloidosis have usually had other inflammatory disorders for a long time, often more than 20 years. Inflammatory disorders include rheumatoid arthritis, osteomyelitis, inflammatory bowel disease, tuberculosis, and Familial Mediterranean Fever.

This type of amyloidosis is caused by a normal blood protein called serum amyloid A protein (SAA). People with inflammatory conditions have very high levels of SAA in their blood which can sometimes lead to formation of amyloid deposits made up of SAA protein.

How is serum amyloid protein produced?

The underlying inflammatory disease causes changes in blood chemistry. Levels of SAA increase from normal levels to extremely high levels. They stay high as long as the inflammatory disease remains active. For a small number of people, after time the SAA proteins begin to deposit as amyloidosis fibrils in tissues and organs of the body. We don't know why this happens in some people and not in others.

Which organs can be involved?

Kidneys and the spleen are the main organs affected by AA amyloidosis. Most people have amyloid deposits in the kidneys and this causes problems. The main problem is protein in the urine (called *proteinuria*). Over time, AA amyloidosis can lead to kidney failure.

AA amyloidosis also causes an enlarged spleen. Your spleen is an organ located in the top left of your belly, near your rib cage. When the spleen is swollen (enlarged), it is called splenomegaly.

Symptoms of AA amyloidosis

Often, symptoms of the underlying disease are more obvious than symptoms of the amyloidosis.

If you have splenomegaly you may feel fullness, discomfort or pain in the upper left belly. You may lose your appetite.

Symptoms related to kidney disease include:

- swelling of the ankles and legs (oedema)
- weight loss
- low appetite
- nausea and vomiting
- breathlessness.

Diagnosis of AA amyloidosis

AA amyloidosis is diagnosed through a tissue biopsy, usually of the kidney. Routine blood tests may show elevated levels of creatinine. It is a sign that the kidneys aren't functioning as they should be. Most people with AA amyloidosis will have high protein levels in their urine when they're diagnosed.

Treatment

AA amyloidosis is managed by controlling the underlying inflammatory disease so treatments will differ according to the nature of the underlying condition. This then reduces production of the amyloid protein SAA. Treatments could involve anti-inflammatory therapies (so called disease modifying agents) for rheumatoid arthritis, antibiotics for chronic infections, or colchicine for Familial Mediterranean Fever.

If the SAA level can be reduced to almost normal and stays there for a long time, there is a chance that the existing amyloid will eventually reduce. This will improve how the organs affected by AA amyloidosis function. Improvement is very slow and can take from months to years.

If you have kidney disease you should:

- be careful with fluid balance – how much fluid you ingest, and how much comes out
- limit salt intake
- weigh yourself daily to check if you're retaining fluid
- use diuretics (fluid tablets) as needed
- see your GP or treatment team for regular blood and urine tests to check kidney function.

What is hereditary amyloidosis?

Hereditary amyloidosis is caused by inheriting an abnormal gene (called a *gene mutation*). The gene mutation leads to the production of the abnormal or variant protein that deposits as amyloid fibrils in the body's organs and tissues. There are a few types of hereditary amyloidosis.

Hereditary amyloidosis is an *autosomal dominant disease*. Someone with the mutation may have inherited it from their father or mother and they can pass the gene to their children, who have a 50% chance of inheriting it. If you haven't inherited the gene you can't pass it to your children.

Symptoms of hereditary amyloidosis

Your symptoms will depend on which type of hereditary amyloidosis you have. Many symptoms are the same as for AL amyloidosis.

Which organs can be affected?

Each type of hereditary amyloidosis and family has its own pattern of organ involvement, although it can affect individual family members differently.

Type	*Usual clinical features*
Transthyretin	Neuropathy, heart failure, diarrhoea
Fibrinogen	Hypertension, kidney failure
Apolipoprotein AI	Kidney failure
Lysozyme	Kidney failure, liver failure
Gelsolin	Corneal changes (eye), occasionally heart and kidney disease
Cystatin C	Intra-cranial (brain) haemorrhage
Apolipoprotein AII	Kidney failure

What tests will I need?

First, you'll usually have a tissue biopsy to confirm you have amyloidosis. Then you'll have more tests to work out which type you have. You'll have DNA testing on your blood to work out which type of hereditary amyloidosis you have.

Types of hereditary amyloidosis

The two main types of hereditary amyloidosis are ATTRv and AFib. The other types of hereditary systemic amyloidosis are very rare.

What is ATTRv?

ATTRv is the most common form of hereditary amyloidosis. The first 'A' in ATTRv stands for amyloidosis and the other letters stand for the protein transthyretin (TTR). The 'v' in ATTRv stands for variant, as the mutation in the transthyretin gene results in the production of a variant protein. This is so it's not confused with amyloid due to the normal or 'wild-type' transthyretin, abbreviated to ATTRwt. You can read more about ATTRwt later in this booklet.

ATTRv is caused by mutations of the TTR gene. TTR is made in the liver and circulates in the blood. In healthy people, normal 'wild-type' transthyretin transports thyroid hormone and vitamin A (retinol) through the bloodstream which is why it's called 'trans-thyretin'. People with mutations in the TTR gene make abnormal or variant TTR throughout their lives. ATTRv is also known as familial amyloid polyneuropathy (FAP) or familial amyloid cardiomyopathy (FAC) depending on which organ was predominantly involved.

Symptoms of ATTRv

ATTRv most commonly causes:

- peripheral neuropathy (loss of feeling, pain or weakness in the limbs)
- autonomic neuropathy (bowel and bladder problems, impotence and dizziness on standing)
- heart failure
- amyloid deposits in the eyes, kidneys, adrenal glands or blood vessels.

Symptoms can come on as early as age 20, or as late as age 80.

Within families, the age it starts, how quickly it progresses and which organs are affected will often be similar for each member. Symptoms vary by family. In some families all affected members only have neuropathy, while in other families all affected members have both neuropathy and heart disease.

Treatment for ATTRv

The main goals of treatments are to:

- reduce the production of the abnormal amyloid-forming protein
- stabilise the amyloid forming protein so it cannot misfold and produce amyloid fibrils
- preserve organ function, and
- improve quality of life.

Transthyretin knock-down therapies: Two drugs, have recently been shown to markedly reduce transthyretin levels and result in stabilising or even improving peripheral neuropathy in patients with ATTRv.

Transthyretin stabilisers: A drug that binds to the amyloid forming transthyretin and stabilises the protein in its normal shape so that it cannot misfold and form amyloid. It has been shown to delay the progression of neuropathy and cardiac failure.

Some people may have a liver transplant to remove the source of the mutated TTR. However, people with amyloid in the heart, especially older people, do not usually benefit.

There are medicines that help with pain caused by peripheral neuropathy. You may be given elastic stockings and/or medication for dizziness on standing. You might need drugs to help with your bowels.

What is AFib amyloidosis?

AFib amyloidosis is the short name for fibrinogen A alpha chain amyloidosis. A number of mutations of a gene called the *fibrinogen A alpha chain cause* amyloid. The first symptoms are of kidney disease around age 50-60. As the abnormal fibrinogen is made only in the liver, a liver transplant can prevent further amyloid deposition. A kidney transplant might be considered for kidney failure.

ApoAI amyloidosis

In ApoAI amyloidosis 'ApoAI' stands for *apolipoprotein A1*. There are a few mutations in the gene for apolipoprotein A1 that cause amyloidosis. Half of the abnormal protein is produced in the liver. The kidneys are the main organs affected but the heart, liver, and other organs can be involved. People with this type of amyloidosis may have a transplant to replace any of these organs.

ALys amyloidosis

ALys amyloidosis is short for *lysozyme amyloidosis*. This type of hereditary amyloidosis is very rare. There is no specific treatment except for liver and kidney transplants to replace failing organs.

What is Wild-type transthyretin amyloidosis (ATTRwt)?

ATTRwt used to be known as *senile systemic amyloidosis*. It is due to amyloid deposits made from the protein transthyretin (TTR). In this case the protein is normal or 'wild-type' transthyretin, which is why it's called ATTRwt.

This type of amyloidosis typically causes heart disease in older men.

Symptoms of ATTRwt

Amyloid deposits in the heart make the heart wall stiff (this is called *restrictive cardiomyopathy*). This leads to heart rhythm problems and to heart failure. These changes happen slowly.

Symptoms may include:

- feeling short of breath when moving about
- swollen legs (oedema)
- fatigue – extreme tiredness that does not improve with rest
- palpitations – sudden fast or hard heartbeat
- dizziness or blackouts, which may happen when you exercise or you eat
- chest pain (called angina).

Almost half the people with ATTRwt amyloidosis experience carpal tunnel syndrome. This is a condition that feels like tingling and pain in the wrists, and pins and needles in the hands. Carpal tunnel syndrome often starts several years before any symptoms of heart disease.

What tests will I need?

To confirm you have this type of amyloidosis, you will either need a bone scan or a tissue biopsy. After that you may have some or all of these tests:

- genetic testing to exclude the hereditary variants of ATTR
- electrocardiography
- echocardiogram
- MRI of the heart
- bone scan (DPD or PYP scan): this is where special molecules (DPD or PYP) are injected into the body and highlight parts of the bones that have increased activity. These molecules get trapped in a heart full of transthyretin amyloidosis so can help find and diagnose TTR amyloid in the heart.

You can read more about most of these tests earlier in this booklet.

Treatment for ATTRwt

The main goals of treatment are to:

- reduce the production of the abnormal amyloid-forming protein
- stabilise the amyloid forming protein so it cannot misfold and produce amyloid fibrils
- support and preserve organ function, and
- improve quality of life.

Transthyretin knock-down therapies: Two drugs have recently been shown to markedly reduce transthyretin levels and result in stabilising or even improving peripheral neuropathy in patients with ATTRv.

Transthyretin stabilisers: A drug that binds to the amyloid forming transthyretin and stabilises the protein in its normal shape so that it cannot misfold and form amyloid. It has been shown to delay the progression of cardiac failure in ATTRwt.

Usually a cardiologist (a heart specialist) manages treatment for people with ATTRwt. Many medicines used for heart failure and heart rhythm problems aren't useful for people with ATTRwt amyloidosis. Some blood pressure-lowering medications should be avoided if there is amyloid affecting the heart.

Some people may have:

- fluid tablets (diuretics) to help with oedema and shortness of breathblood thinners (anticoagulant medicines)
- a pacemaker to help maintain heart rate.

Clinical trials

Clinical trials (also called *research studies*) test new treatments and often provide early access to promising therapies that aren't routinely available. Results from the new agent (or combination of agents) are compared against current (or standard) treatments to see if the new treatment works better. They also check for the new treatment's side effects. Many clinical trials are randomised, which means some patients receive the new treatment and others the current standard of care treatment.

Clinical trials provide important information about how treatments can be improved. Sometimes people on clinical trials (called 'participants') have access to expensive new treatments that aren't available on Australia's PBS.

Your haematologist may suggest you join a clinical trial. Before you can start, you will need to give informed consent. This shows that you understand the risks and benefits of the trial treatment.

Clinical trials are run through hospitals and clinics, just like other treatments. You will have a clinical trial nurse as part of your treatment team.

Clinical trial participation is purely voluntary. If you are offered a clinical trial, make sure you understand how your treatment will be different compared to what is generally given (standard of care treatments). Be sure to ask any questions you have before deciding whether to participate.

You can search current clinical trials at:

Australian Cancer Trials *australiancancertrials.gov.au*

ANZ Clinical Trials Registry: *anzctr.org.au and*

ClinTrial: *clintrial.org.au*

Second opinion

If you feel unsure about your diagnosis or treatment, you are entitled to seek a second opinion from an independent doctor. This may be at the same hospital or clinic, or at a different location. If you feel overwhelmed, then you might benefit from speaking with someone at the Leukaemia Foundation, your GP, or a counsellor for advice.

Complementary therapies

Complementary and alternative medicines (CAM) also known as integrated therapies are not standard medical treatments.

These therapies should 'complement' or be done alongside medical treatment after consultation with your treatment team.

No complementary or alternative or alternative treatment on its own can treat Hodgkin Lymphoma. They may help with some symptoms or side effects.

Managing fatigue

Many people who have blood cancer treatment get fatigue. It's called *cancer-related fatigue*. It can be hard to describe to people who haven't felt it. It's more than being tired, its different to normal everyday tiredness, and is often not resolved with sleep or rest. You will feel tired, but you may also feel weak and also sleepy, drowsy, impatient, or confused. It's hard when you have no get-up-and-go, however, for most people fatigue should improve after you finish treatment.

Tips for managing fatigue

Fatigue is a side effect of your blood cancer or treatment, so managing fatigue is an important part of your overall treatment and care. Make sure you talk to your treating team about it. They

may suggest referral to a psychologist who specialises in sleep management. It's very important to explain how you feel to your carers and support people, and to let them know your priorities and discuss how they can help.

A positive sleep routine, pre-sleep relaxation techniques, napping if tired during the day for up to an hour, regular moderate exercise or movement, eating well, drinking plenty of water, and avoiding use of electronic equipment, such as computers late at night, can be very helpful in improving sleep and reducing fatigue.

While you're managing your fatigue, jot down what time of day you have the most energy and when you feel most tired. That will help you get into a routine and prioritise your energy. Play games, listen to or play music, read, catch up with friends and family; these things might seem difficult, but they will help distract you from the fatigue.

Fertility decisions

Some types of treatment may affect your fertility, which is your ability to conceive a baby. It is important to talk to your treatment team about future fertility before you start treatment. If you are planning on having a child, there are steps you can take.

Make sure you understand:

- the fertility preservation processes
- success rates
- the risks
- side effects of fertility treatments, and
- any costs.

For men

Chemo can stop or lower your sperm production. It can reduce your sperm's ability to move. This can be temporary or permanent. It also affects the hormone testosterone.

The best way to preserve your fertility before treatment is by freezing a semen sample, which contains sperm. This is called *sperm cryopreservation*.

For women

Chemo can reduce your number of available eggs (ova) and can affect your hormones. Tell your treatment team right away if you are pregnant or think you may be.

There are several mainstream fertility *cryopreservation* (freezing) options for women. Egg and embryo freezing are common, less so ovarian tissue freezing. For some young women and their families, it may not be possible to pursue fertility options prior to cancer treatment. Having the opportunity for discussion about your future fertility is important.

Fertility Society of Australia: *fertilitysociety.com.au*

Practical matters

Navigating the health system

The Australian health system may seem large, complicated, and stressful especially when you are also living with a blood cancer. Knowing a bit about how our health system works and who are key people in your care can make navigating the system much easier.

Key people in your health team

Haematologist - A specialist doctor trained in diseases of the blood including blood cancer who leads a team of doctors in your care.

Radiation oncologist - A doctor who specialises in treating cancer using radiotherapy.

Cancer care coordinator (CCC)/ Cancer nurse consultant (CNC) - Specialist cancer nurses who coordinate patient care and provide referrals to allied health professionals if needed.

Cancer nurse - A nurse in an outpatient clinic or cancer ward who supports, educates, and gives you your chemo treatment.

Palliative care physician - A doctor who specialises in controlling symptoms and improving quality of life in people with terminal illnesses and chronic health conditions.

Pharmacist - A health professional who prepares, dispenses medicines (drugs), and supports your understanding of how to manage your side effects with medication prescribed.

Accredited practising dietitian - A university-qualified professional with ongoing training and education programs, who helps to support your recovery and manage challenges in your diet.

Social worker - A health professional who specialises in emotional support, counselling, and advice about practical and financial matters.

Physiotherapist/Exercise physiologist - Health professionals who specialise in treating and rehabilitating patients through physical means.

Psychologist - A health professional who specialises in providing emotional support and difficulties such as anxiety, distress, and depression.

Record your important contact details here:

	Contact name	Phone	Comment
Emergency			
GP			
Haematologist			
CNC/CCC			
Chemo Day Unit			
Pharmacist			
Dietitian			
Social worker			
Psychologist			

Useful website: *https://patients.cancer.nsw.gov.au/diagnosis/navigating-the-health-system*

The new normal – what is it?

For many people, the start of treatment signals changes to life that include the day-to-day managing of a multitude of new activities and changes around treating and monitoring of your blood cancer. Frequent appointments with your health care team and regular follow up can be tiring and stressful for all.

Life is not exactly the same as it was before the blood cancer diagnosis. Everyday life changes for you and the people around you. Things that were once important may no longer be so, or things that weren't important before, now take greater priority.

In essence, a '*new normal*' is about living with your blood cancer, creating and maintaining your new normal to live as good a life as possible while facing changes such as and not limited to:

- Physical/mental/spiritual
- Emotional/relationships/identity/sexuality
- Financial, ability to work/return to productivity

Seeking information, tools, and support, and accepting help to manage challenges that arise throughout a person's cancer experience is very important. Having this support can enable individuals to have a high quality of life throughout their blood cancer journey. It is also important to remember that dealing with the diagnosis and treatment of blood cancer is a big life change and everyone handles it and is affected differently.

Diet and nutrition

During treatment, nutritional goals are designed to prevent or reverse malnutrition, avoiding weight loss (preserving lean body mass/ muscle) and to minimise side effects, such as decreased appetite, nausea, diarrhoea, dry mouth, and taste changes.

Being underweight or malnourished can have a negative effect on your overall quality of life. Poor appetite and weight loss are associated with symptoms such as weakness, fatigue, difficulty sleeping, and pain.

It is likely you will be encouraged to eat a high-energy diet to meet the changing metabolic demands of your body during this time. During chemo treatment, you may experience complications that negatively affect your nutrition and hence your overall wellbeing, such as mucositis (ulcers in the mouth/throat and/or stomach).

You may be given drugs called corticosteroids, as part of your treatment. Steroids can cause weight gain through increased appetite stimulation and fluid retention (oedema) in your limbs, abdomen, and face.

General nutrition recommendations for people receiving cancer treatment:

- **Maintain a healthy weight.** For many people, this means avoiding weight loss by getting enough calories every day. For people who are obese, this may mean losing weight. It's important to get advice from your health care team before you try to lose weight during treatment.
- **Get essential nutrients.** These include protein, carbohydrates, fats, and water.

You can make an appointment to see a hospital dietitian as an outpatient or ask to see one if you are an inpatient. Your treating team may refer you to a dietitian. Community dietitians are also available, and your GP can arrange this through a care plan if your private health insurance doesn’t cover it.

Exercise

With any blood cancer treatment, it is common to experience deconditioning; a physical and/or psychological drop in function. Having cancer doesn't mean you can't be physically active.

People with blood cancer should attempt to avoid inactivity and sedentary behaviour as much as possible and return to normal daily activities as soon as possible following diagnosis.

What are the benefits of exercise/physical activity?

Strong evidence is available to show that exercise and physical activity improves outcomes for people with cancer across a range of dimensions including:

- cancer-related fatigue, pain, psychological distress, anxiety, depression
- in-bone health, cognitive, and cardiovascular function
- health-related quality of life.

Exercise can be tailored to the individual and often around activities of daily living. Before you commence any exercise program it is important to speak with your treatment team first to make sure it is safe to do so and to see who is best placed to help you.

Information on exercise in cancer can be found on the Clinical Oncology Society of Australia (COSA) website: *cosa.org.au*

Mental health and emotions

Your emotional health is a very important aspect of overall wellbeing. Many people being treated for blood cancer experience a range of feelings and it is not uncommon to feel low, depressed, or anxious.

Feeling sad is a normal response to a cancer diagnosis as is worrying about the future. Feelings can be challenging and may include anxiety, grief, guilt, uncertainty, anger, spiritual distress, fear, and feeling isolated or lonely. Worrying about treatment, its success and side effects, or changes in your physical, lifestyle, and family dynamics can also impact your mental health.

Seeking help from your healthcare team is important. They and/or your GP can refer you to someone who can help, such as a psychologist who specialises in blood cancer. The Leukaemia Foundation's Blood Cancer Support Coordinators can also help you to work through what you are feeling and provide information on who might assist you in your local area.

Relationships/carers/family and friends

Undergoing treatment for a blood cancer can affect your role as a parent, partner, friend, and workmate, to name a few. You and all the people in your life will cope differently. Be open with your communication and encourage family and friends to be open with you.

Communicating effectively with family, children, friends, and a carer is essential. Being clear with others about what you want and need allows them to be of greater support. Together you can work as a team to manage and solve problems as they arise.

There are many allied healthcare staff and not-for-profit organisations that can assist with support and information. The Leukaemia Foundation is one of them and can assist you, your carer or family in identifying who can help with different issues and how to contact them.

Carers Australia: *carersaustralia.com.au*

Carer Gateway (Australian Government): *carergateway.gov.au*

Canteen: *canteen.org.au*

Redkite: *redkite.org.au*

Work/finances/legal matters

Finances

People with blood cancer often report a negative impact on their financial situation during treatment. Monthly costs can increase and may be influenced by financial considerations such as travel, childcare, and taking time off work for appointments. Your household income may be reduced due to you or your carer having to stop work, or reduce hours permanently or temporarily, as a result of your diagnosis.

A financial stocktake

When you become aware that you may lose your income or suffer a reduced income as a result of the ill health of you or an immediate family member, the first step should be to run a quick 'financial stocktake'.

First, assess what income you can expect or what financial resources you have available. Possibilities may include:

- Are you or your partner able to work part-time?
- Do you have sick leave or long service leave?
- Do you have income protection or trauma insurance, either as a stand-alone policy or part of a life policy?
- Do you have money in the bank or a line of credit against your mortgage which can be drawn against?

The second step involves checking on important expenses which need paying in the immediate future. Put together a brief budget if you don't have one.

Seeking help

Financial advice around budgeting and what financial assistance is available to you can be discussed with a number of sources. Your local Leukaemia Foundation Blood Cancer Support Coordinator can help point you in the right direction.

A few key other options to consider are:

Centrelink

If you expect to lose all or most of your income or your partner's income, the first organisation to contact is Centrelink. The earlier you make an application, the sooner you may receive some relief payments. If you have employment to return to, this will affect the basis of your benefit. Your partner may also be eligible for a Carer Payment or Carer Allowance, so be sure to enquire about this.

Centrelink online account *(sign in through myGov for instructions): centrelink.gov.au*

Financial institutions

If you are unable to make your regular payments on your mortgage as a result of serious ill health, it is important that you let the relevant organisations know as soon as possible. Most banks and other financial organisations have special arrangements for customers in financial hardship as a result of ill health.

Other sources of help

Do not hesitate to discuss your financial circumstances with your treatment centre social worker or your private insurer. They may be able to assist with advice on deferring payments. Some of your household accounts may also have hardship support programs (like energy providers). It may be possible to access some money from your superannuation fund to help with emergency payments. Don't forget to check if your superannuation has income replacement insurance as one of its features. If you are not sure, give their helpline a call.

Moneysmart: *moneysmart.gov.au*

National Debt Helpline: *ndh.org.au or phone 1800 007 007*

Legal matters

This information applies equally to all members of the community, not just those who have a blood cancer or their carer. The best time to get your affairs in order is when you are in good health. Here we consider some of the most common legal documents you should have and where to get help.

Enduring Power of Attorney/Enduring Guardian

There may be circumstances when a person loses the capacity to make decisions for themself. You can sign a legal document which allows you to choose a trusted person to make decisions on your behalf.

An *Enduring Power of Attorney (EPOA)* is a document that allows your trusted person the power to sign documents on your behalf, make personal, administrative and, if you choose, financial decisions.

An *Enduring Guardian (EG)* is another document where your trusted person can make decisions on your behalf regarding your health matters such as medical treatment, care, and protection, even if this decision overrides your wishes.

Australian Guardianship and Administration council: *agac.org.au*

Wills

It is very important to have specialist legal advice when preparing your Will. There are many questions and options your legal adviser will discuss with you which may not be immediately apparent. This is particularly important when you have dependent family members and you need to determine who will be responsible for them in the event of your death.

If you die without leaving a valid Will, you leave what is known as 'intestacy'. Although you may feel your affairs are very simple and your immediate family will receive your assets, this is not necessarily true. Should you die intestate, distribution of your estate will be determined by a formula set down in legislation. If you already have a Will, you need to consider if it is still current.

Advance Health Directive

This is a document you may complete to clearly indicate your wishes as to medical treatments you may or may not wish to receive in the event of a serious illness or accident. Although this is a lengthy document, it is simple to complete as it mainly consists of a series of optional questions. There are also sections where you make comments in your own words. While this form can be completed on your own, you may wish to discuss it with your family, and you are also required to have a doctor sign the form to certify that you understand the contents of the document.

Advance Care Planning Australia: *advancecareplanning.org.au or phone 1300 208 582.*

Getting help

Help with legal matters is available from several sources including:

- Solicitors
- Trustee companies
- The Public Trustee in your state
- Australian Guardianship and Administration Council

More information & help

Glossary

You can find any **bold** terms in the definitions also defined in this glossary.

allogeneic	Blood, stem cells, bone marrow, or other tissue transferred from one person to another. For example, an allogeneic **stem cell transplant** involves transplanting stem cells harvested from another person.
anaemia	A lower-than-normal number of **red blood cells** in the blood. It causes tiredness, paleness, and sometimes shortness of breath.
antibodies	Also called immunoglobulins. Proteins in the blood that recognise and bind to other substances called targets. Antibodies destroy their target (such as a virus) and prevent infection. Auto-antibodies or self-antibodies target your own cells and can destroy them.
autologous	Blood or other tissue derived from your own body. For example, an autologous **stem cell transplant** involves reinfusing stem cells that have been harvested from your own body.
baseline	A first measurement of a condition taken early on, used to compare over time to look for changes.
basophils	A type of white blood cell. They help fight infection
Bence Jones protein	A type of protein found in the urine of some people with AL amyloidosis or myeloma.
biotherapy	A type of treatment that uses substances made from living organisms to treat disease. These substances may occur naturally in the body or may be made in the laboratory.
blast cells	Immature blood cells normally in the **bone marrow** in small numbers.
bone marrow	Soft, sponge-like tissue in the centre of most bones. It contains **stem cells** that make all blood cells.

bone marrow biopsy	Also called a bone marrow aspirate, bone marrow trephine or BMAT. The removal of a small sample of **bone marrow**. This is sent to the lab for a **pathologist** to look at under a microscope.
bone marrow aspirate	A sample of bone marrow fluid.
bone marrow transplant	Also called a **stem cell transplant**. A procedure where a patient is given healthy stem cells to replace their own damaged stem cells. The healthy stem cells may come from the bone marrow of the patient or a donor. There are three types: autologous (using a patient's own stem cells that were collected from the marrow and saved before treatment), allogeneic (using stem cells donated by someone who is not an identical twin), or syngeneic (using stem cells donated by an identical twin).
bone marrow trephine	A sample of bone marrow tissue.
cancer	Diseases where some of the body's cells become faulty, begin to multiply out of control, can invade and damage the area around them, and can also spread to other parts of the body to cause further damage.
chemotherapy	The use of drugs to treat cancer.
chromosome	Part of a cell that contains genetic information.
coagulation	Process of changing from a liquid blood to a solid. Also called *clotting*. Platelets help with coagulation.
cytogenetic tests	The study of the structure of **chromosomes**. These tests are carried out on samples of blood and **bone marrow**. The results help with diagnosis and getting the most appropriate treatment.
cytopenia	Where there is a lower-than-normal number of a type of blood cell in the blood.
eosinophils	A type of white blood cell. They help fight infection.
erythrocytes	Also called red blood cells. A type of blood cell made in the bone marrow and found in the blood. **Haemoglobin** makes these cells red in colour.

full blood count	Also called FBC or complete blood count. A routine blood test that measures the number and type of cells, and the **haemoglobin** and **haematocrit** in the blood.
granulocytes	A kind of white blood cell. There are three types: eosinophils, basophils and neutrophils. They help fight infection.
growth factors	Proteins that control cell division and cell survival. Some are made in the lab and used as treatments, such as G-CSF.
haematocrit	The amount of blood that is made up of **red blood cells**.
haematologist	A doctor who specialises in diagnosing and treating blood disorders.
haemoglobin	A protein inside **red blood cells** that carries oxygen around the body.
haemopoiesis	The formation of new blood cells.
immune system	The body's defence system against infection and disease.
immunotherapy	Immunotherapy, sometimes called biological therapy, is a type of cancer treatment that works by boosting a person's own **immune system** to fight the **cancer**. Immunotherapy is currently approved in Australia for some types of cancers and is also being trialled for other cancers.
immunoglobulins	Also called **antibodies**. Proteins in the blood that recognise and bind to other substances called targets. Antibodies destroy their target (such as a virus) and prevent infection. Auto-antibodies or self-antibodies target your own cells and can destroy them.
leukocytes	Also called white blood cells. These blood cells are made in the bone marrow and found in the blood and lymph tissue. They help the body fight infection and are part of the **immune system**. Types: granulocytes (neutrophils, eosinophils, and basophils), monocytes, and lymphocytes (T-cells and B-cells).
light chains	Part of an **immunoglobulin**. There are two types: kappa and lambda.

lymphocytes	A type of **white blood cell** that plays a role in the immune system
M protein	Also called monoclonal protein or paraprotein. Abnormal proteins found in AL amyloidosis and myeloma.
megakaryocytes	Very large bone marrow cells that break apart to form **platelets**.
mutation	A harmful change in 'normal' DNA (the building blocks of all cells).
neutropenia	A lower-than-normal number of **neutrophils** in the blood. It increases the risk of infection.
neutrophils	The most common type of **white blood cell**. They help fight infection.
pancytopenia	Where there are lower-than-normal numbers of a type of all blood cells and platelets in the blood.
paraprotein	Also called M protein or monoclonal protein. Abnormal proteins found in myeloma and light chain amyloidosis.
pathology	The study of diseases to understand their nature and their cause. A specialist in this field is called a **pathologist**. In cancer, histopathology/histology involves examining tissue under a microscope. Haematopathology involves blood and lymph.
peripheral neuropathy	Nerve damage to peripheral nerves, including fingers and hand, toes and feet, caused by disease or treatment. Symptoms are pain, tingling and/or numbness.
petechiae	Tiny, unraised, round red spots under the skin caused by bleeding.
platelets	Also called thrombocytes. Tiny pieces of cells (**megakaryocytes**) found in the blood and spleen. They help form blood clots (**coagulation**) to slow or stop bleeding and to help wounds heal.
prognosis	An estimate of the likely course and outcome of a disease.
purpura	Bleeding and bruising under the skin.

red blood cell	Also called an erythrocyte or RBC. A type of blood cell made in the bone marrow and found in the blood. **Haemoglobin** makes these cells red in colour.
relapse	Return of the original disease after it has improved for a time.
rigor	Also called a chill. Feeling cold with shivering or shaking and looking pale, but with a high temperature. A symptom of infection.
stem cells	Young (immature) blood cells that can develop into more than one type of cell. Bone marrow stem cells grow and produce **red blood cells**, **white blood cells** and **platelets**.
stem cell transplant	Also called a SCT or **bone marrow transplant**. A procedure where a patient is given healthy stem cells to replace their own damaged **stem cells**. The healthy stem cells may come from the bone marrow of the patient or a donor. There are three types: autologous (using a patient's own stem cells that were collected from the marrow and saved before treatment), allogeneic (using stem cells donated by someone who is not an identical twin), or syngeneic (using stem cells donated by an identical twin).
thrombocytes	Also called platelets. Tiny pieces of cells (**megakaryocytes**) found in the blood and spleen. They help form blood clots (**coagulation**) to slow or stop bleeding and to help wounds heal.
thrombocytopenia	A lower-than-normal number of **platelets** in the blood. It causes bruising and bleeding.
treatment-related	Also called secondary. Where the disease is caused by previous treatment. For example, treatment-related MDS.
white blood cells	Also called leukocytes or WBCs. Blood cells made in the bone marrow and found in the blood and lymph tissue. They help the body fight infection and are part of the **immune system**. Types: granulocytes (**neutrophils**, eosinophils, and basophils), monocytes, and **lymphocytes** (T-cells and B-cells).

Australian Amyloidosis Network (AAN) Clinic Centres

The AAN clinics provide a multi-disciplinary service to type amyloidosis and provide management advice for all types of amyloidosis. The clinics also run clinical trials.

The day to day provision of therapy and supportive care cannot be administered State-wide for every amyloidosis patient and we share this essential care with local physicians.

Useful websites

Leukaemia Foundation:	leukaemia.org.au
Australian Amyloidosis Network:	amyloidosis.net.au
Australian New Zealand Clinical Trials Registry:	anzctr.org.au
eviQ Cancer Treatments Online:	eviq.org.au
Australian Red Cross Blood Service:	mytransfusion.com.au
Pharmaceutical Benefits Scheme:	pbs.gov.au
ClinTrial Refer:	clintrial.org.au

Question builder

- ☐ Who will be my main contacts? How do I best contact you/ them?
- ☐ What can I do to avoid infections?
- ☐ Can I have the flu shot and other vaccinations?
- ☐ Is it safe to take my supplements or vitamins?
- ☐ Can I eat normally, is there anything I need to avoid or special diets that will help me?
- ☐ Can I exercise and what is the best frequency and type for me?
- ☐ Are there any clinical trials for my type of amyloidosis and am I eligible?
- ☐ Could this treatment affect my sex life? If so, how and for how long?
- ☐ Will my treatment send me into menopause?
- ☐ Where can I or my loved ones get any other support?

How you can help

The Leukaemia Foundation ensures every Australian with blood cancer gets access to the trusted information, best-practice treatment, and essential care they need. With no ongoing government funding, we rely on the generosity of the community to help support all Australians living with blood cancer.

We understand that everyone's personal situation is different, so below are some of the ways you and your family may like to get involved.

Give

Your donations help provide free support services to Australians affected by blood cancer, (like financial and practical assistance, education, counselling and accommodation), and drive some of Australia's most important—and life-saving—cancer research.

Become a Lifeblood Hero

With your regular monthly gift, you can be there every step of every day in every way, for people living with blood cancer. Your ongoing support helps ensure everyone everywhere has access to the life-saving treatment and support they need.

Leave a gift in your Will

After taking care of your loved ones, a gift in your Will is a direct and valuable way of helping transform the future for Australians with blood cancer allowing your support to live on as a lasting legacy.

Fundraise

Get involved in World's Greatest Shave, plan a special Light the Night or celebrate your Best-Birthday-Ever! You can even create your own personal fundraising initiative that is completely unique.

Volunteer

Our wonderful volunteers are a crucial part of our success—helping at our signature fundraising events, maintaining our accommodation centres or providing support with specialised skills.

Become a Leukaemia Foundation Member

You can make a difference to the future of blood cancer by joining a networked community who care about people living with blood cancer. Members are the lifeblood of the Leukaemia Foundation and play an important role in shaping the future of the organisation.

Partner with us

There are many ways your business can help people with blood cancer. Together, we can create a bespoke partnership that aligns with your organisation's objectives and corporate social responsibility.

Give blood, marrow and tissue

Stem cell transplants and blood donations save the lives of many people facing many blood cancer. Visit donateblood.com.au to register today.

To find out more contact us today supporters@leukaemia.org.au 1800 620 420

The Leukaemia Foundation gratefully acknowledges those who assisted in the development of this information: Leukaemia Foundation Blood Cancer Support Coordinators, nursing staff, clinical haematologists, and bone marrow transplant physicians representing the various states and territories of Australia.

The Leukaemia Foundation values feedback. If you would like to make suggestions or tell us about your experience in using this booklet, please contact us via email: **info@leukaemia.org.au** or phone us on **1800 620 420**.

Leukaemia Foundation®

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