AL Amyloidosis

Your guide to best cancer care

About this guide

Being told you have or could have AL amyloidosis can be overwhelming. A lot can happen quickly, and you might have many questions. This resource can help to guide you, your family, and friends through this experience.

Information and support

For more information about AL amyloidosis, visit the Leukaemia Foundation website **www.leukaemia.org.au/blood-cancer/amyloidosis/al-amyloidosis/**. You can also view an educational video on amyloidosis available here **www.youtube.com/watch?v=_l1syuVFLcA**.

Australian Amyloidosis Network: The Australian Amyloidosis Network is a not-for-profit organisation of patients, carers, and health professionals working together to improve the lives of patients with all types of amyloidosis. For more information, visit **www.aan.org.au**.

Leukaemia Foundation: To speak with an experienced healthcare professional who can provide you with disease-specific information, answer your questions, talk through your concerns, and connect you to support groups, call **1800 620 420** or visit **www.leukaemia.org.au**. The team can also help with practical concerns such as accommodation close to treatment, transport to appointments, and financial assistance.

Cancer Council: for information and support, call **13 11 20** to talk to an experienced healthcare professional or visit **www.cancer.org.au**.

More information is available in the 'Resources' section of the optimal care pathway for AL amyloidosis. **www.cancer.org.au/OCP**.

Translating and Interpreting Service (TIS): If you need a translator, call TIS on **13 14 50** or visit **www.tisnational.gov.au**.

Is AL amyloidosis a blood cancer?

Amyloidosis is the name given to a group of rare disorders in which abnormal proteins misfold forming amyloid fibrils. The body cannot break down amyloid proteins easily and they clump together and can deposit and/or accumulate in any tissue and organ of the body. As these deposits progressively build up, they interfere with the working of the body's organs. Without treatment, this will eventually lead to organ failure.

Over 30 types of amyloidosis have been identified. This guide discusses one type only, AL amyloidosis. The prefix A stands for amyloid, and L stands for light chain. Normally the human body produces B cells and plasma cells which are found in the bone marrow. These cells are part of the body's immune system. In AL amyloidosis, these plasma cells, or sometimes B cells, begin to grow abnormally, and begin to produce large amounts of a single type of free light chain.

Many people ask whether AL amyloidosis is a blood cancer. It is not. Although the amyloid forming protein is produced by an underlying bone marrow disorder, this is usually benign (non-cancerous) but in some cases the growth of plasma cells or B cells can be malignant (cancerous). AL amyloidosis is commonly linked with a plasma cell disorder called myeloma, and sometimes it is associated with a B-cell disorder called lymphoma, however, AL amyloidosis is not typically considered to be a cancer.

Initial tests and referral

Symptoms

Your general practitioner (GP) or other primary care physician will do a check-up to see if they can find out what is making you unwell. AL amyloidosis can present with many different symptoms, which will vary from patient to patient (e.g., fatigue and weight loss) and/or blood test abnormalities. These symptoms can mimic other diseases which may delay diagnosis. You might have fatigue, weight loss, feeling faint, bruising of the skin especially around the eyes, a drop in blood pressure after sitting or lying down (postural hypotension), pain, numbness, tingling or weakness in different parts of your body (peripheral neuropathy),

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swelling in the legs and/or abdomen (oedema), diarrhoea or constipation, difficulty breathing or shortness of breath (dyspnoea), kidney and/or liver problems, and an enlarged or painful tongue. Typically, your general practitioner will refer you to an appropriate specialist (e.g., a cardiologist for heart problems, a nephrologist for kidney problems, a neurologist for peripheral neuropathy or a health service with the relevant specialty services). Those specialists will then perform investigations to diagnose amyloidosis.

Initial tests you may have:

Physical exam. Your GP or specialist will check to see the cause of symptoms such as those noted above.

Blood tests. A sample of your blood is collected and sent for a full blood examination and may include a liver function test.

Electrocardiogram (ECG) and echocardiogram. Your doctor may want to check your heart by performing these tests.

Referrals

If your GP or primary care physician thinks you may have the symptoms listed above, they will refer you to an appropriate specialist at a public hospital or in private practice for more tests. You may be referred to different specialists such as a haematologist (blood and bone marrow), cardiologist (heart), nephrologist (kidneys), neurologist (nervous system), or gastroenterologist (gastrointestinal tract) due to the symptoms you experience while the diagnosis is being investigated.

It can help to have a family member, carer, or friend attend appointments with you.

Let your doctor know what is most important to you in a specialist (e.g., cost, location, bedside manner, expertise).

Timeframes

The timing of your haematologist appointment will depend on your initial results of tests performed by your GP or referring specialist. For a small number of patients, an urgent appointment or hospital admission may be required, but for most people with no urgency, an appointment **within four weeks** is appropriate.

If you can't get an appointment within this timeframe, follow up with your GP.

Ask your GP to recommend trusted sources of information and support — you can share these with your family and friends too.

Talk to your GP/specialist about how to manage any other health conditions you may have during your treatment of AL amyloidosis and let them know if you have any concerns.

Questions you might want to ask your GP or specialist

- Are there any symptoms that I should watch out for while I am waiting for my appointment to see a haematologist or other specialist?
- What should I do if any symptoms are worsening while I am waiting for my appointment to see a specialist?
- Can I choose the specialist I see?
- What emotional and mental health support services are available and how do I access them?
- Can I choose whether I go to a public or private hospital?
- What are the differences of being treated in the public versus private system and what are the costs involved?
- What travel assistance schemes are available to me? (If you live in regional or rural areas).

It's a good idea to keep a written diary or digital record of treatment details and appointments with your GP and specialists. You can ask permission to record your appointments. It's also helpful to bring a list of questions and ask for a written treatment plan.

Don't ignore new signs and symptoms. Alert your GP or specialist. Trust yourself. It's OKAY to be persistent.

Travel to access tests, treatment and care

You may have to travel to see a haematologist, undergo tests or access treatment, especially if you live in a rural or regional area. In some cases, you may have to leave home for an extended period of time. Accommodation and transportation support services are available, and it is encouraged to have family support with you whilst you undergo treatment. You can ask about what patient travel subsidy schemes are available to you.

Mental health and emotional wellbeing

A diagnosis of AL amyloidosis can affect your mental and emotional wellbeing. Talking with your GP or your hospital treatment team and being referred to patient support organisations (such as Leukaemia Foundation or the Australian Amyloidosis Network), or other health professionals such as a psychologist or social worker, can help you develop strategies to cope. They can help you access a mental health treatment plan if required.

Diagnosis, staging and treatment planning

If you haven't seen a haematologist, you will be referred to one. Haematologists are trained in conditions that affect the blood and bone marrow. The haematologist will do more tests to confirm if you have AL amyloidosis and help determine the best treatment for you.

You may have one or a combination of these tests:

- **Tissue biopsy** to check for amyloid deposits and further biopsies of other involved organs.
- **Bone marrow biopsy.** A doctor will put a needle into your hip bone. A sample of the bone marrow tissue will be sent to a laboratory for examination. This is usually done with a local anaesthetic, and inhalational pain relief or mild sedation.
- **Cardiac tests** including an echocardiogram to check heart function.
- **Renal tests** including a urine test to check kidney function.
- **Liver tests** to check the size and functioning of the liver.
- **Clinical assessment** for peripheral neuropathy to check for any damage to nerves.
- **Blood pressure test** including testing postural blood pressure (testing your blood pressure while sitting and standing).

The most frequently affected organs by AL amyloidosis are the heart, kidneys, gastrointestinal tract, nervous system, and liver.

Timeframes

You should have most results back **within four weeks**.

Questions you might want to ask

- What is AL amyloidosis and where can I find more information about it?
- What tests will I need and why?
- Will my follow-up appointments be face-to-face or by tele/video health or a combination?
- What's my prognosis?
- How much will appointments and tests cost and how much will I have to pay myself?
- Can I choose where I have treatment?
- Will I need to start treatment straight away?
- How long will the treatment last?
- How will my disease be monitored?
- Is there information that I can share with my family or friends?
- What support services are available to me?
- Are there support groups available?

Treatment

Your haematologist will talk to you about your treatment options for AL amyloidosis. You may need more than one treatment type to get the best results, and you may require ongoing treatment to manage your AL amyloidosis for the rest of your life. Treatment for AL amyloidosis focuses on stopping or slowing the production of amyloid and subsequently its deposition in the body's organs and tissues, to preserve organ function, and to improve quality of life.

You may have one or a combination of these treatments:

- **Proteasome inhibitors (PIs).** Drugs that are responsible for killing the affected plasma cells that are causing AL amyloidosis.
- **Chemotherapy.** Drugs that are used to kill the affected plasma cells that are causing AL amyloidosis.
- **Monoclonal antibodies (mAbs)** work in the same way as natural antibodies that your body produces; they fight infection and target the affected plasma cells that are causing AL amyloidosis.
- **Corticosteroids.** Drugs that help kill the affected plasma cells that are causing AL amyloidosis.
- **Autologous stem cell transplant.** Your haematologist will discuss whether this is an option for you.

Your treatment team will monitor you closely. If the treatment is not controlling the production of the amyloid quite quickly, treatment may be changed after discussion with you.

For more information visit **www.cancer.org.au/cancer-information/treatment**, and **https://aan.org.au/patients-and-carers/al/**.

Supportive care is very important and includes treatment for your symptoms to improve your quality of life and psychosocial help to support you through your amyloidosis journey. This may be delivered through your multidisciplinary team including nurses, pharmacists, psychologists, physiotherapists, and dietitians. You can also ask to be contacted by the Cancer Nurse Consultant/Cancer Care Navigator at your hospital or by a social worker if you have not yet seen one.

Timeframes

By evaluating your symptoms and test results, your specialist will determine when your treatment should start. For most people this will generally be **within four weeks** of the decision being made. If your heart has been affected, treatment should start **within two weeks**. There may be instances where your treatment could be delayed beyond this timeframe. If this occurs your specialist will discuss this with you.

Clinical trials

Your specialist may recommend participating in a clinical trial (or you can ask if you are eligible for any clinical trials). Clinical trials often provide access to promising new treatments that are not yet available to the general public. Many people with blood cancer related disorders are now living longer, with a better quality of life, because of clinical trials.

For more information visit **www.australiancancertrials.gov.au** or search the Australian New Zealand Clinical Trials Register **www.anzctr.org.au or https://aan.org.au/clinical-trials/**.

Complementary therapies and other medications

Speak to your healthcare team about any dietary supplements, vitamins, herbal remedies, and other medications that you use or would like to use, including prescription and over-the-counter medicines (e.g., paracetamol or ibuprofen). Some medicines, complementary therapies, vitamins, and foods might not work well with your treatment.

Fertility and sexual health

Blood cancer related disorders such as AL amyloidosis and its treatment may cause fertility problems for both men and women. This will depend on age, the type of disorder, and the treatment received. If this is relevant, get advice from your treating team about contraception before, during, and after treatment. Discuss having children whilst undergoing treatment and the need for referral to a fertility specialist and options for fertility preservation before starting treatment. Diagnosis and treatment may affect your sex life or overall sexual health. For support and, if necessary, referral to counselling services, contact the Leukaemia Foundation on **1800 620 420** or visit **www.leukaemia.org.au** and type 'relationships and sex' in the search bar.

You can ask for more time to think about your treatment options.

You can ask your GP for a referral to another specialist for a second opinion.

Questions you might want to ask

- What treatment do you recommend and why?
- Are there alternatives?
- What will happen if I don't have treatment?
- How long will I need to be on treatment?
- What will treatment cost and how much of the cost will I have to pay myself, or through health insurance?
- What activities/exercise will help me during and after treatment?
- Is there any specific diet I should follow or foods or drink to avoid during treatment?
- How will the treatment affect my day-to-day life, and can I still work?
- Who is in my treatment team and who is the main contact person if I have any questions or feel unwell?
- Who should I contact in an emergency?
- What side effects could I have from treatment and how should I manage them?
- Will treatment affect my ability to have children, and if yes, what options should I consider?

Decisions about cost

You may have to pay for some appointments, tests, medications, accommodation, travel, or parking. Speak with your GP, specialist, or private health insurer (if you have one) to understand what is covered and what your out-of-pocket costs may be.

If you have concerns about costs, talk to your healthcare team or a social worker about:

- being bulk-billed or being treated in the public system
- help with accommodation and/or transport during treatment
- the possible financial impact of your treatment and financial support schemes you may be able to access
- how your treatment might impact your ability to work.

For more information about financial and practical support call the Cancer Council on **13 11 20** or visit **www.cancer.org.au/support-and-services/practical-and-financial-assistance**, or contact the Leukaemia Foundation on **1800 620 420** or visit **www.leukaemia.org.au/how-we-can-help/practical-support-services/**.

Care during treatment

Patients with AL amyloidosis will need to be followed up by their medical team for the rest of their lives. Although there is a good chance that many patients with AL amyloidosis will, with treatment, achieve remission (the amyloid is no longer being produced and deposited in any organs) they may still require maintenance treatment or further treatment if the amyloid production starts again. Comprehensive supportive care including emotional and social support should be part of routine ongoing care.

Survivorship care plan

Current therapies mean that most people with AL amyloidosis will go on to live long lives. You may need regular ongoing follow-up appointments and care, so your specialist and healthcare team will work with you to make a survivorship care plan for you and your GP. Your survivorship care plan may need to be updated over time if your needs change. This plan will explain:

- who your main contact person is after treatment
- how often you should have check-ups and what tests this will include
- understanding and dealing with potential side effects of treatment
- how to get help quickly if you have an urgent problem
- how to look after your overall health and wellbeing
- what healthcare and community support services are available to you and how to access them.

Your specialist and healthcare team will talk to you about your needs and can refer you to other health professionals and community support services. Other information you get might be about:

- the side effects of treatment and the specialists you may need to see
- how to make healthy lifestyle choices to give you the best chance of staying well.

For more information visit **www.cancer.org.au/cancer-information/after-a-diagnosis/after-cancer-treatment**.

Questions you might want to ask

- Who should I contact if I'm feeling unwell?
- Who should I contact if I'm concerned about symptoms or side effects?
- What can I do to be as healthy as possible?
- Who should I contact if I need advice?
- Where can I get more help?

Living with relapsed or progressive disease

Over time, most patients with AL amyloidosis will relapse after responding to initial treatment. Relapse is not certain and may not occur soon after treatment. As people respond differently, many patients can live in long-term remission. Access to the best available therapies, including clinical trials and treatment with a multidisciplinary team, are crucial to achieving the best outcomes for relapsed disease.

Treatment will depend on how far your amyloidosis has progressed and the different organs that may be affected.

AL amyloidosis patients will be closely monitored throughout treatment. If treatment is stopped because it is not controlling the amyloid production or the amyloid starts depositing again and the patient comes out of remission, a full assessment by the treatment team might be called for to determine the present status of the disease.

Options may include:

- using a different drug regimen that contains drugs of a different class or a different drug of the same class
- 'Watch and Wait'
- retreatment with a previous regimen
- clinical trial, if appropriate
- stem cell transplantation in some patients
- supportive and/or palliative care.

Questions you might want to ask

- What is the stage of my disease?
- What is happening with my disease?
- What are my treatment options?
- What are the chances that the treatment will work this time?
- Is there a clinical trial available?
- What financial, practical, or emotional support is available?

Advance care planning

Your GP or healthcare team may talk with you, your family, and carer about future treatment and medical needs.

Advance care directive

Sometimes known as a living will, an advance care directive is a legally binding document that you prepare to let your family and medical team know about the treatment and care you might want or not want in case you become too unwell to make those decisions yourself. For more information visit **www.advancecareplanning.org.au**.

Symptom and palliative care management

Your specialist may refer you to palliative care services, but this doesn't always mean end-of-life care. Today people can be referred to these services much earlier if they're living with AL amyloidosis or if their AL amyloidosis returns. Palliative care can help you to live as well as you can including managing pain and symptoms, psychosocial support, and assistance in identifying care goals. This care may be at home, in a hospital, or at another location you choose. Speak to your GP or haematologist/specialist or visit **www.palliativecare.org.au**.

Making treatment decisions

You may decide not to have treatment at all, or to only have some treatment to reduce pain and discomfort. You can always change your mind about your treatment. Just talk to your specialist. For more information visit **www.cancer.org.au/cancer-information/treatment/advanced-cancer-treatment**.

Questions you might want to ask

- What can be done to reduce my symptoms, if any?
- What extra support can I get if my family and friends care for me at home?
- Can you help me to talk to my family about what is happening?
- What support is available for my family or carer?
- Can I be referred to a community support service?
- What support services are available to me to help manage the ongoing nature of my AL amyloidosis?

Published in May 2024.

This resource is based on information from the optimal care pathway for people with AL Amyloidosis (1st edition), available at **www.cancer.org.au/OCP**.