AL amyloidosis
An introduction

An Introduction | Essentials

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Disclaimer: The information in this Introduction is not meant to replace the advice of your medical team. They are the best people to ask if you have questions about your individual situation.

This publication is intended for a UK audience. It therefore may not provide relevant or accurate information for a non-UK setting.

About this Introduction

This Introduction has been written for AL amyloidosis patients. It may also be helpful for their families and friends.

It aims to:

- Provide an overview of AL amyloidosis and its treatment
- Answer some of the common questions about AL amyloidosis

For more detailed information about AL amyloidosis and living with AL amyloidosis, see **AL amyloidosis – Your Essential Guide** and **Living well with AL amyloidosis – Your Essential Guide.**

Myeloma UK also provides Infoguides and Infosheets covering a range of specific topics about AL amyloidosis, its treatment and management.

To order your free copies call our **Myeloma Infoline** on **0800 980 3332**. This information is also available on our website at **www.myeloma.org.uk/amyloidosis**

To talk to one of our Myeloma Information Specialists about any aspect of myeloma, call the **Myeloma Infoline** on **0800 980 3332**. The Myeloma Infoline is open from Monday to Friday, 9am to 5pm and is free to phone from anywhere in the UK and Ireland. From outside the UK, call + 44 (0)131 557 9988 (charged at normal rate). Information and support about myeloma is also available around the clock at **www.myeloma.org.uk/amyloidosis**

What is AL amyloidosis?

The term 'amyloidosis' is a general term used for a group of diseases where an abnormal protein, called amyloid, accumulates in the tissues. The build-up of amyloid protein is called an 'amyloid deposit' which can occur in various organs or tissues and cause problems.

Different types of amyloidosis are named according to the type of amyloid protein which is produced. All begin with the initial **'A'** which stands for amyloidosis, followed by another letter(s) which identifies the particular amyloid protein, for example: AL amyloidosis, AA amyloidosis and ATTR amyloidosis.

In AL amyloidosis it is abnormal plasma cells in the bone marrow that produce the amyloid protein. In AL amyloidosis the amyloid proteins are light chains (the 'L' in 'AL' stands for 'light chain'). Light chains are normally part of healthy antibodies, also known as immunoglobulins, produced by healthy plasma cells.

The amyloid protein is only broken down very slowly by the body and starts to build up in the tissues and organs - gradually damaging their function and causing symptoms. This build-up can happen almost anywhere in the body. Each patient has a different pattern of amyloid deposition, with different organs affected.

Basic facts

- Amyloid can build up in the kidneys, heart, liver, spleen, nerves, or digestive system
- Amyloid can affect two or more organs at the same time
- AL amyloidosis does not affect the brain
- AL amyloidosis is a relatively rare disorder, with approximately 500 – 600 people diagnosed in the UK each year

AL amyloidosis and myeloma

Although the amyloid deposits in AL amyloidosis are not themselves cancerous, the disease may occasionally be associated with myeloma (a plasma cell cancer).

You may have been diagnosed with AL amyloidosis alone or, less commonly; you may have developed AL amyloidosis after being diagnosed with myeloma. It is rare, but possible, for someone diagnosed with AL amyloidosis to later develop myeloma in addition to their AL amyloidosis.

Regardless of whether or not a patient has myeloma associated with their amyloidosis, the treatments for AL amyloidosis are similar to those for myeloma.

For more information about myeloma, see Myeloma - Your Essential Guide from Myeloma UK.

What causes AL amyloidosis

The causes of AL amyloidosis are not known, but it is not contagious or known to be inherited.

Typically AL amyloidosis affects people in their 60s and 70s, although younger people have been diagnosed.

Symptoms and complications of AL amyloidosis

AL amyloidosis affects every patient differently. The symptoms experienced depend on which organs are affected by amyloid deposits and the degree to which their function is impaired.

Most patients will have more than one organ affected by amyloid and the organ that is most affected will be referred to as the 'dominant organ'. You may experience some or all of the symptoms listed below.

Symptoms involving:

The kidneys

Chronic kidney disease is common in patients with AL amyloidosis. Amyloid deposits in the kidneys affect their filtering system and can result in a condition called nephrotic syndrome in which the lower legs typically become swollen (oedema). In some cases the amyloid deposits will cause the kidneys to lose the ability to purify the blood. This is called kidney or renal failure.

The heart

Amyloid deposits in the heart can cause it to become unusually stiff and therefore unable to function effectively. This results in shortness of breath, which may become apparent with only the slightest exertion. Amyloid can also affect the electrical system of the heart causing the heart beat to be disturbed (arrhythmia).

The nervous system

Amyloid deposits can affect the nerves of the hands, feet and lower legs and may cause pain, numbness and tingling. This is called peripheral neuropathy.

Nerves that control blood pressure, heart rate, gut motility (movement) and other body functions can also be affected causing a variety of symptoms

including dizziness when standing too quickly, nausea and diarrhoea. This is called autonomic neuropathy.

The digestive system

Amyloid in the digestive system, also called the gastrointestinal tract (GI tract), can cause nausea, diarrhoea, weight loss, a feeling of fullness in the stomach after eating small amounts and an enlarged tongue (called macroglossia).

Non-specific symptoms

These are common and may have been present for some time before diagnosis. They include fatigue, weakness, weight loss and loss of appetite.

How is AL amyloidosis diagnosed and monitored?

In order to diagnose and monitor AL amyloidosis, several tests and investigations need to be carried out. These are outlined below.

Tests and investigations are done in order to:

- Establish a diagnosis
- Determine a treatment plan and to monitor progress
- Detect complications of the disease so they can be monitored and treated

Serum Free Light Chain Assay

As well as being important in diagnosing AL amyloidosis, changes in the level of free light chains in the blood are usually a fairly good indicator of changes in the activity of the disease (the higher the free light chain level, the more quickly amyloid is deposited). For this reason free light chain measurements are done regularly to see how well treatment is working and to check that the AL amyloidosis is remaining stable during periods when you are not receiving treatment.

For more information see the AL amyloidosis - Serum Free Light Chain Assay Infosheet from Myeloma UK.

Bone marrow and tissue biopsies

A biopsy involves the removal of a small sample of tissue for microscopic examination. In AL amyloidosis this is done to look for evidence of amyloid deposits. In order to diagnose AL amyloidosis this may be taken from the bone marrow, liver, heart, nerves, kidneys or simply an area of fat. Regardless of the organ(s) predominately

affected, microscopic amyloid deposits can usually be detected throughout the body.

The SAP scan

This investigation is available at the NHS National Amyloidosis Centre in London and is performed routinely for most patients who are referred there for evaluation of their AL amyloidosis.

The SAP scan is able to show where the amyloid is located within the body and which organs are affected. It has therefore reduced the need for organ biopsies as a way of monitoring the disease. The scan is likely to be repeated every 6 – 12 months to monitor the amount and location of the amyloid deposits and therefore help guide the need for ongoing treatment.

Through the use of the SAP scan it has been found that amyloid deposits often decrease and disperse when the underlying abnormal plasma cell disease is controlled. Over time this is usually accompanied by an improvement in organ function and general health.

AL amyloidosis is an individual disease and the results from these tests and investigations may vary from patient to patient. It is not enough just to make a diagnosis of AL amyloidosis; it is critical to have an accurate picture of the disease in each patient before an appropriate treatment plan can be developed.

For more information see the AL amyloidosis - SAP scan Infosheet from Myeloma UK.

Treatment of AL amyloidosis – the basics

Treatments for AL amyloidosis can be effective at controlling the disease, reducing symptoms and improving quality of life but as yet, they are not curative.

In general, treatment is given to:

- Reduce the levels of abnormal plasma cells responsible for producing the amyloid protein, as far as possible
- Prevent further tissue or organ damage
- Control the AL amyloidosis if it has come back again (relapse)
- Improve quality of life
- Prolong life

Treatment for AL amyloidosis is often most effective when two or more drugs, with different but complementary mechanisms of action, are given together.

In the past the number of treatment options for AL amyloidosis was somewhat limited but with the development of newer treatments in the last decade, there are now more options available.

Before starting treatment, each option must be considered carefully so that the benefits of treatment are weighed against the possible risks of side-effects. In most patients, overall health, age, fitness and any previous treatments will be taken into account.

The length of treatment varies depending on the type of treatment(s) being used and the stage at which the treatment is being given. Treatment is usually given over a number of weeks which may or may not be followed by a rest period. This pattern constitutes one cycle of treatment and a series of treatment cycles is referred to as a course of treatment.

Future directions

Much research is taking place to find more effective treatments for AL amyloidosis and finding the best ways of using them. Many of these are currently being tested in clinical trials taking place around the world.

For example, Imnovid® (pomalidomide) – another immunomodulatory drug similar to Revlimid and thalidomide – is currently undergoing clinical trials in both newly diagnosed and relapsed AL amyloidosis patients.

Since AL amyloidosis is related to myeloma, much of the research undertaken in the field of myeloma can be applied to AL amyloidosis and several new treatments are in the pipeline. These treatments may therefore also become available for AL amyloidosis patients in the near future.

About Myeloma UK

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Ireland: 1800 937 773

Find your nearest **AL amyloidosis Support Group** to meet up and talk to other people face to face.

AL amyloidosis **Matters**

Read ***AL amyloidosis Matters*** newsletter which offers a mix of the latest news in research and development for AL amyloidosis, and patient experiences.

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Learn about AL amyloidosis from experts and meet others at our **Patient and Family AL amyloidosis Infodays.**

Visit **www.myeloma.org.uk**, a one-stop-shop for information on AL amyloidosis; from news on the latest research and drug discovery to articles on support, treatment and care.

Watch **Myeloma TV** which hosts videos about AL amyloidosis presented by experts, patients and family members.

Myeloma TV

Use the **Discussion Forum** for the opportunity to share experiences and advice about living with AL amyloidosis.

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Information available from Myeloma UK

Our information covers all aspects of AL amyloidosis.
For a full publication list visit **www.myeloma.org.uk/publications**

To fill in a short survey about our patient information online, please go to **www.myeloma.org.uk/pifeedback**

Essentials	Gives an overview of AL amyloidosis, its treatment and management. Particularly useful for newly diagnosed patients and their families.
Treatments and tests	Provides information about the range of treatments and tests used in AL amyloidosis.
Symptoms and complications	Information about the most common symptoms and complications of AL amyloidosis.
Living well with AL amyloidosis	Provides information relating to living well with AL amyloidosis, such as diet, managing finances, travel insurance and caring for someone with AL amyloidosis.
Related conditions	Information on conditions related to AL amyloidosis, including MGUS, plasmacytoma, smouldering myeloma.

Other publications

Patient diary

This diary helps patients keep a track of hospital appointments and key test results in a practical, simple way.

The small things that make all the difference

Hints and tips written for people affected by myeloma, by people affected by myeloma, many may also relate to AL amyloidosis.

Myeloma A – Z

A booklet which explains key terms relating to myeloma, many also relate to AL amyloidosis.

Our information and publications are free and available to order by phone. You can also download or read online.
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  Or by posting a cheque payable to **Myeloma UK, 22 Logie Mill, Beaverbank Business Park, Edinburgh, EH7 4HG**
- **Fundraising** – fundraising is a positive way of making a difference and every pound raised helps. However you decide to raise funds, our Fundraising Team is here to support you. Contact us on **0131 557 3332** or email **fundraising@myeloma.org.uk**
- **Leaving a legacy** – gifts from Wills are an important source of income for Myeloma UK and will help us to continue providing practical support and advice to myeloma patients and their families. They also help us to undertake research into the causes of AL amyloidosis and investigate new treatments.

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Myeloma UK publications are extensively reviewed by patients and healthcare professionals prior to publication.

Nobody ever forgets the moment they are diagnosed. Myeloma UK advances the discovery of effective treatments, with the aim of finding a cure. That is what patients want, it's what they deserve and it's what we do.

Judy Dewinter – Chairman, Myeloma UK

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