In AL amyloidosis, an abnormal protein called amyloid is produced and accumulates in tissues and organs. The build-up of amyloid protein is called an amyloid deposit. These deposits can occur in various organs or tissues, where they can cause damage. If this occurs in the heart, it is known as cardiac amyloidosis.

What does the heart do?
The heart is the muscle at the centre of our circulatory system. It pumps blood around the body to provide tissues with the oxygen and nutrients they need from the blood and then removes waste products from tissues. The heart has four chambers formed by walls of muscles that contract and relax to pump blood in and out.
What is cardiac amyloidosis?
Cardiac amyloidosis occurs when amyloid deposits build up in the heart. These deposits cause the heart to become stiff and enlarged, which impairs its ability to pump blood efficiently around the body. When amyloid deposits accumulate they damage the heart and can cause heart damage and failure.

What are the symptoms and complications of cardiac amyloidosis?
One of the most common symptoms of cardiac amyloidosis is oedema (the retention of abnormally large amounts of fluid in the body), which causes:
- Swelling in the lower legs
- Swelling in the abdomen
- Rapid increase in weight, e.g. more than three pounds in 2 days
- Coughing and/or trouble breathing, especially at night/when lying flat

Other common symptoms and complications associated with cardiac amyloidosis include:
- Shortness of breath – which can occur even after minimal movement
- Arrhythmia (irregular heartbeat) – causes breathlessness, dizziness or fainting after exertion or eating
- Low blood pressure – particularly when standing up
- Congestive heart failure
- Embolism (e.g. blood clot or blockage)
- Stroke

Diagnosing and monitoring cardiac AL amyloidosis
There are several tests and investigations used to diagnose and monitor cardiac amyloidosis and assess the extent of amyloid deposits in the heart.

Blood Tests
Blood tests are used at diagnosis and during treatment to monitor heart damage. Blood is tested for NT-proBNP and cTnT proteins which are released into the blood when there is damage to the heart muscle. Higher than normal levels of these proteins in AL amyloidosis patients indicate that there is probably cardiac damage.

However, if you have renal (kidney) damage, your kidneys are less able to filter these proteins from the blood, which
can cause them to build up. This means that measuring these proteins may not be reliable in patients with renal damage.

**Echocardiogram (echo)**

An echo uses high frequency sound waves (ultrasound) to create images of the heart and surrounding tissues.

**Magnetic Resonance Imaging (MRI)**

An MRI scan is a scanning procedure which involves a combination of radio waves, a powerful magnetic field and a computer to produce images of organs or tissues in the body. An MRI scan generates more detailed images than an echo and allows doctors to look at the heart from different angles.

**Serum Amyloid P Component Scintigraphy (SAP scan)**

A SAP scan is a scanning procedure which shows the distribution and amount of amyloid in the organs, without the need for biopsies. This scan is only performed at the National Amyloidosis Centre in London.

**Heart Biopsy**

A biopsy involves the removal of a small sample of the heart tissue for examination. The sample is taken by inserting a thin wire into a vein in the neck and threading it into the right ventricle of the heart, using x-ray and ultrasound to guide the wire. This procedure is painless but quite invasive. This is only required to diagnose cardiac amyloidosis when other less invasive tests are inconclusive.

**How is cardiac amyloidosis treated and managed?**

The most effective way to treat cardiac amyloidosis is to treat the underlying condition, while controlling the symptoms and complications. By treating the AL amyloidosis itself, the amount of abnormal amyloid produced is reduced or controlled, resulting in fewer deposits in the heart which cause damage. The symptoms and complications associated with cardiac amyloidosis can be managed to improve quality of life.

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

For an overview of the treatment for AL amyloidosis see AL amyloidosis – Your Essential Guide from Myeloma UK
Shortness of breath

Shortness of breath can be managed by breathing techniques that help you to focus on your breathing. Try finding a comfortable position where you can rest and relax your neck and shoulders. Breathe in through your nose and out through your mouth. If you are feeling short of breath it is important to let your doctor or nurse know.

Arrhythmia

It may be necessary to be fitted with a pacemaker if your cardiac amyloidosis is causing your heart to beat irregularly. A pacemaker regulates the heart rhythm and can improve symptoms of low blood pressure.

Low blood pressure

If you have low blood pressure, stand up slowly, especially first thing in the morning. Try stretching and making small physical movements before standing up from lying or sitting. Eat small meals more frequently rather than having a few large ones and avoid drinking alcohol. Also try to avoid standing for long periods of time.

Most patients with cardiac amyloidosis have low blood pressure and drugs routinely used to treat high blood pressure, like ACE inhibitors or beta-blockers, may worsen symptoms of cardiac amyloidosis. Some treatments for other conditions, such as diabetes, can also worsen symptoms. You should speak to your doctor or nurse if you are taking any drugs for any other conditions.

Oedema (fluid retention)

Patients with cardiac amyloidosis can retain abnormal amounts of fluid and are very sensitive to changes in the sodium (salt) levels in the body. Diuretics (a type of drug used to promote the excretion of fluids by increasing urine production) can be prescribed to help regulate the amount of fluid in your body. It is important to maintain a steady fluid intake, so speak to your doctor or nurse about the amount of fluid you need to have every day, as both too little and too much fluid can worsen your symptoms.
Some tips for self-management

It is important that you manage and monitor the symptoms and complications of cardiac amyloidosis at home. You can do this by:

- Controlling your fluid intake. Fluid intake should be steady and limited to your doctor or nurse’s recommendation.

- Controlling your salt intake. Following a healthy, balanced diet and limiting the amount of salt in your food can help reduce water retention. A dietician can be helpful for personalised dietary advice.

- Monitoring your weight with a set of accurate digital scales. Digital scales are recommended as small weight changes can be monitored more precisely. Keep a diary of your weight and make a note of any changes. If you have a weight gain of more than three pounds in 2 days, let your doctor or nurse know as soon as possible as this could indicate that you are retaining too much fluid.

- Exercising gently and regularly to increase general well-being and improve circulation.

Summary

Amyloid deposits in the heart can cause it to enlarge, stiffen and prevent it from working properly. Treatment is focused on treating the underlying condition while controlling the symptoms and complications to improve your overall quality of life. It is important for you to be aware of the symptoms and complications associated with cardiac amyloidosis and report them to your doctor or nurse as soon as possible.

About this Infosheet

The information in this Infosheet is not meant to replace the advice of your medical team. They are the people to ask if you have questions about your individual situation. All Myeloma UK publications are extensively reviewed by patients and healthcare professionals prior to publication.

For more information see the Diet and nutrition in AL amyloidosis Infosheet from Myeloma UK.
Other information available from Myeloma UK

Myeloma UK provides a wide range of information covering all aspects of the treatment and management of AL amyloidosis.

For a full publication list visit www.myeloma.org.uk/publications

To order your free copies contact Myeloma UK. Our information is also available to download at www.myeloma.org.uk

To talk to one of our Myeloma Information Specialists about any aspect of AL amyloidosis, call the Myeloma Infoline on 0800 980 3332 or 1800 937 773 from Ireland.

Information and support about AL amyloidosis is also available around the clock at www.myeloma.org.uk/amyloidosis