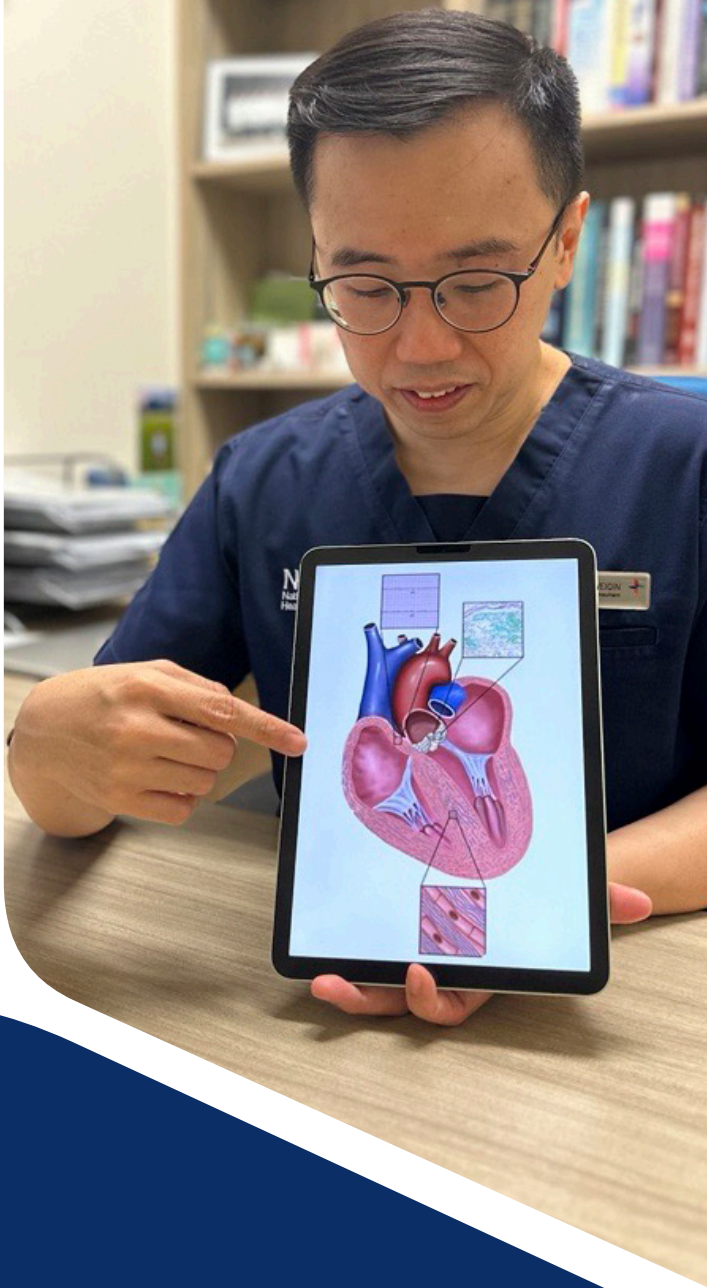
National University Heart Centre Singapore



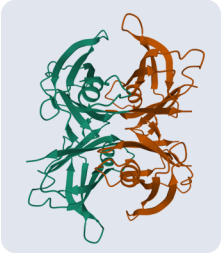
Amyloid Cardiomyopathy

What is Amyloid Cardiomyopathy?

Amyloidosis is a disorder caused by misfolding of normal body proteins. These misfolded proteins are insoluble and are termed 'amyloid proteins.' Amyloid proteins can deposit in various body organs, resulting in organ dysfunction. When these amyloid proteins deposit in the heart, it causes heart dysfunction, termed 'Amyloid Cardiomyopathy' or 'Cardiac Amyloidosis'.

The 2 proteins responsible for most Amyloid Cardiomyopathy cases are:

- **Immunoglobulin light chains**: protein subunits of antibodies produced by bone marrow cells
- **Transthyretin**: a transporter protein produced by the liver



Light Chain Amyloidosis is caused by an abnormal multiplication of bone marrow cells that produce immunoglobulin light chain proteins. On the other hand, Transthyretin Amyloidosis is caused by either a mutation in the gene coding the Transthyretin protein ('hereditary Transthyretin Amyloidosis') or due to aging ('wild-type Transthyretin Amyloidosis').

What are the common symptoms?

- Shortness of breath
- Dizziness and low blood pressure upon standing up
- Difficulty breathing when lying flat
- Swelling of ankles/legs/abdomen
- Unexplained weight loss

Other common conditions associated with Amyloid Cardiomyopathy include:

- Bilateral carpal tunnel syndrome (pain/ tingling sensation in the palms of the hand)
- Spinal canal stenosis
- Aortic valve stenosis (calcific obstruction of the valve separating the aorta from the heart)

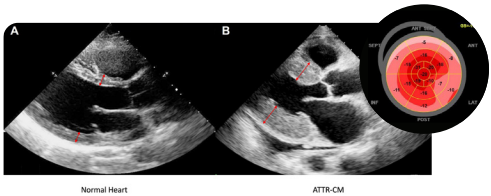
How is Amyloid Cardiomyopathy diagnosed?

Amyloid Cardiomyopathy is diagnosed by cardiologists, by integrating various clinical information, including:

- **Patient symptoms**

- **Clinical examination**
  - Signs of fluid retention
  - Supporting signs of underlying amyloidosis

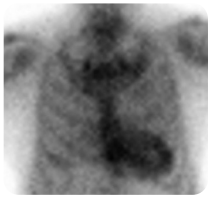
- **Echocardiogram**
  - Ultrasound examination looking at supporting signs of Amyloid Cardiomyopathy (such as severely thickened heart muscles)



- **Blood and urine tests**
  - To look for abnormal light chain protein levels, in Light Chain Amyloid Cardiomyopathy (AL-CM)

- **Electrocardiogram (ECG)**
  - Electrical tracing of the heart, looking for heart rhythm abnormalities, as well as signs of underlying structural heart abnormalities

- **Cardiovascular Magnetic Resonance Imaging (Cardiac MRI)**
  - Specialised scan looking specifically at the heart muscles, for signs of Amyloid Cardiomyopathy



- **Technetium Pyrophosphate (Tc-PYP) scintigraphy**
  - Specialised nuclear scan which allows doctors to diagnose Transthyretin Cardiac Amyloidosis (ATTR-CM) non invasively

- **Endomyocardial biopsy**
  - Invasive procedure to sample heart muscles, to look for amyloid proteins in the heart

What is the treatment for Amyloid Cardiomyopathy?

Management of symptoms

- Exercise prescription
- Lifestyle advice
- Patients will be put on diuretics ('water pills') to reduce the chance of fluid retention. Medications that can potentially lower the blood pressure or heart rate excessively, will be also discontinued.

Disease-specific treatment

Light Chain Amyloid Cardiomyopathy (AL-CM)

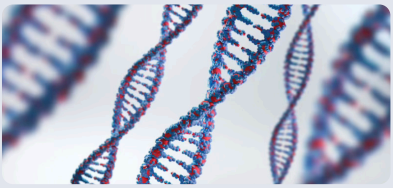
- Treated by our haematology colleagues with chemotherapy, aim at removing the abnormal bone marrow cells producing excessive light chain proteins.

Transthyretin Amyloid Cardiomyopathy (ATTR-CM)

- Disease-specific treatment are emerging such as:
  - Medications:
    - Tafamidis, Acoramidis
    - Patisiran, Inotersen, Eplontersen
  - CRISPR-Cas9 gene editing therapy
- Liver transplantation can be considered for patients with hereditary ATTR-CM

Genetic Counselling

Since ATTR-CM is a potentially inheritable condition, a detailed family history will be obtained. ATTR-CM patients will be encouraged to undergo genetic counselling and genetic testing, to identify if their condition is caused by a gene mutation. Family screening will also be encouraged.



The Cardiomyopathy Clinic at the National University Heart Centre, Singapore (NUHCS) provides a comprehensive evaluation and treatment service for patients with Amyloid Cardiomyopathy. Our multi-disciplinary amyloidosis team consists of of cardiologists, haematologists and neurologists specialised in managing amyloidosis patients.

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