ST VINCENT'S HOSPITAL SYDNEY

Amyloidosis Clinic Heart Failure Guide

Cardiac amyloidosis is a disorder caused by amyloid fibril deposition in the extracellular space of the heart. Among the different types of amyloidosis, nearly all cases of cardiac amyloidosis are caused by light chain amyloidosis (AL) or transthyretin amyloidosis (ATTR). Infiltration of amyloid within the heart leads to progressive dysfunction of the cardiac muscle and development of a **restrictive cardiomyopathy**. The conduction system of the heart can also be affected, with manifestations including **arrythmias and conduction block**. Amyloid cardiomyopathy is an **important and underdiagnosed cause of heart failure and cardiac arrhythmias**.

Managing heart failure in cardiac amyloidosis can be challenging as patients **may not tolerate standard heart failure therapy**. Patients often have a **narrow window of euvolaemia** and can **require careful titration** of heart failure therapy. This simplified guide has been developed from the National and International Cardiac Amyloid and Heart Failure guidelines.[1,2]

Heart Failure Pharmacotherapy in Amyloid

Diuretics

- Loop diuretics are first line for managing fluid overload.
- Retrospective trials suggest mineralocorticoid receptor antagonists may have benefit.
- Monitoring for renal impairment and postural hypotension is required.
- Thiazides should be used with caution due to increased risk of adverse effects.

SGLT2i

- SGLT2 inhibitors are generally well tolerated and early observational[3] data suggests they may reduce heart failure hospitalisations and mortality.

Beta blockers

- Beta blockers should be used with caution and may worsen heart failure symptoms and outcomes.

ACEi / ARBs / ARNIs

- ACEi / ARBs / ARNIs are often poorly tolerated, particularly as disease progresses and are not recommended.

Arrythmia Management in Amyloid

- If **atrial fibrillation** is detected, **anticoagulation is recommended regardless of CHA2DS2-VASc score** as patients with amyloidosis are highly thrombogenic.
- **Prior to cardioversion, a transesophageal echocardiogram should always be performed** due to the high risk of thrombus regardless of anticoagulation.
- Clinicians should have a **high index of suspicion for conduction disease and arrhythmias particularly atrial fibrillation**, which are common in amyloid cardiomyopathy and a frequent cause for heart failure exacerbations.

Patient Centered Care in Amyloid

- **Empower your patient to drive their heart failure care**:
  - ✓ Educate your patient regarding fluid/weight monitoring and fluid restrictions (1.5 – 2L) if needed.
  - ✓ Encourage your patient to seek medical attention if weight increases by more than 2kg in 2 days or they have new breathlessness or dependent swelling.
- **Encourage regular exercise to prevent frailty:**
  - ✓ Moderate daily exercise is encouraged.
  - ✓ Develop an exercise plan with your patient or in coordination with cardiac rehabilitation.
- **Engage community resources:**
  - ✓ Refer to a local heart failure service.
  - ✓ Refer to a specialist amyloid centre to support care including diagnosis, multi-organ screening, genetic testing and clinical trial access.

References:

1. Bart NK, Fatkin D, Gunton J, Hare JL, Korczyk D, Kwok F, Lam K, Russell D, Sidiqi H, Sutton T, Gibbs SDJ, Mollee P, Thomas L. 2024 Australia-New Zealand Expert Consensus Statement on Cardiac Amyloidosis. Heart Lung Circ. 2024 Apr;33(4):420-442. doi: 10.1016/j.hlc.2023.11.027. Epub 2024 Apr 2. PMID: 38570258.
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3. Porcari A, Cappelli F, Nitsche C, Tomasoni D, Sinigiani G, Longhi S, Bordignon L, Masri A, Serenelli M, Urey M, Musumeci B, Cipriani A, Canepa M, Badr-Eslam R, Kronberger C, Chimenti C, Zampieri M, Allegro V, Razvi Y, Patel R, Ioannou A, Rauf MU, Petrie A, Whelan C, Emdin M, Metra M, Merlo M, Sinagra G, Hawkins PN, Solomon SD, Gillmore JD, Fontana M. SGLT2 Inhibitor Therapy in Patients With Transthyretin Amyloid Cardiomyopathy. J Am Coll Cardiol. 2024 Jun 18;83(24):2411-2422. doi: 10.1016/j.jacc.2024.03.429. PMID: 38866445.

For more information or individual patient advice:

1. Visit our website via the QR code
2. Email us at amyloidreferrals@svha.org.au
3. Telephone the St Vincent's Hospital switchboard (p) 8382 1111 and ask for the Cardiac Amyloidosis Fellow