Cardiac Amyloidosis: An AHP Guide

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Cardiac amyloidosis is an infiltrative cardiomyopathy caused by the deposition of misfolded protein fibrils—most commonly light chains (AL) or transthyretin (ATTR)—within the myocardial extracellular space. This progressive accumulation leads to increased myocardial stiffness, impaired diastolic function, and ultimately heart failure. Unlike hypertrophic cardiomyopathy, the increased wall thickness in cardiac amyloidosis is due to infiltrative expansion rather than true hypertrophy. For the CMR Technologist or Allied Health Professional (AHP), understanding the imaging features of this condition is essential for accurate diagnosis, prognostication, and therapeutic planning.

Cardiac MRI is a pivotal tool in the diagnosis of amyloidosis, offering superior tissue characterization via T1 mapping and late gadolinium enhancement (LGE). Extracellular volume (ECV) mapping provides additional quantification of amyloid burden. Familiarity with these imaging biomarkers allows AHPs to tailor protocols effectively and contribute to patient care pathways.

1. Patient Preparation and Set up

- Field strength: 1.5 T preferred; 3 T acceptable with optimized sequences.
- ECG gating: Reliable R-wave detection is crucial due to potential conduction abnormalities. Consider prospective triggering or real time cine imaging in arrhythmias
- Coils: ≥ 8-element phased-array coil for optimal SNR.
- Contrast: Confirm no contraindications; obtain haematocrit for ECV quantification.
- Breath-holds: Use acceleration techniques to maintain ≤10–12 s where possible, especially in dyspnoeic patients.

2. Imaging Workflow

Set	Sequence/Technique	Notes/Purpose
1	Anatomy/Localisers	Scout in standard planes to define cardiac orientation
2	LV Function – Cine SSFP	Full short-axis stack and long-axis views (2CH, 3CH, 4CH) to evaluate wall motion and EF. Note diffuse thickening.
3	Atrial Volumes + Function	Assess biatrial enlargement—a key clue in amyloidosis
4	T1 Mapping	Native T1 values are elevated in both AL and ATTR; critical for early detection

5	ECV Mapping	Quantifies amyloid burden—use haematocrit-adjusted maps pre- and post-contrast.
6	LGE Imaging	Inversion time–scouted LGE sequences in SA and long-axis planes. Adjust TI carefully—may require phase-sensitive IR (PSIR).

3. Reporting Essentials

- Indexed Dimensions, mass and function
- LV: EDV, SV, EF, longitudinal function , mass
- RV: EDV, SV, EF, longitudinal function
- Regional wall motion abnormalities
- Valve regurgitation
- Thickness of interatrial septum
- Report wall thickness (often concentric), indexed mass, and diastolic function
- Biatrial size—commonly enlarged

Tissue Characterization

- Native T1 and ECV:
  - Elevated native T1: >1,050 ms at 1.5T.
  - ECV often >0.40 in advanced disease.
- LGE Pattern:
  - Subendocardial or transmural LGE in a non-coronary distribution.
  - "Zebra pattern" or global subendocardial enhancement common in ATTR.
  - Difficulty nulling myocardium is a red flag.

Additional Findings

- Pericardial /pleural effusions (common, especially in ATTR).
- Thickened interatrial septum and valves.

4. Key Diagnostic Criteria for Cardiac Amyloidosis

Wall Thickness & Chamber Sizes

- Increased LV wall thickness, typically concentric
- Restrictive LV pattern – non dilated ventricles, preserved LV function, restrictive filling pattern, enlarged LA/LA
- Atrial septal hypertrophy - >6mm in 20% cases

Tissue Markers

- Native T1: Elevated in both AL and ATTR.
- ECV: Strongly elevated (>0.45 highly specific).
- LGE: Myocardial nulling difficult to achieve despite optimum technique
- Predominantly global subendocardial or transmural patter, PISR often necessary for visualisation
- Patchy subendocardial or transmural enhancement can also occur

Functional Indicators

- Diastolic dysfunction.

5. Tips & Tricks for Technologists

Difficulty Nulling Myocardium on LGE:

- Strong indicator of diffuse amyloid infiltration—use PSIR to avoid TI sensitivity.
    - On Look-locker images blood pool and myocardium cross zero (appear black) at similar timepoints
    - Cross-check skeletal muscle signal intensity while choosing optimal inversion time

Pre-contrast T1 & ECV Mapping:

- Ensure precise haematocrit measurement (same-day sample).
- Acquire pre- and post-contrast T1 maps in identical locations.

Look Beyond the LV:

- Biatrial enlargement, valve thickening, and pericardial, pleural effusion are supporting features.
- Right ventricular thickening and dysfunction may also be present.

6. Distinguishing AL vs ATTR:

- AL LV mass mildly increased, ATTR LV mass markedly increased

- AL higher native T1; often progresses more rapidly
- ATTR typically has more dramatic LGE and higher ECV. AL LGE often less extensive and often has a global subendocardial pattern. ATTR LGE often more extensive and more diffuse and transmural pattern.

7. Red Flags for Amyloidosis (vs HCM):

- Concentric vs asymmetric hypertrophy.
- Low voltage on ECG despite thick myocardium.
- Difficult LGE nulling, global subendocardial enhancement.

Reference:

Herzog, B. A., Greenwood, J. P., Plein, S., Garg, P., Haaf, P., & Onciul, S. (2017). Cardiovascular magnetic resonance pocket guide. Eur Soc Cardiol.