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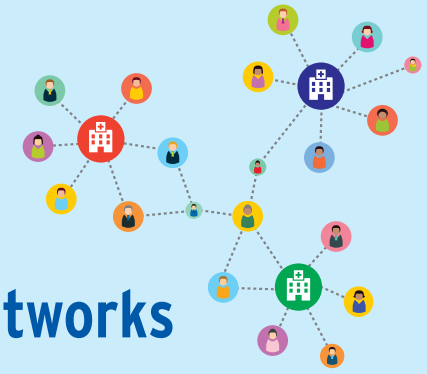
Building Effective Amyloidosis Care Networks



Cardiac Imaging in Amyloidosis: Tools for Early and Accurate Detection

Role of Imaging in Cardiac Amyloidosis[1]

- Imaging is a requisite component of the diagnostic algorithm for cardiac amyloidosis (CA) to identify patients who may have CA, confirm diagnosis, differentiate between amyloid types (AL vs ATTR), and monitor disease progression and treatment response
- Imaging findings differ between early and later stages of disease
  - **Early:** increased left ventricular ejection fraction (LVEF) to global strain (GLS) ratio, grade 1 scintigraphy
  - **Later:** wall thickness > 12 mm, cherry-on-top apical sparing pattern, diffuse late gadolinium enhancement (LGE), and grade 2-3 scintigraphy

Imaging Options[1,2,3]

Imaging Option	Findings	Strengths/Caveats
Electrocardiography (ECG)[4]	• Low voltage (46%–56%) and pseudoinfarct pattern (47%–60%) are common but nonspecific • Atrial fibrillation and conduction abnormalities are frequent	• Findings lack sensitivity and specificity
Echocardiography[5]	• Biventricular wall thickening, biatrial enlargement, thickened valves/interatrial septum, and pericardial effusion • Apical sparing strain pattern helps differentiate CA from other cardiomyopathies	• Widely available, first-line test to identify patients for further evaluation • Quantifies structure and function • Non-specific without strain imaging, particularly in early disease
Scintigraphy with SPECT[6,7]	• Detects myocardial uptake characteristic of CA • Heart-to-contralateral lung ratio > 1.5 or grade 2-3 is highly sensitive and specific for ATTR CA when AL is ruled out	• Must confirm uptake with SPECT to avoid blood pool artifact • Scintigraphically silent genetic variants exist (L58H, V30M, F84L, S97Y)
Cardiac Magnetic Resonance (CMR)[8]	• Increased native T1 and extracellular volume (ECV) • Diffuse subendocardial or transmural LGE	• Excellent functional and structural assessment • Identifies early disease and provides prognostic data • Contraindicated or limited use in patients with renal insufficiency or devices • Access may be limited

SPECT, single photon emission computed tomography

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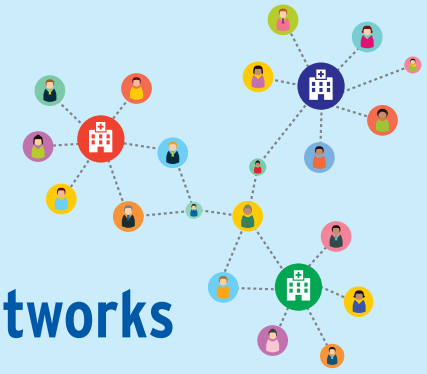

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Differentiating AL vs ATTR[6]

Signs & symptoms, ECG, echo or CMR suggestive of cardiac amyloidosis

^{99m}Tc-DPD/PYP/HMDP Scintigraphy with SPECT **&** **Haematologic tests** (serum free-light chain quantification & serum and urine immunofixation)

Scintigraphy grade 0, Haematologic tests -

AL/ATTR cardiac amyloidosis unlikely

If suspicion persists consider CMR followed by biopsy

Scintigraphy grade 1-3, Haematologic tests -

Grade 2-3

Cardiac ATTR amyloidosis

TTR genetic testing

ATTRwt / ATTRv

Grade 1

Histological confirmation (cardiac/extracardiac) **to diagnose**

Scintigraphy grade 0, Haematologic tests +

AL amyloidosis?

CMR negative

Amyloidosis unlikely

CMR + or inconclusive

Histological confirmation (cardiac/extracardiac) **to diagnose**

Scintigraphy grade 1-3, Haematologic tests +

Histological confirmation (usually cardiac) **to subtype**

- Positive scintigraphy, in combination with a negative monoclonal antibody screen, is diagnostic of ATTR-CM
- Biopsy with mass spectrometry is required to differentiate ATTR from AL in the presence of monoclonal protein in serum or urine
- When imaging findings are equivocal and clinical suspicion is high, biopsy is recommended

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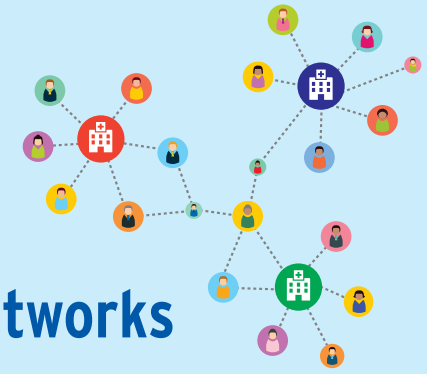

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Key Takeaways

- **ECG and echocardiography** are useful for initial screening but are not diagnostic and do not distinguish AL vs ATTR
- **Scintigraphy** with SPECT is highly specific for ATTR when AL is ruled out
- **CMR** provides tissue characterization and prognostic information as well as ability to monitor disease activity; does not differentiate AL vs ATTR
- **Always screen for monoclonal protein** before interpreting scintigraphy results
- **Educate patients** on the role of multimodal imaging for diagnosis and follow-up

References

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