

ATTR-CM NEWS-BEAT

Key steps to achieving a confirmed diagnosis of ATTR-CM

1. Building a suspicion of ATTR-CM¹⁻³

65+ year old Patient with HF and LV wall thickness ≥ 12mm



Clinical signs: (AL symptoms are

excluded)

- Cardiac:
- HF symptoms · Hypotension or normotensive if previous

- **Extracardiac:**
- hypertensive

- Dysautoniomia
- Polyneuropathy Bilateral carpal
- tunnel syndrome Ruptured biceps tendon
- Lumbar spinal stenosis
- Vitreous deposits
- · Family history



- between QRS voltage on ECG and LV wall thickness Pseudo-infarct
- ECG pattern Conduction
- abnormalities, and arrhythmias



Disproportionally elevated NT-

- proBNP to degree of HF Persisting
- elevated troponin levels



RV wall thickness

- increased atrial
- septum thickness Biatrial
- enlargement
- Thickened valves Diastolic
- dysfunction Granular sparkling of myocardium
- Pericardial
- effusion Reduced
- longitudinal strain with apical sparing pattern

Cardiac MRI: Subendocardial Late Gadolinium Enhancement

T1 values Increased extracellular

Elevated native

- volume Abnormal
- gadolinium kinetics



Early recognition of these signs should trigger diagnostic work-up for ATTR-CM

Amyloidosis (CA) Once CA is suspected, a timely, definitive diagnosis should be obtained. A diagnostic approach for patients with suspected cardiac amyloidosis should include testing for

2. Achieving a definitive diagnosis of Cardiac

monoclonal protein and scintigraphy or biopsy². Invasive diagnostic criteria apply to all forms of CA, whereas non-invasive criteria are for ATTR-CM only².

NONINVASIVE INVASIVE

Nuclear Scintigraphy



Nuclear scintigraphy in combination

with testing to rule out AL Amyloidosis

can diagnose ATTR-CM with high sensitivity and specificity.^{1,2} When signs and symptoms are suggestive of CA, diagnosis may be made noninvasively with nuclear

scintigraphy and testing to rule out AL amyloidosis*.4,5 * Ruling Out AL Amyloidosis should be made by three

tests screening for presence of monoclonal gammopathy:

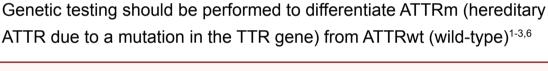
Serum Free Light-Chain (FLC) Assay, Serum and Urine Electrophoresis with immunofixation (SPIE, UPIE)²

Endomyocardial Biopsy (EMB)



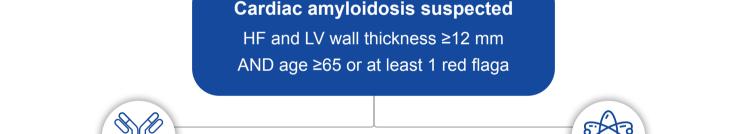
The gold standard for the diagnosis

of ATTR-CM with high sensitivity and specificity if specimens are collected from multiple sites and tested for amyloid deposits by Congo red staining. Identification of amyloid should be followed by classification of the amyloid protein by mass spectrometry, immunohistochemistry, or Immunoelectron Microscopy. However, EMB is not needed with a grade 2-3 positivity of scintigraphy with SPECT and normal hematologic tests. 1,2



Genetic Testing



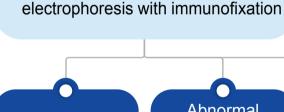


Normal

haematology

Grade 1 uptake

treatment of chronic heart failure



Both abnormal

Hematologic tests

Serum free light-chain assay

Serum and urine protein



Grade 2-3 uptake

ATTR

99mTc-PYP, DPD or HMDP

scintigraphy with SPECT

All normal

Histological

confirmation

to subtype

(usually cardiac)b

Positive or Negative inconclusive

Amyloidosis

unlikely

Abnormal

haematology

No myocardial

uptake

Cardiac amyloidosis on CMR imaging

> Histological confirmation of diagnosis

> > (cardiac/

extracardiac)c

Genetic counselling

In this video, Dr García-Pavía takes us on a deep dive into the diagnostic pathway for ATTR-

Amyloidosis

unlikely

Dr Pablo García-Pavía Hospital Universitario Puerta de Hierro Madrid, Spain

ATTR-CM: from suspicion to diagnosis

CM, highlighting four key scenarios in achieving a confirmed diagnosis.

Stay tuned for our upcoming "Cardiac imaging for ATTR-CM: the key to raising suspicion and diagnosing

Thank you for taking time to review this information. Please reach out if you have any questions or would like to discuss further.

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Heart failure vol. 12,9 (2019): e006075. doi:10.1161/CIRCHEARTFAILURE.119.006075

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