PLACE 2026
PLATFORM OF LABORATORIES FOR ADVANCES IN CARDIAC EXPERIENCE

MINICORSI
ROMA, 24 Giugno 2026

DIAGNOSI E GESTIONE DELL'AMILOIDOSI CARDIACA

Fisiopatologia ed inquadramento clinico (ATTRwt, ATTRv, AL).
Quando sospettare l'amiloidosi ATTR

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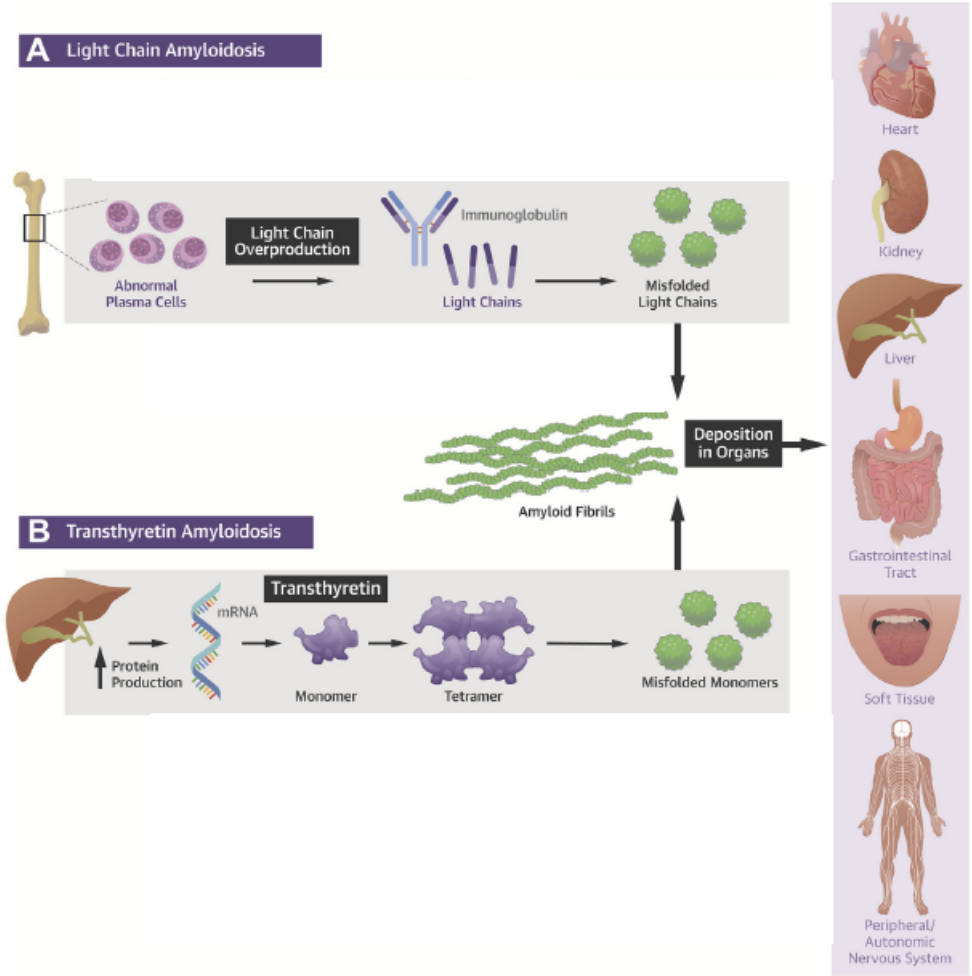
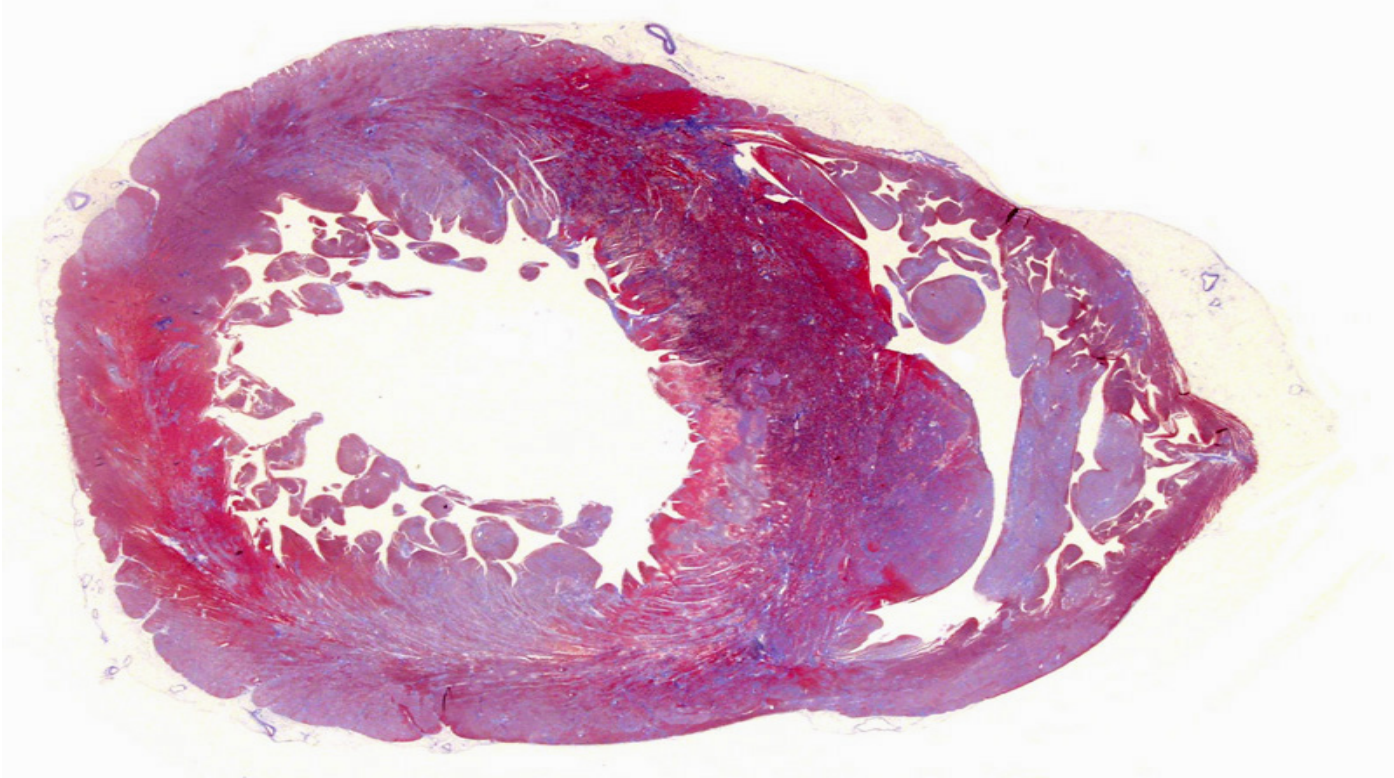
Pisa



Zhang, K.W. et al. J Am Coll Cardiol Basic Trans Science. 2019;4(3):438-48.



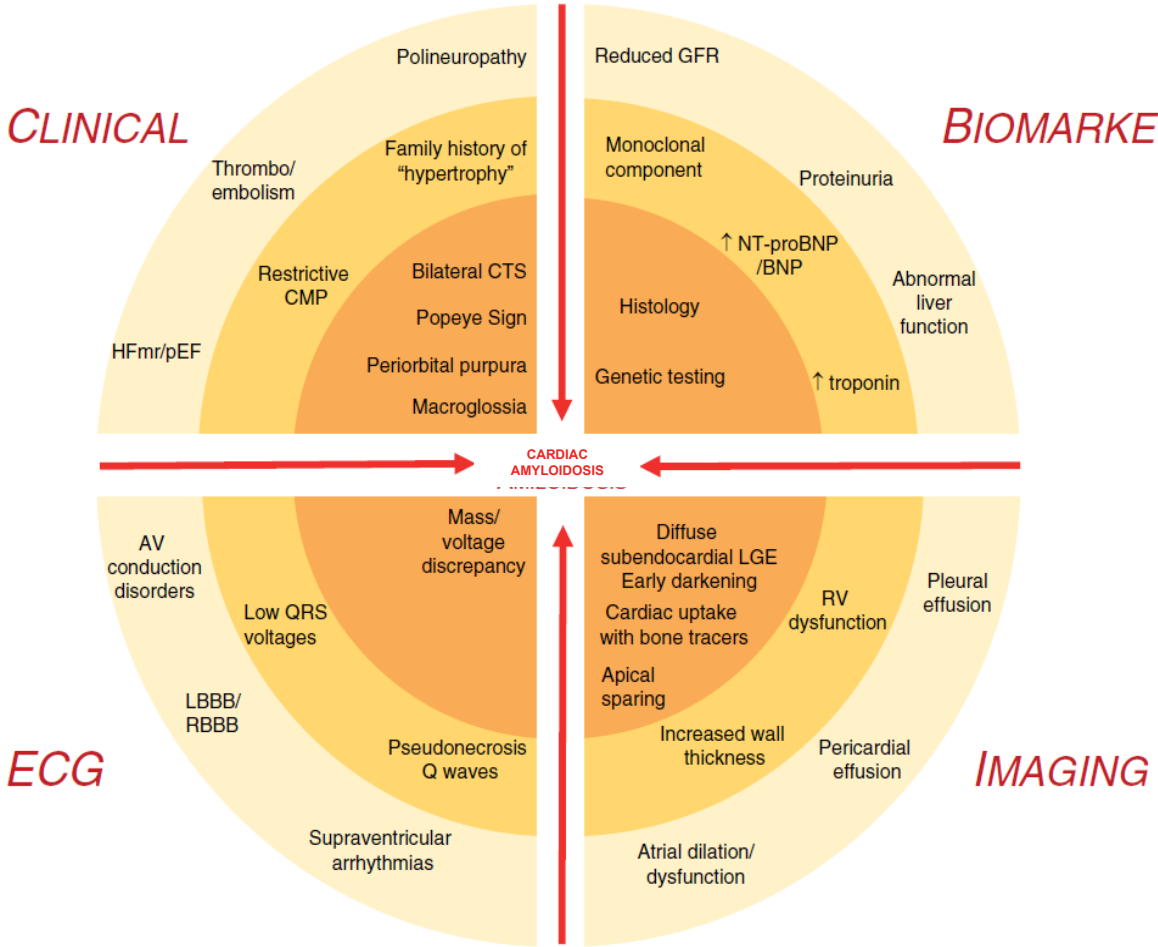
Left Ventricular Wall Thickness ≥ 12 mm

or ≥11 mm (W)

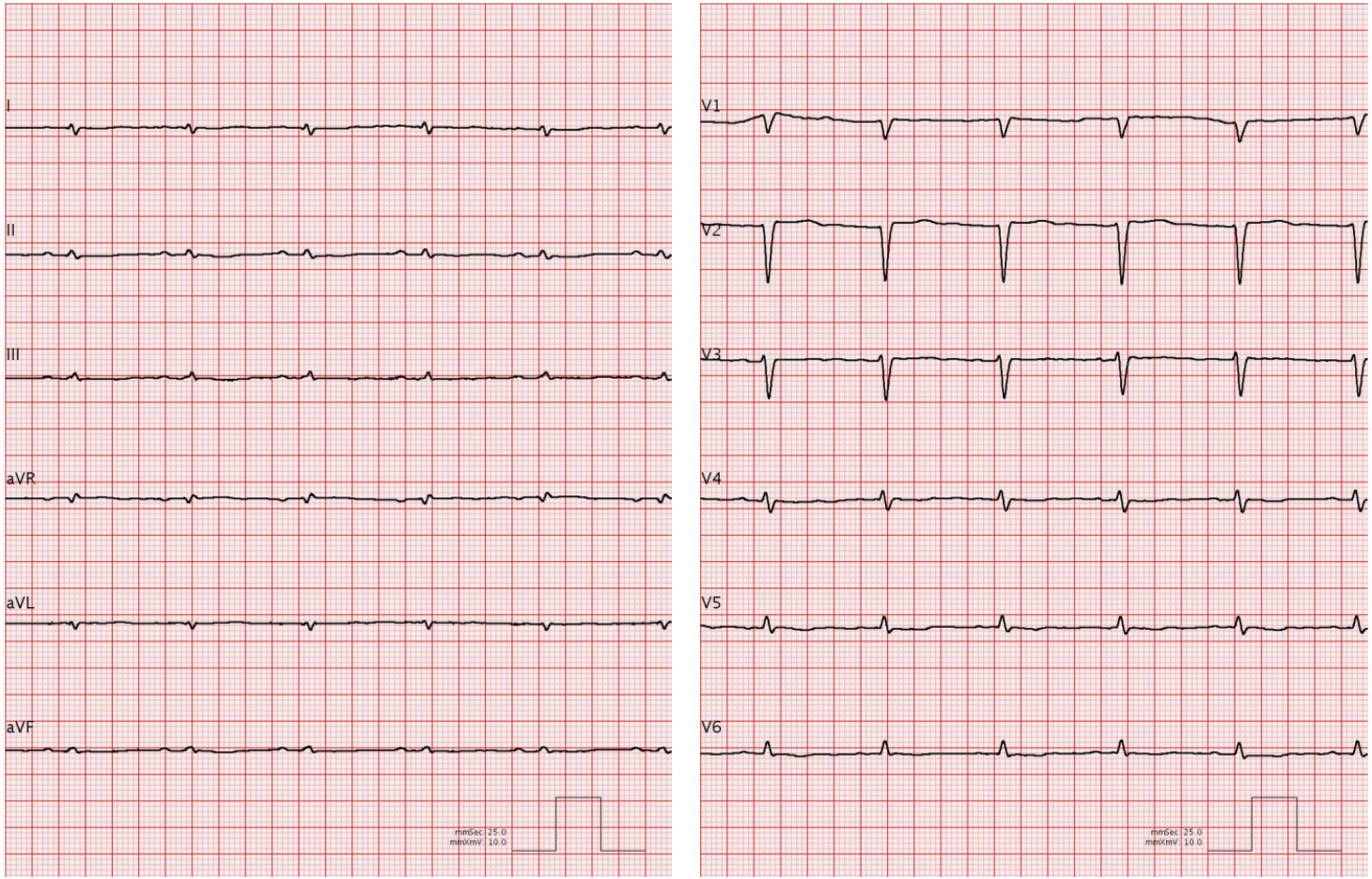
+ ≥1 of

- Heart failure in ≥ 65 years
- Aortic stenosis in ≥ 65 years
- Hypotension or normotensive if previously hypertensive
- Sensory involvement, autonomic dysfunction
- Peripheral polyneuropathy
- Proteinuria
- Skin bruising
- Bilateral carpal tunnel syndrome
- Ruptured biceps tendon
- Subendocardial/transmural LGE or increased ECV
- Reduced longitudinal strain with apical sparing
- Decreased QRS voltage to mass ratio
- Pseudo Q waves on ECG
- AV conduction disease
- Possible family history

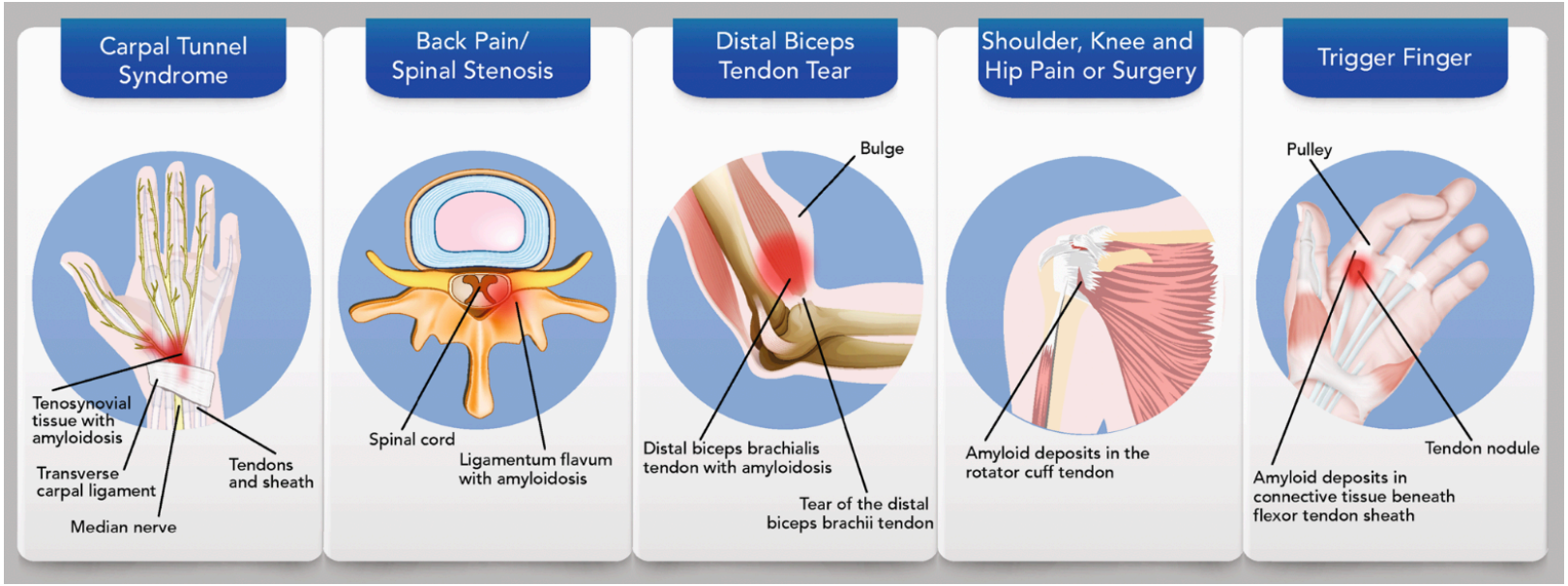
Garcia-Pavia P et al., *EJHF* 2021



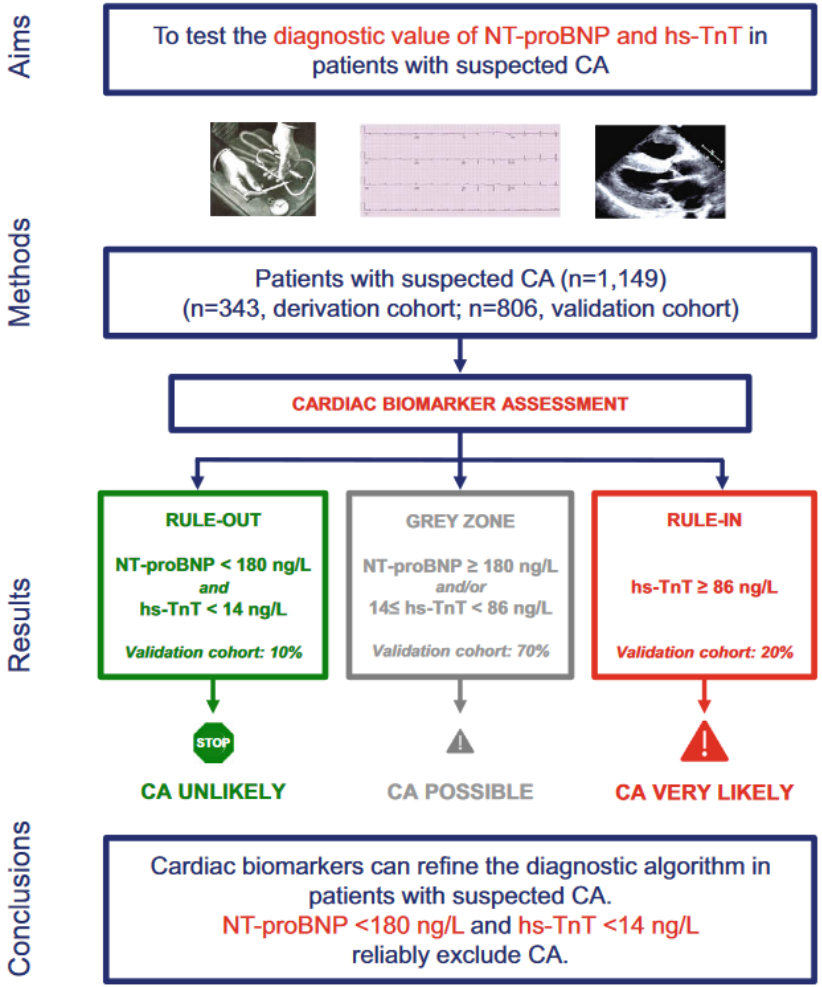
Vergaro G, Aimo A et al., *EJPC* 2020


I
II
III
aVR
aVL
aVF
V1
V2
V3
V4
V5
V6
mmSec: 25.0
mmXmV: 10.0
mmSec: 25.0
mmXmV: 10.0

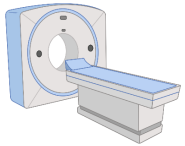
Musculoskeletal manifestations associated with CA



Nativi-Nicolau JN et al, 2021

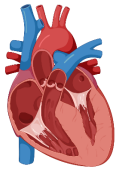


Vergaro G et al, 2022

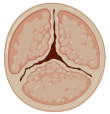


Bone scintigraphy
for non-cardiac reasons:
≥81 years: ~1.3% M, ~0.4% W

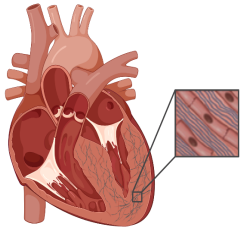
Autopsy in unselected
elderly individuals: 21%
(95% CI 7-39%)

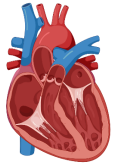


HFpEF: 12%
(95% CI 6-20%)
M 73% (39-100%)
77 years (66-86)
AL-CA 10% (0-40%)



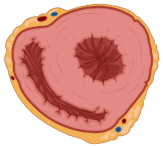
Aortic stenosis: 8%
(95% CI 5-13%)
M 67% (50-89%)
84 years (75-88)
AL-CA 2% (0-6%)





HFrEF/HFmrEF: 10%
(95% CI 6-15%)
M 100%
81 years (76-85)
AL-CA 0%

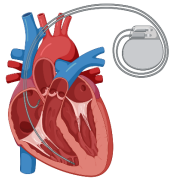
Prevalence of cardiac amyloidosis in screening studies



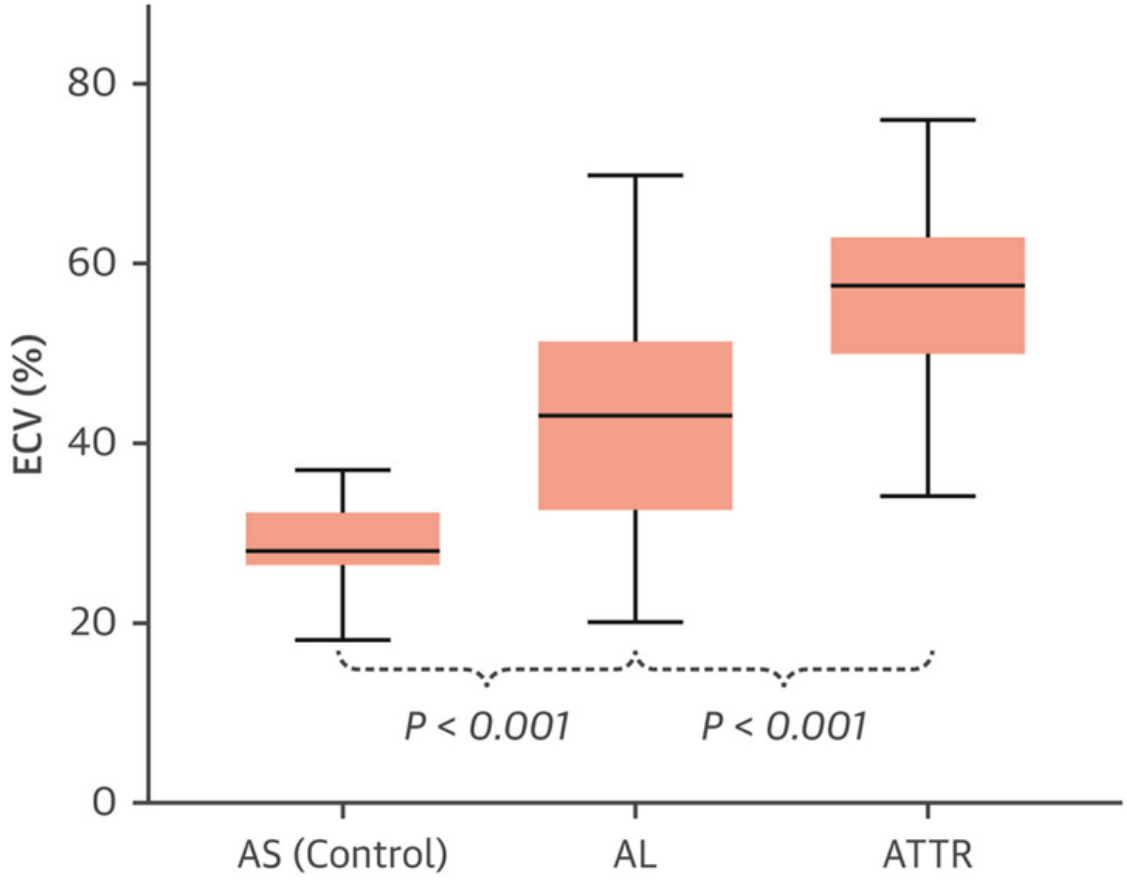
HCM: 7%
(95% CI 5-9%)
M 80% (73-87%)
74 years
AL-CA 0-9%

Surgery for carpal tunnel
syndrome: 7%
(95% CI 5-10%)
M 64% (33-100%)
76 years (73-79)
AL-CA 18% (0-33%)

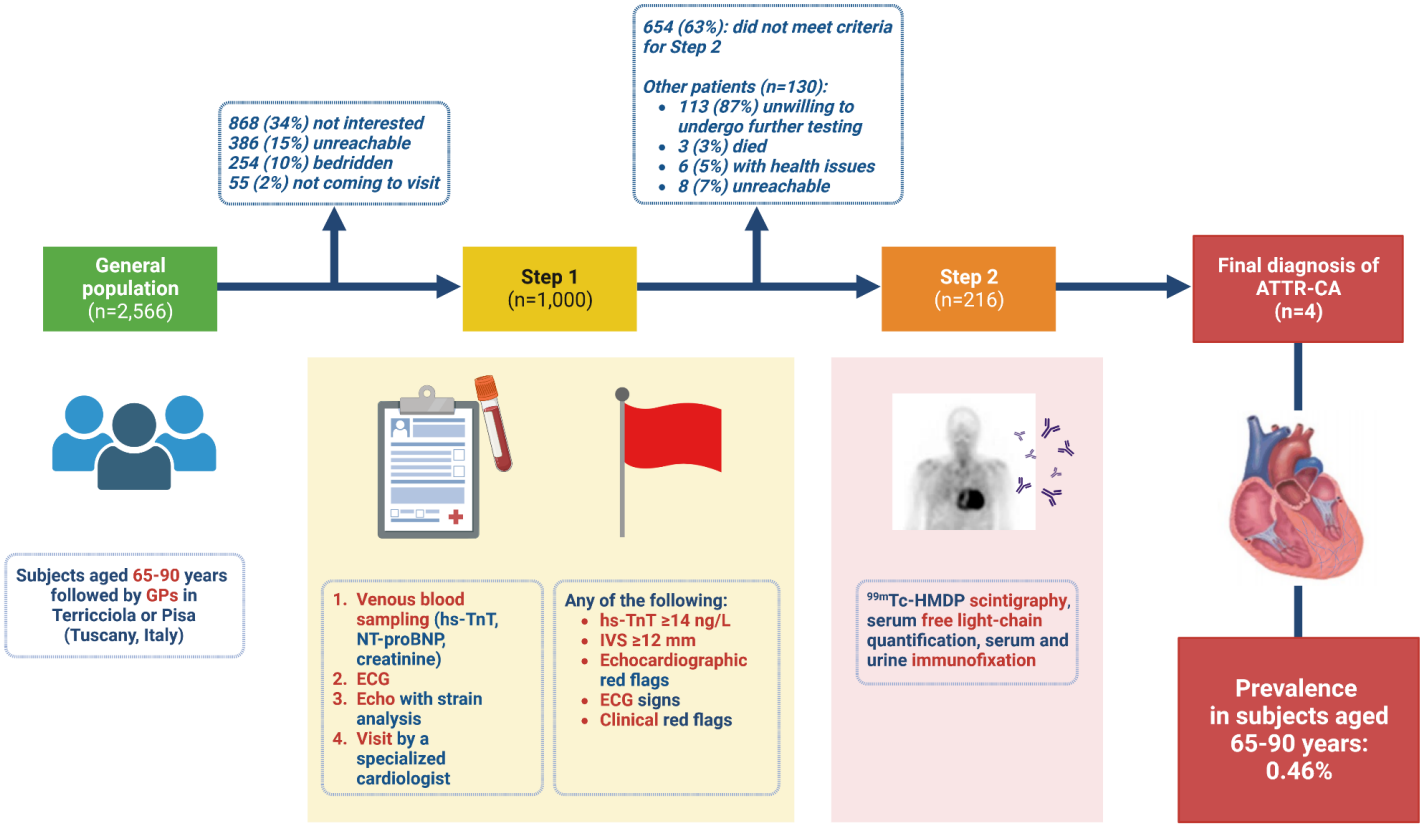
Conduction disorders: 2%
(95% CI 0-4%)
M 50%
90 years
AL-CA 0%



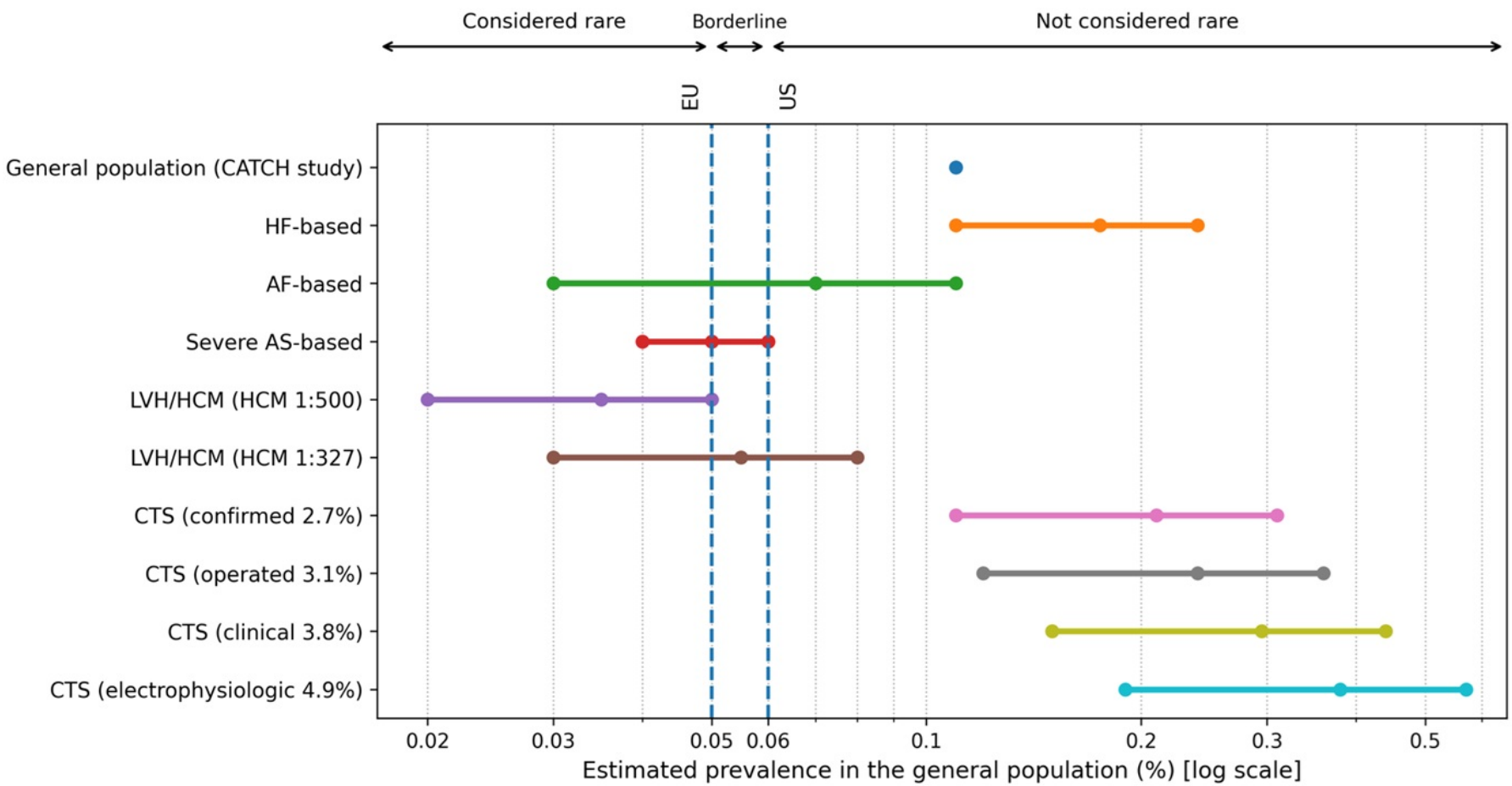
Rapezzi C, Aimo A et al., *EJHF* 2022



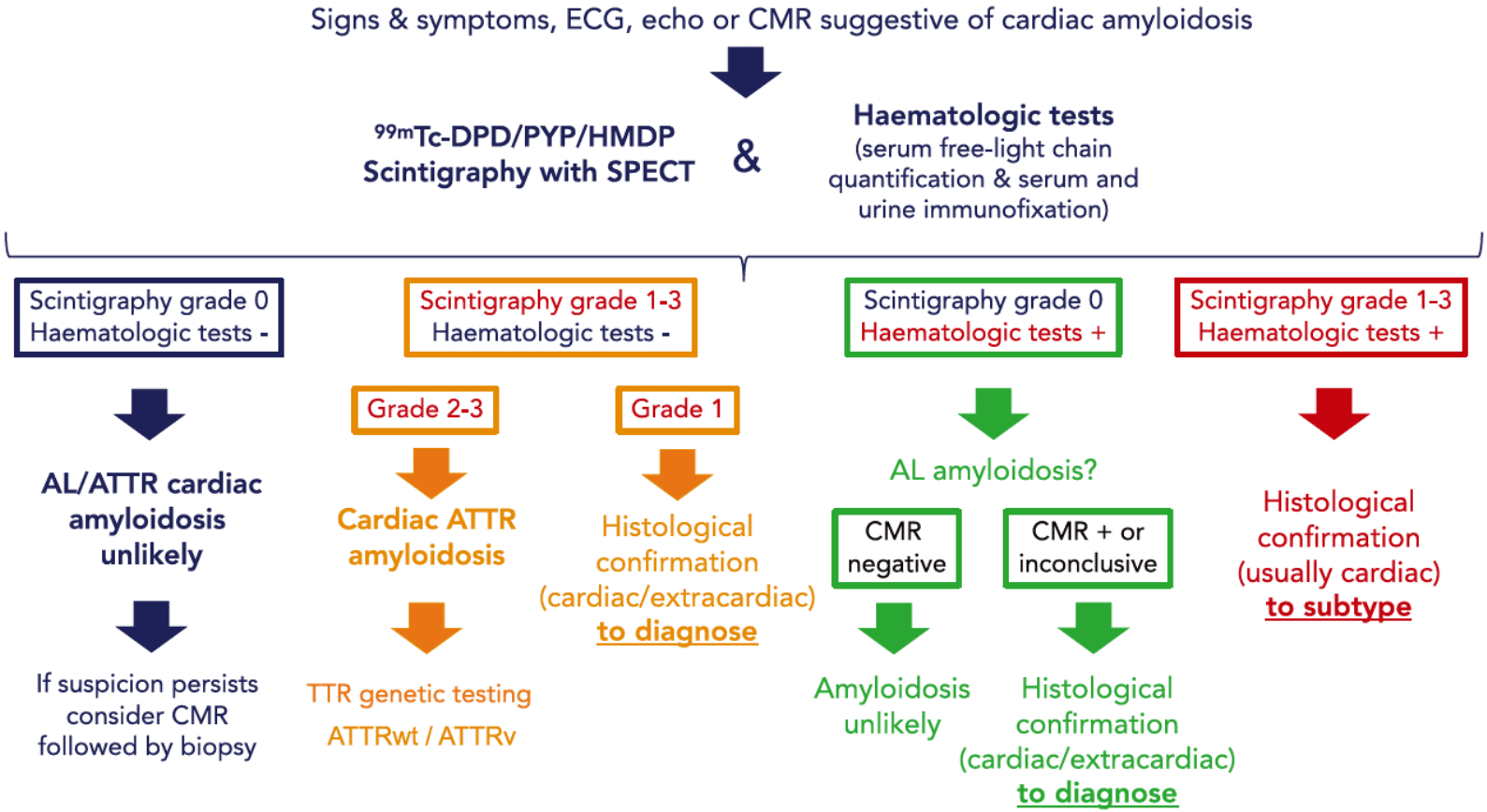
Gama F et al., *JACC CV Imag* 2022



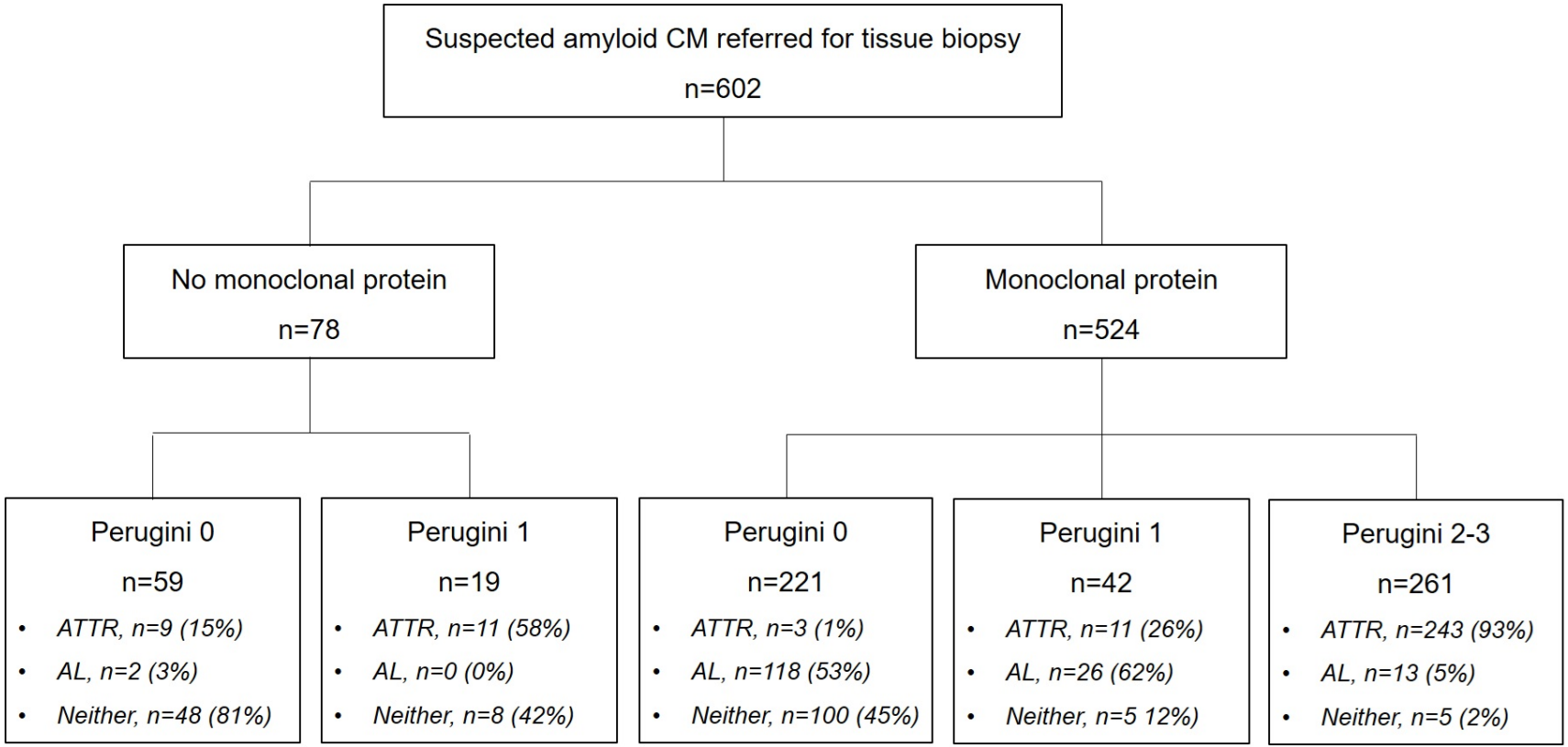
Aimo A et al., *EJPC* 2024



Aimo A et al., submitted

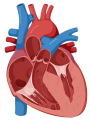


Garcia-Pavia P et al., *EJHF* 2021



Aimo A et al., under review

A pragmatic algorithm for prioritizing tissue biopsy in patients with suspected amyloid cardiomyopathy



Patients with suspected amyloid CM and a monoclonal protein

AGE

≤73 years → AL-CM more likely → Tissue biopsy ASAP

74-83 years
Requires integration of CTS and risk modifiers

AL score (0-7 points):
- Abnormal FLC ratio (+2)
- Positive urine immunofixation (+1)
- Proteinuria (+1)
- hs-troponin T ≥70 ng/L (+1)
- NT-proBNP ≥4,000 ng/L (+1)
- Perugini grade 0-1 (+1)

ATTR score (0–7 points):
- CTS: unilateral = **1**, bilateral = **2**
- Male sex = **1**
- IVS ≥15 mm = **1**
- Perugini 2–3 = 3

AL-favored: AL score ≥4 AND ATTR score ≤3
ATTR-favored: ATTR score ≥4 AND AL score ≤2

Need for tissue biopsy (urgency guided by AL- vs ATTR-favored pattern)

≥84 years → **Biopsy still required; AL-CM very unlikely (timing more flexible)**

Tissue biopsy and amyloid typing are required in all patients with a monoclonal protein; this framework guides *urgency* and *most likely type*.

Aimo A et al., under review

Take-home messages

- L'amiloidosi cardiaca va sospettata in caso di spessori parietali aumentati (soprattutto, ma non esclusivamente, in assenza di chiare cause) e red flag, tra cui: voltaggi ECG bassi o sproporzionati, biomarcatori cardiaci cronicamente elevati, sindrome del tunnel carpale (++ bilaterale), stenosi del canale lombare, rottura del tendine del bicipite, neuropatia periferica/autonomica, proteinuria o storia familiare.
- L'amiloidosi cardiaca è relativamente comune in pazienti con scompenso cardiaco a frazione d'eiezione preservata o stenosi aortica.
- In caso di sospetto diagnostico è fondamentale eseguire *sia* scintigrafia con difosfonati *che* ricerca della componente monoclonale.
- In assenza di componente monoclonale, una scintigrafia ossea positiva di grado 2–3 consente la diagnosi non invasiva di ATTR-CM; in tutti gli altri casi è necessaria una biopsia. Il test genetico è sempre necessario per distinguere ATTRwt da ATTRv.
- L'iter diagnostico deve concludersi con la conferma o l'esclusione della diagnosi e la tipizzazione (ATTRwt, ATTRv, AL).

Grazie per la vostra attenzione!

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