



AMI REVEALING?

AI-enhanced early diagnosis and referral of patients with unknown ATTR Amyloidosis

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ASUGI e Università di Trieste

Segni e sintomi dell'amiloidosi da Transtiretina ereditaria

• **LE RED FLAGS CARDIOLOGICHE**

Il sottoscritto **GIANFRANCO SINAGRA**
in qualità di (moderatore, relatore, formatore, tutor, docente)

ai sensi dell'art. 76 sul Conflitto di Interessi, comma 4 dell'Accordo Stato-Regioni del 2 febbraio 2017 e del paragrafo 4.5. del Manuale nazionale di accreditamento per l'erogazione di eventi ECM

dichiara

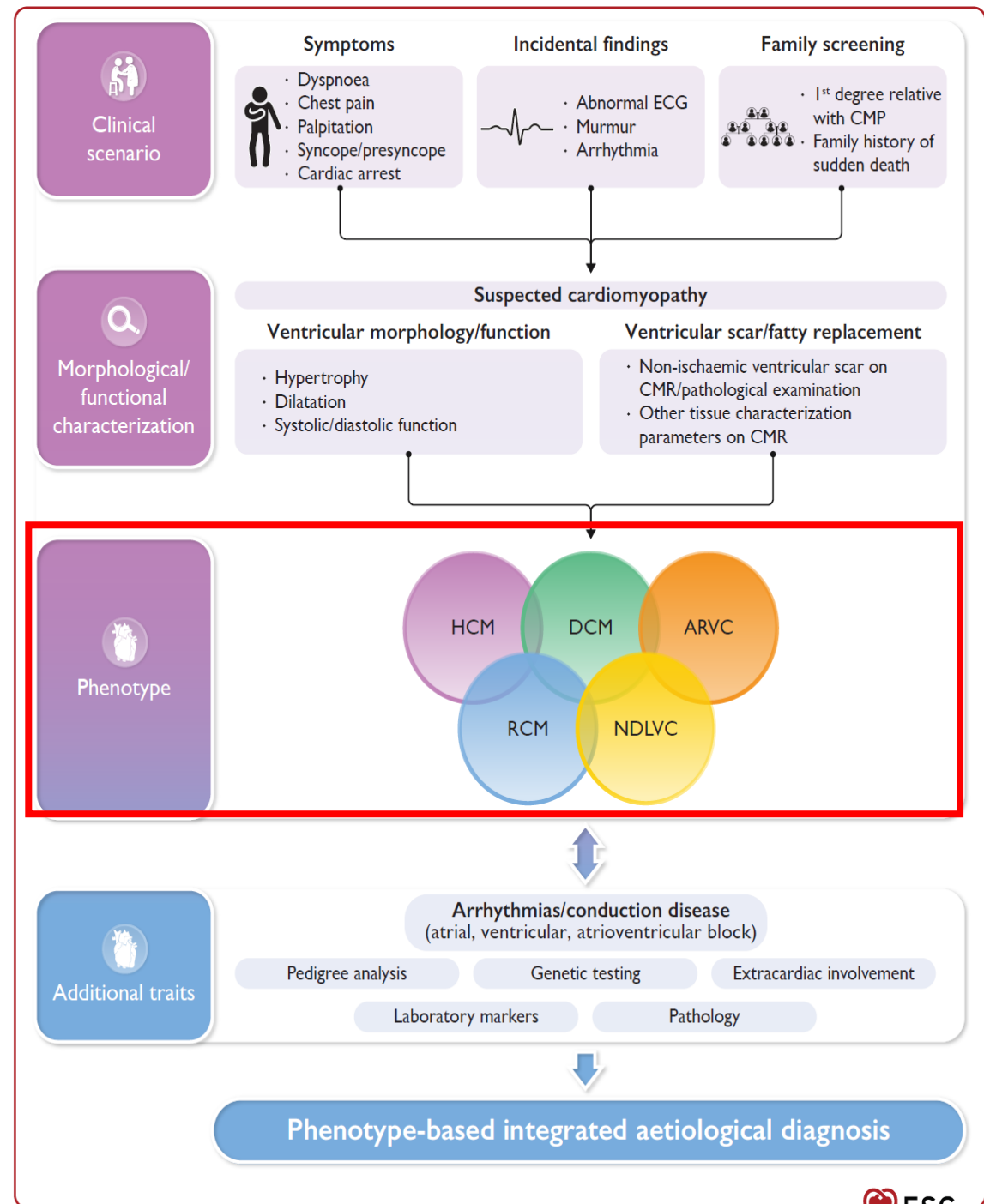
che negli ultimi due anni ha avuto i seguenti rapporti con soggetti portatori di interessi
in campo sanitario:

Novartis, Bayer, Astrazeneca, Bruno Farmaceutici, Boston Scientific, Biotronik, Menarini, Amgen,
Boehringer Ingelheim (**Relatore a congressi**)

Novartis, Impulse Dynamics, BMS (**Consulenze e Collaborazione scientifica occasionale**)

2023 ESC Guidelines for the management of cardiomyopathies

Developed by the task force on the management of cardiomyopathies of the European Society of Cardiology (ESC)



The patient pathway and clinical method

Recommendation Table 2 — Recommendations for diagnostic work-up in cardiomyopathies

Recommendations	Class ^a	Level ^b
It is recommended that all patients with suspected or established cardiomyopathy undergo systematic evaluation using <u>a multiparametric approach</u> that includes clinical evaluation, pedigree analysis, ECG, Holter monitoring, laboratory tests, and multimodality imaging. ⁶³	I	C
It is recommended that all patients with suspected cardiomyopathy undergo <u>evaluation of family history</u> and that a three- to four-generation family tree is created to aid in diagnosis, provide clues to underlying aetiology, determine inheritance pattern, and identify at-risk relatives. ^{64–66}	I	C

The **cardiomyopathy-oriented approach** is based on *interpreting clinical and instrumental findings to suspect and ultimately generate a phenotype-based aetiological diagnosis* to guide disease-specific management.

The **identification of red flags** is a crucial aspect of the initial work-up.

The aims are:

- (i) establishing and characterizing the presence of a cardiomyopathy phenotype;
- (ii) identifying the underlying *aetiological diagnosis*.

Transthyretin amyloid cardiomyopathy: An uncharted territory awaiting discovery

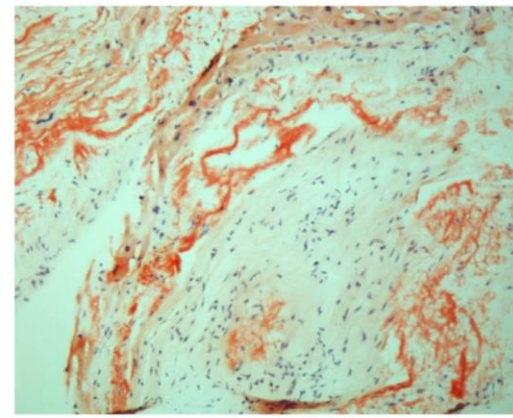
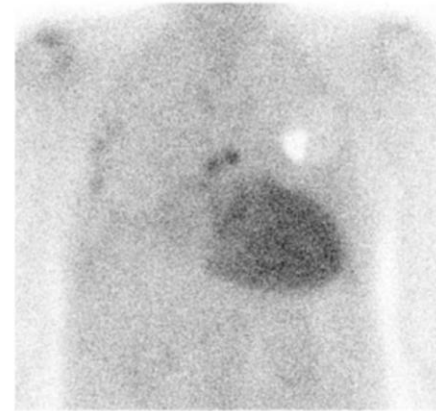
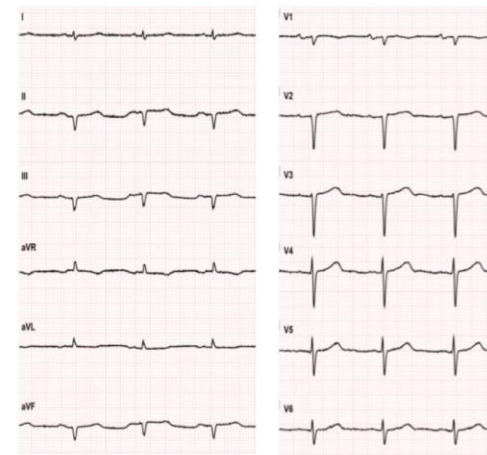
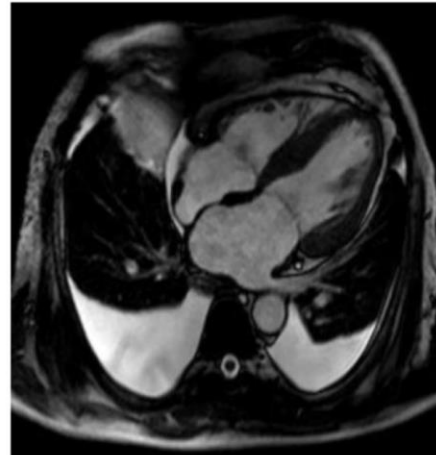
Aldostefano Porcari^a, Marco Merlo^a, Claudio Rapezzi^{b,c}, Gianfranco Sinagra^{a,*}

European Journal of Internal Medicine 82 (2020) 7–15

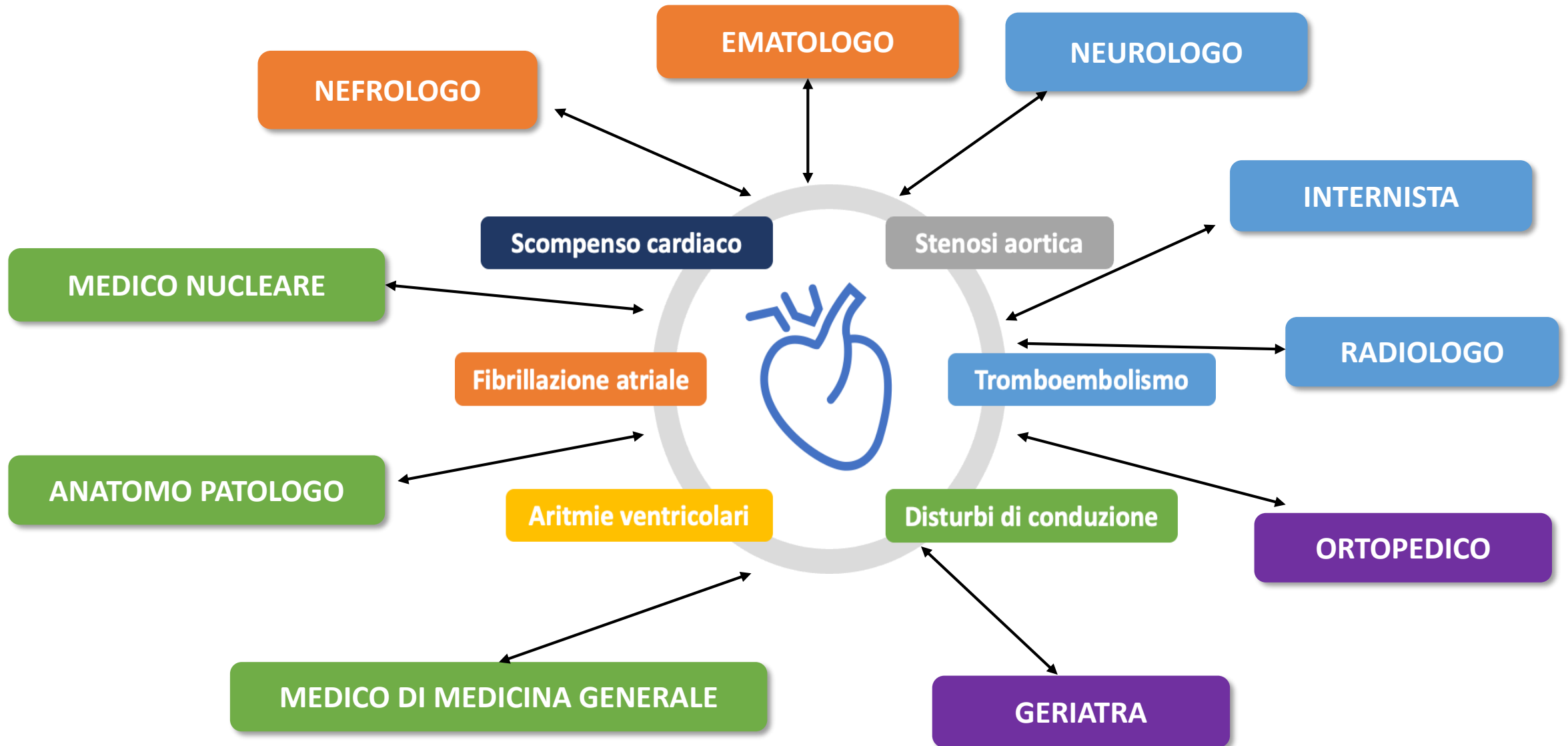


The comprehensive ‘red flag’ approach to TTR-AC.

Cardiological Evaluation	Findings of high suspicion for amyloid disease
Clinical Signs	HCM diagnosed after the sixth decade of life Supposed ‘hypertensive cardiomyopathy’ in elderly patients with normal BP values and no valvular disease Recurrent syncope Need for down-titration or discontinuation of antihypertensive therapy due to poor tolerability Intolerance of β -blockade in newly diagnosed HF Bowel dysfunction (constipation, diarrhoea) HF with systemic venous congestion Aortic valve stenosis in the elderly Periorbital purpura Macroglossia Bilateral CTS, atraumatic rupture of biceps tendon and LS stenosis Unexplained neuropathic pain, mostly in non-diabetic patients Orthostatic hypotension and erectile dysfunction due to autonomic neuropathy Vitreous deposits
	Persistent elevation in serum troponin values Increased NT-proBNP values (> 5000 pg/mL), disproportional to the clinical severity of HF Monoclonal gammopathy of undetermined significance (frequently coexisting) Low QRS voltages (peripheral and/or precordial leads) Discrepancies between severity of LV wall thickness and QRS voltages AV delay and/or blocks Pseudonecrosis
Laboratory	
ECG	
Echocardiography	Diffuse LV cardiac hypertrophy with non-dilated LV Coexisting RV hypertrophy Restrictive diastolic filling pattern Paradoxical low-flow low-gradient aortic valve stenosis Pericardial effusion Increased thickness of the interatrial septum and AV valves Granular sparkling appearance of the myocardium Reduced LV longitudinal strain with ‘apical sparing’ pattern



L'APPROCCIO ALL'AMILOIDOSI CARDIACA E' FONDATAO SUL "SOSPETTO" DI MALATTIA E RICHIEDE UN LAVORO DI SQUADRA



Uomo 57->64 anni

APR: ipercolesterolemia, IRC (**GFR 53 ml/min**), **tendinopatia** capo lungo del bicipite, sindrome del **tunnel carpale bilaterale** operato, **ipotensione ortostatica** (mai iperteso)

- **2016** Accesso in PS per **palpitazioni e discomfort toracico** → indagini «negative»;

*ECG: RS, **BAV I, EAS**; *Ecocardio: FE 60%, **SIV 13 mm**

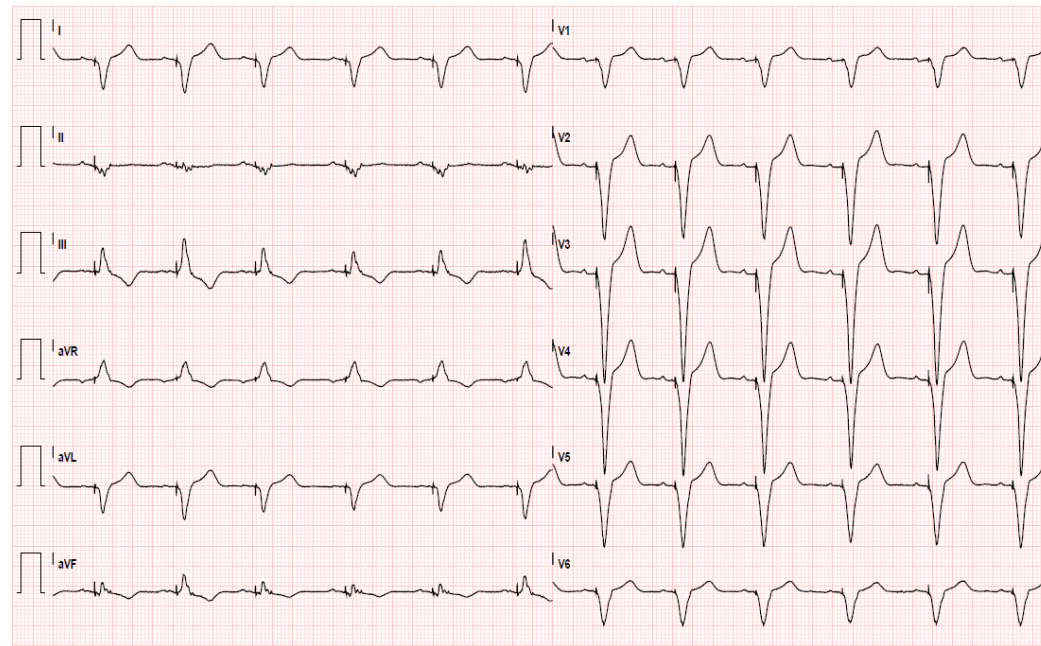
- Successivamente sintomatico per **dispnea** da sforzo e astenia; non diagnosi eziologica;

- **01/2023** NYHA III, *Eco: **SIV 20 mm, PP 18 mm, FE 40%**, pattern restrittivo

64 anni

- ***CoroTC**: NON malattia coronarica significativa (IVA media 40%, CDx prox 30%).
- ***CardioRM**: **SIV 29 mm**, ipocinesia globale, LGE diffuso transmurale nei segmenti basali e subendocardico in sede medio-apicale e dei m. papillari → *suggestiva per AC*
- ***Scintigrafia con traccianti ossei**: **Perugini 2** (NON componente monoclonale)
- * **Test genetico**: negativo per mutazioni TTR

AMILOIDOSI DA TRANSTIRETINA WILD-TYPE



Scintigrafia: Perugini 2

SIV 30 mm



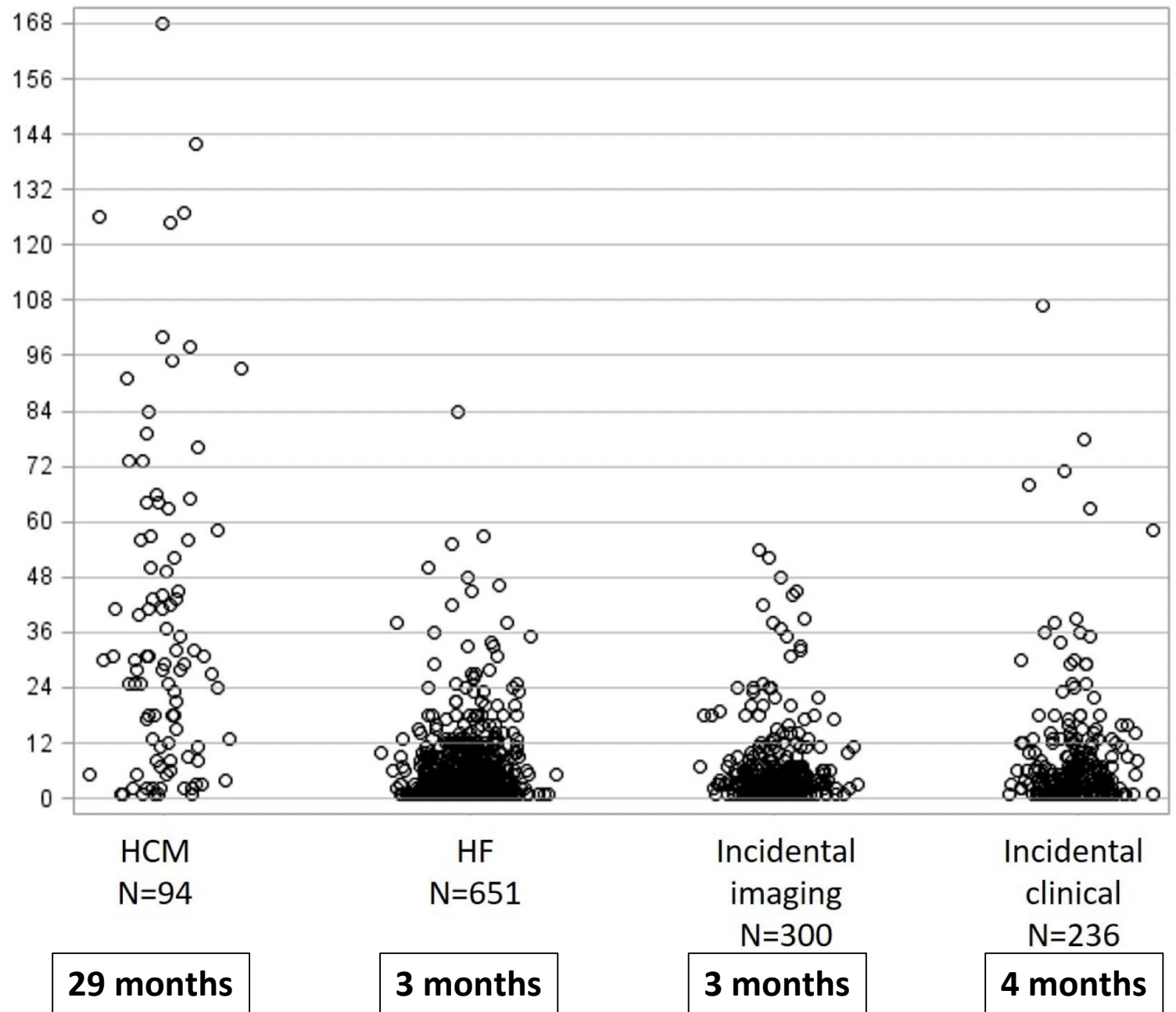
Severa disfunzione biventricolare, pattern restrittivo

Diagnostic pathways to wild-type transthyretin amyloid cardiomyopathy: a multicentre network study

Giacomo Tini^{1,2}, Paolo Milani^{3,4}, Mattia Zampieri⁵, Angelo G. Caponetti^{6,7}, Francesca Fabris^{3,4}, Andrea Foli⁴, Alessia Argirò⁵, Carlotta Mazzoni⁵, Christian Gagliardi^{6,8}, Simone Longhi^{6,8}, Giulia Saturi^{6,7}, Giuseppe Vergaro^{9,10}, Alberto Aimò^{9,10}, Domitilla Russo², Guerino G. Varrà¹¹, Matteo Serenelli¹², Gioele Fabbri¹², Laura De Michieli¹³, Giuseppe Palmiero¹⁴, Giuseppe Ciliberti^{15,16}, Samuela Carigi¹⁷, Eugenio Sessarego¹, Giulia E. Mandoli¹⁸, Giulia Ricci Lucchi¹⁹, Valeria Rella²⁰, Enrico Monti²¹, Elisa Gardini²¹, Michela Bartolotti²², Lia Crotti^{20,23}, Elisa Merli¹⁹, Roberta Mussinelli⁴, Pier Filippo Vianello¹, Matteo Cameli¹⁸, Francesca Marzo¹⁷, Federico Guerra^{15,16}, Giuseppe Limongelli^{8,14}, Alberto Cipriani^{13,24}, Stefano Perlini^{3,4}, Laura Obici⁴, Federico Peretto⁵, Camillo Autore^{2,25}, Italo Porto^{1,26}, Claudio Rapezzi^{12,27}, Gianfranco Sinagra^{8,11}, Marco Merlo^{8,11}, Beatrice Musumeci², Michele Emdin^{9,10}, Elena Biagini^{6,8}, Francesco Cappelli⁵, Giovanni Palladini^{3,4}, and Marco Canepa^{1,26*}



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doi:10.1002/ejhf.2823



Time to diagnosis

29 months

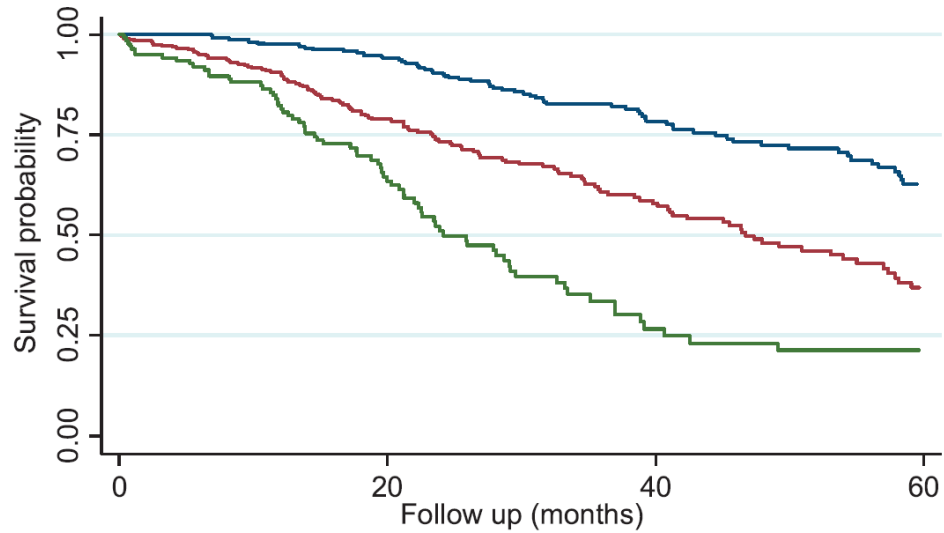
3 months

3 months

4 months

A new staging system for cardiac transthyretin amyloidosis

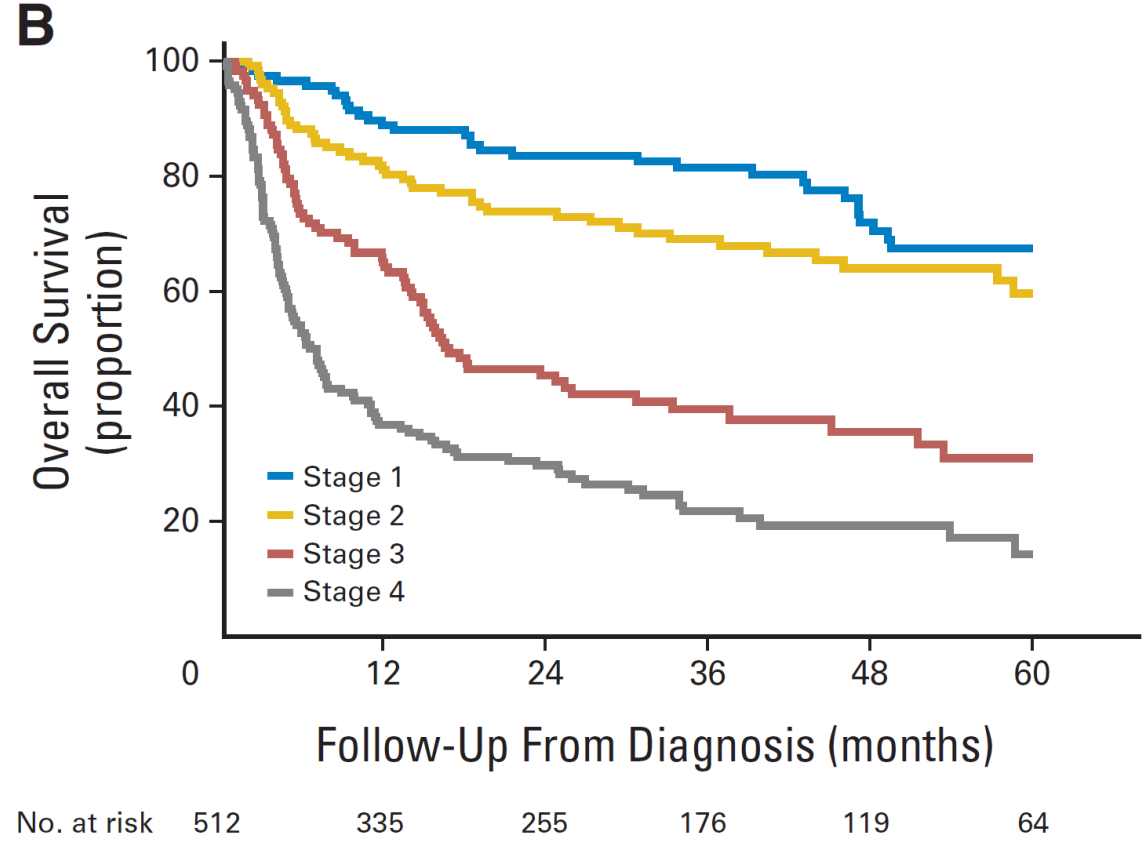
Staging of cardiac ATTR amyloidosis at diagnosis using NT-proBNP and eGFR



Number at risk	0	20	40	60
Stage I	393	254	117	58
Stage II	334	181	78	30
Stage III	142	60	15	8

- eGFR < 45 ml/min/1.73m²
- NT-proBNP > 3000 ng/L

Revised Prognostic Staging System for Light Chain Amyloidosis Incorporating Cardiac Biomarkers and Serum Free Light Chain Measurements



No. at risk	512	335	255	176	119	64
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Table 2. Results of Univariate and Multivariate Analyses of Various Prognostic Factors

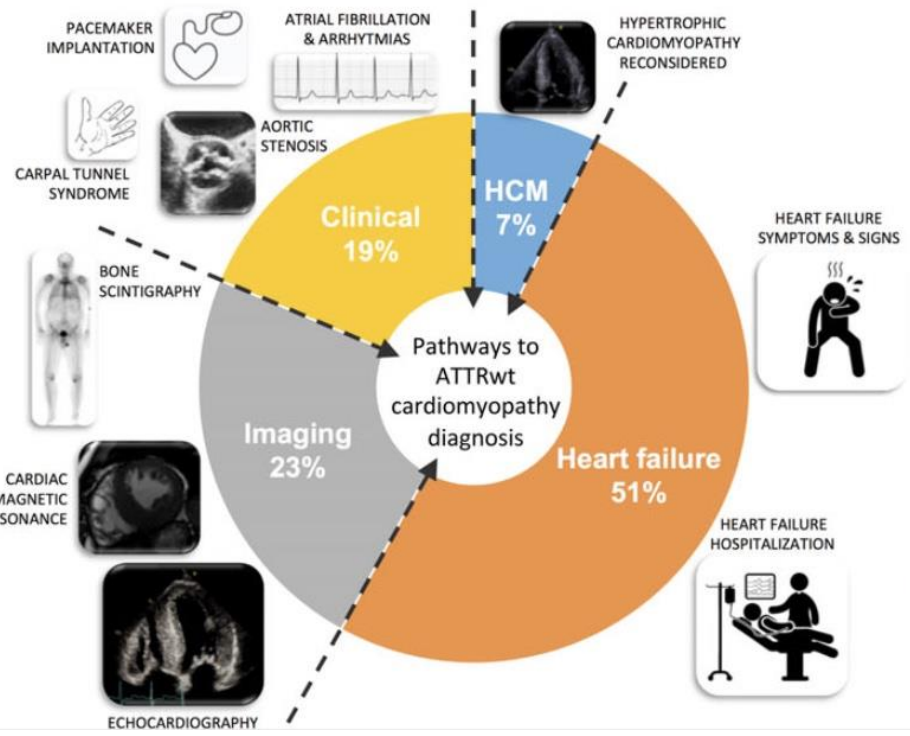
Prognostic Factor	Comparison	No. of Patients*	Univariate		Multivariate 1†		Multivariate 2‡	
			Risk Ratio	P	Risk Ratio	P	Risk Ratio	P
FLC-diff, mg/dL	> 18 v ≤ 18	758	1.6	< .001	1.4	.01	1.4	.002
Plasma cells, %	> 10 v ≤ 10	707	1.5	< .001	1.2	.2	NI	NA
PCLI, %	> 0 v 0	541	1.3	.009	1.3	.09	NI	NA
β2-microglobulin, mg/dL	> 3 v ≤ 3	720	1.9	< .001	1.5	< .01	NI	NA
Circulating plasma cells	Yes v no	293	1.5	.08	NI	NA	1.2	.1
cTnT, ng/mL	> 0.03 v ≤ 0.03	810	3.0	< .001	NI	NA	2.4	< .001
NT-proBNP, pg/mL	> 1,800 v ≤ 1,800	810	2.3	< .001	NI	NA	1.4	.004

Diagnostic pathways to wild-type transthyretin amyloid cardiomyopathy: a multicentre network study

Giacomo Tini,, Claudio Rapezzi, Gianfranco Sinagra, ...Marco Canepa

1,281 patients (mean age 78 years, 89% males) from 17 Italian centers between Jan 2017 and Dec 2021

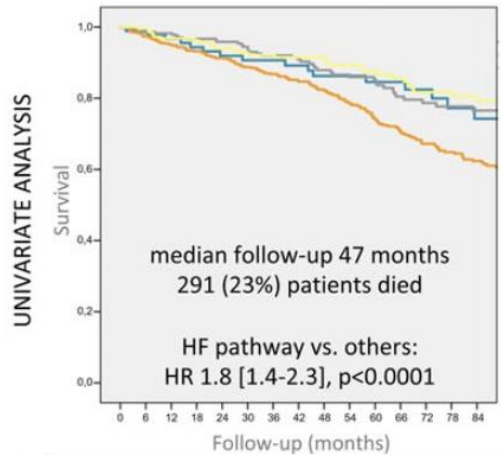
1. Prevalence



2. Characteristics

- HCM pathway (n=94):**
 - 75±7 years, 93% males
 - Red flags lead to case re-evaluation
 - 37% previous HF hospitalization
 - 16% NYHA III-IV, 33% CKD, 64% AFib
- Heart failure pathway (n=651):**
 - 79±7 years, 87% males
 - 61% during HF hospitalization
 - 52% had LVEF≥50%
 - 44% NYHA III-IV, 50% CKD, 65% AFib
- Incidental imaging pathway (n=300):**
 - 77±7 years, 92% males
 - 74% during echocardiography for routine evaluation
 - 13% NYHA III-IV, 38% CKD, 51% AFib
- Incidental clinical pathway (n=236):**
 - 78±7 years, 90% males
 - 94% cardiovascular evaluation including AFib, PM, arrhythmias
 - 15% NYHA III-IV, 38% CKD, 70% AFib

3. Prognosis



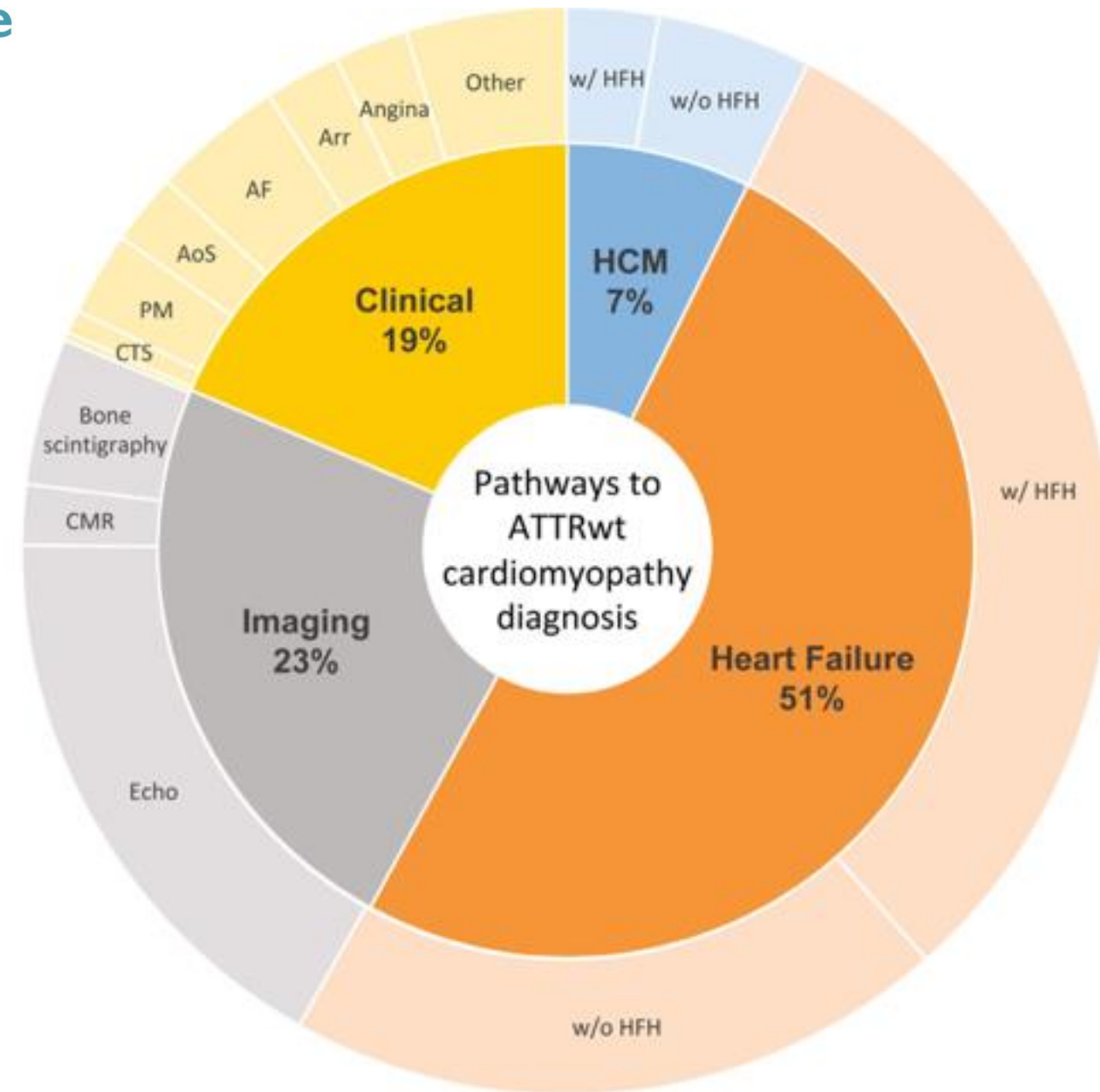
MULTIVARIATE ANALYSIS

	HR	p value
HF vs. other pathways	-	-
Age at diagnosis	1.1 [1.0-1.1]	<0.0001
NYHA III-IV	2.2 [1.7-2.7]	<0.0001
Hypertension	1.4 [1.1-1.9]	0.01
Diabetes mellitus	1.5 [1.1-2.0]	0.01
CKD	1.4 [1.1-1.8]	0.01
Cancer (active)	1.9 [1.2-2.9]	0.01

Diagnostic pathways to wild-type transthyretin amyloid cardiomyopathy: a multicentre

network study

Giacomo Tini^{1,2}, Paolo Milani^{3,4}, Mattia Zampieri⁵, Angelo G. Caponetti^{6,7}, Francesca Fabris^{3,4}, Andrea Foli⁴, Alessia Argirò⁵, Carlotta Mazzoni⁵, Christian Gagliardi^{6,8}, Simone Longhi^{6,8}, Giulia Saturi^{6,7}, Giuseppe Vergaro^{9,10}, Alberto Aimò^{9,10}, Domitilla Russo², Guerino G. Varrà¹¹, Matteo Serenelli¹², Gioele Fabbri¹², Laura De Michieli¹³, Giuseppe Palmiero¹⁴, Giuseppe Ciliberti^{15,16}, Samuela Carigi¹⁷, Eugenio Sessarego¹, Giulia E. Mandoli¹⁸, Giulia Ricci Lucchi¹⁹, Valeria Rella²⁰, Enrico Monti²¹, Elisa Gardini²¹, Michela Bartolotti²², Lia Crotti^{20,23}, Elisa Merli¹⁹, Roberta Mussinelli⁴, Pier Filippo Vianello¹, Matteo Cameli¹⁸, Francesca Marzo¹⁷, Federico Guerra^{15,16}, Giuseppe Limongelli^{8,14}, Alberto Cipriani^{13,24}, Stefano Perlini^{3,4}, Laura Obici⁴, Federico Perfetto⁵, Camillo Autore^{2,25}, Italo Porto^{1,26}, Claudio Rapezzi^{12,27†}, Gianfranco Sinagra^{8,11}, Marco Merlo^{8,11}, Beatrice Musumeci², Michele Emdin^{9,10}, Elena Biagini^{6,8}, Francesco Cappelli⁵, Giovanni Palladini^{3,4}, and Marco Canepa^{1,26*}



Diagnosis and treatment of cardiac amyloidosis: a position statement of the ESC Working Group on Myocardial and Pericardial Diseases



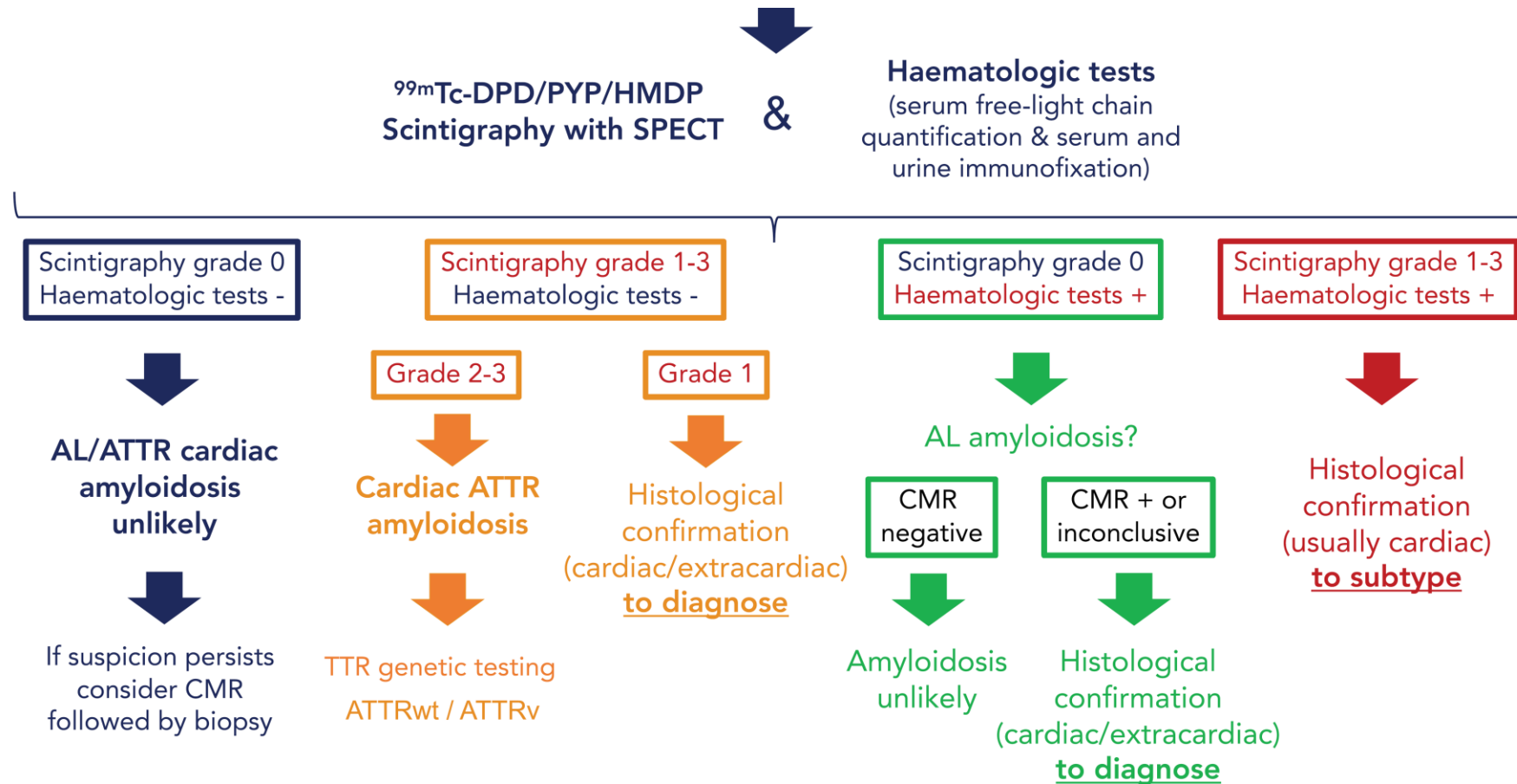
ESC

European Society
of Cardiology

European Heart Journal (2021) 42, 1554–1568

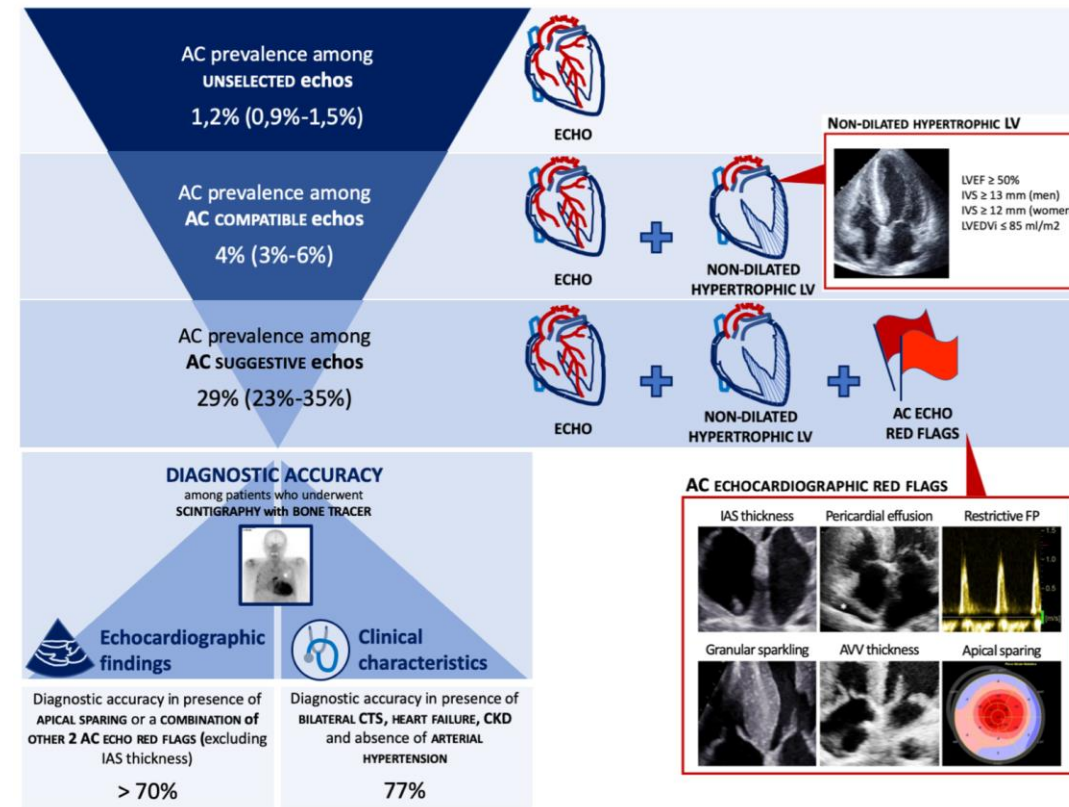
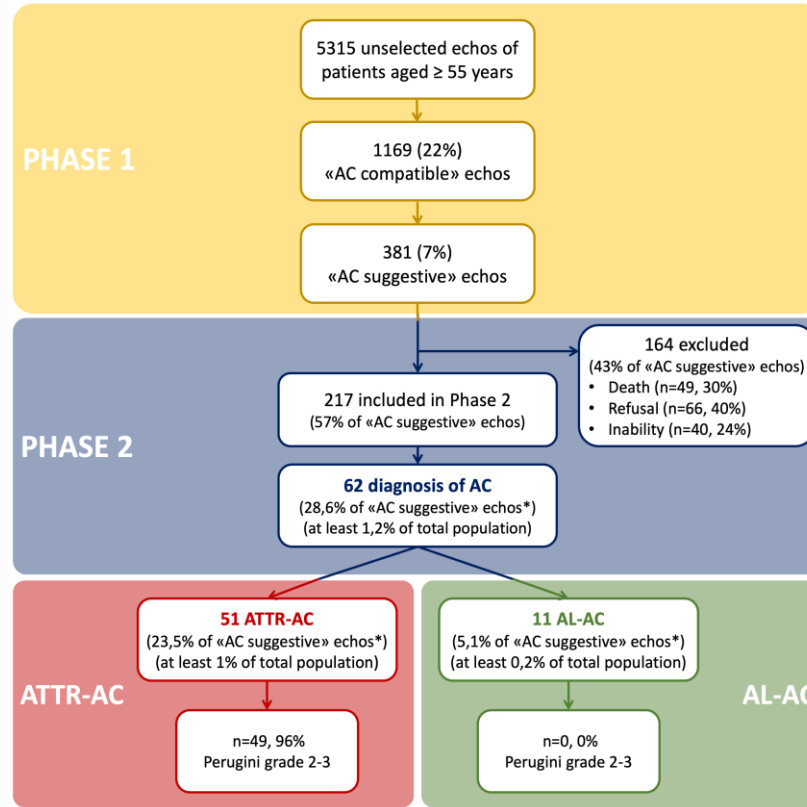
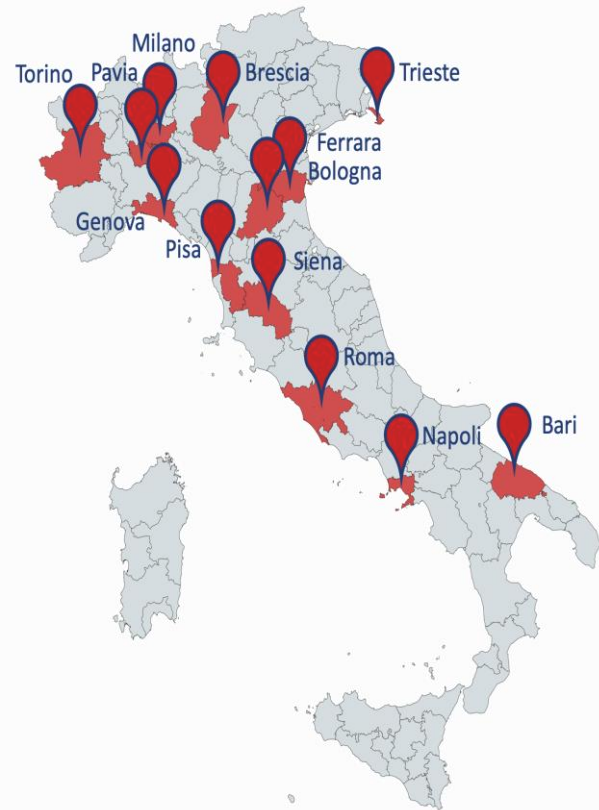
doi:10.1093/eurheartj/ehab072

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



Unmasking the prevalence of amyloid cardiomyopathy in the real world: results from Phase 2 of the AC-TIVE study, an Italian nationwide survey

Marco Merlo, Aldostefano Porcari, ... Claudio Rapezzi, Gianfranco Sinagra



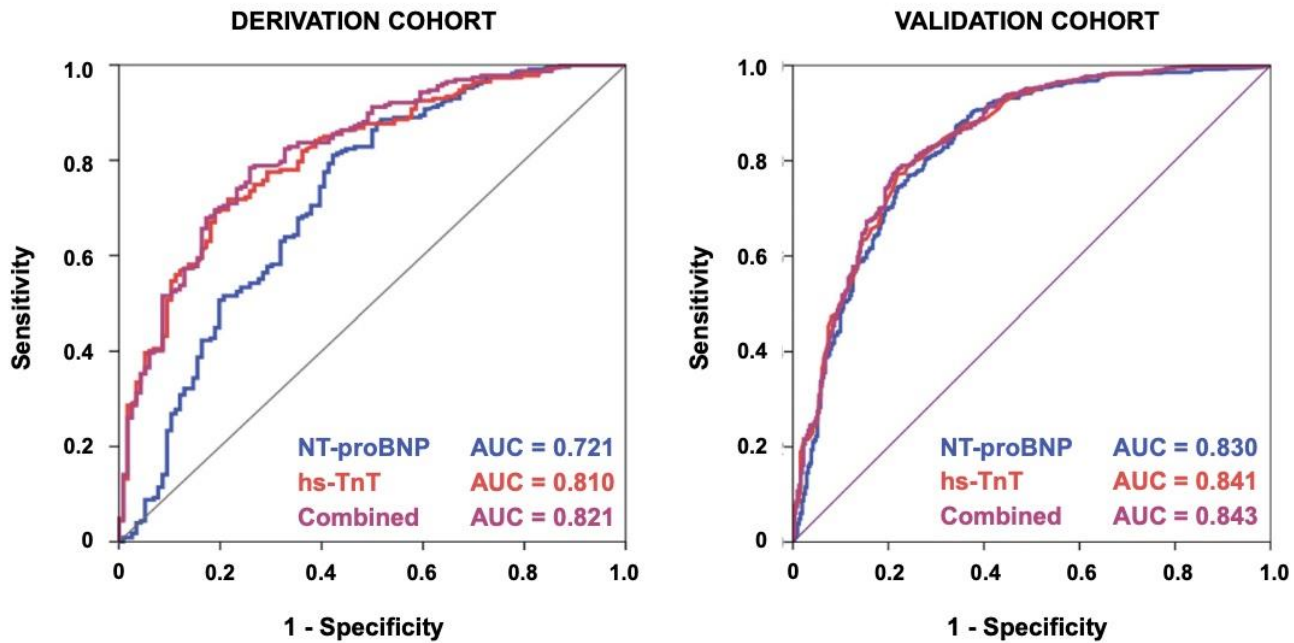
Prevalence of 29% in Italy among adults with echo red flags of AC

N-terminal pro-B-type natriuretic peptide and high-sensitivity troponin T hold diagnostic value in cardiac amyloidosis

Giuseppe Vergaro^{1,2*}, Vincenzo Castiglione¹, Alberto Aimo^{1,2}, Concetta Prontera², Silvia Masotti², Veronica Musetti², Martin Nicol³, Alain Cohen Solal³, Damien Logeart³, Georgios Georgiopoulos^{4,5}, Vladyslav Chubuchny², Alberto Giannoni^{1,2}, Aldo Clerico^{1,2}, Gabriele Buda⁶, Kiara N. Patel⁷, Yousuf Razvi⁷, Rishi Patel⁷, Ashutosh Wechalekar⁷, Helen Lachmann⁷, Philip N. Hawkins⁷, Claudio Passino^{1,2}, Julian Gillmore⁷, Michele Emdin^{1,2}, and Marianna Fontana⁷

European Journal of Heart Failure (2023)
doi:10.1002/ejhf.2769

Diagnosis of CA in patients with clinical suspicion



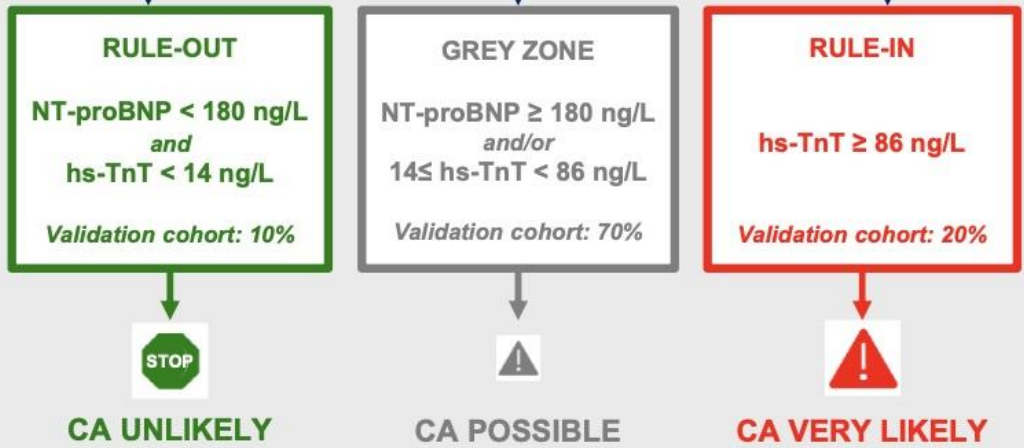
Aims
Methods
Results
Conclusions

To test the **diagnostic value of NT-proBNP and hs-TnT** in patients with suspected CA



Patients with suspected CA (n=1,149)
(n=343, derivation cohort; n=806, validation cohort)

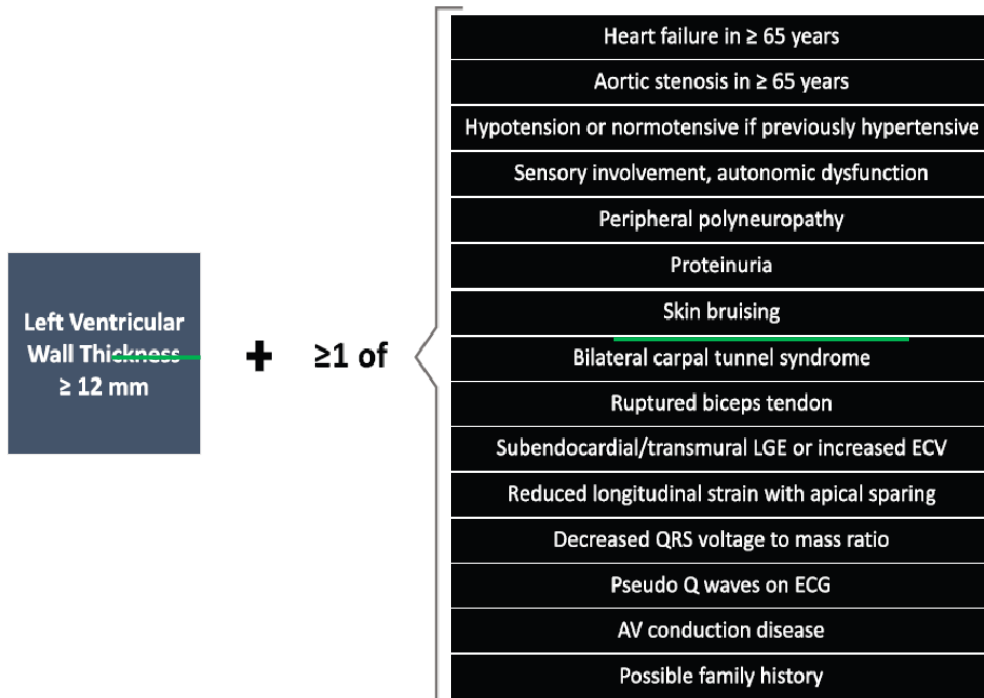
CARDIAC BIOMARKER ASSESSMENT



Cardiac biomarkers can refine the diagnostic algorithm in patients with suspected CA.
NT-proBNP <180 ng/L and hs-TnT <14 ng/L reliably exclude CA.



Diagnosis and treatment of cardiac amyloidosis: a position statement of the ESC Working Group on Myocardial and Pericardial Diseases



Type

Red flag

Extracardiac

Clinical

Polyneuropathy
Dysautonomia
Skin bruising
Skin discoloration
Cutis laxa
Macroglossia
Deafness
Bilateral carpal tunnel syndrome
Ruptured biceps tendon
Lumbar spinal stenosis
Vitreous deposits
Corneal lattice dystrophy
Family history
Renal insufficiency
Proteinuria

Laboratory

Cardiac

Clinical

Hypotension or normotensive if previous hypertensive
Pseudoinfarct pattern
Low/decreased QRS voltage to degree of LV thickness
AV conduction disease

Laboratory

Disproportionally elevated NT-proBNP to degree of HF
Persisting elevated troponin levels

Echocardiogram

Granular sparkling of myocardium
Increased right ventricular wall thickness
Increased valve thickness
Pericardial effusion

CMR

Reduced longitudinal strain with apical sparing pattern
Subendocardial late gadolinium enhancement
Elevated native T1 values
Increased extracellular volume
Abnormal gadolinium kinetics

Figure 3 Screening for cardiac amyloidosis. AV, atrio-ventricular; ECG, electrocardiogram; ECV, extracellular volume; LGE, late gadolinium enhancement.

Cardiac amyloidosis: do not forget to look for it

Aldostefano Porcari¹, Luca Falco¹, Veronica Lio², Marco Merlo¹, Enrico Fabris¹,
Rossana Bussani³, and Gianfranco Sinagra^{1*}

Table 1 Clinical manifestations of ATTR and AL amyloidosis

	ATTR	AL
Heart failure	+	+
Coronary disease (often microvascular)	+	++
Atrial fibrillation/flutter	+	+
Valvular leaflets thickening	++	+
Pericardial effusion	+	+
Decreased GLS (con apical sparing)	+	+
Low-voltage QRS	+	++
AVB	+	+
Capillary friability	+	+
Coagulation abnormalities (liver dysfunction)	+	+
Hepatomegalia	++	++
Splenomegalia	+	+
Macroglossia	+	++
Periorbital purpura	–	++
Dysfonia	+	+
Renal damage	+	++
Lymphadenopathy	–	++
Bone pain	–	++
Immune deficit	–	++
Urinary incontinence	+	+
Lower bowel dysfunction	+	+
Sensory neuropathy	++ (ATTRm)	+
Motor neuropathy	+	+
Carpal tunnel syndrome	++	+

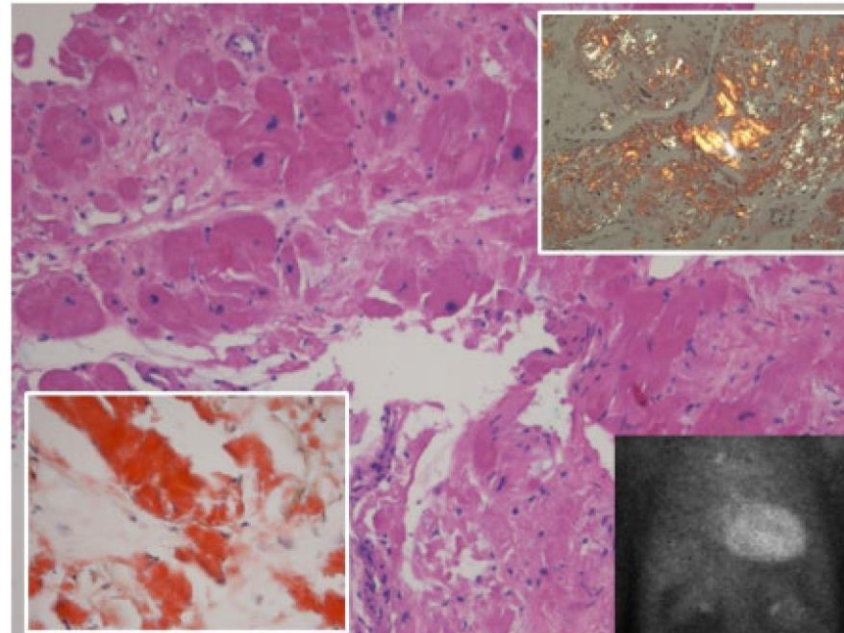


Table 2 ‘Red Flags’ in the cardiac amyloidosis

The diagnosis of AC must always start from a high degree of clinical suspicion
 In patients with heart failure with conserved systolic function and hypertrophy of the walls, the diagnosis of AC must be considered
 Patients with suspected HCM should always be critically evaluated to search for clinical-instrumental elements that can direct them towards an accumulation of heart disease (VP, ABV, etc.)
 Elderly patients with IVS in the absence of hypertension or aortic valve stenosis should be considered for AC
 Low QRS voltages to the ECG in the presence of IVS are suspected for AC
 AC from TTR and MGUS can coexist: an AC in the presence of monoclonal gammopathy is not necessarily AL
 GLS in AC is often significantly reduced (typically with apical savings), even in the presence of normal LVEF
 Magnetic resonance in both T1 signalling and extracellular oedema in IVS patients are very suggestive of AC. The distribution of the LGE is heterogeneous and the subendocardial distribution is not the only possible pattern
 The history of bilateral carpal tunnel syndrome in a man with IVS is very suggestive of AC ATTRwt

Left ventricular wall thickness and severity of cardiac disease in women and men with transthyretin amyloidosis

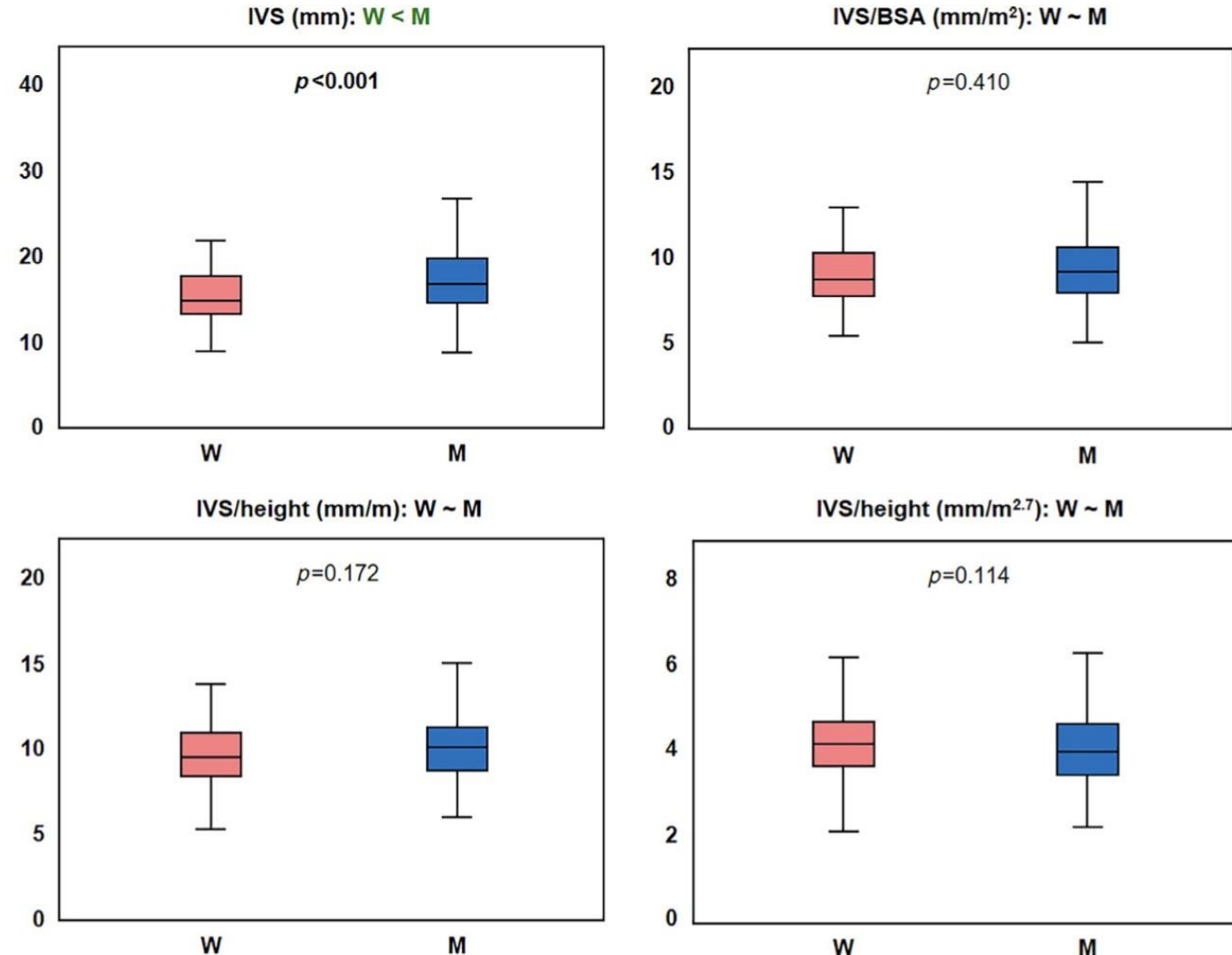
Alberto Aimo^{1,2*}, Daniela Tomasoni³, Aldostefano Porcari⁴, Giuseppe Vergaro^{1,2}, Vincenzo Castiglione^{1,2}, Claudio Passino^{1,2}, Marianna Adamo³, Maria Giulia Bellicini³, Carlo Mario Lombardi³, Matilde Nardi³, Gloria Palamara³, Guerino Giuseppe Varrà⁴, Riccardo Saro⁴, Valentina Allegro⁴, Marco Merlo⁴, Gianfranco Sinagra⁴, Marco Metra³, Michele Emdin^{1,2}, and Claudio Rapezzi^{5,6†}

FINDING: Women may reach the 12 mm threshold when their **disease is more advanced**, possibly leading to underdiagnosis or delayed diagnosis.

PROPOSAL: To replace the single diagnostic cut-off of 12mm with **LV wall thickness values indexed by height^{2,7}**.

Alternative would be to calculate a cut-off for women based on the mean height of men and women in Europe (1.77m and 1.65m, respectively)

Given 12mm the cut-off for men, the corresponding cut-off for women would be $12\text{mm} \times (1.65/1.77) = 11\text{mm}$.



Screening for Cardiac Amyloidosis 5 to 15 Years After Surgery for Bilateral Carpal Tunnel Syndrome



Oscar Westin, MD,^a Emil L. Fosbøl, MD, PhD,^a Mathew S. Maurer, MD,^b Birgitte P. Leicht, MD,^c Philip Hasbak, MD, DMSc,^d Anne Kærsgaard Mylin, MD, PhD,^e Sara Rørvig, MD,^f Thomas Hartvig Lindkær Helle Hjorth Johannesen, MD,^d Finn Gustafsson, MD, PhD, DMSc^a

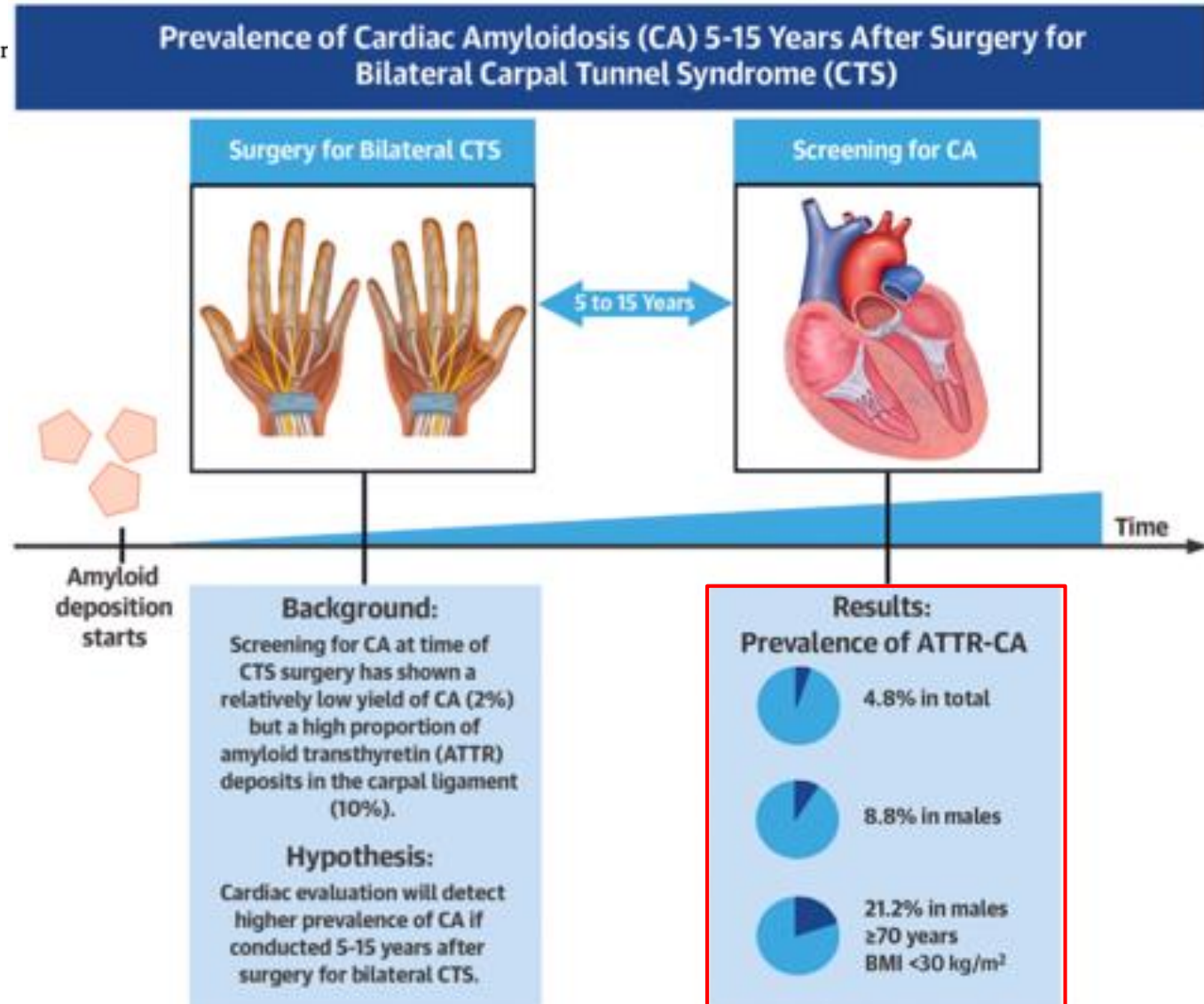
125 women and 125 men who had surgery for bilateral carpal tunnel syndrome in the previous 5 to 15 years underwent diagnostic work up for amyloidosis

Overall **4.8%** had ATTRwt-CM

8.8% of males aged 60 to 85 years had undiagnosed ATTRwt-CM

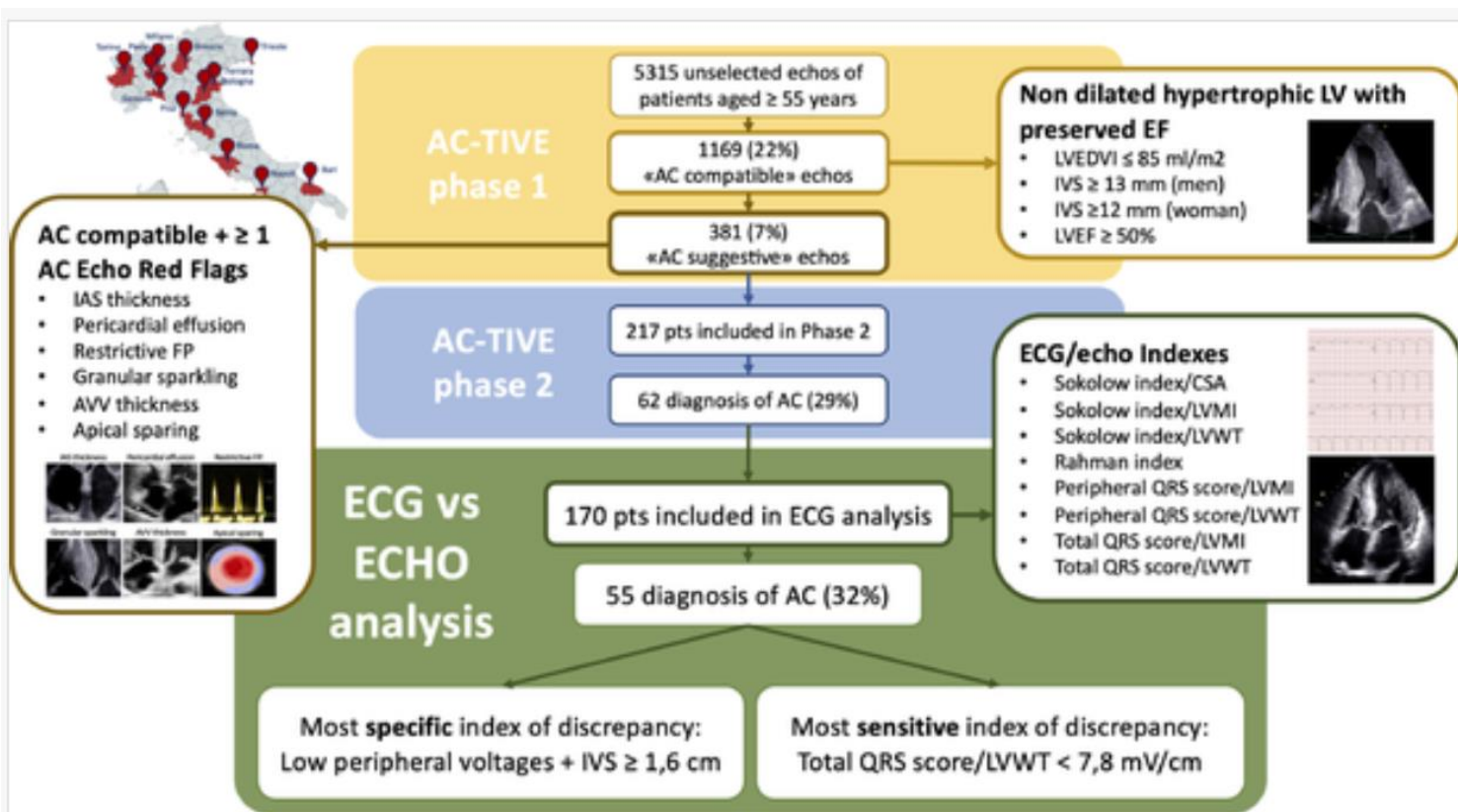
21.2% of men over 70 years with a BMI <30 Kg/m² had undiagnosed ATTRwt-CM

No case of AL amyloidosis was identified



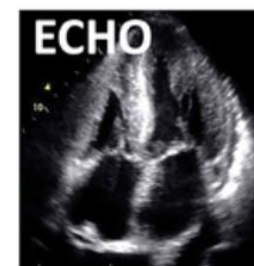
ECG/echo indexes in the diagnostic approach to amyloid cardiomyopathy: A head-to-head comparison from the AC-TIVE study

Linda Pagura^{a,b,1}, Aldostefano Porcari^a, Matteo Cameli^c, Elena Biagini^{d,1}, Marco Canepa^{e,f}, Lia Crotti^{g,h,1}, Massimo Imazio^{i,j}, Cinzia Forleo^k, Rita Pavasini^l, Giuseppe Limongelli^{m,1}, Stefano Perlini^{n,1}, Marco Metra^o, Giuseppe Boriani^p, Michele Emdin^q, Gianfranco Sinagra^{a,2}, Marco Merlo^{a,2,*}, ACTIVE study group³



1 In the echo-lab: identify patients with an **echocardiogram** suggestive of AC

Non-dilated hypertrophic heart with preserved LVEF



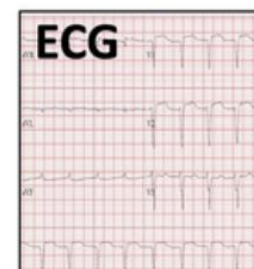
- LVEDVI ≤ 85 ml/m²
- IVS ≥ 12 mm (♀)
- IVS ≥ 13 mm (♂)
- LVEF $\geq 50\%$



≥ 1 red flag for AC

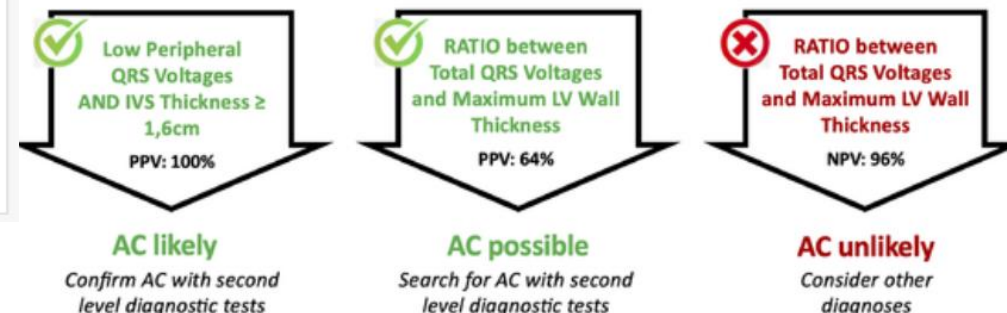
- IAS thickness
- Pericardial effusion
- Restrictive FP
- Granular sparkling
- AVV thickness
- Apical sparing

2 Add information from an **electrocardiogram**: look for ECG/echo indexes



- Low Peripheral QRS Voltages AND IVS Thickness $\geq 1,6$ cm
- RATIO** between Total QRS Voltages and Maximum LV Wall Thickness $< 7,8$ mV/cm

3 Decide for second level diagnostic tests: **integrate echo and ECG** parameters and check whether ECG/echo indexes are satisfied (\checkmark) or not (\times)



Low QRS Voltages in Cardiac Amyloidosis

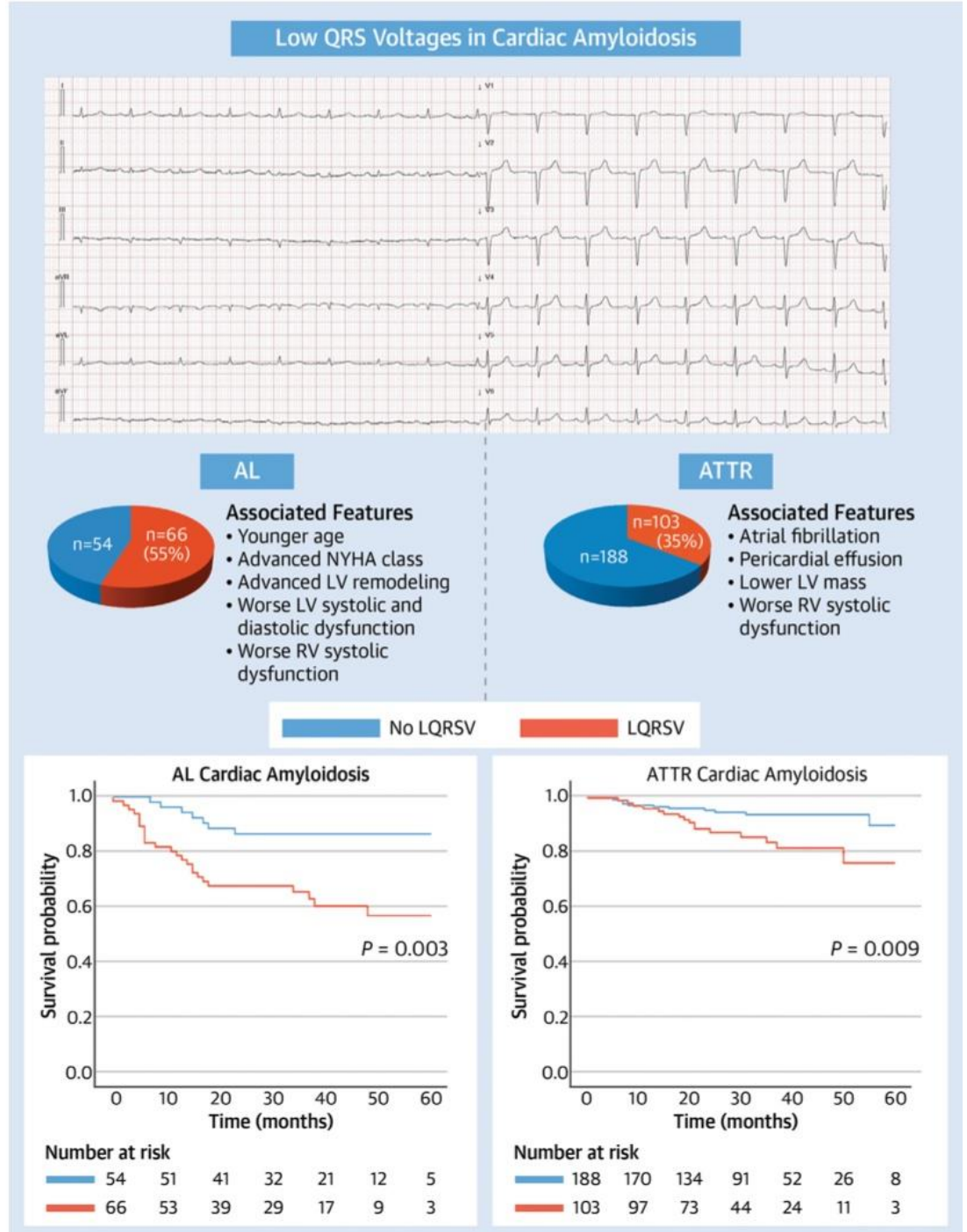
Clinical Correlates and Prognostic Value

Alberto Cipriani, MD,^{a,*} Laura De Michieli, MD,^{a,*} Aldostefano Porcari, MD, FISC,^b Luca Licchelli, MD,^a Giulio Sinigiani, MD,^a Giacomo Tini, MD,^c Mattia Zampieri, MD,^d Eugenio Sessarego, MD,^e Alessia Argirò, MD,^d Carlo Fumagalli, MD,^d Monica De Gaspari, MD,^a Roberto Licordari, MD,^f Domitilla Russo, MD,^c Gianluca Di Bella, MD,^f Federico Perfetto, MD,^{d,g} Camillo Autore, MD,^c Beatrice Musumeci, MD,^c Marco Canepa, MD,^{e,h} Marco Merlo, MD,^b Gianfranco Sinagra, MD, FESC,^b Dario Gregori, MD,^a Sabino Iliceto, MD,^a Martina Perazzolo Marra, MD, PhD,^a Francesco Cappelli, MD,^{d,g,i} Claudio Rapezzi, MD,^{i,j,i†}

Cipriani A, et al. J Am Coll Cardiol CardioOnc. 2022;4(4):458-470.

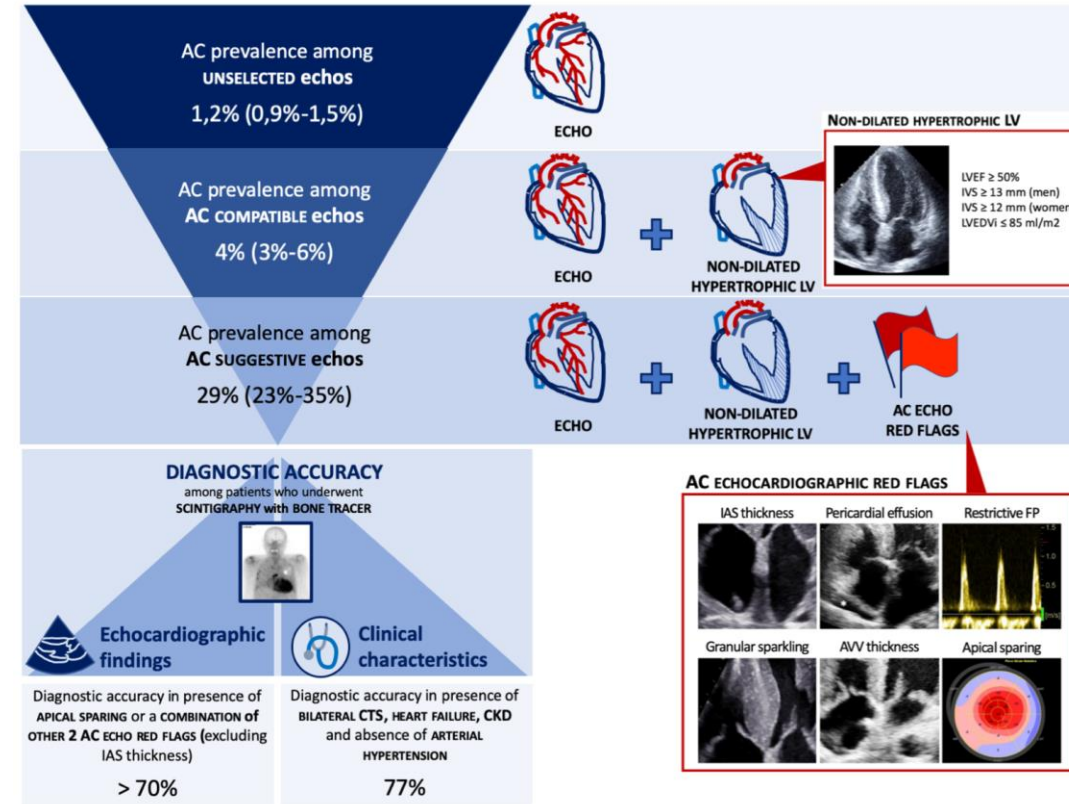
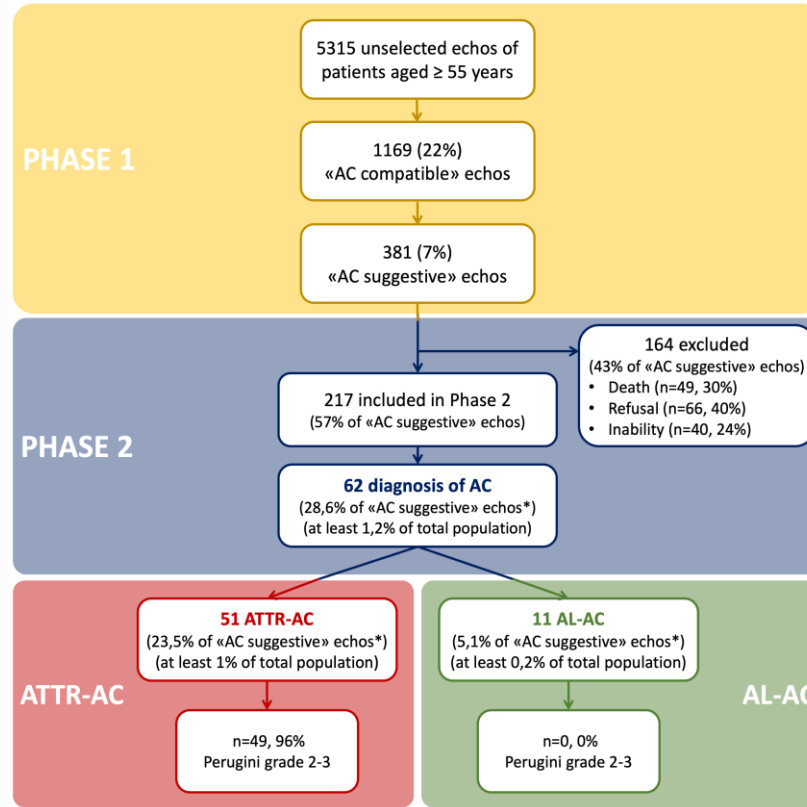
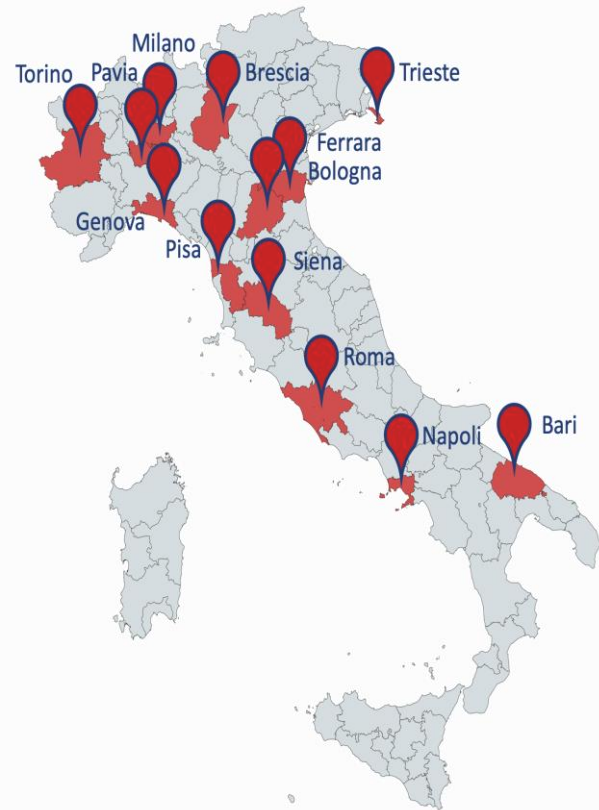
411 patients with CA: 120 AL-CA; 291 ATTR-CA

During a median FUP of 33 months (21-46), **LQRSVs independently predicted CV death** in both AL CA (HR: 1.76; P =0.031) and ATTR CA (HR: 2.64; P=0.005).

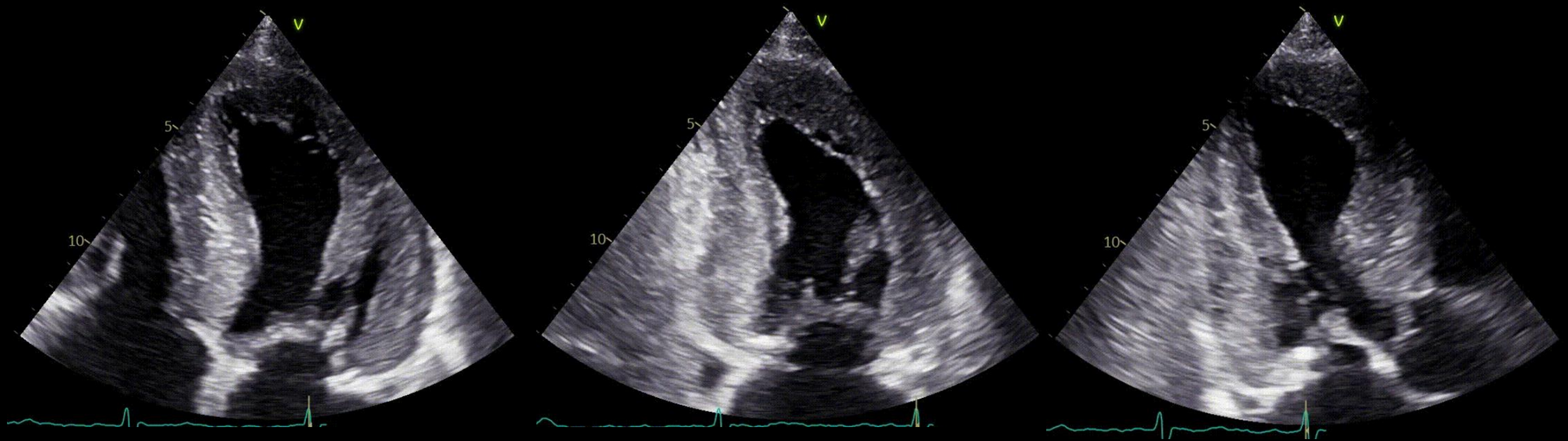


Unmasking the prevalence of amyloid cardiomyopathy in the real world: results from Phase 2 of the AC-TIVE study, an Italian nationwide survey

Marco Merlo, Aldostefano Porcari, ... Claudio Rapezzi, Gianfranco Sinagra

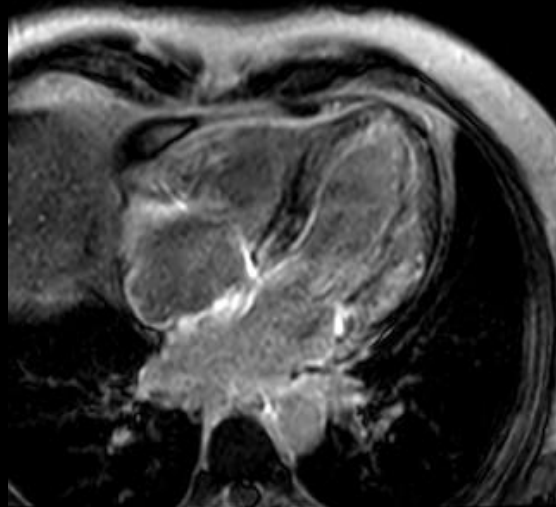
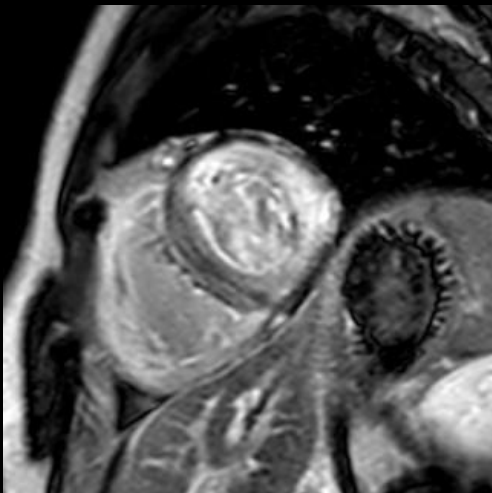
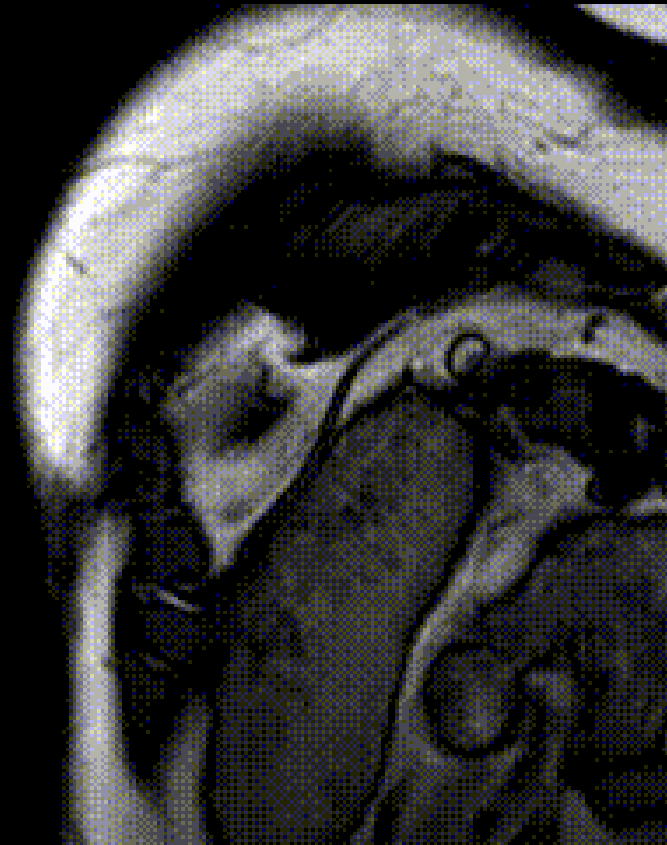
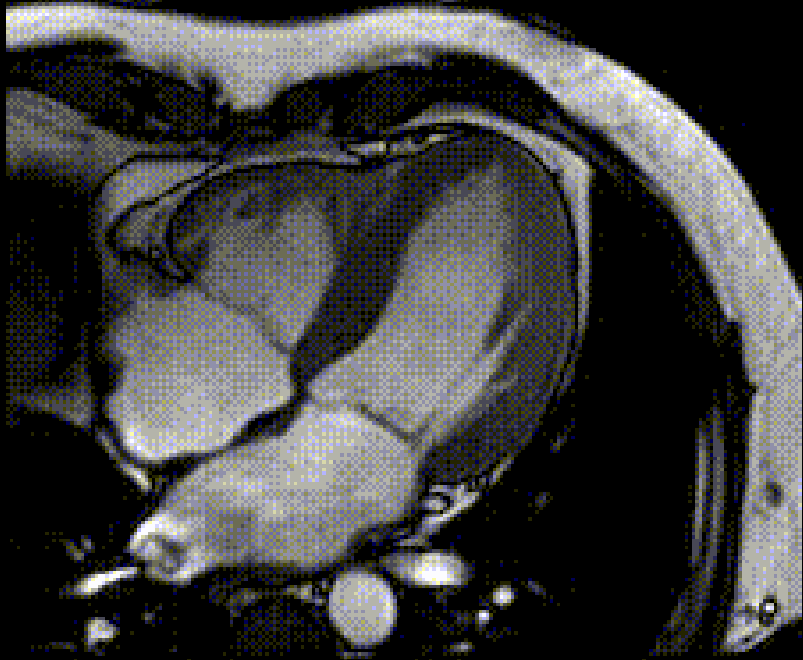


Prevalence of 29% in Italy among adults with echo red flags of AC



FE Vsin 45%.

Deposizione di amiloide secondo uno schema a "doppio gradiente":
basale-apicale e subendocardico-subepicardico.



- VSin VTDi 57 ml/m², FE 53%
- SIV 17 mm
- VDx ipertrofico, FE 55%
- Diffuso incremento del segnale T₂
- LGE subendocardico diffuso con inadeguato T.I., interessamento delle pareti atriali
- Incremento di T₁ mapping e ECV (50%)

Razionale e valore fondamentale della Rete Italiana integrata dell'Amiloidosi Cardiaca

Gianfranco Sinagra^{1,2}, Michele Emdin^{3,4}, Marco Merlo^{1,2}, Giuseppe Vergaro^{3,4}, Alberto Aimo^{3,4}, Elena Biagini^{2,5}, Massimo Imazio⁶, Aldostefano Porcari^{1,2}, Giuseppe Limongelli^{2,7}, Alberto Cipriani^{2,8}, Marco Canepa⁹, Beatrice Musumeci¹⁰, Matteo Cameli¹¹, Lia Crotti^{2,12,13}, Gianluca Di Bella¹⁴, Andrea Di Lenarda¹⁵, Francesco Cappelli¹⁶, Cristina Chimenti¹⁷, Laura Obici^{2,18}, Massimo Iacoviello¹⁹, Stefano Perlini^{2,20}, Maurizio Pieroni²¹, Marco Metra²², Fabrizio Oliva²³, Pasquale Perrone Filardi²⁴, Furio Colivicchi²⁵, Ciro Indolfi^{26,27}

FINALITA' DELLA RETE ITALIANA DELL'AMILOIDOSI CARDIACA

- Creare **contatti stabili tra centri** disegnando e supportando **PDTA condivisi** che assicurino a tutti i pazienti **equità ed una diagnosi e cura allo stato dell'arte**;
- Garantire un **accesso dedicato alla diagnostica avanzata** con RM cardiaca, medicina nucleare, BEM (esecuzione e interpretazione), tipizzazione dei depositi di amiloide (immuno-microscopia elettronica e spettrometria di massa);
- **Promuovere la ricerca**;
- Facilitare i processi di **rimborsabilità dei nuovi farmaci** da parte degli enti regolatori (AIFA), consentendo alle **Società Scientifiche di divenire interlocutrici degli enti regolatori anche rispetto a scenari controversi (es NYHA III)**; Redigere documenti nazionali finalizzati alla **standardizzazione dei processi di cura** e alla **descrizione dei modelli organizzativi** complessi legati alla malattia **(PDTA) elaborati con il contributo di Centri con consolidata esperienza di referenza ed attività in Rete.**



ISA 2022
XVIII. INTERNATIONAL SYMPOSIUM
ON AMYLOIDOSIS
4TH - 8TH
SEPTEMBER 2022



Razionale e valore fondamentale della Rete Italiana integrata dell'Amiloidosi Cardiaca

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