CAMPUS Cuore SUMMIT

Napoli, 1-2 ottobre 2021
Hotel Excelsior

12.00 **FOCUS ON 4**
CARDIOLOGIA CLINICA
Coordinatori: **Edoardo Bossone, Carmine Riccio**

- Triplice terapia con acido bempedoico e statina/ezetimibe per il rischio evitabile nel paziente con malattia aterosclerotica cardiovascolare - **Arturo Cesaro**
- Terapia farmacologica per la lotta al rischio evitabile nel paziente con ipertrigliceridemia - **Plinio Cirillo**
- Trattamento farmacologico integrato per il rischio evitabile nella sindrome coronarica cronica - **Ciro Indolfi**
- Terapia antitrombotica bilanciata tra rischio emorragico ed ischemico nel paziente con sindrome coronarica acuta - **Leonardo De Luca**
- Amiloidosi cardiaca: update 2020-2021 - **Giuseppe Limongelli**
- Take home message - **Carmine Riccio**

CENTRO COORDINAMENTO MALATTIE RARE REGIONE CAMPANIA

What is new?

Definition

Epidemiology

Clinical Presentation - Diagnosis

Imaging

Management

What is new?

Definition

One name, many disease…

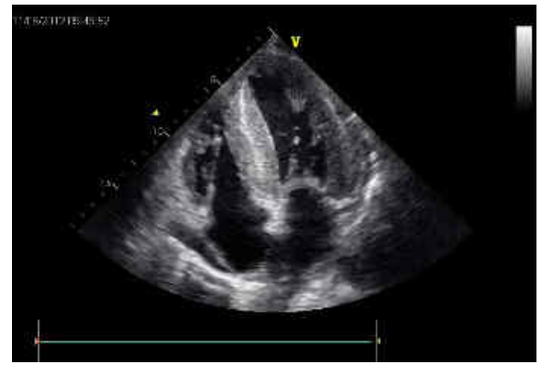
Terminology and Classification of Amyloidoses

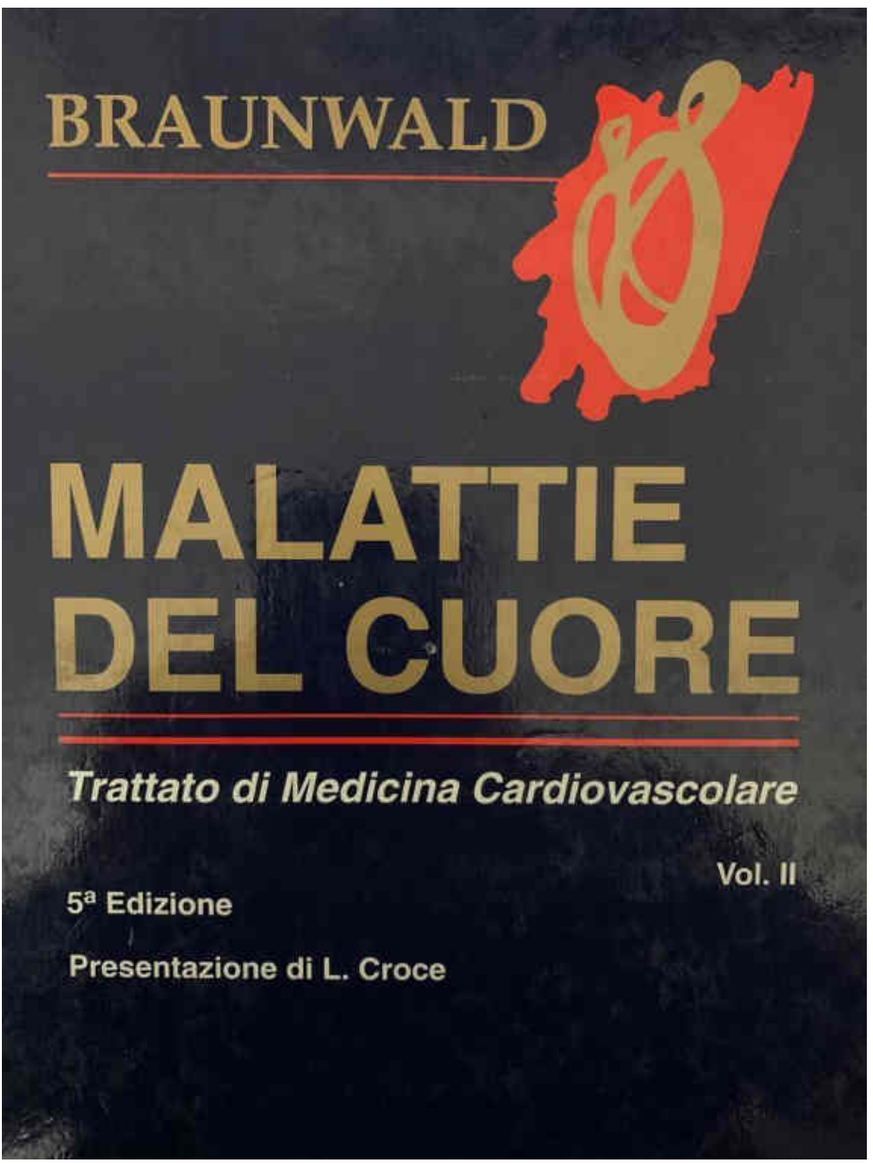
Amyloid protein	Precursor	Syndrome or involved tissue
AL	immunoglobulin light chain	Primary / Myeloma associated
ATTR	Transthyretin	Familial (PAF) Senile (wilde type TTR)
AA	Serum AA	Secondary, reactive
A $ß_2$M	$ß_2$ microglobulin	Hemodyalisis associated
AApo AII AFib Alys	Apolipoprotein AII Fibrinogen α chain Lysozyme	Familial Familial Familial
…. Aß APrP	…. Aß protein precursor Prion protein	…. Alzheimer's desease, aging Spongioform encephalopathies

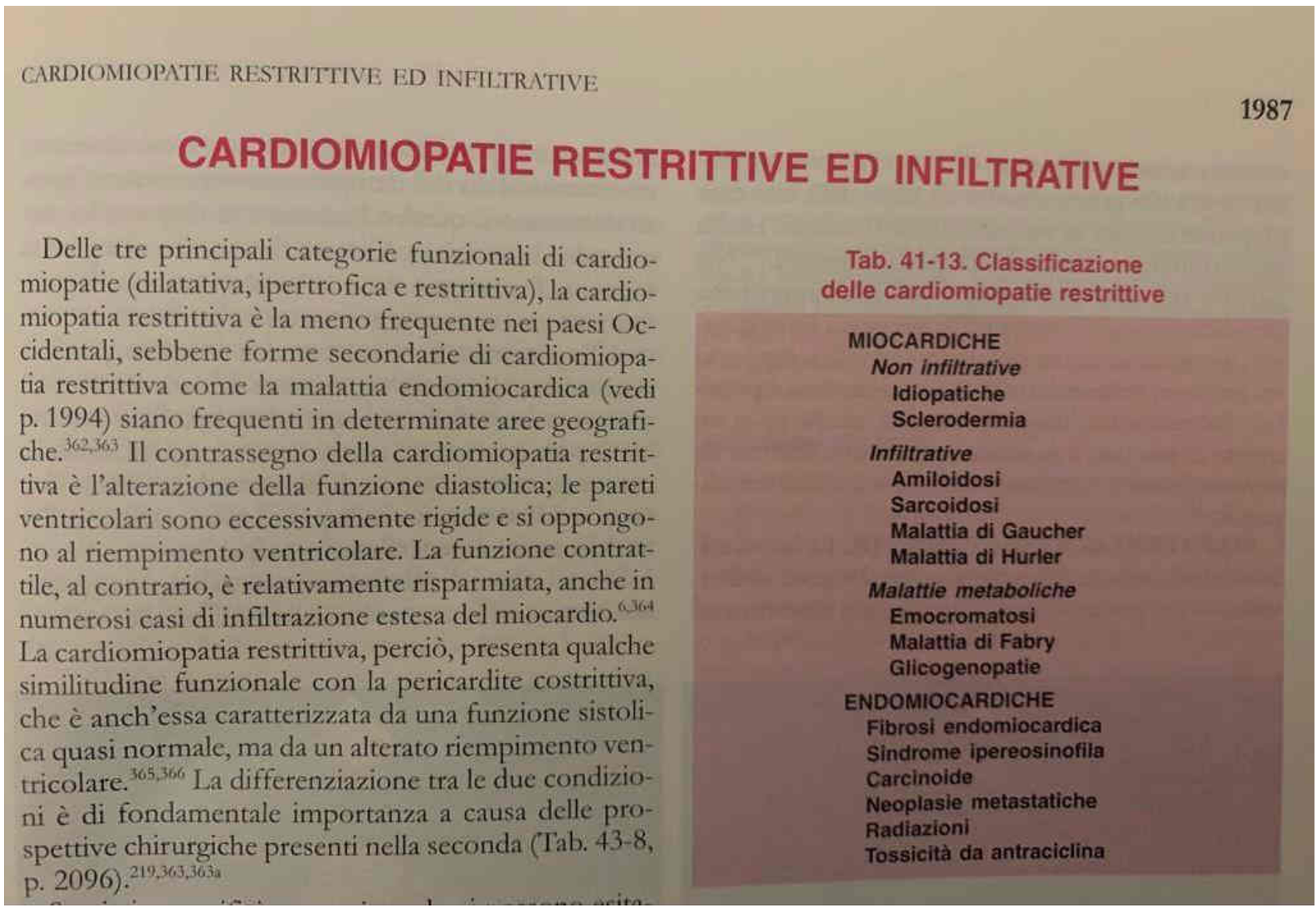
-More than 30 different types of Amyloidosis (each due to a specific protein)

-Cardiac Amyloidosis: AL + ATTr (up to 95%)

A restrictive cardiomyopathy?







CARDIOMIOPATIE RESTRITTIVE ED INFILTRATIVE 1987

CARDIOMIOPATIE RESTRITTIVE ED INFILTRATIVE

Delle tre principali categorie funzionali di cardiomiopatie (dilatativa, ipertrofica e restrittiva), la cardiomiopatia restrittiva è la meno frequente nei paesi Occidentali, sebbene forme secondarie di cardiomiopatia restrittiva come la malattia endomiocardica (vedi p. 1994) siano frequenti in determinate aree geografiche.[362,363] Il contrassegno della cardiomiopatia restrittiva è l'alterazione della funzione diastolica; le pareti ventricolari sono eccessivamente rigide e si oppongono al riempimento ventricolare. La funzione contrattile, al contrario, è relativamente risparmiata, anche in numerosi casi di infiltrazione estesa del miocardio.[6,364] La cardiomiopatia restrittiva, perciò, presenta qualche similitudine funzionale con la pericardite costrittiva, che è anch'essa caratterizzata da una funzione sistolica quasi normale, ma da un alterato riempimento ventricolare.[365,366] La differenziazione tra le due condizioni è di fondamentale importanza a causa delle prospettive chirurgiche presenti nella seconda (Tab. 43-8, p. 2096).[219,363,363a]

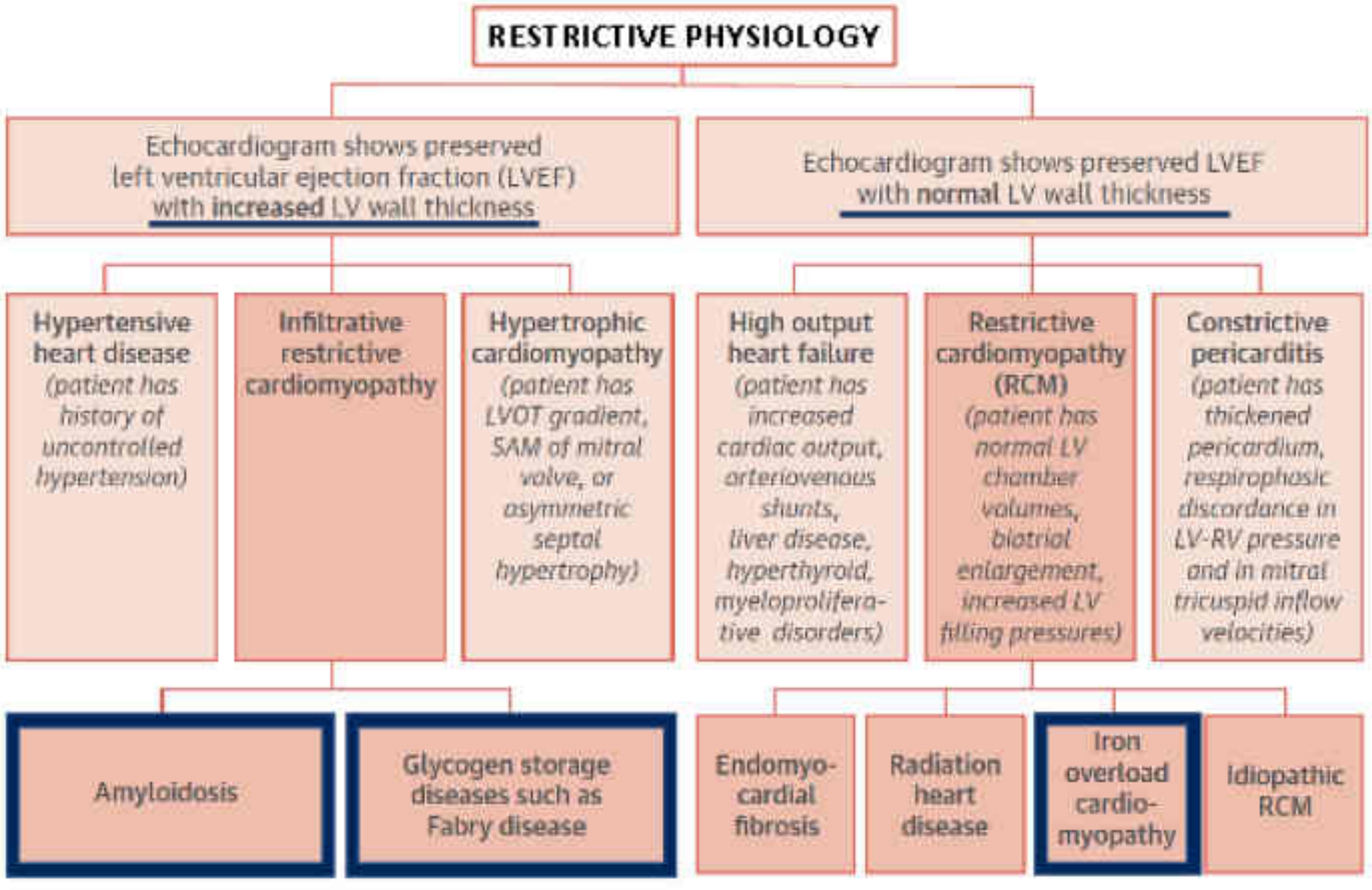
Tab. 41-13. Classificazione delle cardiomiopatie restrittive

MIOCARDICHE
Non infiltrative
Idiopatiche
Sclerodermia
Infiltrative
Amiloidosi
Sarcoidosi
Malattia di Gaucher
Malattia di Hurler
Malattie metaboliche
Emocromatosi
Malattia di Fabry
Glicogenopatie

ENDOMIOCARDICHE
Fibrosi endomiocardica
Sindrome ipereosinofila
Carcinoide
Neoplasie metastatiche
Radiazioni
Tossicità da antraciclina

CENTRO COORDINAMENTO MALATTIE RARE REGIONE CAMPANIA

Infiltrative cardiomyopathies: phenotypic classification



Modified from Pereira, N.L. et al. J Am Coll Cardiol. 2018;71(10):1130–48.

What is new?

Epidemiology

Increased awarness on Cardiac Amyloidosis

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cardiac amyloidosis

Search

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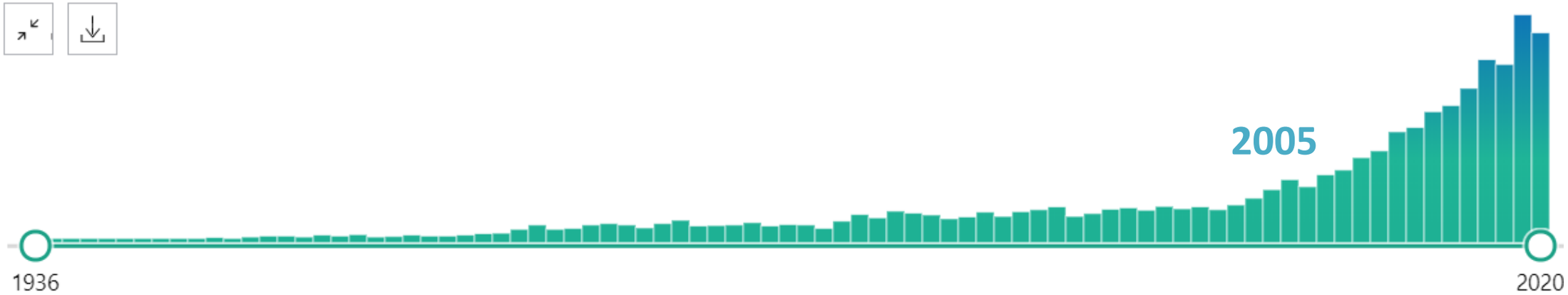
RESULTS BY YEAR

5,330 results



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Cite

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Cardiac amyloidosis.

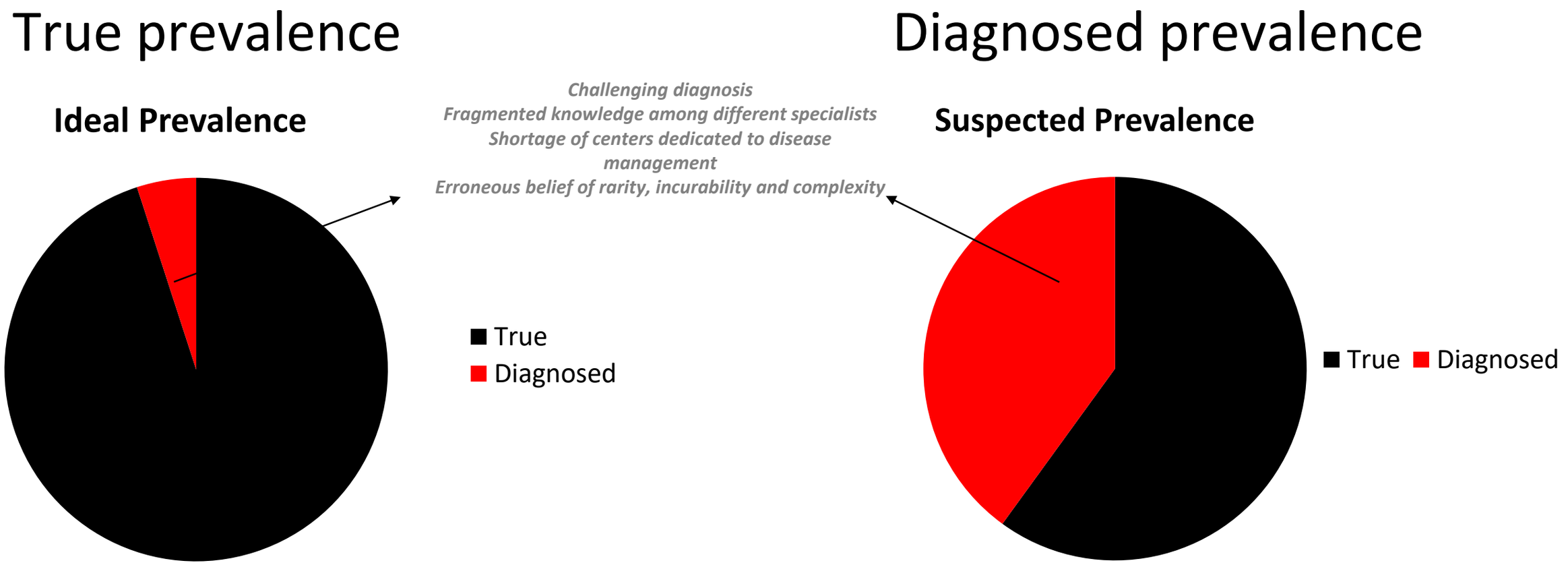
Martinez-Naharro A, Hawkins PN, Fontana M.

Clin Med (Lond). 2018 Apr 1;18(Suppl 2):s30-s35. doi: 10.7861/clinmedicine.18-2-s30.

PMID: 29700090 Free PMC article. Review.

Cardiac amyloid deposition, causing an infiltrative/restrictive cardiomyopathy, is a frequent feature of **amyloidosis** and a major determinant of survival. ...This article aims to review recent developments in diagnosis and management of **cardiac amyloidosis** ...

Epidemiologia e prevalenza:
l'amiloidosi è una patologia rara o poco diagnosticata?



International Journal of Cardiology xxx (xxxx) 1–5

Contents lists available at ScienceDirect

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journal homepage: http://ees.elsevier.com

Editorial

Cardiac amyloidosis: Watching the tip of the iceberg emerging from the "heart of the sea"

Cristiano Amarelli [a,*], Giuseppe Limongelli [b]

[a] Department of Cardiac Surgery and Transplants, Monaldi, Azienda dei Colli, 80131 Naples, Italy
[b] Inherited and Rare Cardiovascular Diseases, Department of Translational Medical Sciences, University of Campania "Luigi Vanvitelli", Monaldi Hospital, 80131 Naples, Italy

What is new?

Clinical Presentation

Clinical Scenarios - Cardiac Red Flags

-HFpEF

-HCM/LVH

-Aortic Stenosis

-Arrhythmias (AVB, AF)

-Syncope, orthostatic hypotension

wtATTR

13%

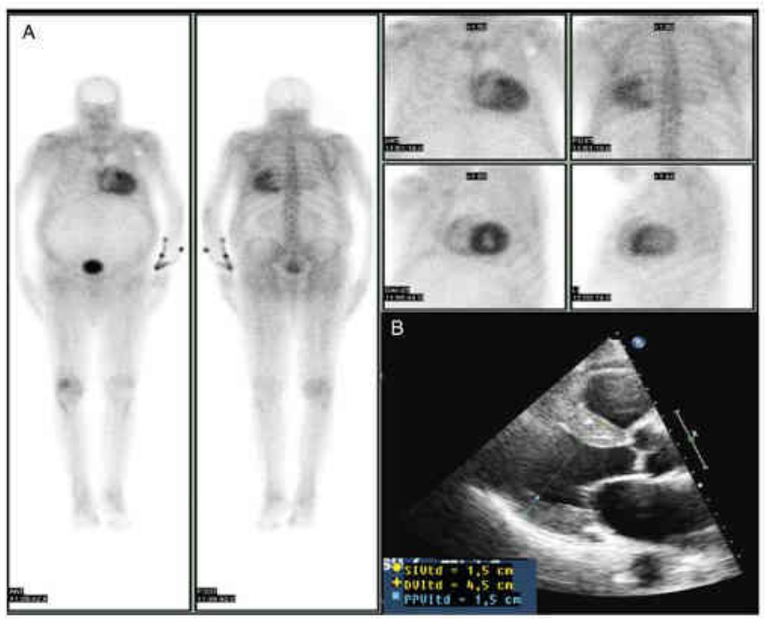
HFPEF!

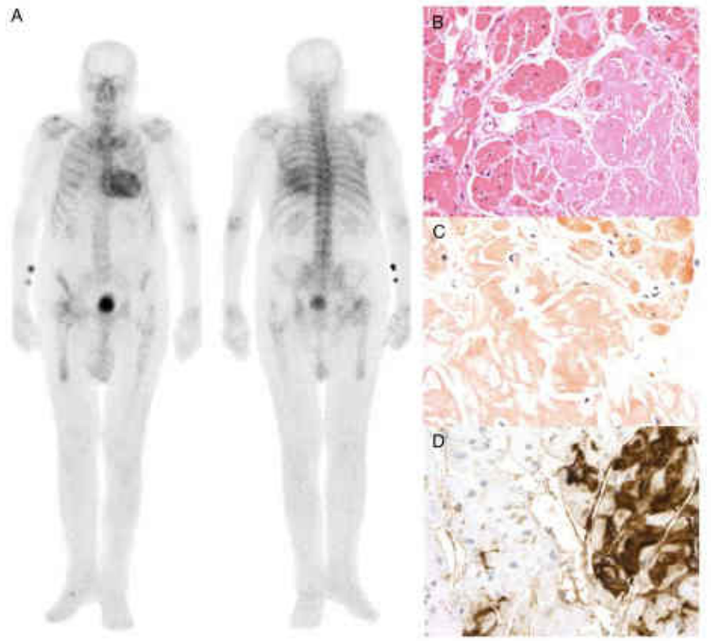
European Heart Journal (2015) **36**, 2585–2594
doi:10.1093/eurheartj/ehv338

CLINICAL RESEARCH
Heart failure/cardiomyopathy

Wild-type transthyretin amyloidosis as a cause of heart failure with preserved ejection fraction

Esther González-López[1], Maria Gallego-Delgado[1], Gonzalo Guzzo-Merello[1], F. Javier de Haro-del Moral[2], Marta Cobo-Marcos[1], Carolina Robles[1], Belén Bornstein[3,4,5], Clara Salas[6], Enrique Lara-Pezzi[7], Luis Alonso-Pulpon[1], and Pablo Garcia-Pavia[1,7]*





ESC European Society of Cardiology

European Heart Journal (2017) **38**, 2879–2887
doi:10.1093/eurheartj/ehx350

CLINICAL RESEARCH
Heart failure/cardiomyopathy

Unveiling transthyretin cardiac amyloidosis and its predictors among elderly patients with severe aortic stenosis undergoing transcatheter aortic valve replacement

Adam Castaño[1,2]*, David L. Narotsky[1], Nadira Hamid[3], Omar K. Khalique[3], Rachelle Morgenstern[2], Albert DeLuca[2], Jonah Rubin[1], Codruta Chiuzan[4], Tamim Nazif[3], Torsten Vahl[3], Isaac George[3], Susheel Kodali[3], Martin B. Leon[3], Rebecca Hahn[3], Sabahat Bokhari[2], and Mathew S. Maurer[1]

Aims Transthyretin cardiac amyloidosis (ATTR-CA) has been reported in patients with aortic stenosis (AS) but its prevalence and phenotype are not known. We examine elderly patients with severe symptomatic AS undergoing transcatheter aortic valve replacement (TAVR) and determine the prevalence and phenotype of ATTR-CA non-invasively.

Methods and results We performed technetium-99m pyrophosphate (^{99m}Tc-PYP) cardiac scintigraphy prospectively on patients who underwent TAVR, to screen for ATTR-CA. Transthoracic echocardiography and speckle-strain imaging were performed. We assessed the association of several parameters with ATTR-CA using multivariable logistic regression and constructed receiver operating curves to evaluate the best predictors of ATTR-CA. Among 151 patients (mean age 84 ± 6 years, 68% men), 16% ($n = 24$) screened positive for ATTR-CA with ^{99m}Tc-PYP scintigraphy. Compared with patients without ATTR-CA, ATTR-CA patients had a thicker interventricular septum (1.3 vs. 1.1 cm, $P = 0.007$), higher left ventricular (LV) mass index (130 vs. 98 g/m^2, $P = 0.002$), and lower stroke volume index (30 vs. 36 mL/m^2, $P = 0.009$). ATTR-CA patients had advanced diastolic dysfunction with higher *E/A* ratio (2.3 vs. 0.9, $P = 0.001$) and lower deceleration time (176 vs. 257 ms, $P < 0.0001$); impairment in systolic function with lower ejection fraction (48% vs. 56%, $P = 0.011$), myocardial contraction fraction (26 vs. 41, $P < 0.0001$), and average of lateral and septal mitral annular tissue Doppler S' (4.0 vs. 6.6 cm/s, $P < 0.0001$). While ATTR-CA patients had more impaired global longitudinal strain (-12 vs. -16%, $P = 0.007$), relative apical longitudinal strain was the same regardless of ATTR-CA diagnosis (0.98 vs. 0.98, $P = 0.991$). Average S' best predicted ATTR-CA in multivariable logistic regression (odds ratio 16.67 per 1 cm/s decrease with AUC 0.96, 95% confidence interval 0.90–0.99, $P = 0.002$) with a value ≤6 conferring 100% sensitivity for predicting a positive ^{99m}Tc-PYP amyloid scan.

Conclusions Transthyretin cardiac amyloidosis is prevalent in 16% of patients with severe calcific AS undergoing TAVR and is associated with a severe AS phenotype of low-flow low-gradient with mildly reduced ejection fraction. Average tissue Doppler mitral annular S' of < 6 cm/s may be a sensitive measure that should prompt a confirmatory ^{99m}Tc-PYP scan and subsequent testing for ATTR-CA. Prospective assessment of outcomes after TAVR is needed in patients with and without ATTR-CA.

wtATTR

16%

AS!

European Heart Journal (2016) **37**, 1826–1834
doi:10.1093/eurheartj/ehv583

FASTTRACK CLINICAL RESEARCH
Heart failure/cardiomyopathy

Prevalence and clinical phenotype of hereditary transthyretin amyloid cardiomyopathy in patients with increased left ventricular wall thickness

Thibaud Damy[1]*, Bruno Costes[2], Albert A. Hagège[3], Erwan Donal[4], Jean-Christophe Eicher[5], Michel Slama[6], Aziz Guellich[1], Stéphane Rappeneau[1], Jean-Pierre Gueffet[7], Damien Logeart[8], Violaine Planté-Bordeneuve[9], Hélène Bouvaist[10], Olivier Huttin[11], Geneviève Mulak[12], Jean-Luc Dubois-Randé[1], Michel Goossens[2], Florence Canoui-Poitrine[13]†, and Joel N. Buxbaum[14]†

Aims Increased left ventricular wall thickness (LVWT) is a common finding in cardiology. It is not known how often hereditary transthyretin-related familial amyloid cardiomyopathy (mTTR-FAC) is responsible for LVWT. Several therapeutic modalities for mTTR-FAC are currently in clinical trials; thus, it is important to establish the prevalence of TTR mutations (mTTR) and the clinical characteristics of the patients with mTTR-FAC.

Methods and results In a prospective multicentre, cross-sectional study, the TTR gene was sequenced in 298 consecutive patients diagnosed with increased LVWT in primary cardiology clinics in France. Among the included patients, median (25–75th percentiles) age was 62 [50;74]; 74% were men; 23% were of African origin; and 36% were in NYHA Class III–IV. Median LVWT was 18 (16–21) mm. Seventeen (5.7%; 95% confidence interval [CI]: [3.4;9.0]) patients had mTTR of whom 15 (5.0%; 95% CI [2.9;8.2]) had mTTR-FAC. The most frequent mutations were V142I ($n = 8$), V50M ($n = 2$), and I127V ($n = 2$). All mTTR-FAC patients were older than 63 years with a median age of 74 [69;79]. Of the 15 patients with mTTR-FAC, 8 were of African descent while 7 were of European descent. In the African descendants, mTTR-FAC median age was 74 [72;79] vs. 55 [46;65] years in non-mTTR-FAC ($P < 0.001$). In an adjusted multivariate model, African origin, neuropathy, carpal tunnel syndrome, electrocardiogram (ECG) low voltage, and late gadolinium enhancement (LGE) at cardiac-magnetic resonance imaging were all independently associated with mTTR-FAC.

Conclusion Five per cent of patients diagnosed with hypertrophic cardiomyopathy have mTTR-FAC. Mutated transthyretin genetic screening is warranted in elderly subjects with increased LVWT, particularly, those of African descent with neuropathy, carpal tunnel syndrome, ECG low voltage, or LGE.

Keywords Hypertrophic cardiomyopathy • Cardiac amyloidosis • Transthyretin

mATTR

5.7%

IVS!

Figura 1. Spettro di correlazioni genotipo-fenotipo nell'amiloidosi transtiretino-relata. L'espressione fenotipica varia da un interessamento quasi esclusivamente neurologico (mutazione Val30Met "early onset") ad uno esclusivamente cardiaco (mutazioni Thr60Ala, Leu111Met, Ile68Leu e Val122Ile). Altre mutazioni sono legate ad un fenotipo misto con gradi variabili di coinvolgimento neurologico e cardiaco.
SSA, amiloidosi sistemica senile.

When To Suspect Cardiac Amyloidosis?

Contents lists available at ScienceDirect

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Prevalence and clinical significance of red flags in patients with hypertrophic cardiomyopathy

Giuseppe Limongelli [a,b,c,d,*], Emanuele Monda [a,b], Stefania Tramonte [b], Felice Gragnano [g], Daniele Masarone [b], Giulia Frisso [e,f], Augusto Esposito [b], Rita Gravino [b], Ernesto Ammendola [b], Gemma Salerno [b], Marta Rubino [b], Martina Caiazza [b], Mariagiovanna Russo [a], Paolo Calabrò [g], Perry Mark Elliott [d], Giuseppe Pacileo [b]

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[c] European Reference Network - GUARD HEART, UK
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[g] UOC Cardiologia Vanvitelli - Osp. Sant'Anna e San Sebastiano - Caserta, Italy

International Journal of Cardiology 299 (2020) 186–191

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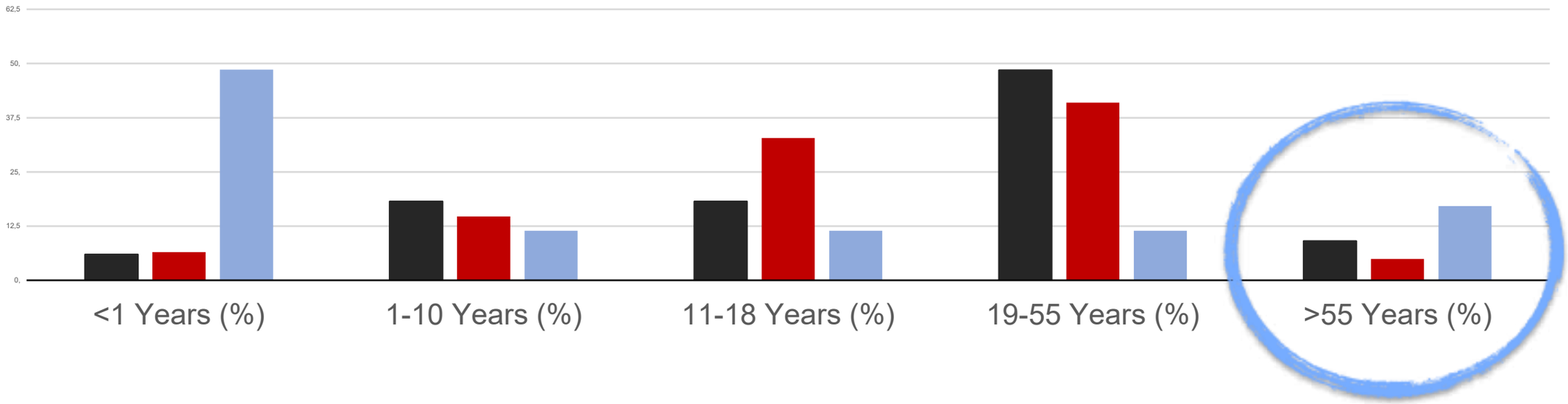
Prevalence and clinical significance of red flags in patients with hypertrophic cardiomyopathy

Giuseppe Limongelli [a,b,c,d,*], Emanuele Monda [a,b], Stefania Tramonte [b], Felice Gragnano [g], Daniele Masarone [b], Giulia Frisso [e,f], Augusto Esposito [b], Rita Gravino [b], Ernesto Ammendola [b], Gemma Salerno [b], Marta Rubino [b], Martina Caiazza [b], Mariagiovanna Russo [a], Paolo Calabrò [g], Perry Mark Elliott [d], Giuseppe Pacileo [b]



■ Idiopathic HCM ■ Sarcomeric HCM ■ Specific causes of HCM

"Prevalence of specific disease was higher:

-in infants (78%; i.e. Pompe, RAS-Mapk)

-in adults >55 years old (58%; Amyloydosis & Fabry) **Limongelli et al. 2019**

When to suspect cardiac amiloydosis?

ESC European Society of Cardiology

European Heart Journal (2021) 00, 1–128
doi:10.1093/eurheartj/ehab368

ESC GUIDELINES

2021 ESC Guidelines for the diagnosis and treatment of acute and chronic heart failure

Developed by the Task Force for the diagnosis and treatment of acute and chronic heart failure of the European Society of Cardiology (ESC)

With the special contribution of the Heart Failure Association (HFA) of the ESC

Authors/Task Force Members: Theresa A. McDonagh* (Chairperson) (United Kingdom), Marco Metra* (Chairperson) (Italy), Marianna Adamo (Task Force Coordinator) (Italy), Roy S. Gardner (Task Force Coordinator) (United Kingdom), Andreas Baumbach (United Kingdom), Michael Böhm (Germany), Haran Burri (Switzerland), Javed Butler (United States of America), Jelena Čelutkienė (Lithuania), Ovidiu Chioncel (Romania), John G.F. Cleland (United Kingdom), Andrew J.S. Coats (United Kingdom), Maria G. Crespo-Leiro (Spain), Dimitrios Farmakis (Greece), Martine Gilard (France), Stephane Heymans

* Corresponding author: The two chairpersons contributed equally to the document.

Red Flags

Table 35 "Red flags" for most common forms of cardiac amyloidosis

Type	Red Flag	TTR	AL
Extracardiac	Polyneuropathy	X	X
	Dysautonomia	X	X
	Skin bruising		X
	Macroglossia		X
	Deafness	X	
	Bilateral carpal tunnel syndrome	X	
	Ruptured biceps tendon	X	
	Lumbar spinal stenosis	X	
	Vitreous deposits	X*	
	Family history	X*	
	Renal insufficiency		X
	Proteinuria		X
Cardiac	**Clinical**		
	Hypotension or normotensive if previously hypertensive	X	X
	ECG		
	Pseudo-infarct ECG pattern	X	X
	Low/decreased QRS voltage to degree of LV thickness	X	X
	AV conduction disease	X	X
	Laboratory		
	Disproportionally elevated NT-proBNP to degree of HF	X	X
	Persisting elevated troponin levels	X	X
	Echocardiography		
	Granular sparkling of myocardium	X	X
	Increased right ventricular wall thickness	X	X
	Increased AV valve thickness	X	X
	Pericardial effusion	X	X
	Reduced longitudinal strain with apical sparing pattern	X	X
	CMR		
	Subendocardial LGE	X	X
	Elevated native T1 values	X	X
	Increased extracellular volume	X	X
	Abnormal gadolinium kinetics	X	X

ORIGINAL RESEARCH

Multiparametric Echocardiography Scores for the Diagnosis of Cardiac Amyloidosis

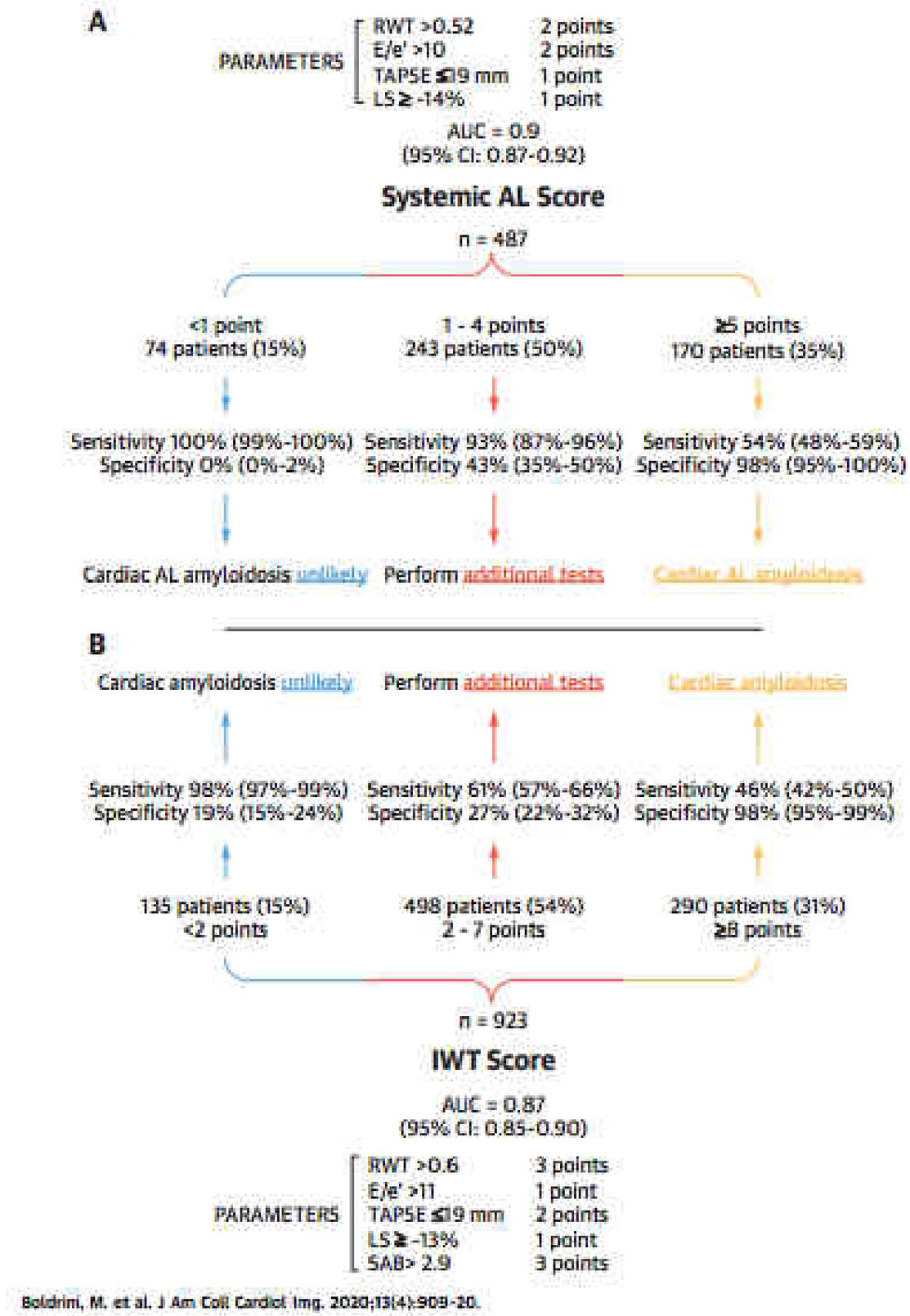
Michele Boldrini, MD,[a,b] Francesco Cappelli, MD,[c] Liza Chacko, MD,[a] Maria Alejandra Restrepo-Cordoba, MD,[d,e] Angela Lopez-Sainz, MD, PhD,[d,e] Alberto Giannoni, MD, PhD,[f,g] Alberto Aimo, MD, MRCP,[a,g] Andrea Baggiano, MD,[h] Ana Martinez-Naharro, MD,[a] Carol Whelan, MD,[a] Cristina Quarta, MD, PhD,[a] Claudio Passino, MD,[f,g] Vincenzo Castiglione, MD,[g] Vladyslav Chubuchnyi, MD,[f] Valentina Spini, MD,[f] Claudia Taddei, BSc,[f] Giuseppe Vergaro, MD, PhD,[f,g] Aviva Petrie, BSc (Hons),[i] Luis Ruiz-Guerrero, MD,[d] Vanessa Moñivas, MD, PhD,[j] Susana Mingo-Santos, MD, PhD,[j] Jesus G. Mirelis, MD, PhD,[d,e] Fernando Dominguez, MD, PhD,[d,e] Esther Gonzalez-Lopez, MD, PhD,[d,e] Stefano Perlini, MD, PhD,[b] Gianluca Pontone, MD, PhD,[b] Julian Gillmore, MD, PhD,[a] Philip N. Hawkins, PhD, FMedSci,[a] Pablo Garcia-Pavia, MD, PhD,[d,e,j] Michele Emdin, MD, PhD,[f,g] Marianna Fontana, MD, PhD[a]

Here, we provide a multiparametric echocardiographic approach for the 2 challenging clinical scenarios of patients with systemic AL amyloidosis or a hypertrophic phenotype.

By using highly sensitive and highly specific cutoffs in patients with systemic AL amyloidosis, amyloidosis can be excluded or confirmed in 50% of patients without the need for further tests.

In patients with a hypertrophic phenotype, highly specific or highly sensitive cutoffs can be used to guide the diagnostic algorithm, avoid unnecessary tests, and limit the time to diagnosis

CENTRAL ILLUSTRATION Diagnostic Algorithms With Echocardiographic Scores in 2 Different Clinical Scenarios



Boldrini, M. et al. J Am Coll Cardiol Img. 2020;13(4):909-20.

International Journal of CARDIOLOGY

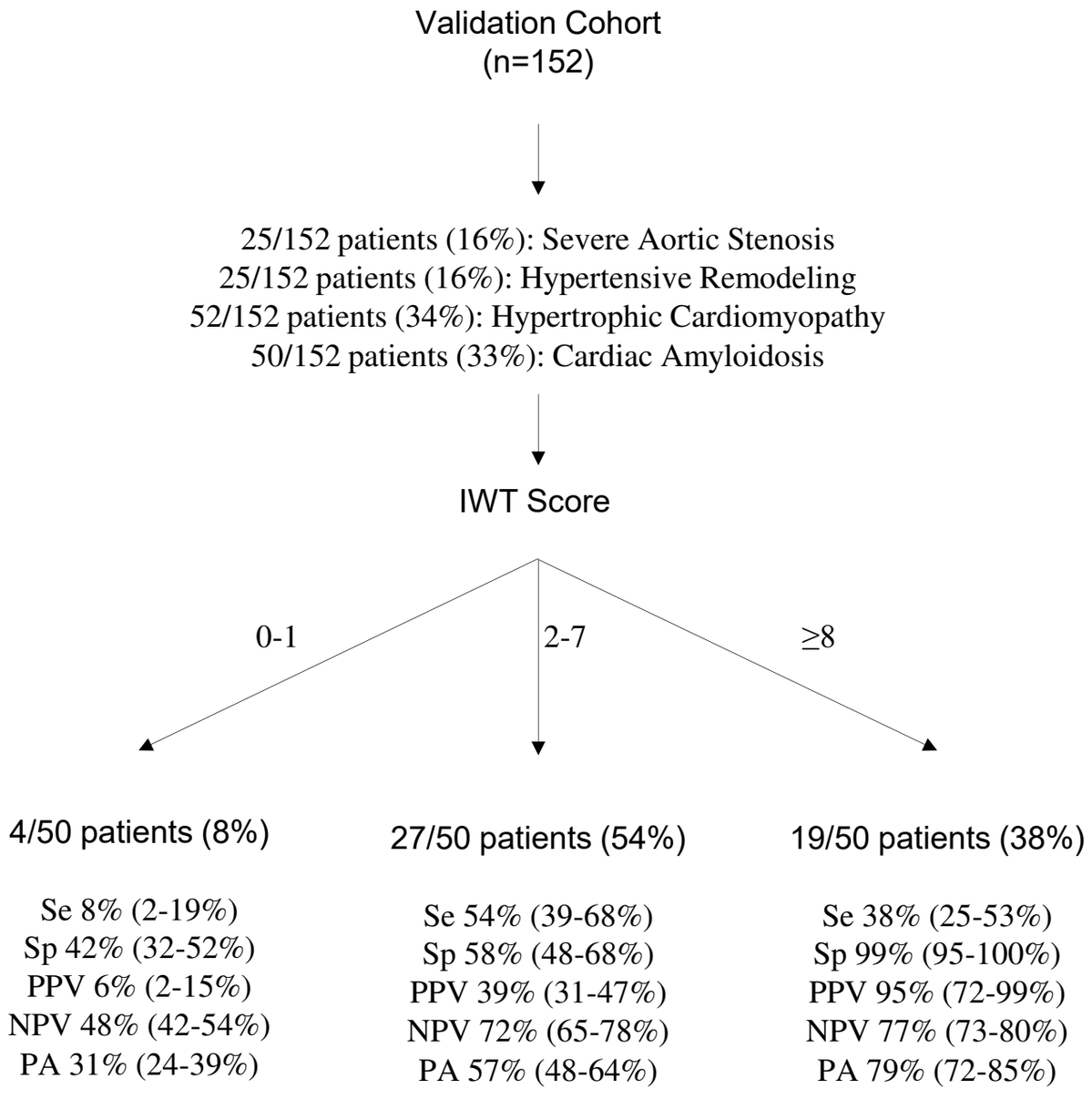
SHORT COMMUNICATION | VOLUME 339, P99-101, SEPTEMBER 15, 2021

External validation of the increased wall thickness score for the diagnosis of cardiac amyloidosis

Emanuele Monda [1] • Giuseppe Palmiero [1] • Michele Lioncino • ... Martina Caiazza • Francesca Dongiglio • Giuseppe Limongelli • Show all authors • Show footnotes

Published: July 18, 2021 • DOI: https://doi.org/10.1016/j.ijcard.2021.07.035 •

A

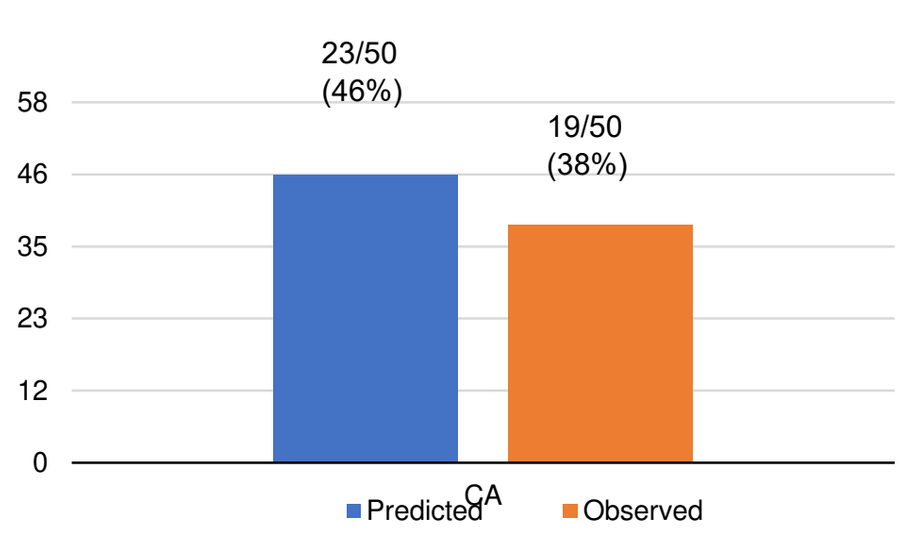


B

IWT Score (0-10 Points)

Parameters	Cut-Off	Points
RWT	>0.6	3
E/e’	>11	1
TAPSE, mm	≤19	2
GLS, %	≥-13	1
SAB	>2.9	3

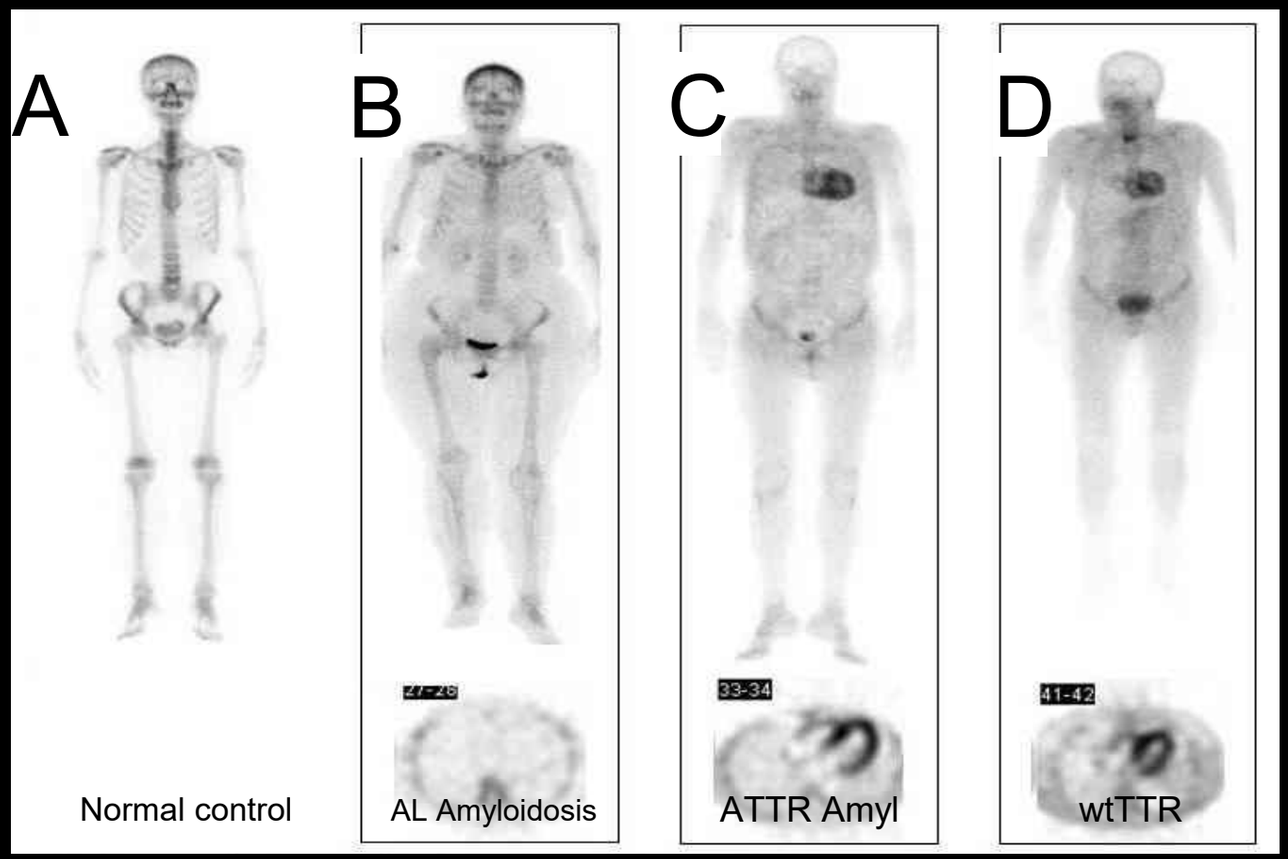
C



What is new?

Imaging

TcDPD Scintigraphy



Perugini et al, J Am Coll Cardiol 2005;46:1076–84

ESC European Society of Cardiology

European Heart Journal (2021) **42**, 1554–1568
doi:10.1093/eurheartj/ehab072

SPECIAL ARTICLE
Heart failure and cardiomyopathies

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

Pablo Garcia-Pavia [1,2,3]*, Claudio Rapezzi[4,5], Yehuda Adler[6], Michael Arad[7], Cristina Basso [3,8,9], Antonio Brucato [10], Ivana Burazor [11], Alida L.P. Caforio [3,12], Thibaud Damy [3,13], Urs Eriksson [14], Marianna Fontana [15], Julian D. Gillmore [15], Esther Gonzalez-Lopez[1,3], Martha Grogan[16], Stephane Heymans[17,18,19], Massimo Imazio [20], Ingrid Kindermann[21], Arnt V. Kristen [22,23], Mathew S. Maurer[24], Giampaolo Merlini [25,26], Antonis Pantazis[27], Sabine Pankuweit[28], Angelos G. Rigopoulos[29], and Ales Linhart [30]

Essential concepts

- Although nine types of cardiac amyloidosis are known, AL and ATTR currently account for the vast majority of cardiac amyloidosis.
- Both invasive and non-invasive diagnostic criteria are accepted to diagnose cardiac amyloidosis. While invasive diagnostic criteria apply to all forms of cardiac amyloidosis, non-invasive criteria are accepted only for ATTR.

Bone & Blood

Diagnostic Pathway

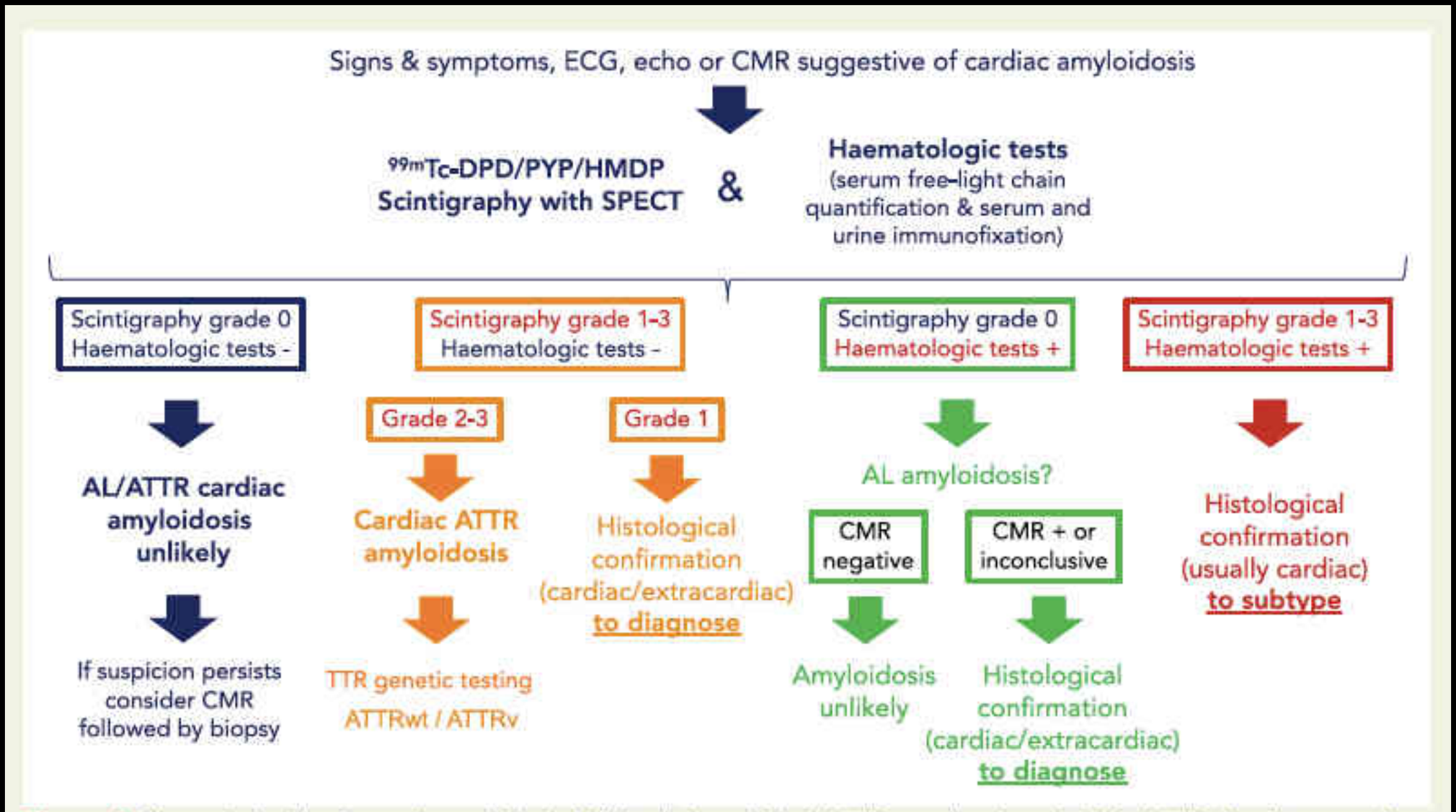


Figure 4 Diagnostic algorithm for cardiac amyloidosis. AL, light-chain amyloidosis;ATTR, transthyretin amyloidosis; ATTRv, hereditary transthyretin amyloidosis; ATTRwt, wild-type transthyretin amyloidosis; CMR, cardiac magnetic resonance; ECG, electrocardiogram; SPECT, single photon emission computed tomography; TTR, transthyretin.

ESC European Society of Cardiology
European Heart Journal (2021) **42**, 1554–1568
doi:10.1093/eurheartj/ehab072

Position Paper Amy 2021

ESC European Society of Cardiology
European Heart Journal (2021) 00, 1–128
doi:10.1093/eurheartj/ehab368

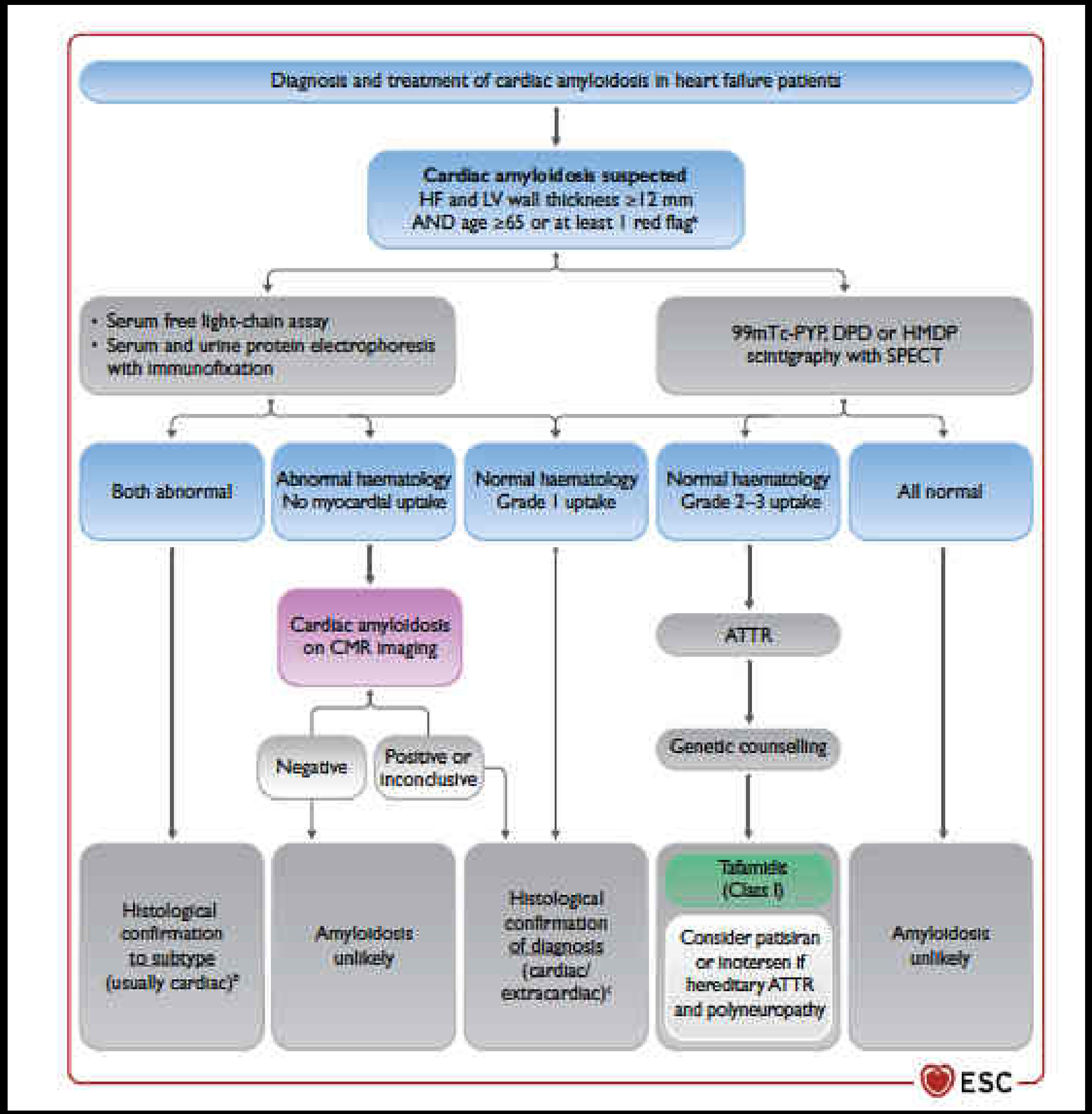
ESC GUIDELINES

2021 ESC Guidelines for the diagnosis and treatment of acute and chronic heart failure

Developed by the Task Force for the diagnosis and treatment of acute and chronic heart failure of the European Society of Cardiology (ESC)

With the special contribution of the Heart Failure Association (HFA) of the ESC

Authors/Task Force Members: Theresa A. McDonagh* (Chairperson) (United Kingdom), Marco Metra* (Chairperson) (Italy), Marianna Adamo (Task Force Coordinator) (Italy), Roy S. Gardner (Task Force Coordinator) (United Kingdom), Andreas Baumbach (United Kingdom), Michael Böhm (Germany), Haran Burri (Switzerland), Javed Butler (United States of America), Jelena Čelutkienė (Lithuania), Ovidiu Chioncel (Romania), John G.F. Cleland (United Kingdom), Andrew J.S. Coats (United Kingdom), Maria G. Crespo-Leiro (Spain), Dimitrios Farmakis (Greece), Martine Gilard (France), Stephane Heymans



Bone & Blood

HF Guidelines 2021

JACC: CARDIOVASCULAR IMAGING
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VOL. ■, NO. ■, 2020

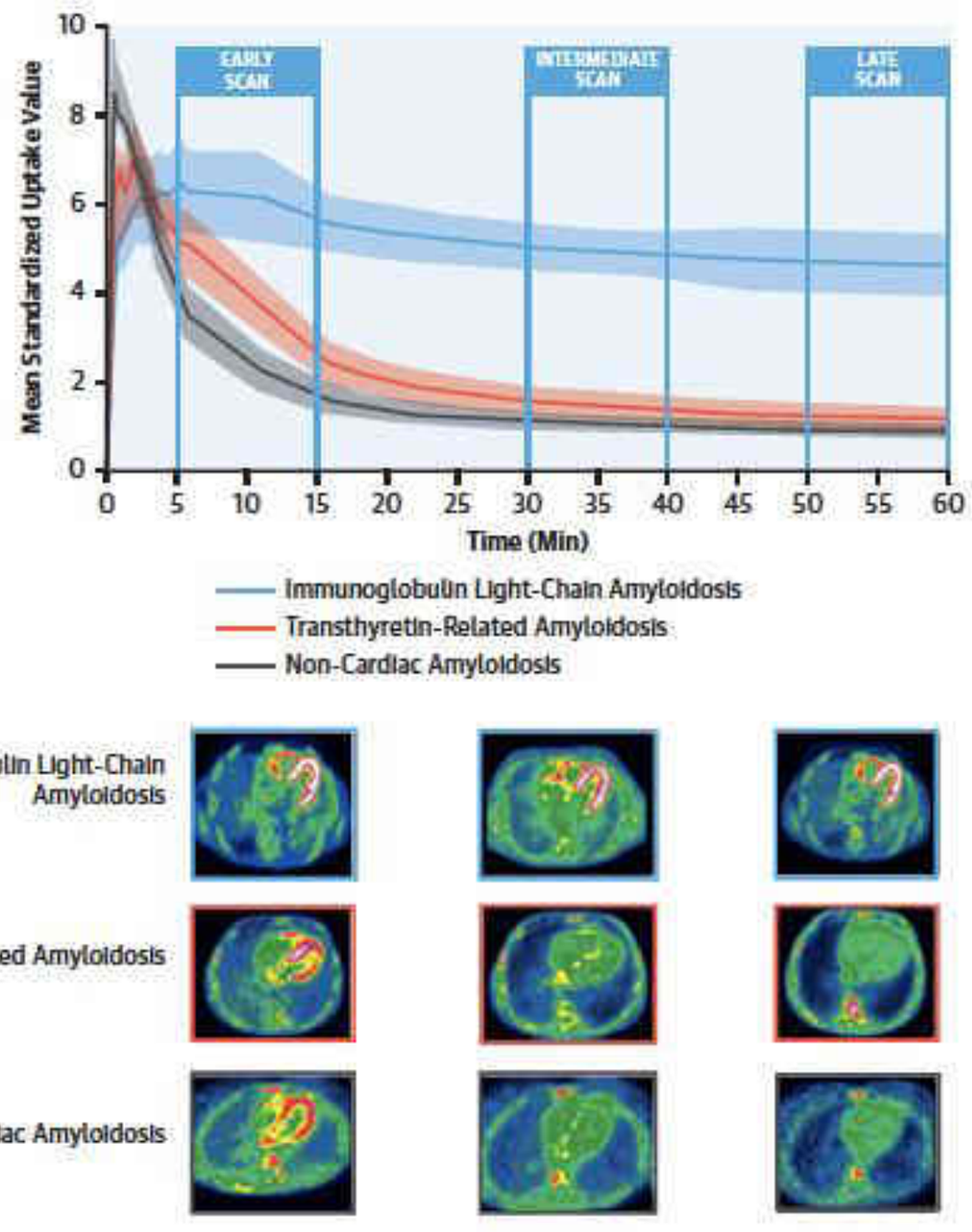
ORIGINAL RESEARCH

[18F]-Florbetaben PET/CT for Differential Diagnosis Among Cardiac Immunoglobulin Light Chain, Transthyretin Amyloidosis, and Mimicking Conditions

Dario Genovesi, MD,[a,*] Giuseppe Vergaro, MD, PhD,[b,c,*] Assuero Giorgetti, MD,[a,*] Paolo Marzullo, MD,[a] Michele Scipioni, EngD,[d] Maria Filomena Santarelli, EngD,[e] Angela Pucci, MD,[f] Gabriele Buda, MD,[g] Elisabetta Volpi, BSc, PhD,[h] Michele Emdin, MD, PhD[b,c]

What next?

CENTRAL ILLUSTRATION Cardiac [18F]-Florbetaben Uptake in Patients With and Without CA



Genovesi, D. et al. J Am Coll Cardiol Img. 2020;■(■):■-■.

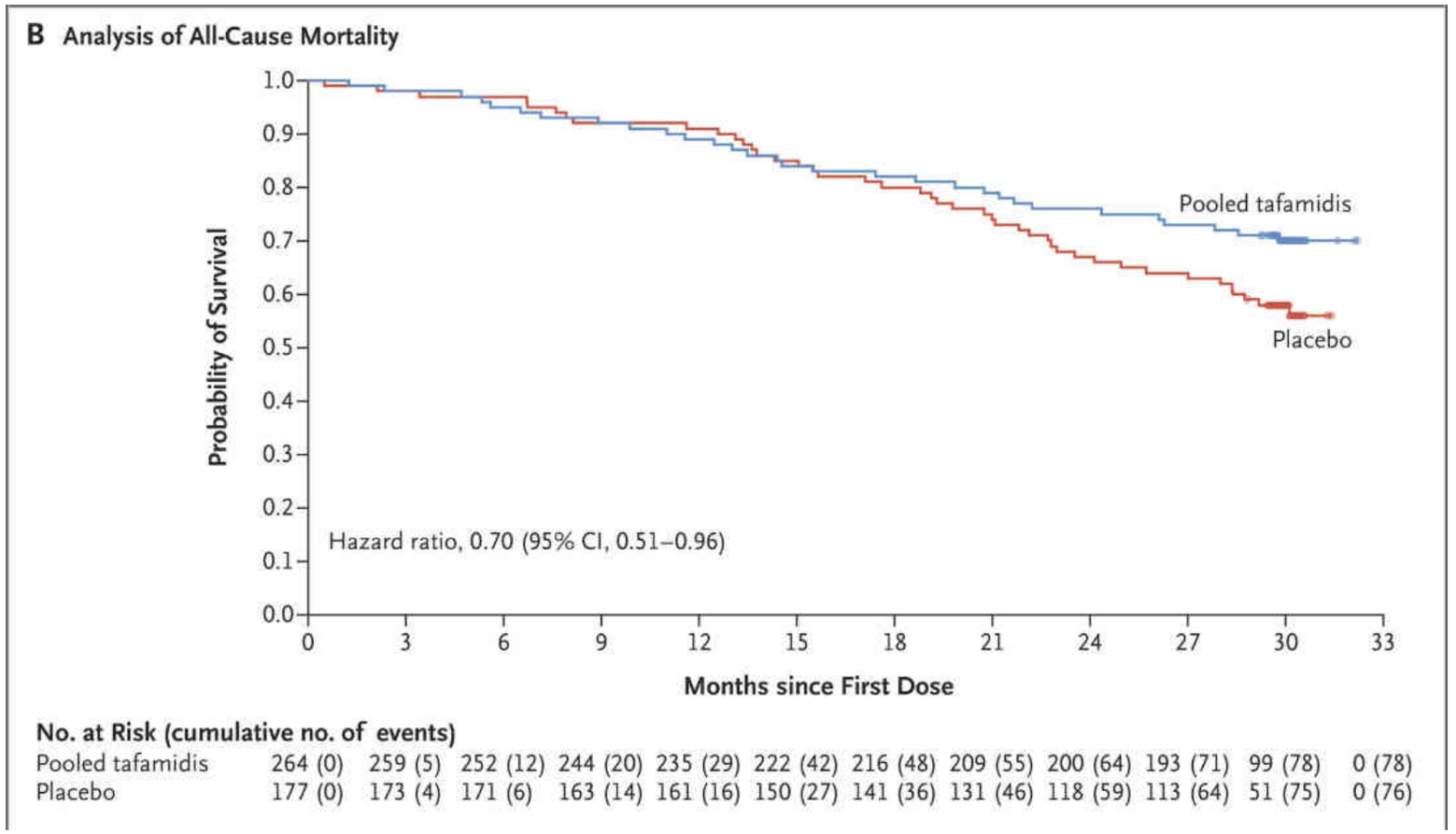
What is new?

Management

ORIGINAL ARTICLE FREE PREVIEW

Tafamidis Treatment for Patients with Transthyretin Amyloid Cardiomyopathy

Mathew S. Maurer, M.D., Jeffrey H. Schwartz, Ph.D., Balarama Gundapaneni, M.S., Perry M. Elliott, M.D., Giampaolo Merlini, M.D., Ph.D., Marcia Waddington-Cruz, M.D., Arnt V. Kristen, M.D., Martha Grogan, M.D., Ronald Witteles, M.D., Thibaud Damy, M.D., Ph.D., Brian M. Drachman, M.D., Sanjiv J. Shah, M.D., et al., for the ATTR-ACT Study Investigators[a]



ESC European Heart Journal (2021) 00, 1–128
European Society of Cardiology doi:10.1093/eurheartj/ehab368

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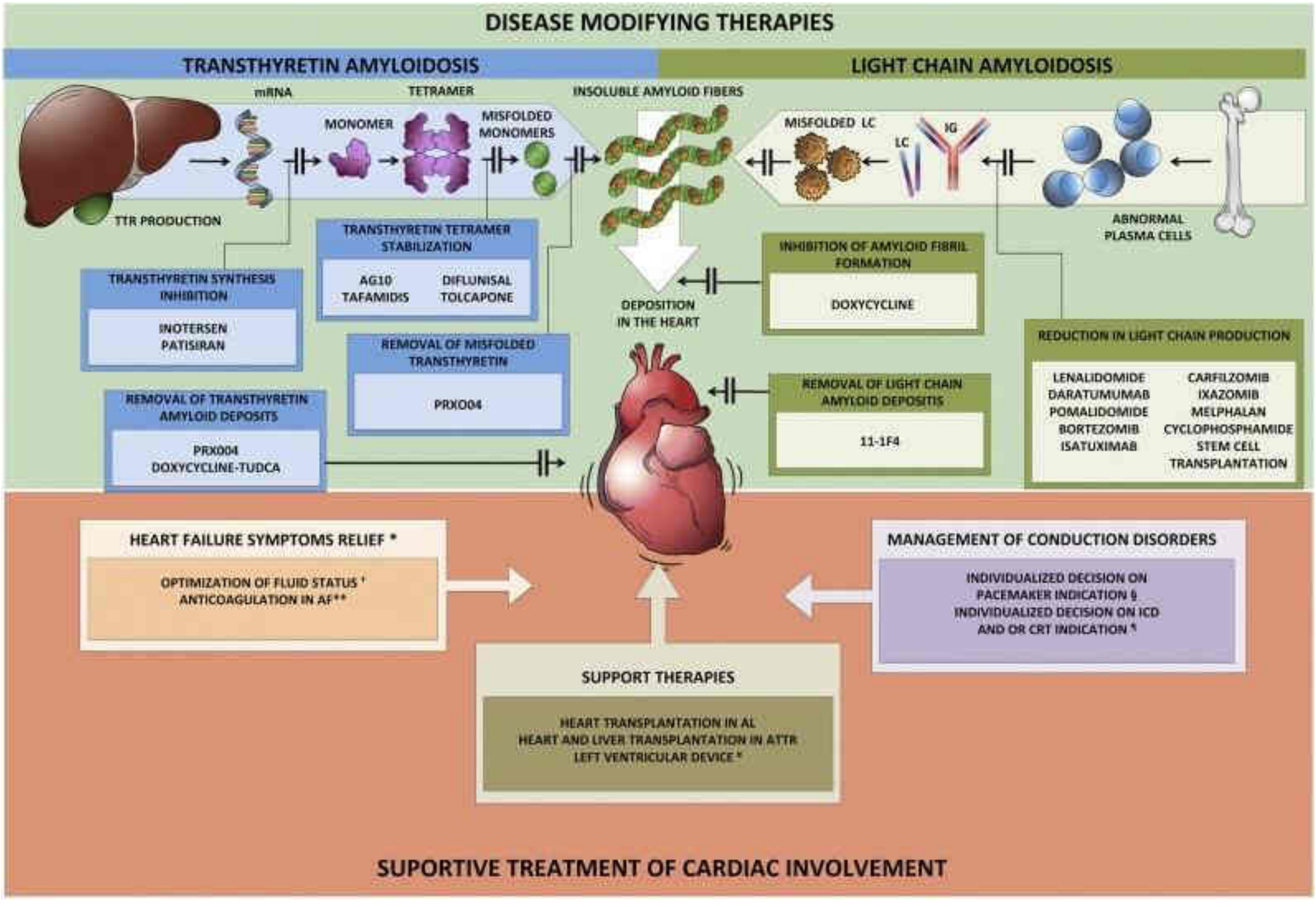
Recommendations for treatment of patients with HF and amyloidosis	
Tafamidis is recommended in patients with genetic testing proven hereditary hTTR-CMP and NYHA class I or II symptoms to reduce symptoms, CV hospitalization and mortality.	I
Tafamidis is recommended in patients with wtTTR-CA and NYHA class I or II symptoms to reduce symptoms, CV hospitalization and mortality.	I


DISEASE MODIFYING THERAPIES
TRANSTHYRETIN AMYLOIDOSIS
LIGHT CHAIN AMYLOIDOSIS
mRNA
TETRAMER
INSOLUBLE AMYLOID FIBERS
MONOMER
MISFOLDED MONOMERS
MISFOLDED LC
LC
IG
TTR PRODUCTION
ABNORMAL PLASMA CELLS
TRANSTHYRETIN TETRAMER STABILIZATION
AG10
TAFAMIDIS
DIFLUNISAL
TOLCAPONE
TRANSTHYRETIN SYNTHESIS INHIBITION
INOTERSEN
PATISIRAN
INHIBITION OF AMYLOID FIBRIL FORMATION
DOXYCYCLINE
DEPOSITION IN THE HEART
REMOVAL OF MISFOLDED TRANSTHYRETIN
PRX004
REDUCTION IN LIGHT CHAIN PRODUCTION
LENALIDOMIDE
DARATUMUMAB
POMALIDOMIDE
BORTEZOMIB
ISATUXIMAB
CARFILZOMIB
IXAZOMIB
MELPHALAN
CYCLOPHOSPHAMIDE
STEM CELL TRANSPLANTATION
REMOVAL OF TRANSTHYRETIN AMYLOID DEPOSITS
PRX004
DOXYCYCLINE-TUDCA
REMOVAL OF LIGHT CHAIN AMYLOID DEPOSITS
11-1F4
HEART FAILURE SYMPTOMS RELIEF *
OPTIMIZATION OF FLUID STATUS †
ANTICOAGULATION IN AF**
MANAGEMENT OF CONDUCTION DISORDERS
INDIVIDUALIZED DECISION ON PACEMAKER INDICATION §
INDIVIDUALIZED DECISION ON ICD AND OR CRT INDICATION ¶
SUPPORT THERAPIES
HEART TRANSPLANTATION IN AL
HEART AND LIVER TRANSPLANTATION IN ATTR
LEFT VENTRICULAR DEVICE ‡
SUPORTIVE TREATMENT OF CARDIAC INVOLVEMENT

TAKE HOME

ESC SPECIAL ARTICLE

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

Pablo Garcia-Pavia, Claudio Rapezzi, Yehuda Adler, Michael Arad, Cristina Basso, Antonio Brucato, Ivana Burazor, Alida L.P. Caforio, Thibaud Damy, Urs Eriksson, Marianna Fontana, Julian D. Gillmore, Eloy Gonzalez-Lopez, Martha Grogan, Stephane Heymans, Maurizio Imazio, Ingrid Kindermann, Arnt V. Kristen, Mathew S. Maurer, Giampaolo Merlini, Antonis Pantazis, Sabine Pankuweit, Angelos G. Rigopoulos, and Alex Linhart

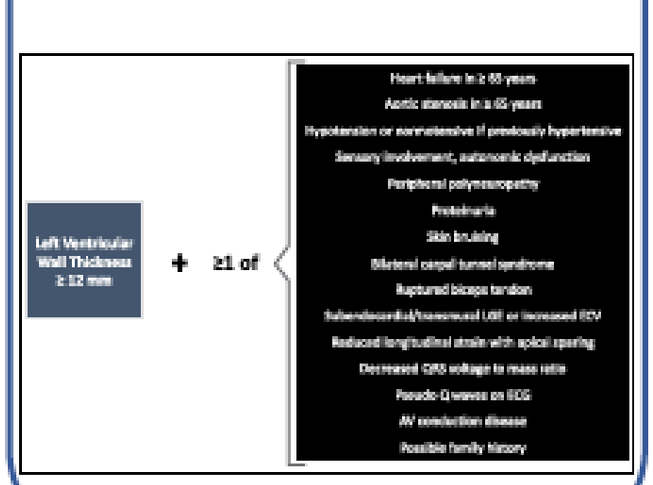
Cardiac amyloidosis

ESC Myocardial WG position paper

SUSPECT

Screen if

Left ventricle wall thickness ≥ 12 mm
&
≥ 1 Red Flag or Clinical Scenario



DIAGNOSIS

Diagnostic criteria

Invasive (all types)	Non-Invasive (only for ATTR)
Cardiac Biopsy positive for amyloid or Extracardiac Biopsy positive for amyloid + Echocardiographic/CMR criteria	Grade 2 or 3 cardiac uptake at diphosponate Scintigraphy + Negative serum free light chains & negative serum and urine immunofixation (SPIE & UPIE) + Echocardiographic/CMR criteria

Diagnostic algorithm

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

Diagnosis made or proceed to **CMR and/or biopsy** according to results

TREATMENT

Cardiac complications and comorbidities

- Heart Failure
- Thromboembolism
- Atrial fibrillation
- Conduction disorders
- Ventricular arrhythmias
- Aortic stenosis

Disease modifying treatment

- **ATTR**: genetic silencers, stabilizers and removers.
- **AL**: chemotherapy and ASCT.
- **AA**: anti-inflammatory, anti-infective and immunosuppressive drugs.

Essential concepts

- Cardiac amyloidosis should be considered in patients with increased wall thickness in the presence of cardiac or extracardiac red flags and/or in specific clinical situations.
- A diagnostic algorithm based initially on the use of bone scintigraphy coupled to assessment for monoclonal proteins allows appropriate diagnosis in patients with suggestive signs/symptoms.

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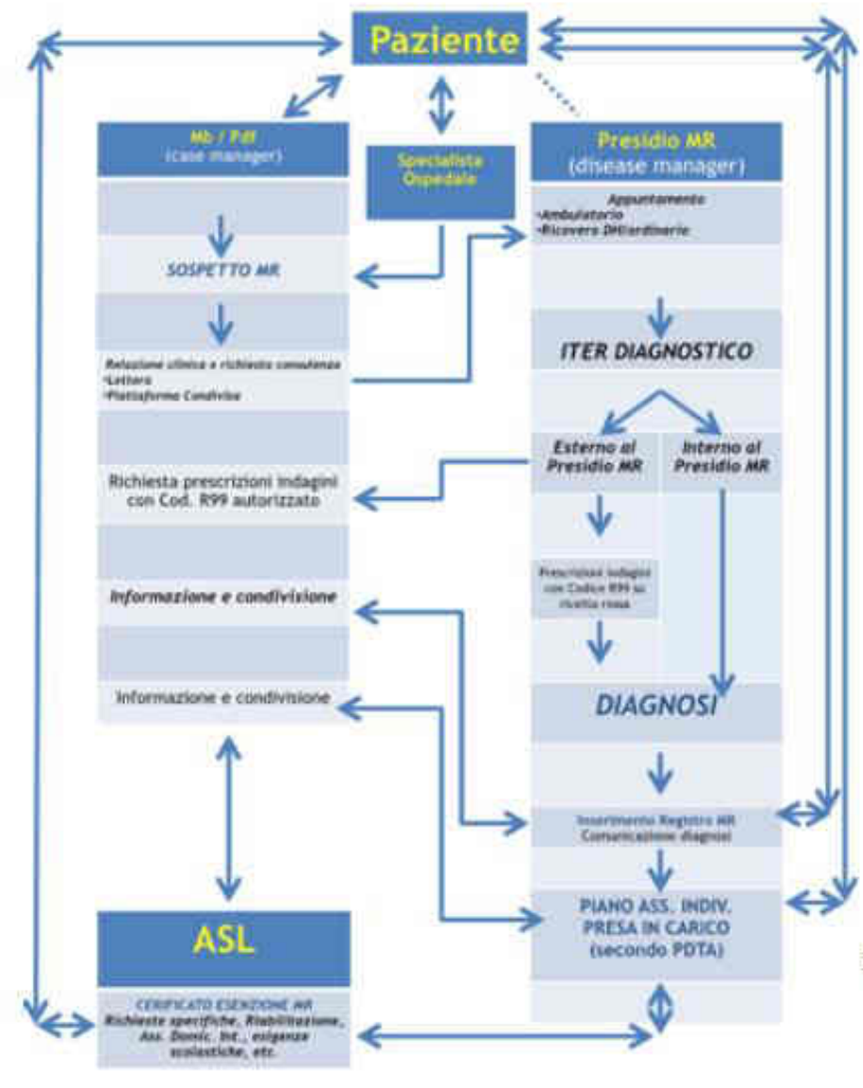
doi:10.1093/eurheartj/ehab072



FASE I
Sospetto

FASE II
Iter Diagnostico

FASE III
Diagnosi

FASE IV
Presa in Carico

Regione Campania
Il Commissario ad Acta per l'attuazione
del Piano di rientro dai disavanzi del SSR campano
(Deliberazione Consiglio dei Ministri 10/07/2017)

DECRETO N. 48 DEL 27/10/2017

OGGETTO: *Approvazione Piano Regionale Malattie Rare e del Documento Percorso Diagnostico Assistenziale del paziente raro (acta vii)*

Giunta Regionale della Campania

DECRETO DIRIGENZIALE

DIRETTORE GENERALE/
DIRIGENTE UFFICIO/STRUTTURA — **dott. Postiglione Antonio**
DIRIGENTE UNITA' OPERATIVA DIR. / DIRIGENTE STAFF — **Dott.ssa Romano Maria Rosaria**

DECRETO N°	DEL	DIREZ. GENERALE / UFFICIO / STRUTT.	UOD / STAFF
143	**21/05/2020**	**4**	**0**

Oggetto:
Adozione Percorso Diagnostico Terapeutico Assistenziale per la Gastroenterite Eosinofila del paziente raro (RI0030)

Giunta Regionale della Campania

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446	**29/12/2020**	**4**	**0**

Oggetto:
Adozione Percorsi Diagnostico. Terapeutici Assistenziali per le seguenti patologie rare: Retinite Pigmentosa (RFG110), Ipertensione Polmonare Arteriosa idiopatica (RGO120), Fibrosi Polmonare (RHG010) e Pemfigo (RLO030).

Percorso diagnostico terapeutico assistenziale
PDTA- Regione Campania

Grazie per l'attenzione...