31 GIORNATE CARDIOLOGICHE TORINESI

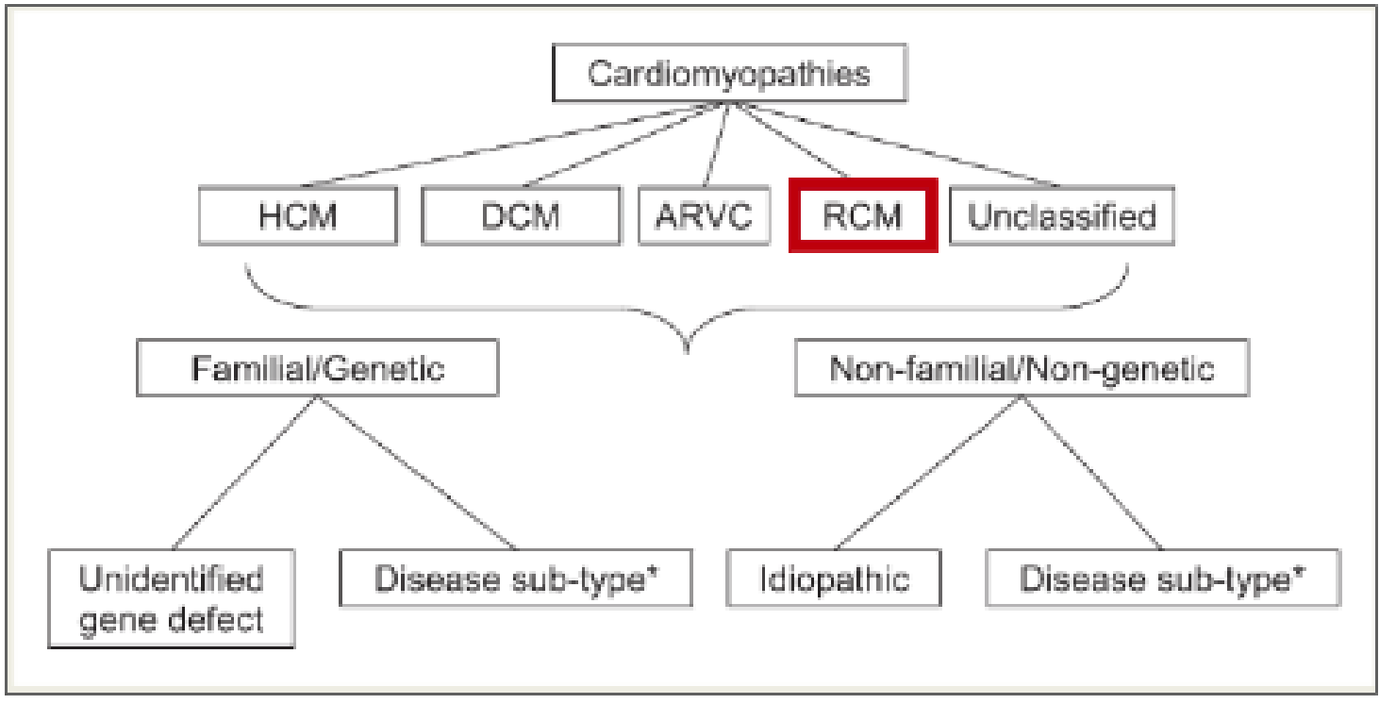
TURIN
October
24th-26th
2019

AMYLOIDOSIS AND INFILTRATIVE CARDIOMYOPATHIES

ANTONELLA FAVA
CARDIOLOGIA UNIVERSITARIA
CITTÀ DELLA SALUTE E DELLA SCIENZA
TORINO

INFILTRATIVE CARDIOMYOPATHIES

Cardiomyopathies characterized by the deposition of substances (iron, proteins or glycogen) that cause the ventricular walls to become progressively rigid, thereby impeding ventricular filling.



Classification of the cardiomyopathies: a position statement from the european society of cardiology working group on myocardial and pericardial diseases

European Heart Journal (2008) **29**, 270–276
doi:10.1093/eurheartj/ehm342

Infiltrative cardiomyopathies: phenotypic classification

RESTRICTIVE PHYSIOLOGY

- Echocardiogram shows preserved left ventricular ejection fraction (LVEF) with **increased** LV wall thickness
  - Hypertensive heart disease *(patient has history of uncontrolled hypertension)*
  - **Infiltrative restrictive cardiomyopathy**
    - **Amyloidosis**
    - **Glycogen storage diseases such as Fabry disease**
  - Hypertrophic cardiomyopathy *(patient has LVOT gradient, SAM of mitral valve, or asymmetric septal hypertrophy)*
- Echocardiogram shows preserved LVEF with **normal** LV wall thickness
  - High output heart failure *(patient has increased cardiac output, arteriovenous shunts, liver disease, hyperthyroid, myeloproliferative disorders)*
  - Restrictive cardiomyopathy (RCM) *(patient has normal LV chamber volumes, biatrial enlargement, increased LV filling pressures)*
    - Endomyocardial fibrosis
    - Radiation heart disease
    - **Iron overload cardiomyopathy**
    - Idiopathic RCM
  - Constrictive pericarditis *(patient has thickened pericardium, respirophasic discordance in LV-RV pressure and in mitral tricuspid inflow velocities)*

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

AMYLOIDOSIS

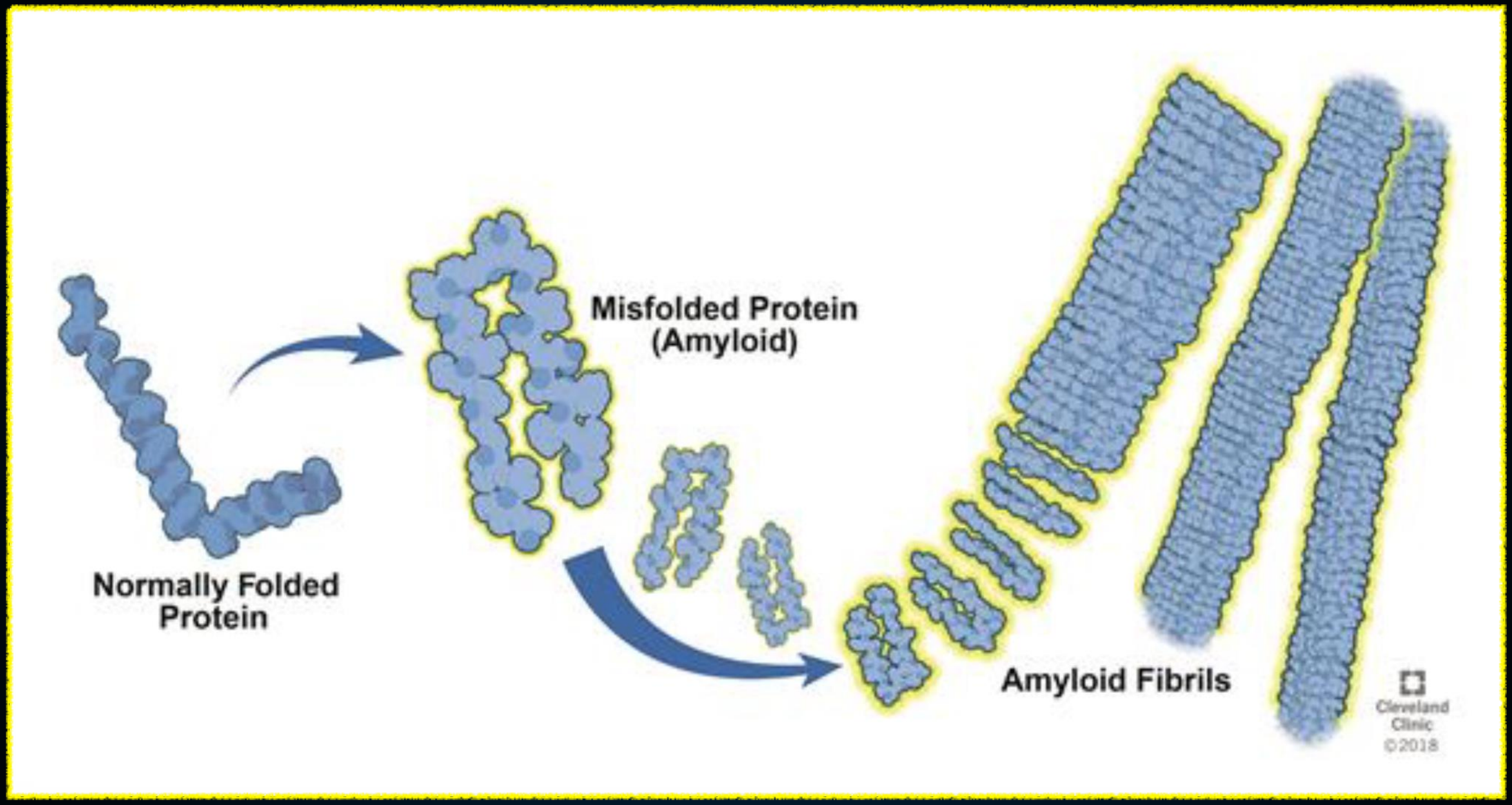
- **AMYLOIDOSIS** is a protein disorder by **accumulation of pathogenic amyloids**: **aggregates of misfolded proteins** accumulate in a variety of organs, **disrupt** their tissue **architecture** and **impair their function**

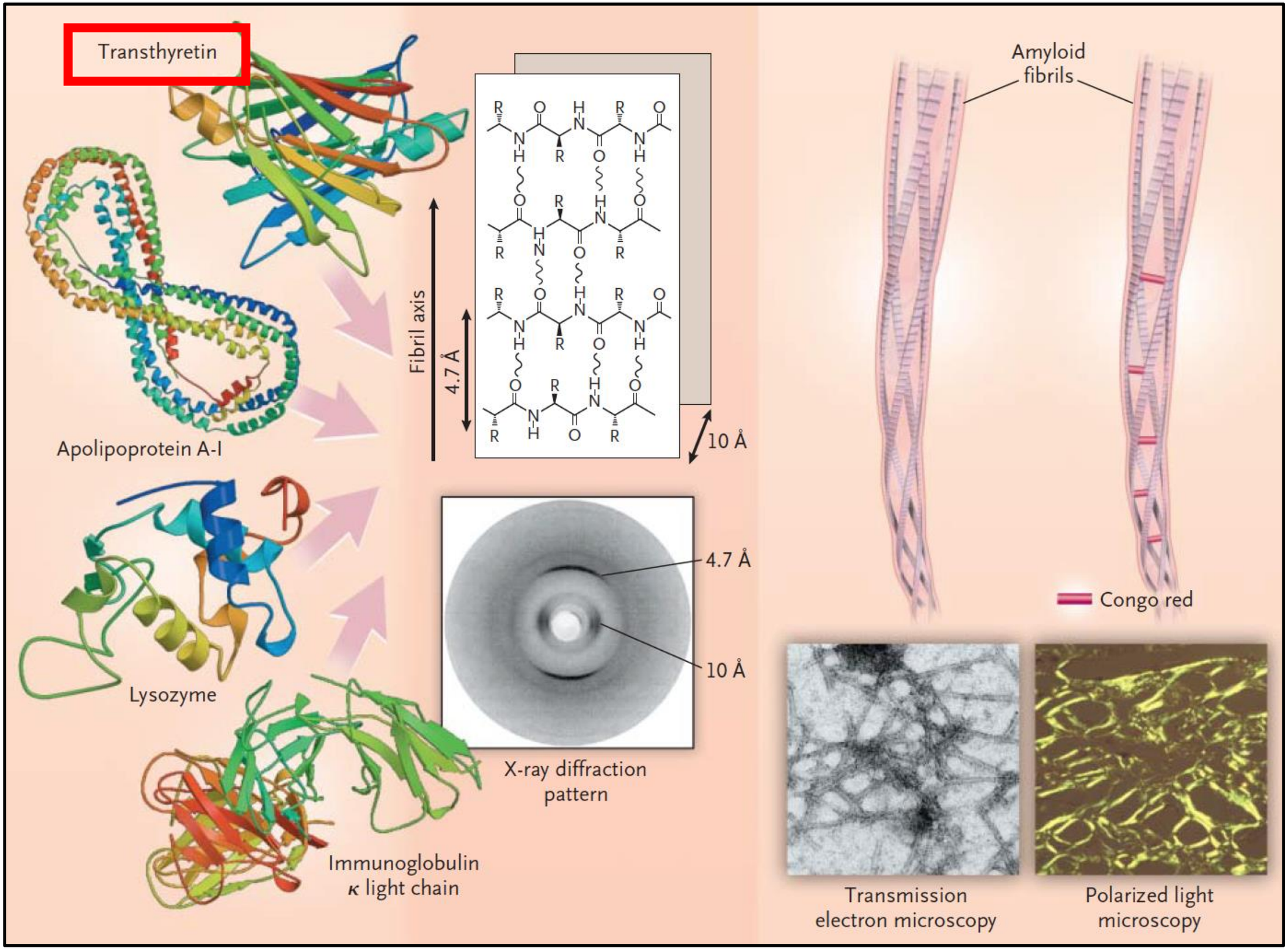
- The **clinical manifestations and prognosis** vary **widely** depending on the specific type of the affected protein.

- **Incidence = 14 / million / year**

- **Prevalence = < 5 / 10,000 inhabitants → RARE DISEASE !**

Amyloid Fibrils Formation





Merlini G, Bellotti V. Molecular mechanisms of amyloidosis. N Engl J Med. 2003;349:583–596. doi: 10.1056/NEJMra023144

Table 1. Amyloid fibril proteins and their precursors in human[a].

Fibril protein	Precursor protein	Systemic and/or localized	Acquired or hereditary	Target organs
AL	Immunoglobulin light chain	S, L	A, H	All organs, usually except CNS
AH	Immunoglobulin heavy chain	S, L	A	All organs except CNS
AA	(Apo) Serum amyloid A	S	A	All organs except CNS
ATTR	Transthyretin, wild type	S	A	Heart mainly in males, Lung, Ligaments, Tenosynovium
	Transthyretin, variants	S	H	PNS, ANS, heart, eye, leptomen.
Aβ2M	β2-Microglobulin, wild type	S	A	Musculoskeletal System
	β2-Microglobulin, variant	S	H	ANS
AApoAI	Apolipoprotein A I, variants	S	H	Heart, liver, kidney, PNS, testis, larynx (C terminal variants), skin (C terminal variants)
AApoAII	Apolipoprotein A II, variants	S	H	Kidney
AApoAIV	Apolipoprotein A IV, wild type	S	A	Kidney medulla and systemic
AApoCII	Apolipoprotein C II, variants	S	H	Kidney
AApoCIII	Apolipoprotein C III, variants	S	H	Kidney
Agel	Gelsolin, variants	S	H	PNS, cornea
ALys	Lysozyme, variants	S	H	Kidney
ALECT2	Leukocyte Chemotactic Factor-2	S	A	Kidney, primarily
AFib	Fibrinogen α, variants	S	H	Kidney, primarily
ACys	Cystatin C, variants	S	H	PNS, skin
ABri	ABriPP, variants	S	H	CNS
ADan*	ADanPP, variants	L	H	CNS
Aβ	Aβ protein precursor, wild type	L	A	CNS
	Aβ protein precursor, variant	L	H	CNS
AαSyn	α-Synuclein	L	A	CNS
ATau	Tau	L	A	CNS
APrP	Prion protein, wild type	L	A	CJD, fatal insomnia
	Prion protein variants	L	H	CJD, GSS syndrome, fatal insomnia
	Prion protein variant	S	H	PNS
ACal	(Pro)calcitonin	L	A	C-cell thyroid tumors
AIAPP	Islet amyloid polypeptide**	L	A	Islets of Langerhans, insulinomas
AANF	Atrial natriuretic factor	L	A	Cardiac atria
APro	Prolactin	L	A	Pituitary prolactinomas, aging pituitary
AIns	Insulin	L	A	Iatrogenic, local injection
ASPC***	Lung surfactant protein	L	A	Lung
AGal7	Galectin 7	L	A	Skin
ACor	Corneodesmosin	L	A	Cornified epithelia, hair follicles
AMed	Lactadherin	L	A	Senile aortic media
AKer	Kerato-epithelin	L	A	Cornea, hereditary
ALac	Lactoferrin	L	A	Cornea
AOAAP	Odontogenic ameloblast-associated protein	L	A	Odontogenic tumors
ASem1	Semenogelin 1	L	A	Vesicula seminalis
AEnf	Enfurvitide	L	A	Iatrogenic
ACatK****	Cathepsin K	L	A	Tumor associated

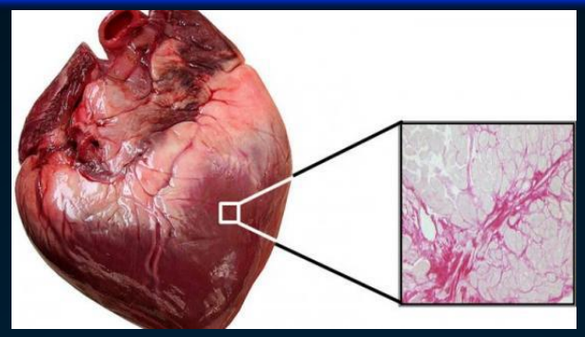
[a]Proteins are listed, when possible, according to relationship. Thus, apolipoproteins are grouped together, as are polypeptide hormones.
*ADan is the product of the same gene as ABri.
**Also called amylin.
***Not proven by amino acid sequence analysis.
****Full amino acid sequence to be established.

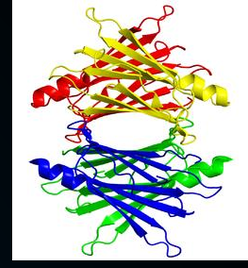
} 85%

→ 15%

More than 30 different types of amyloidosis, each due to a specific protein. Some are **genetic**, while others are **acquired**.

AMILOIDOSI CARDIACA





- **AL (light chain amyloidosis)**
- **ATTR (transtiretina amyloidosis)**
  - **wild-type (senile SSA)**
  - **hereditary (mutata H-ATTR)**

AMILOIDOSI AL (LIGHT CHAIN AMYLOIDOSIS)

- Clonal population of bone marrow **plasma cells** that produces a **clonal light chain** of **κ** or **λ** type as either an intact molecule or a fragment

- The clonal plasma cells express light chains of the **λ isotype more frequently than the κ**, with a ratio of approximately **3:1**, despite the greater proportion of κ than λ expressing PC in a normal bone marrow

- **Prevalence** of AL amyloidosis in **Multiple Myeloma**: **12-15%**

- **Median age at diagnosis = 63 years** and only 1.3% of Pts < 34 y

- There is a **male predominance** (55% of Pts).

- AL-A occurs in all races and geographic locations, but data are limited regarding the incidence of AL-A across different ethnic groups

AMILOIDOSI AL

Heart
- Heart failure with preserved ejection fraction
- Thickened ventricular walls and low voltages on electrocardiography
- Dysponea at rest or exertion, fatigue
- Hypotension or syncope
- Peripheral oedema

71%

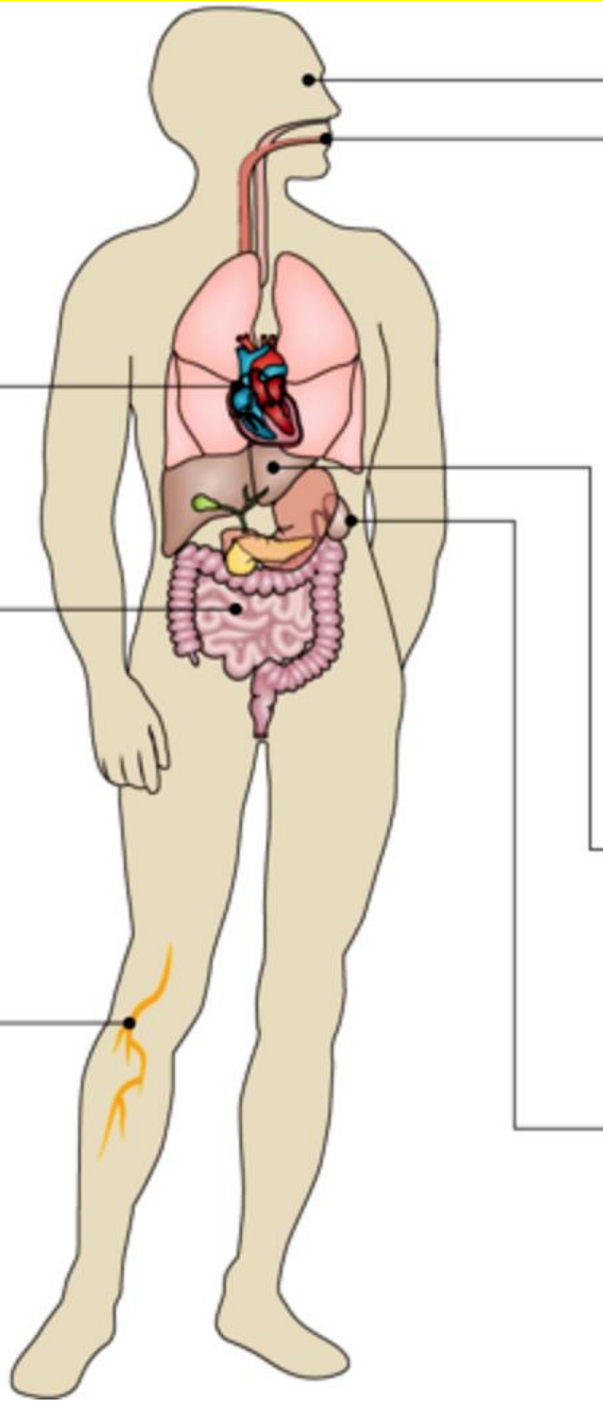
Gastrointestinal tract
- Malabsorption and weight loss
- Bleeding (factor X)

22%

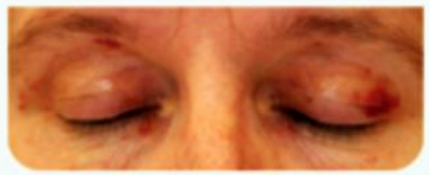
Nervous system
Peripheral
- Symmetric lower extremity sensorimotor polyneuropathy

Carpal tunnel syndrome (bilateral)
Autonomic
- Postural hypotension
- Erectile dysfunction (males)
- Gastrointestinal motility alterations

23%

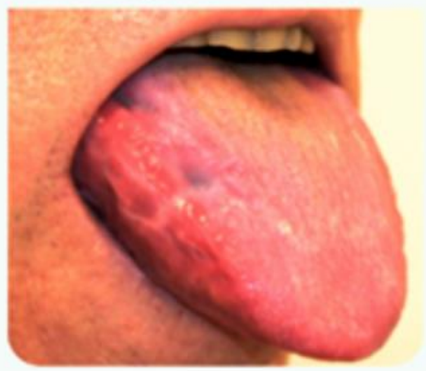


Periorbital purpura



10%

Macroglossia



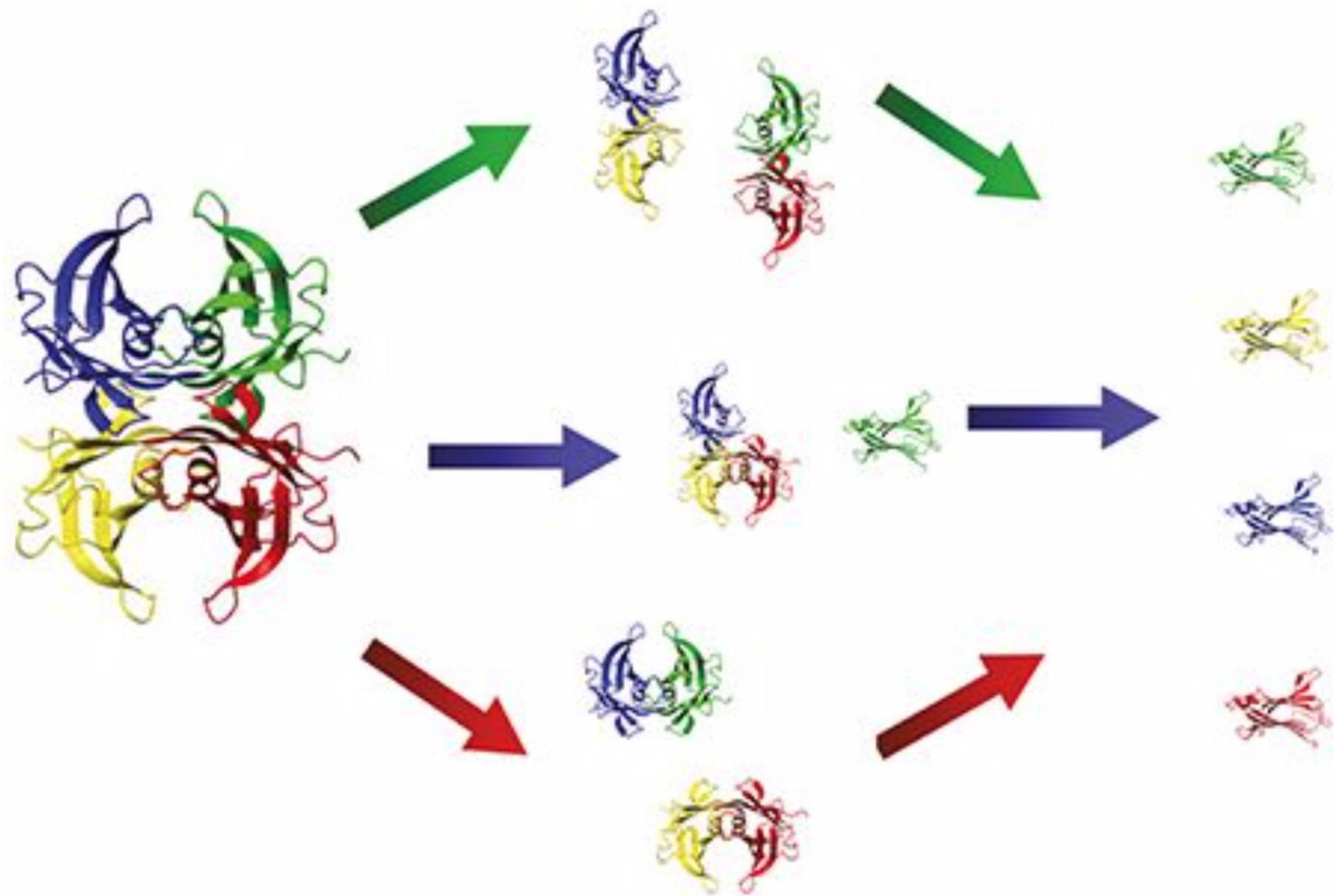
16%

Liver
- Increased alkaline phosphatase
- Hepatomegaly

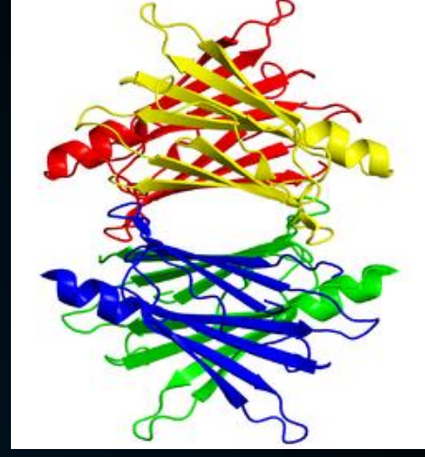
58%

Kidney
- Nephrotic range proteinuria
- Renal failure
- Peripheral oedema

TTR-AMYLOIDOSIS

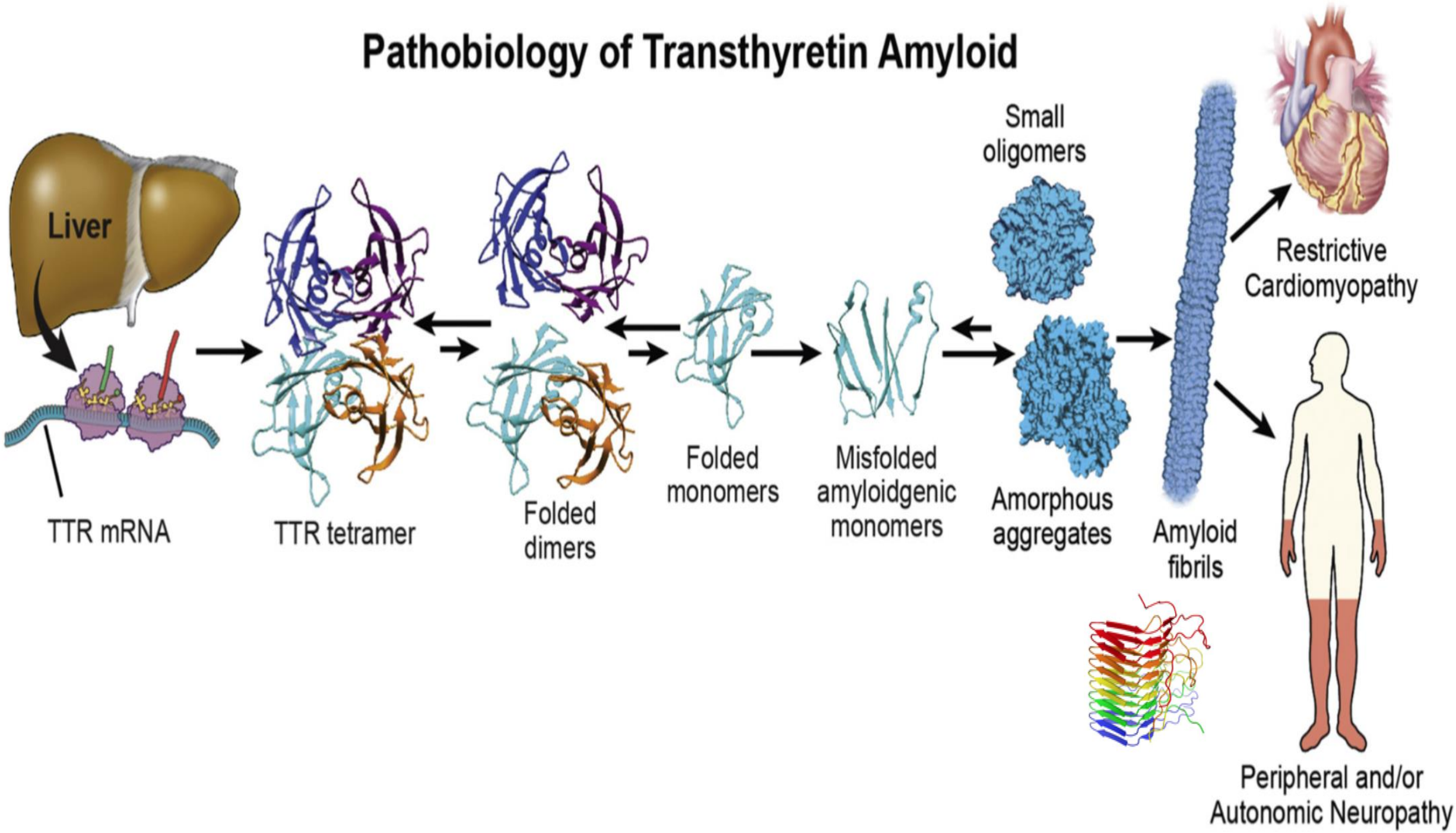


TRANSTIRETINA

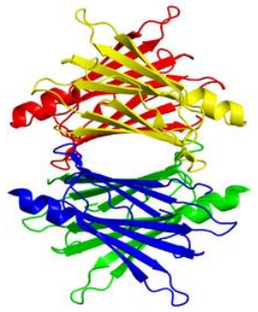


- PROTEINA SERICA (“PRE-ALBUMINA”)
- COMPOSTA DA 4 ***β***-SHEET MONOMERI E CIRCOLA COME **TETRAMERO SOLUBILE**
- DEPUTATA AL TRASPORTO DELLA TIROXINA E DELLA PROTEINA LEGANTE IL RETINOLO
- PRODOTTA PRINCIPALMENTE DAL **FEGATO** ED IN PICCOLA PARTE DAL PLESSO CORIOIDEO E DALLA RETINA

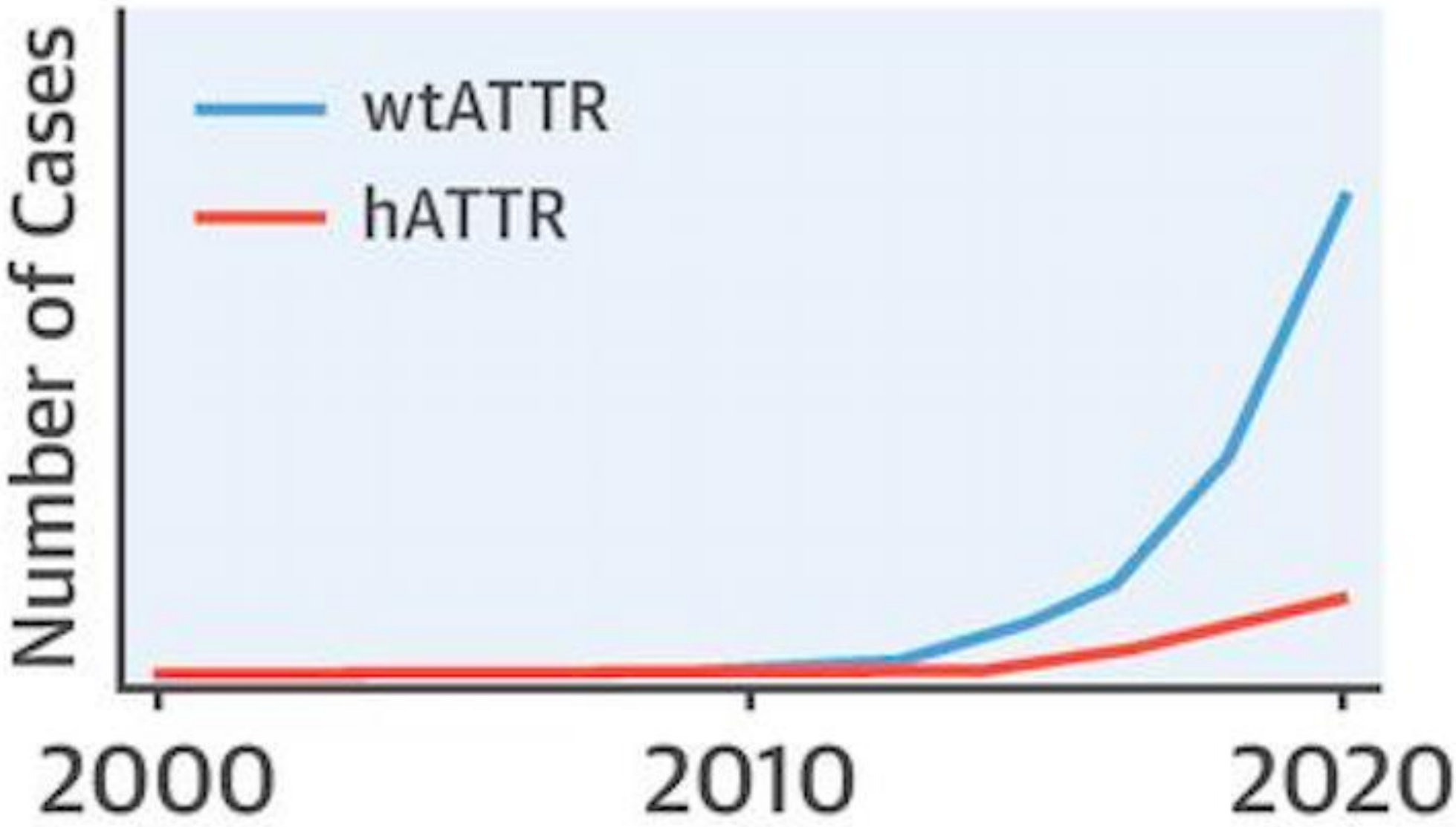
Pathobiology of Transthyretin Amyloid



Condizione sottodiagnosticata!



…un fenomeno in espansione…



J Am Coll Cardiol 2019;73:2872-91

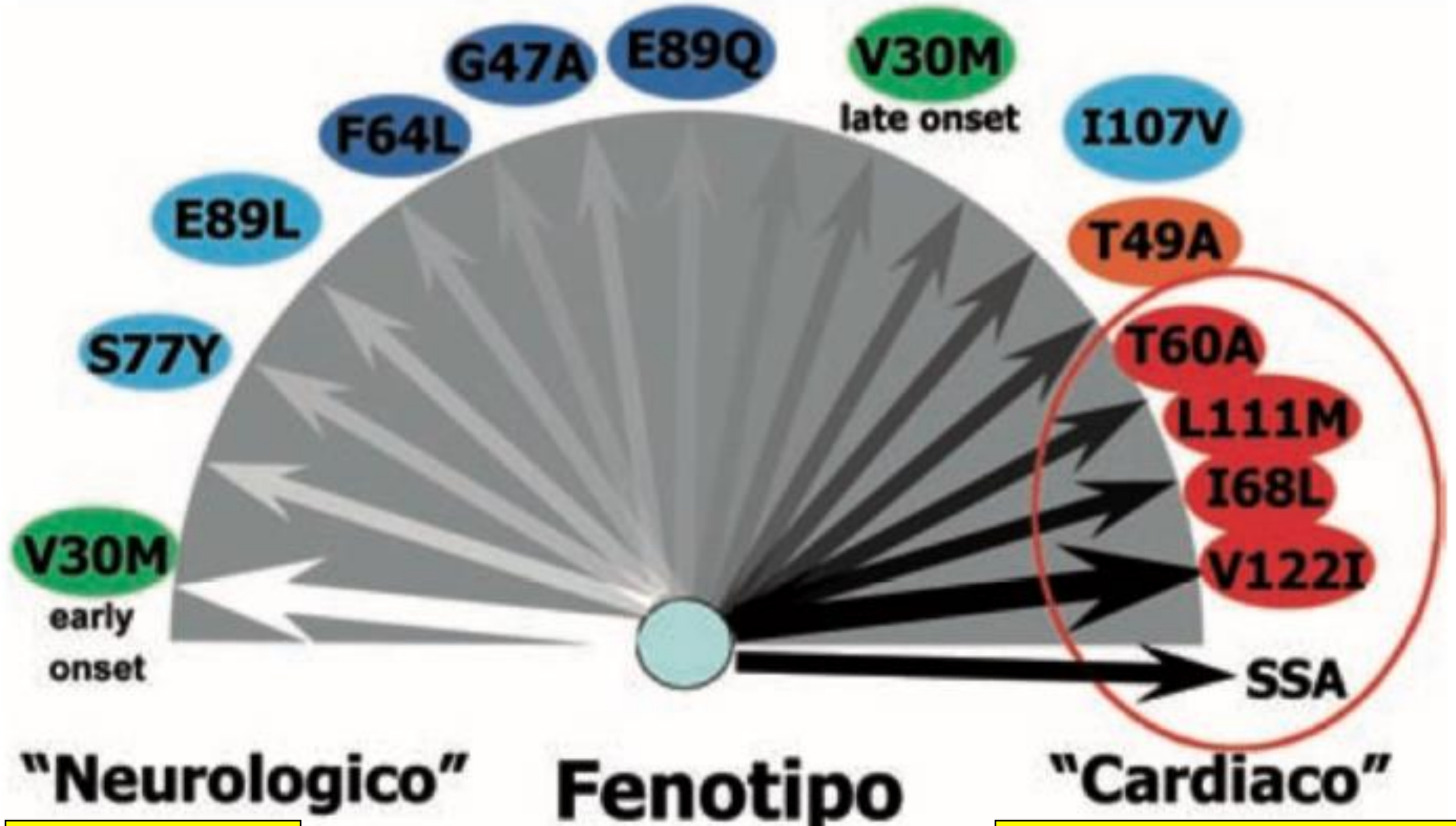
WT-ATTR (SENILE)

- Anche detta **SSA** (**S**ystemic **S**enile **A**myloidosis)
- La sequenza genetica della TTR è normale
- Non è chiaro perchè la proteina wt diventi instabile e aggreghi
- Correlata ai processi di **invecchiamento**
- **Fenotipo** generalmente e prevalentemente **cardiologico**
- Neuro: **sindrome tunnel carpale bilaterale** (può anticipare anche di 10 anni le manifestazioni cardiologiche)
- Frequente riscontro **post-mortem >80 aa**

H-ATTR (EREDITARIA O MUTATA)

- Il **gene TTR** si trova sul **cromosoma 18**
- Nella hATTR ci sono **mutazioni** di singoli aminoacidi nella **sequenza 127**
- Nomenclatura: aa normale - posizione - aa sostituito (es. **Val30Met**)
- **Autosomica dominante**
- Particolare tropismo per **tessuto nervoso e cardiaco**
- “**Case-mix**”

Correlazione genotipo-fenotipo nell'ATTr



Val30Met "early onset"

Thr60Ala - Leu111Met - Ile68Leu - Val122Ile

THAOS database

Rapezzi EHJ 2013

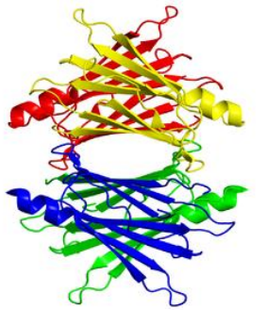
ATTR: clinical features at presentation

	AL[39,47]	ATTRwt[34,37,39]	Val122Ile[34]	Ile 68Leu[31]	Thr60Ala[33]
Median age at diagnosis, y	62	76	70	70	62
Males, %	66	95	75	75	70
Common ethnicity	Variable	White	African American Caribbean	White (Italy)	White (United States, Ireland)
Cardiac referral route, %	65	>80	>80	>80	30
IVS/PW (median values)	15/14	18/17	17/17	17/16	17/17
LVEF, %	56	50	50	50	53
Low QRS voltages, %	45	33	45	30	16
Peripheral sensory-motor neuropath, %	10–20	<10	15	25	54
History of carpal tunnel syndrome, %	<10	30–45	30	37	Unknown
Autonomic symptoms, %	24	12–20	10	<10	75
Median survival, y	Depends on stage	3.5	2–3	4–5	3.5
NT-pro-BNP, pg/L (median)	↑↑↑	↑↑	↑	↑	↑

AL, immunoglobulin light chain; ATTR, amyloid transthyretin; ATTRwt, wild-type ATTR; CA, cardiac amyloidosis; IVS, interventricular septum; LVEF, left ventricular ejection fraction; NT-pro-BNP, N-terminal pro brain natriuretic peptide; and PW, posterior wall.

Circulation 2017

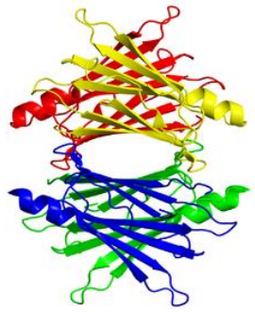
ATTr - CM?



1) Pz già con dg di hATTr inviato dal neurologo -> cercare i SEGNI di amiloidosi cardiaca:

- **ECG**
- **ECOCARDIOGRAMMA**

2) Pz con problema cardiologico senza apparente malattia neurologica (o compromissione neurologica ad etiologia sconosciuta) -> DIAGNOSI DIFFICILE !

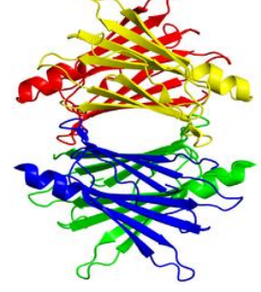


ETEROGENEITÀ CLINICA: UNA SFIDA!

L'AMILOIDE PUÒ INFILTRARE QUALSIASI STRUTTURA CARDIOVASCOLARE:

- **SISTEMA DI CONDUZIONE**
- **PARETI VENTRICOLARI E SETTI (SIA) -> FENOTIPO "IPERTROFICO"**
- **APPARATI VALVOLARI**
- **PERICARDIO**

ATTR - CM?



- **SEGNI e SINTOMI di INSUFFICIENZA CARDIACA SINISTRA senza causa evidente**
- **INSUFFICIENZA CARDIACA DESTRA in CARDIOPATIA IPERTENSIVA**
- **ASTENIA, SINCOPE,IPOTENSIONE ORTOSTATICA**
- **VERSAMENTO PERICARDICO ndd**
- **BLOCCHI (BB, BAV, BSA)**
- **SY TUNNEL CARPALE BILATERALE**
- **STENOSI AORTICA SEVERA LF/LG con EF lievemente ridotta**

OXFORD JOURNALS European Heart Journal

Eur Heart J. 2017 Oct 7; 38(38): 2879–2887.

Published online 2017 Aug 1. doi: 10.1093/eurheartj/ehx350

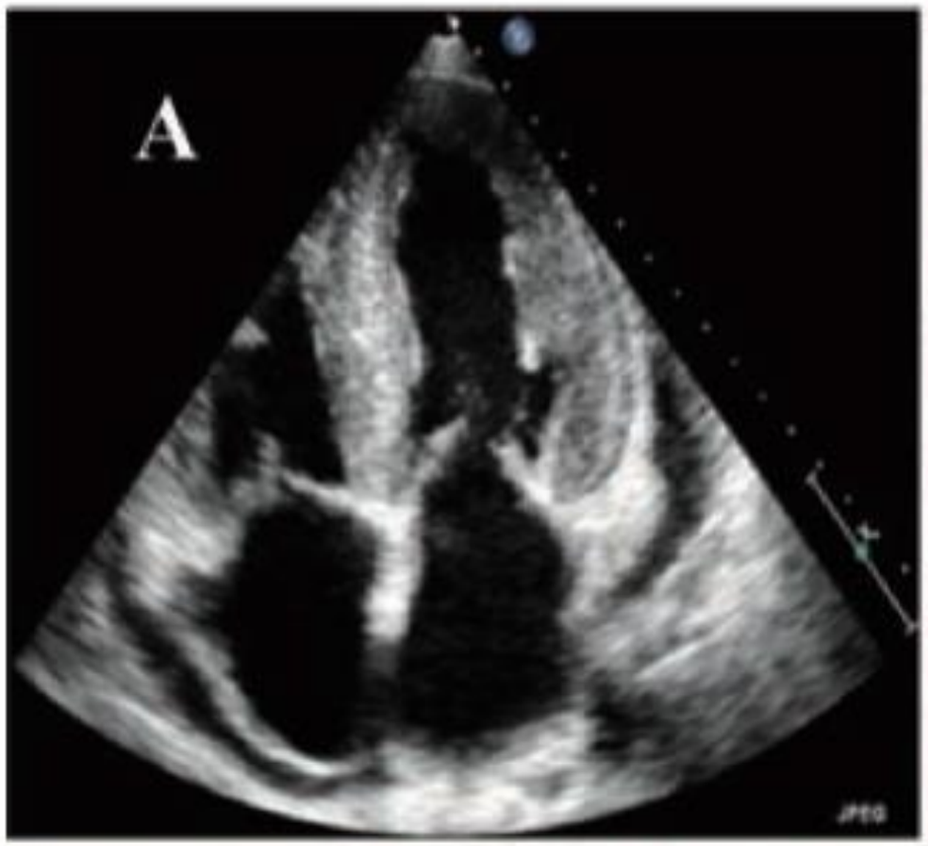
Unveiling transthyretin cardiac amyloidosis and its predictors among elderly patients with severe aortic stenosis undergoing transcatheter aortic valve replacement

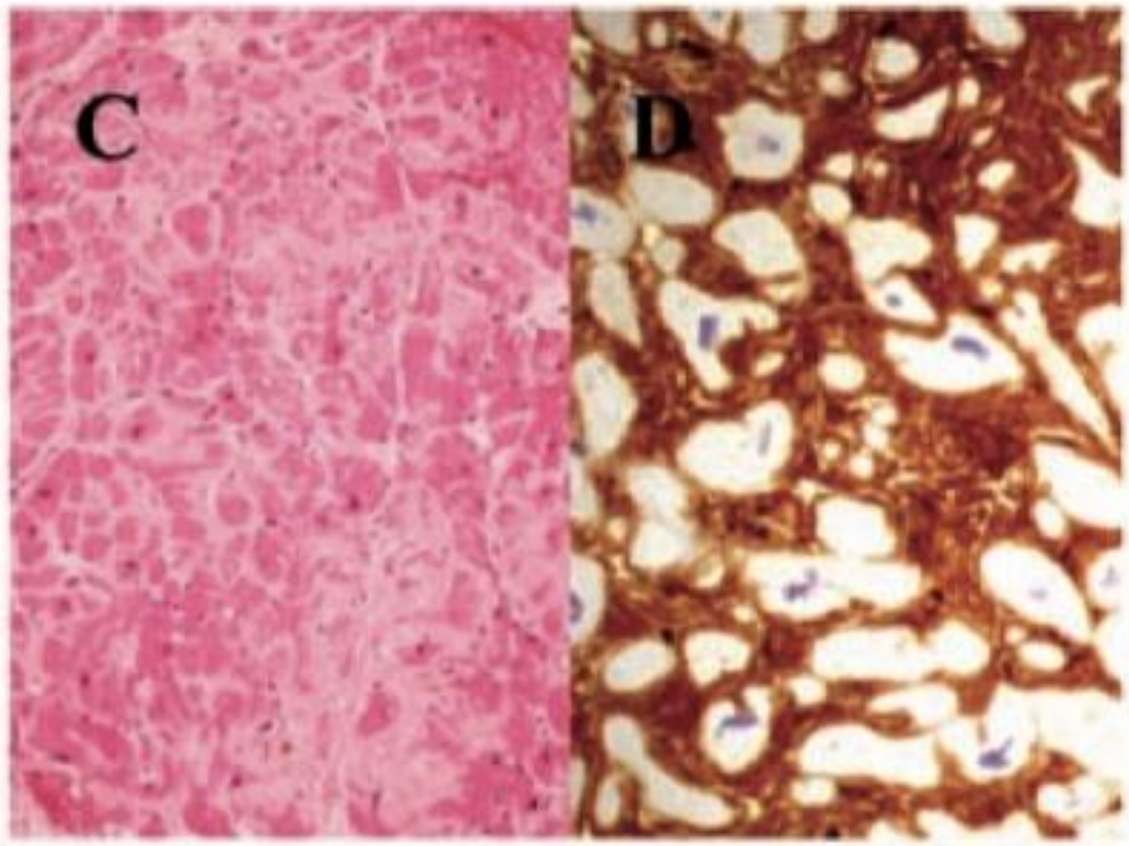
Adam Castaño,[1,2] David L Narotsky,[1] Nadira Hamid,[3] Omar K Khalique,[3] Rachelle Morgenstern,[2] Albert DeLuca,[2] Jonah Rubin,[1] Codruta Chiuzan,[4] Tamim Nazif,[3] Torsten Vahl,[3] Isaac George,[3] Susheel Kodali,[3] Martin B Leon,[3] Rebecca Hahn,[3] Sabahat Bokhari,[2] and Mathew S Maurer[1]

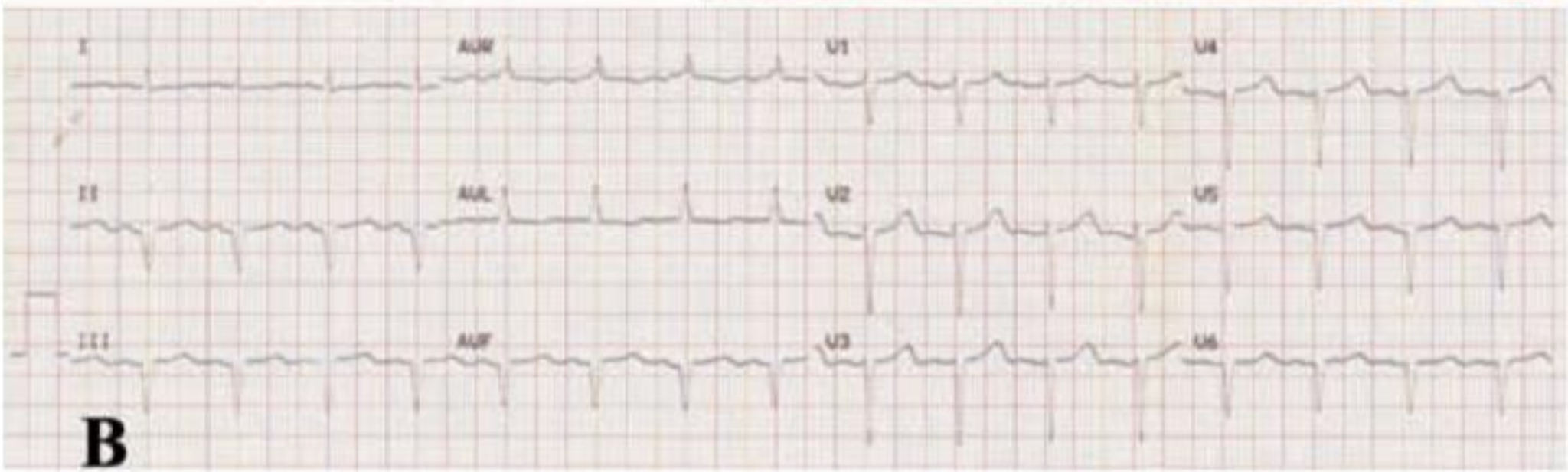
Conclusions

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.

PRINCIPALI ESAMI DIAGNOSTICI

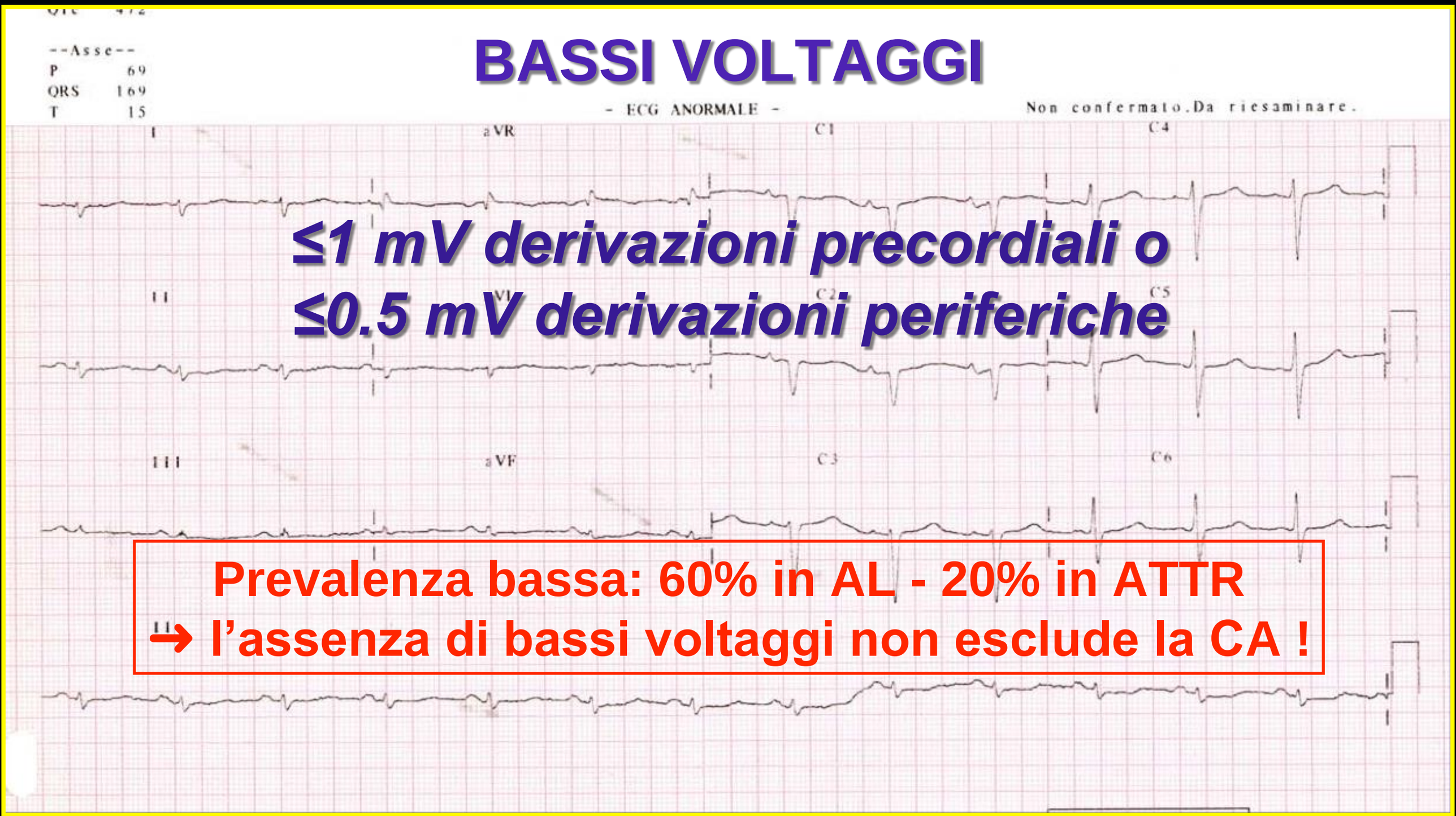






ECG

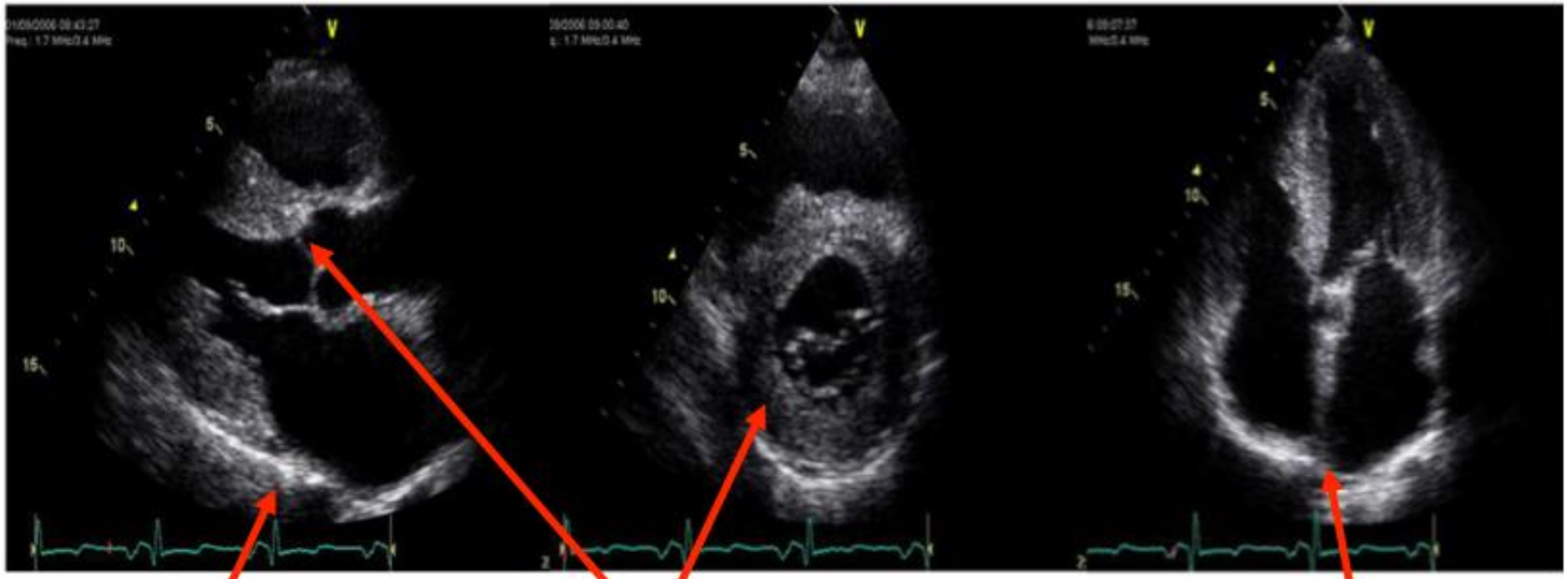
BASSI VOLTAGGI



≤1 mV derivazioni precordiali o
≤0.5 mV derivazioni periferiche

Prevalenza bassa: 60% in AL - 20% in ATTR
➔ l'assenza di bassi voltaggi non esclude la CA !

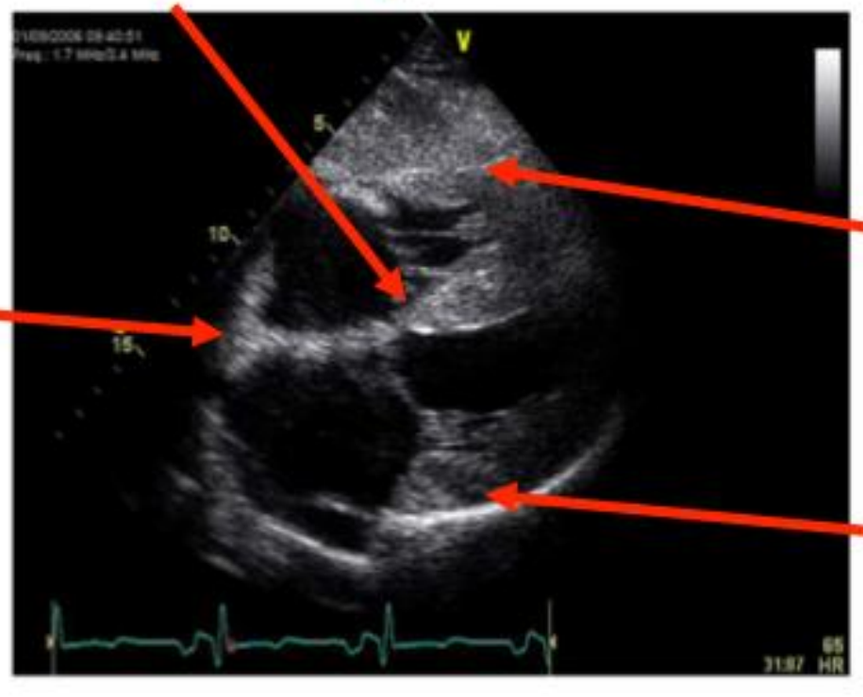
Echocardiogram in ATTR-CA



Left atrial dilatation

Bright myocardium and concentric symmetrical LVH

Biatrial dilatation



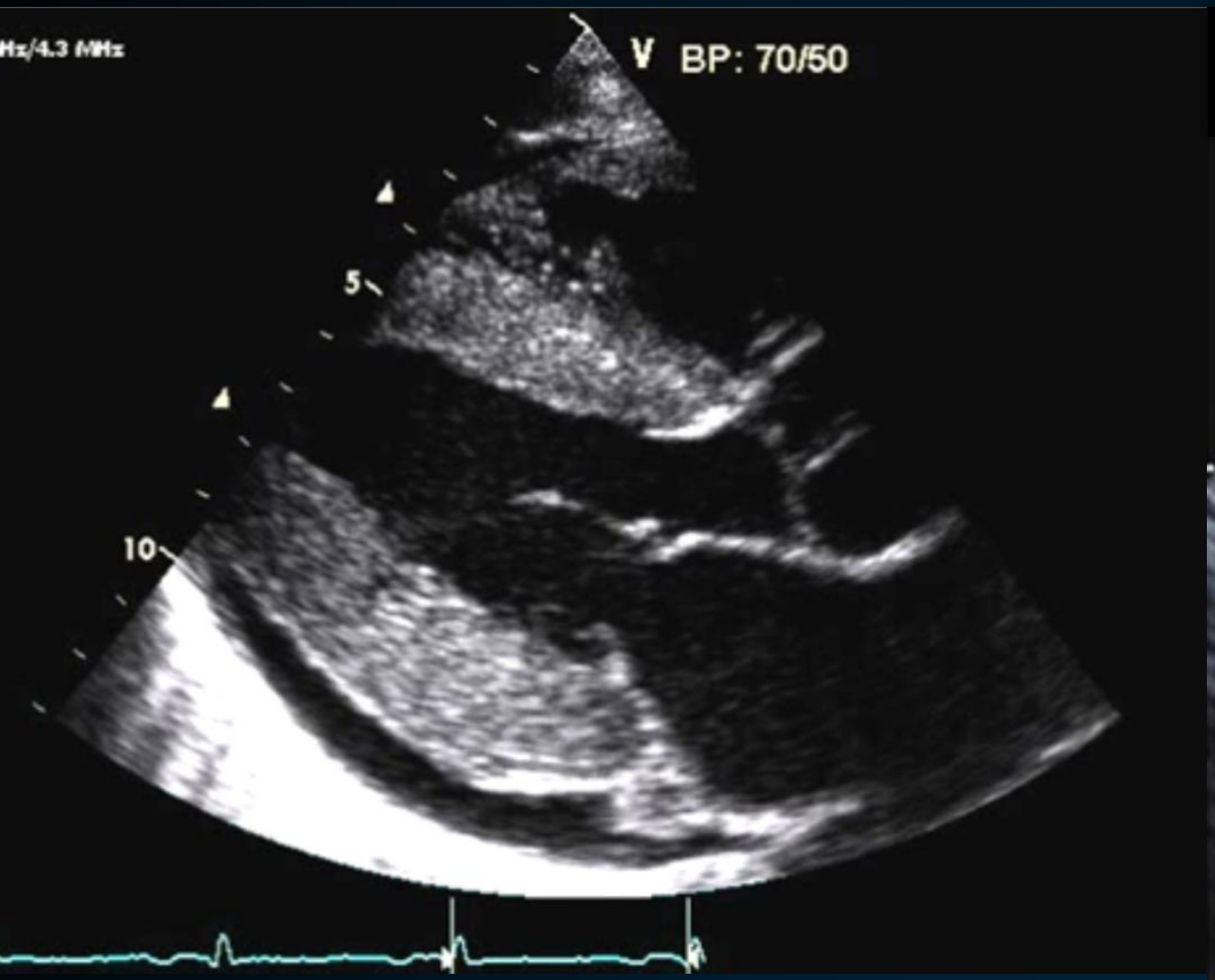
Thickened interatrial septum

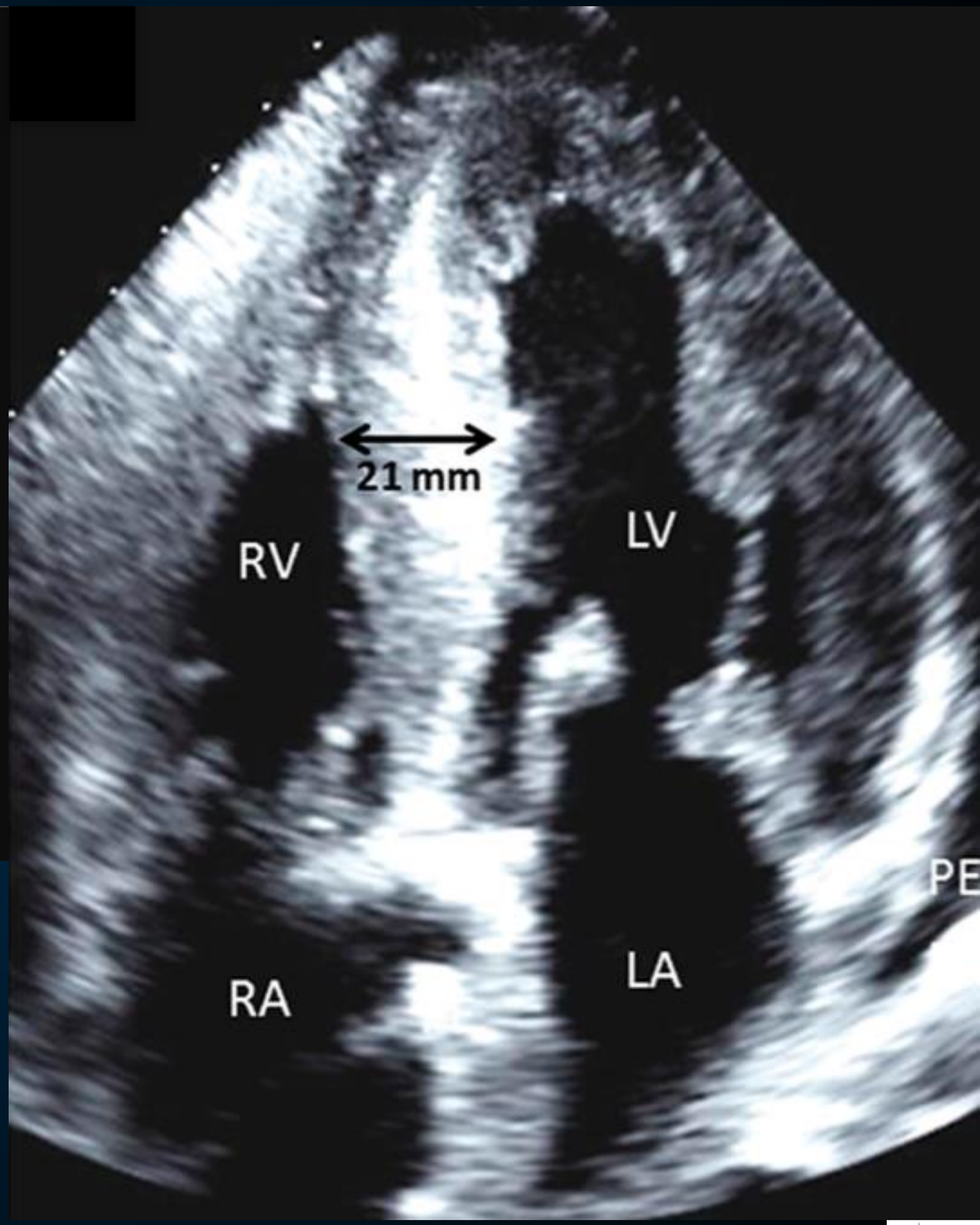
Thickening of RV free wall

Thickened valves

Circulation. 2017;135:1357–1377. DOI: 10.1161/CIRCULATIONAHA.116.024438

“SPARKLING” MYOCARDIUM



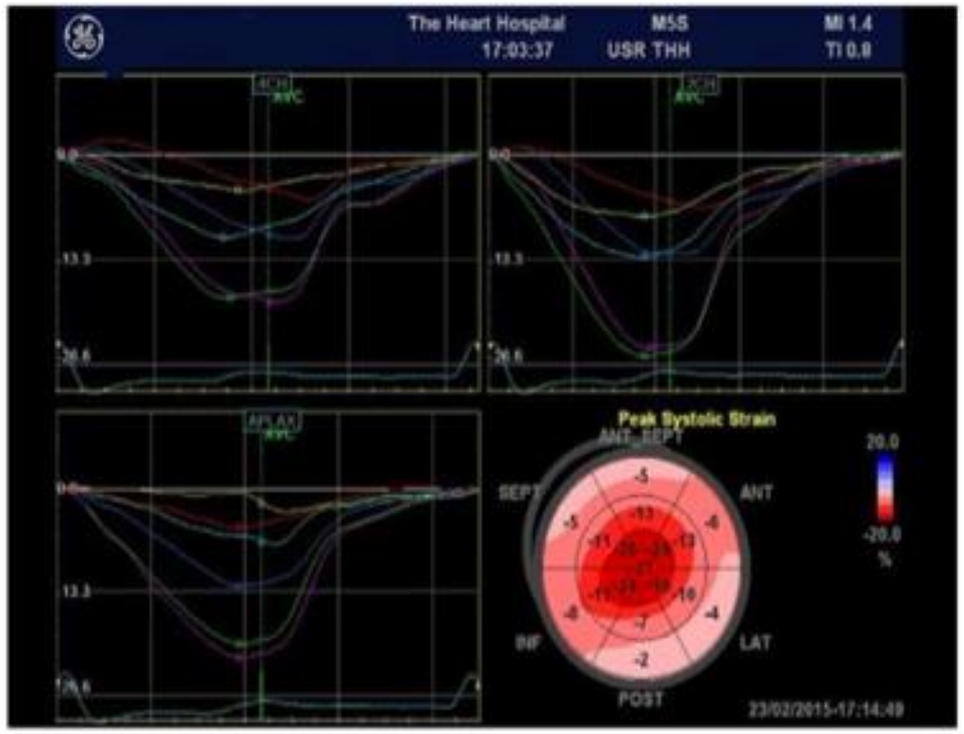


Presente nel 25% dei casi

N.B.: meglio evidente **senza armonica**

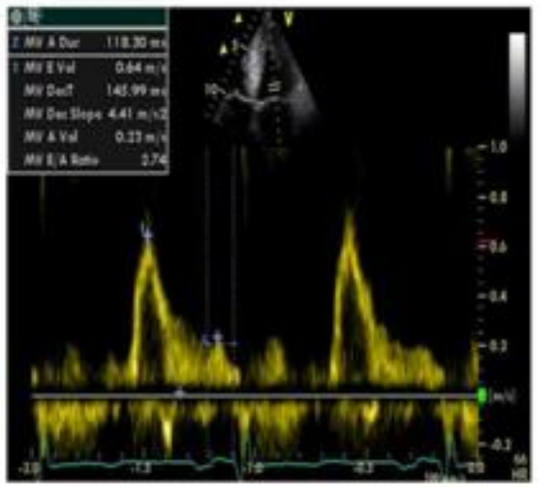
Echocardiogram in ATTR-CA

STRAIN ECHO



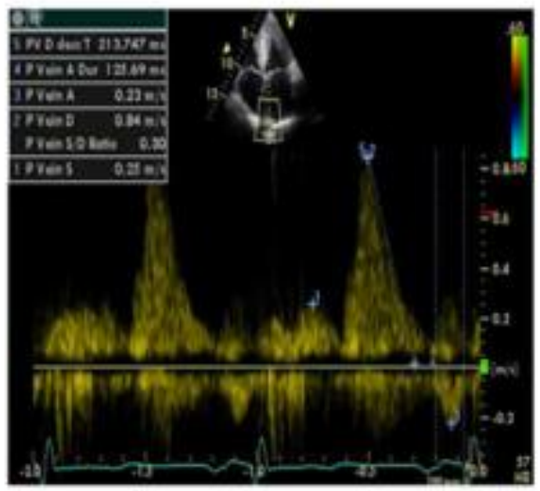
Impaired sistolic longitudinal shortening, with a base-apex gradient

DOPPLER WAVES



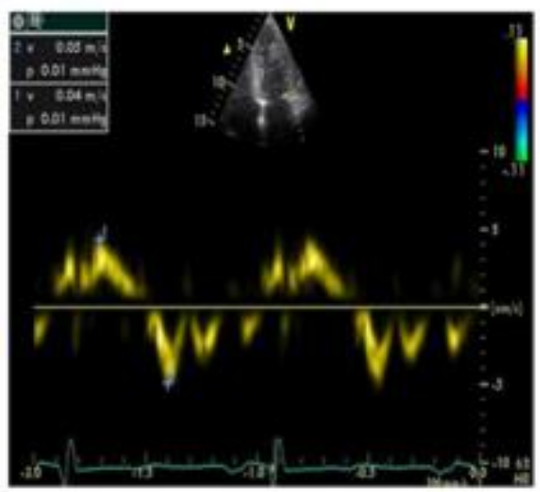
Mitral inflow PW Doppler

- Increased E/A ratio
- Normal E wave decT time
- Marked reduction in transmitral A-wave velocity



Pulm. vein inflow PW Doppler

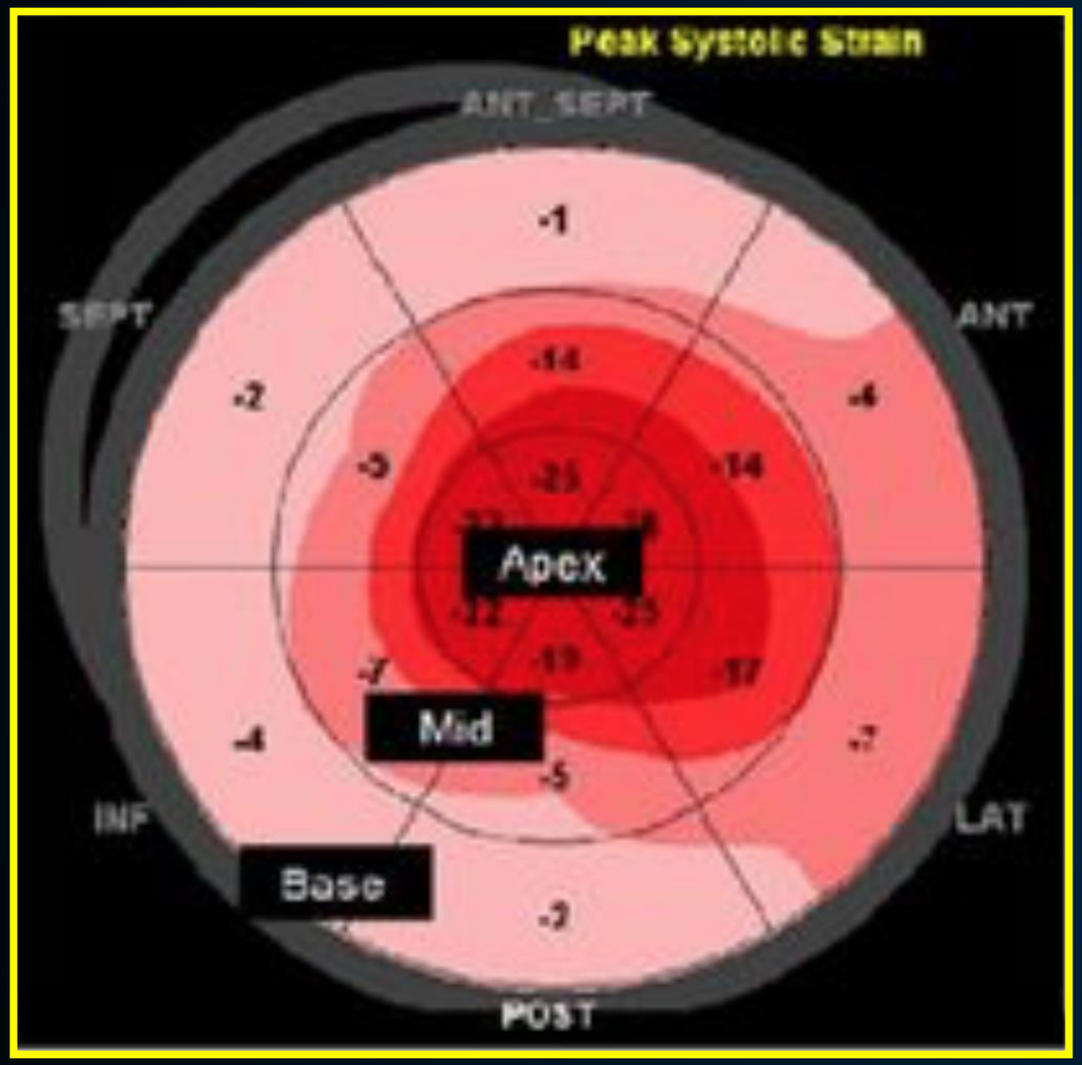
- Diastolic atrial reversal with increased duration and peak velocity as compared to transmitral signal



Mitral annulus tissue Doppler

- Marked reduction in apical systolic and diastolic velocities

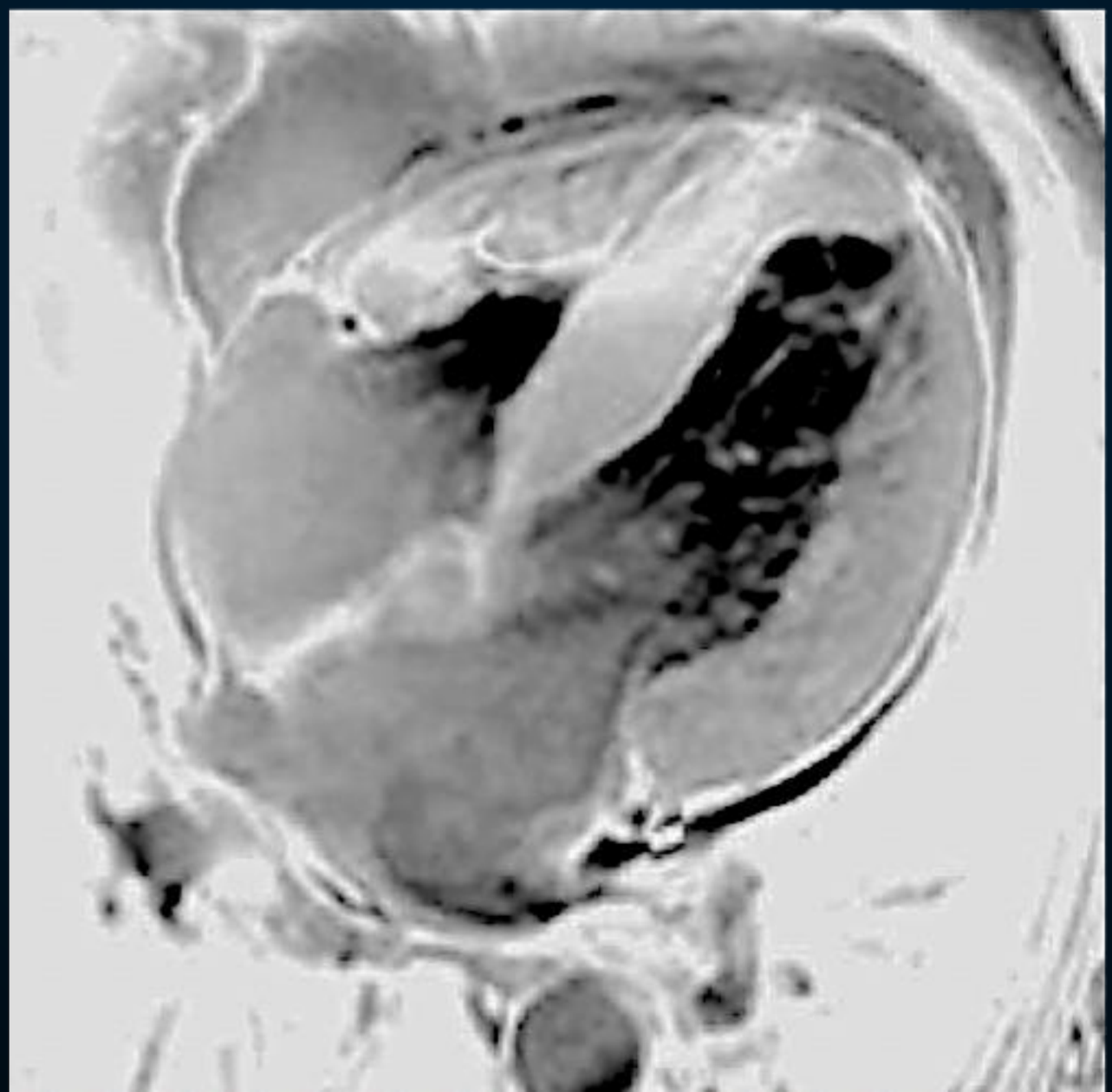
RELATIVE APICAL SPARING



$$\text{RELAPS} = \frac{\text{AVG APICAL LS}}{\text{SUM OF AVG BASAL and MID LS}}$$

CUT OFF : > 1

RISONANZA MAGNETICA



Modalità di captazione del gadolinio

Ischemica

Subendocardica

Transmurale

Non-ischemica

Intramiocardica
(CMP ipertrofica
CMP dilatativa
Sovraccarico dx)

Intramiocardica-
Subepicardica
(Miocardite, Sarcoidosi)

Subendocardica diffusa
(Amiloidosi)

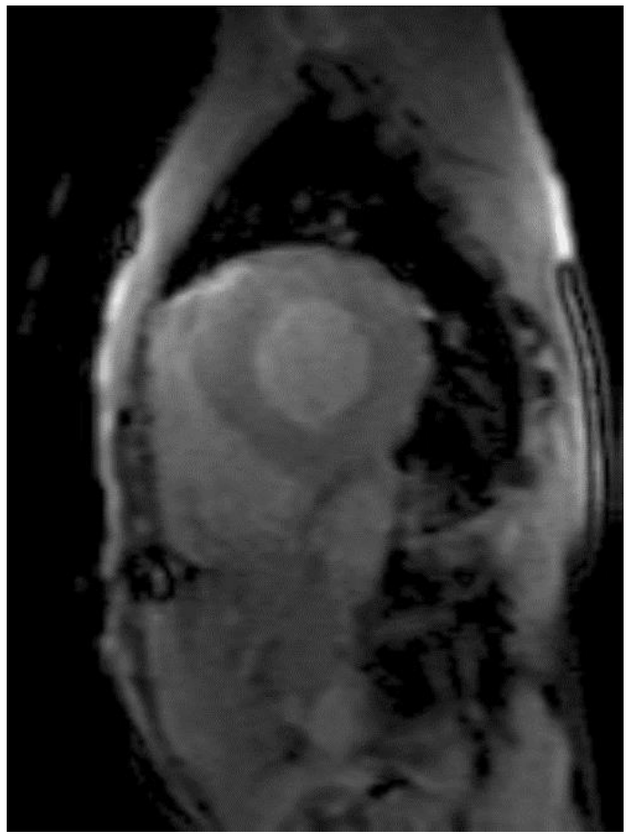
Sarcoidosi

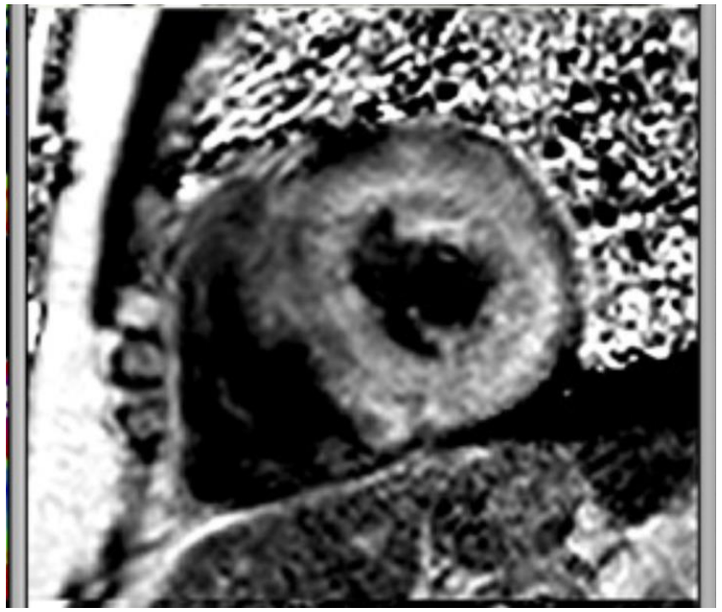
Subendocardica Transmurale Coinvolgimento dx

Courtesy of Dr. Imazio

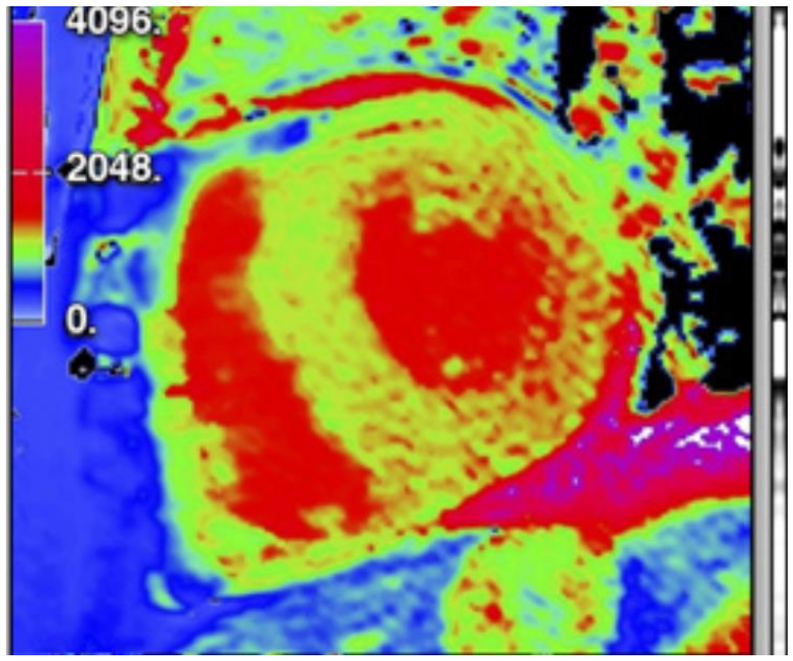
CMR in AMYLOIDOSIS

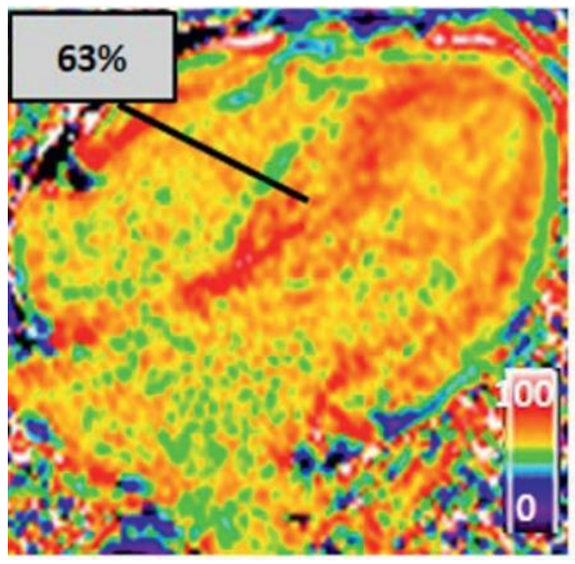
NO NULL





LE



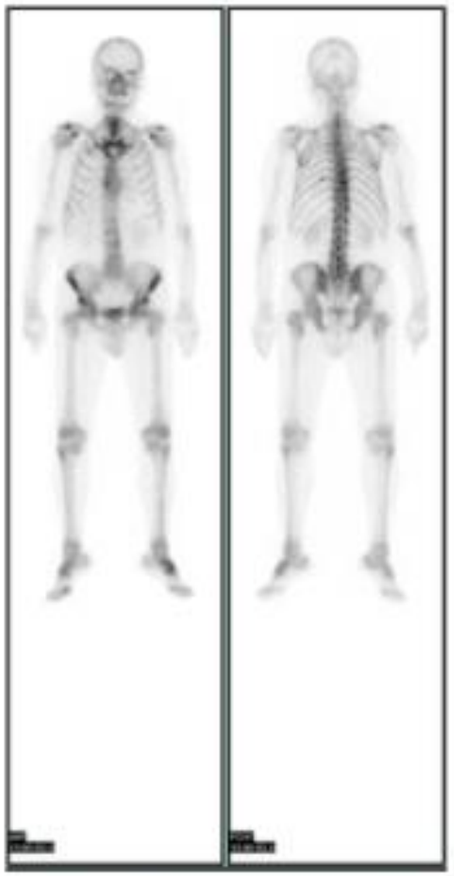


ECV

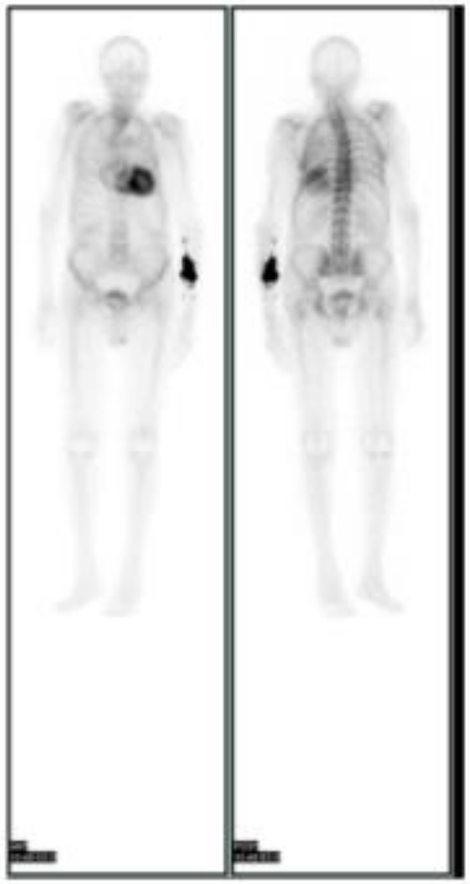
T1
NATIVE

Courtesy of Dr. Faletti

Bone tracer scintigraphy in ATTR-CA



Healthy control

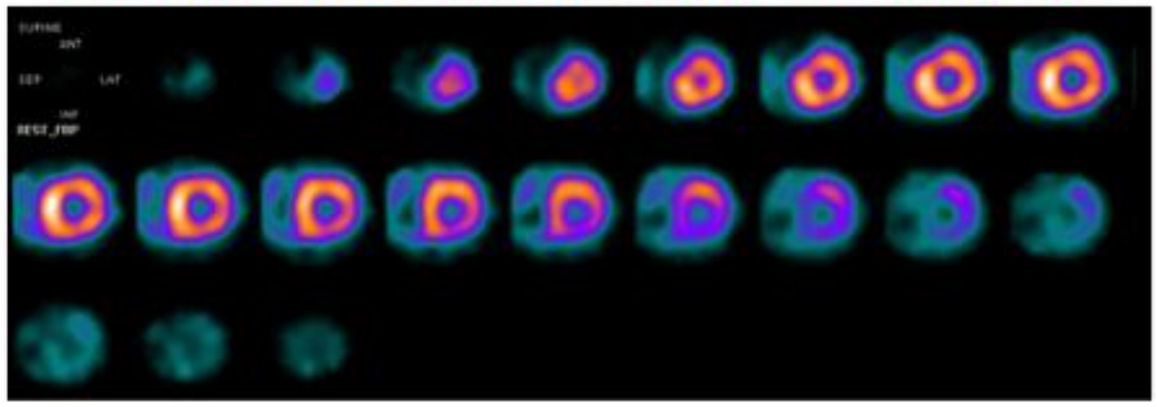


ATTR-CA

Perugini Score

Visual Cardiac Score
0 Absent Myocardial Uptake
1 Myocardial Uptake < Bone
2 Myocardial Uptake = Bone
3 Myocardial Uptake > Bone

Quantitative and regional assessment



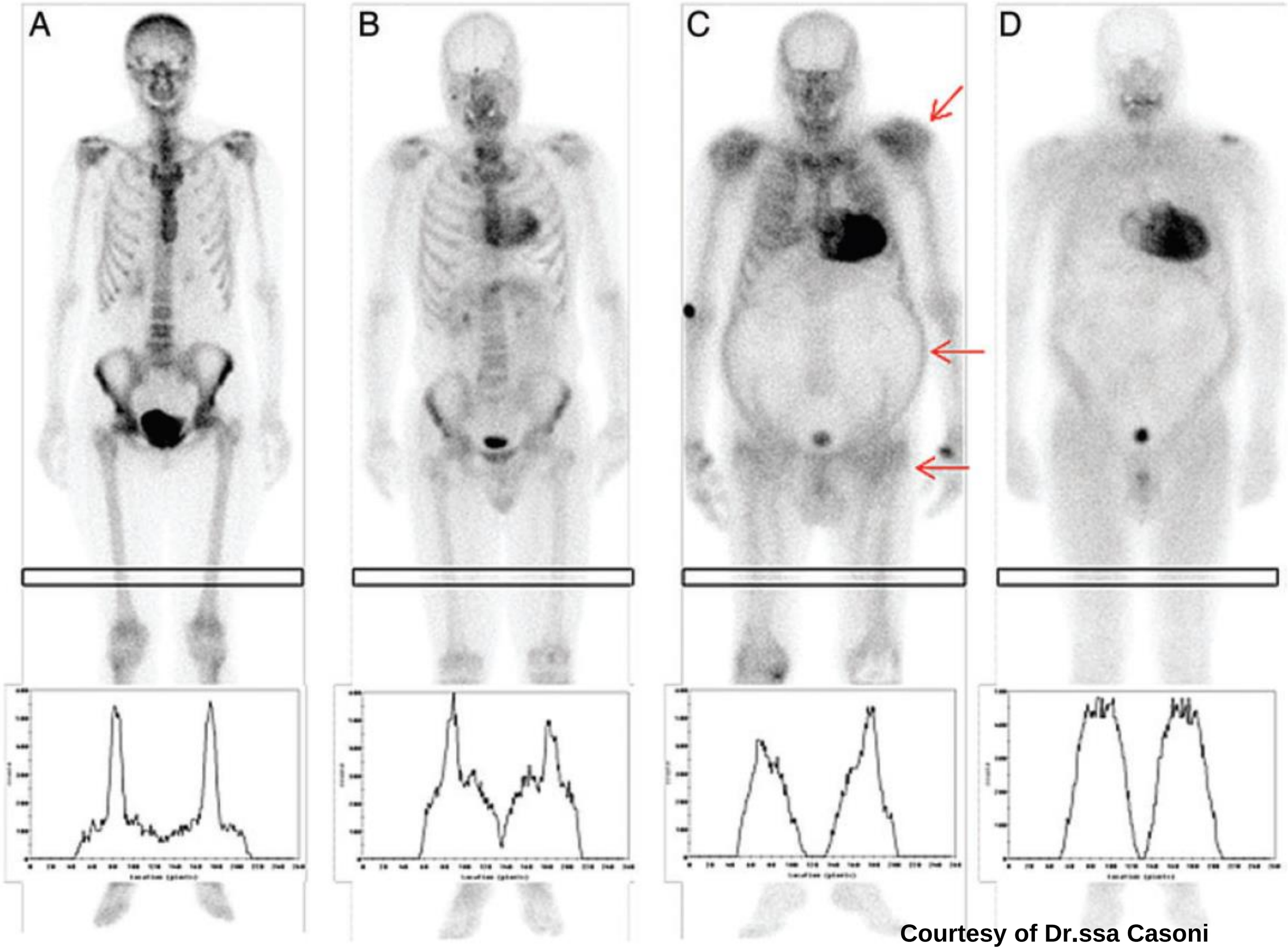
Courtesy of Dr.ssa Casoni

- **>99% sensitive and 86% specific for ATTR-CA**
- **false positives almost exclusively from uptake in patients with cardiac AL amyloidosis**
- **Grade 2-3 Perugini score + the absence of a monoclonal protein: 100% Sp and PPV**

Nonbiopsy Diagnosis of Cardiac Transthyretin Amyloidosis

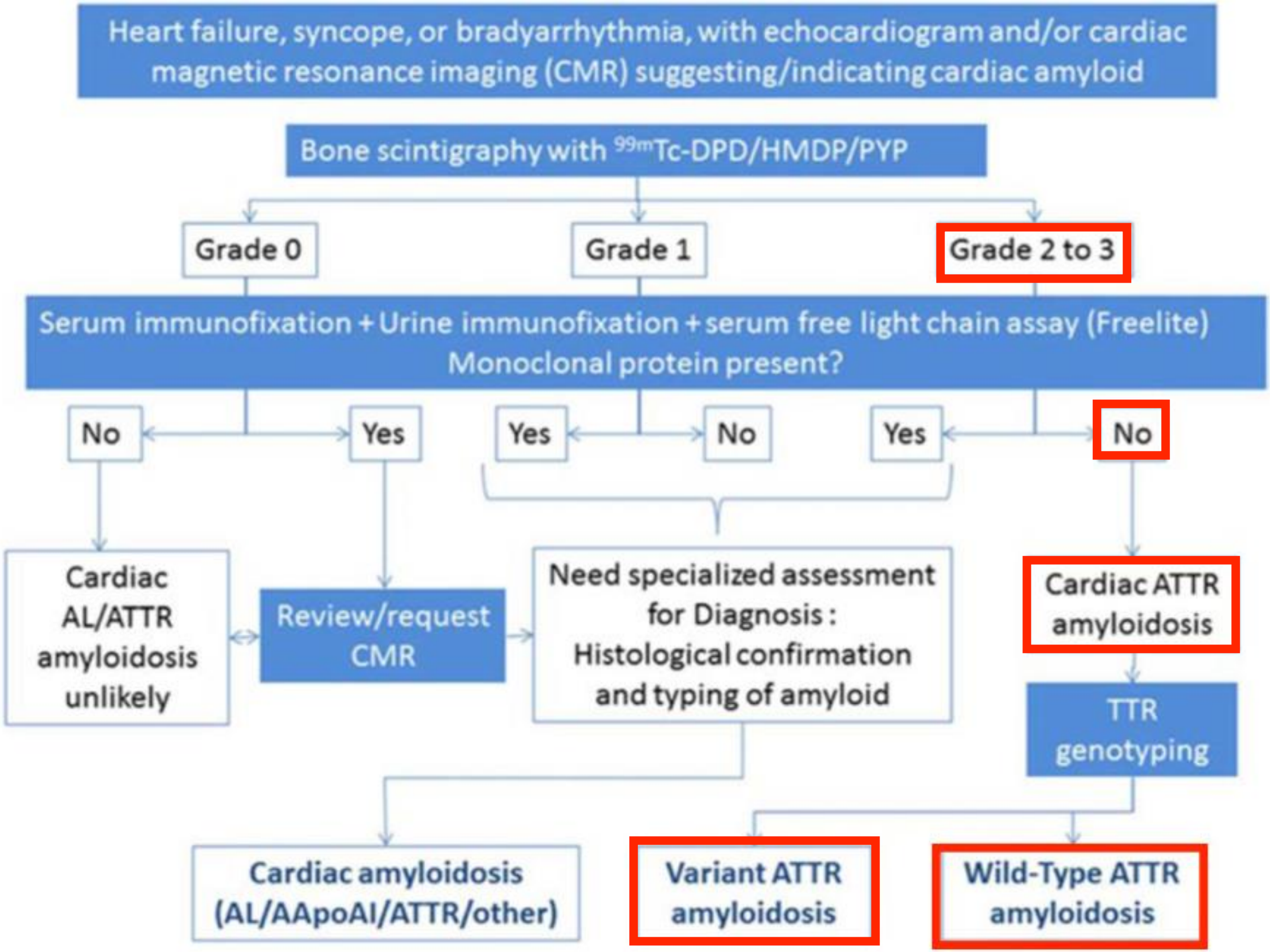
Julian D. Gillmore, MD, PhD; Mathew S. Maurer, MD; Rodney H. Falk, MD; Giampaolo Merlini, MD; Thibaud Damy, MD; Angela Dispenzieri, MD; Ashutosh D. Wechalekar, MD, DM; John L. Berk, MD; Candida C. Quarta, MD, PhD; Martha Grogan, MD; Helen J. Lachmann, MD; Sabahat Bokhari, MD; Adam Castano, MD; Sharmila Dorbala, MD, MPH; Geoff B. Johnson, MD, PhD; Andor W.J.M. Glaudemans, MD, PhD; Tamer Rezk, BSc; Marianna Fontana, MD; Giovanni Palladini, MD, PhD; Paolo Milani, MD; Pierluigi L. Guidalotti, MD; Katarina Flatman; Thirusha Lane, MSc; Frederick W. Vonberg, MBBS; Carol J. Whelan, MD; James C. Moon, MD; Frederick L. Ruberg, MD; Edward J. Miller, MD, PhD; David F. Hutt, BApSc; Bouke P. Hazenberg, MD, PhD; Claudio Rapezzi, MD; Philip N. Hawkins, PhD, FMedSci

Circulation. 2016;133:2404–2412.
doi: 10.1161/ CIRCULATIONAHA.116.021612.

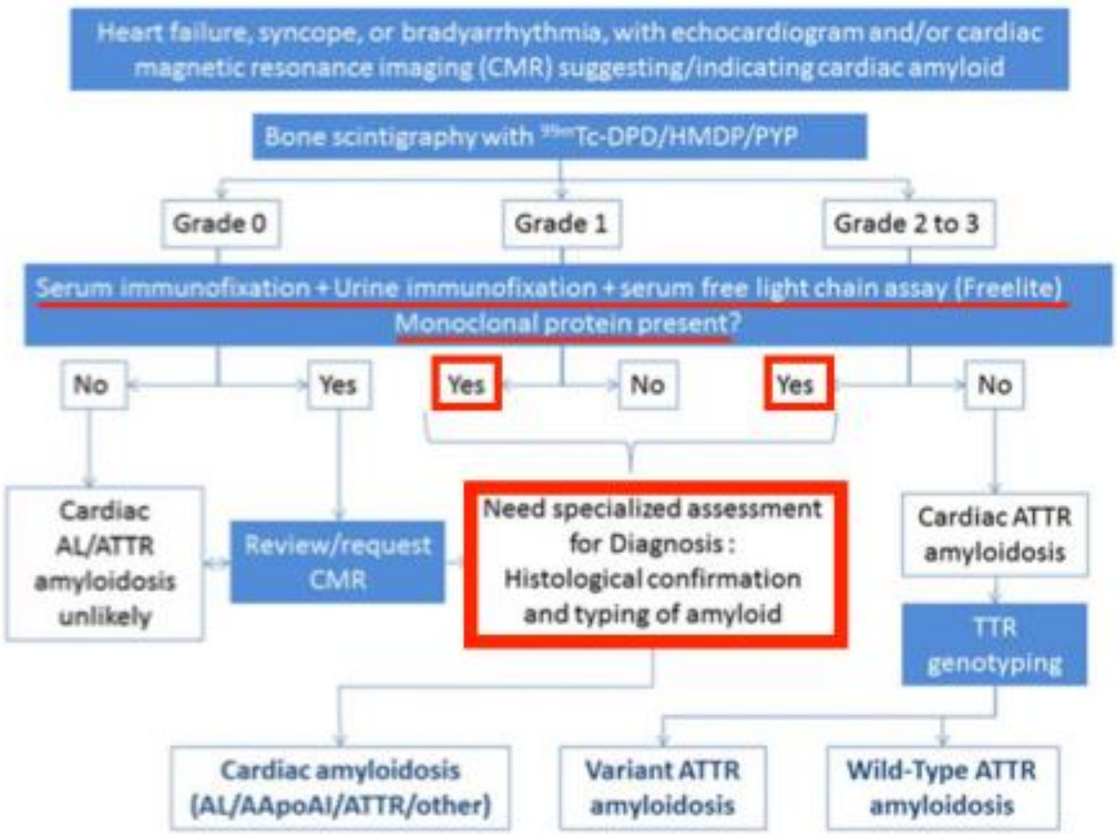


Courtesy of Dr.ssa Casoni

Nonbiopsy diagnosis of ATTR-CA



When and How Should Tissue Biopsy BeUndertaken?



- Positive 99mTc-phosphate scan without evidence for plasma clone on blood and urine testing -> **diagnosis of ATTR-CA without a biopsy**

- Positive 99mTc-phosphate scan with evidence for plasma clone on blood and urine testing -> **histological diagnosis is still required because the uptake on a 99mTc-phosphate scan is not 100% specific for ATTR-CA.**

EXTRACARDIAC BIOPSY *(abdominal fat pad, gingiva, skin, salivary gland, or gastrointestinal tract*

Diagnostic accuracy:

- 70% for AL-CA / 67% for ATTRm / 14% for ATTRwt

Fine NM et al. Yield of noncardiac biopsy for the diagnosis of transthyretin cardiac amyloidosis. Am J Cardiol. 2014;113:1723–1727. doi: 10.1016/j. amjcard.2014.02.030.

although a fat pad biopsy is a preferred initial site, a negative result is insufficient to exclude the diagnosis

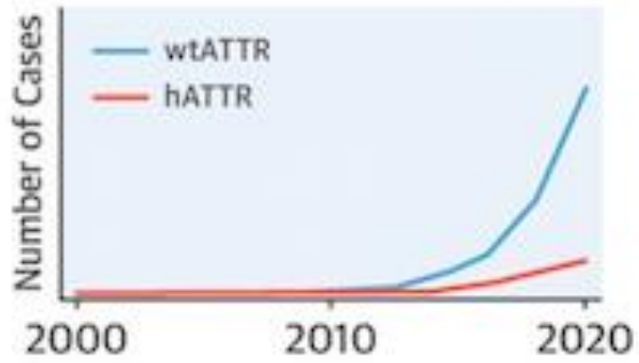
CARDIAC BIOPSY

Tissue diagnosis is required for AL amyloid and endomyocardial biopsy should be pursued if the index of suspicion is high despite a negative fat pad.

Present/Future

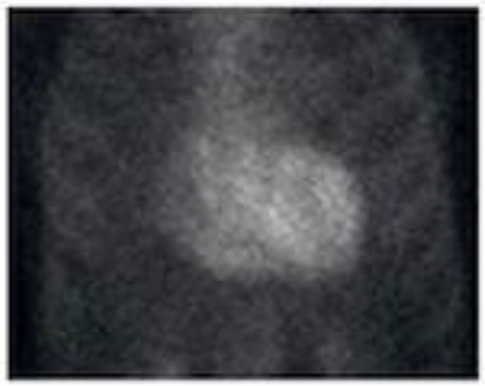
Epidemiology →

Recognition of ATTR-CM



Noninvasive-Scintigraphy

Diagnosis →

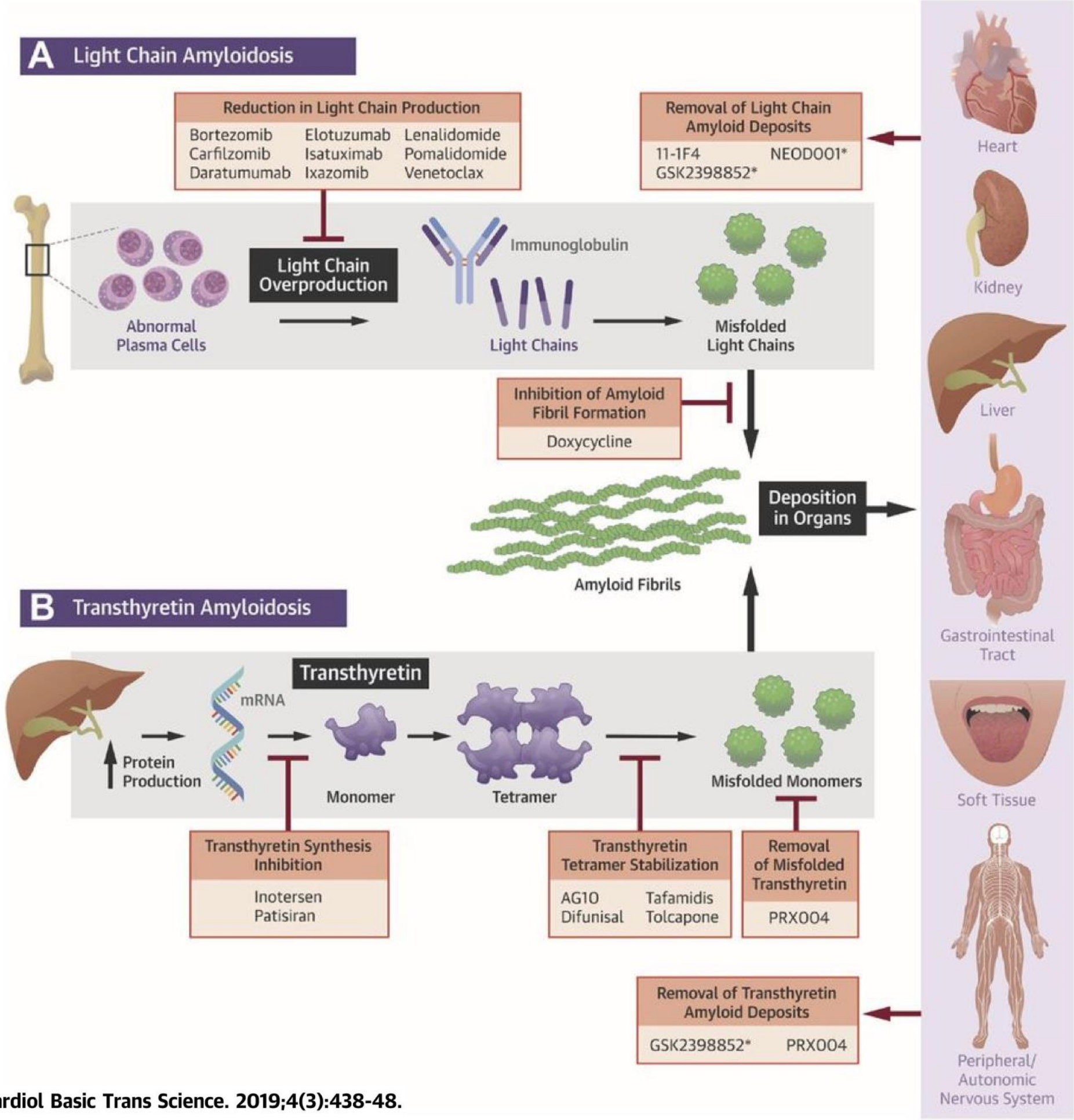


Treatment →

Emerging Treatment Options

wtATTR-CM	hATTR		
	Cardiomyopathy ONLY	*Cardiomyopathy AND Neuropathy*	*Neuropathy ONLY*
TAFAMIDIS Under review by FDA	TAFAMIDIS Under review by FDA	TAFAMIDIS Under review by FDA	PATISIRAN
DIFLUNISAL Off-label usage	DIFLUNISAL Off-label usage	PATISIRAN	INOTERSEN
		INOTERSEN	DIFLUNISAL Off-label usage
		DIFLUNISAL Off-label usage	

J Am Coll Cardiol 2019;73:2872-91

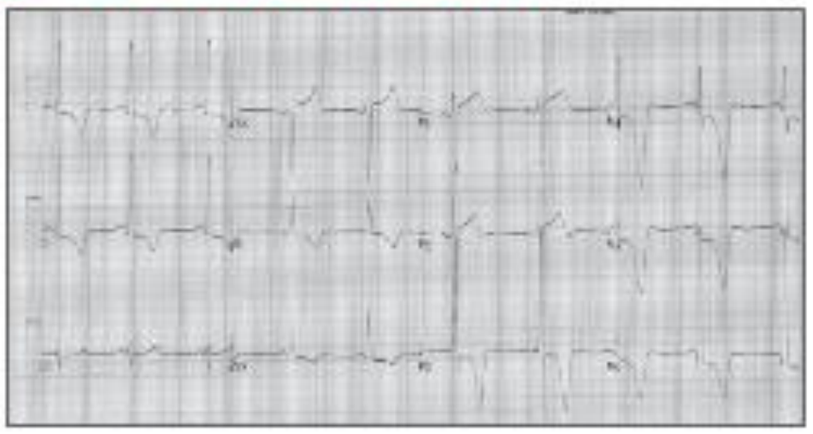


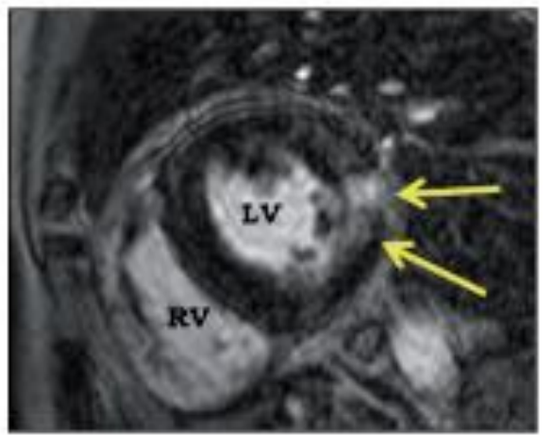
Zhang, K.W. et al. J Am Coll Cardiol Basic Trans Science. 2019;4(3):438-48.

31 GIORNATE CARDIOLOGICHE TORINESI
TURIN October 24th-26th 2019

Fabry's Disease: key messages

1. HCM phenocopy





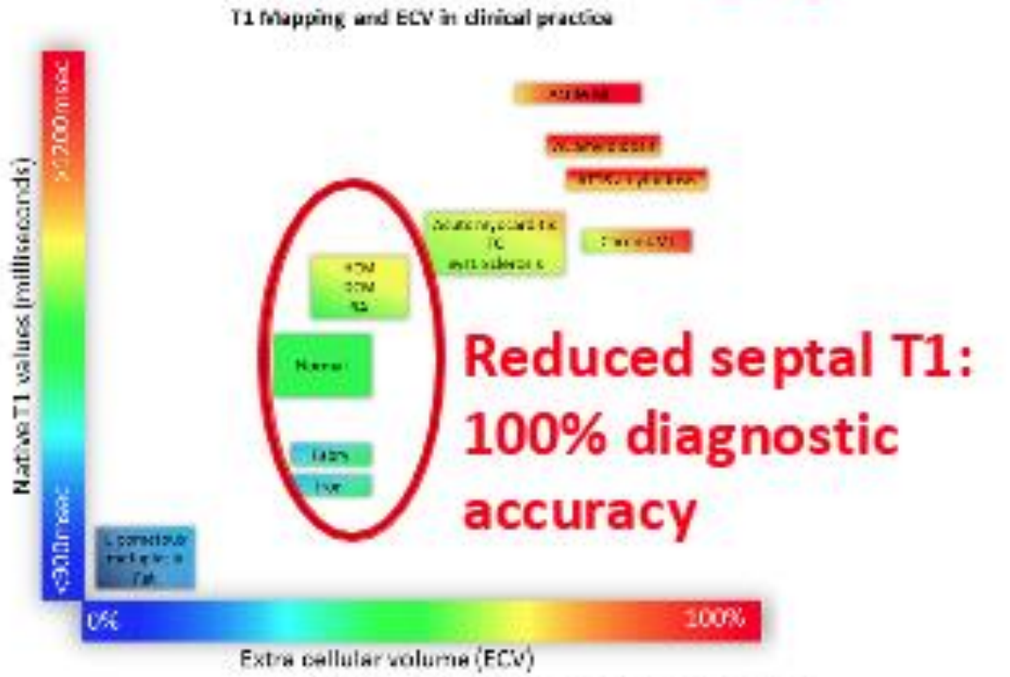
1% prevalence among patients diagnosed with HCM

(Monserrat et al. JACC Vol. 50, No. 25, 2007 18/25, 2007:2399–403)

Condition	Age at Presentation	History and Clinical Presentation	Echocardiography	ECG Profile	CMR LGE
Fabry disease	Male: 11 ± 7 yrs; female: 33 ± 16 yrs	Neuropathic pain, impaired sweating, skin rashes	Symmetrical increase in LV and RV wall thickness, normal EF	Increased or normal QRS complex voltage, short or prolonged PR interval	Focal, midwall, inferolateral wall
Differential diagnosis					
Hypertrophic cardiomyopathy	17–18 yrs	Maybe asymptomatic, dyspnea, angina, syncope, sudden death	Asymmetrical hypertrophy, small LV cavity, LVOT obstruction, normal EF	Increased QRS complex voltage, pseudo-delta wave, giant T-wave inversion	Patchy, midwall, junctions of the ventricular septum and RV
Hypertensive heart disease	Adults	History of hypertension	Symmetrical increase in LV wall thickness, mild LV dilation, normal EF	Increased QRS complex, nonspecific ST-T-wave changes	No pattern, predominantly subendocardial

Seward et al. JACC Vol. 55, No. 17, 2010 April 27, 2010:1769–79

2. CMR parametric imaging



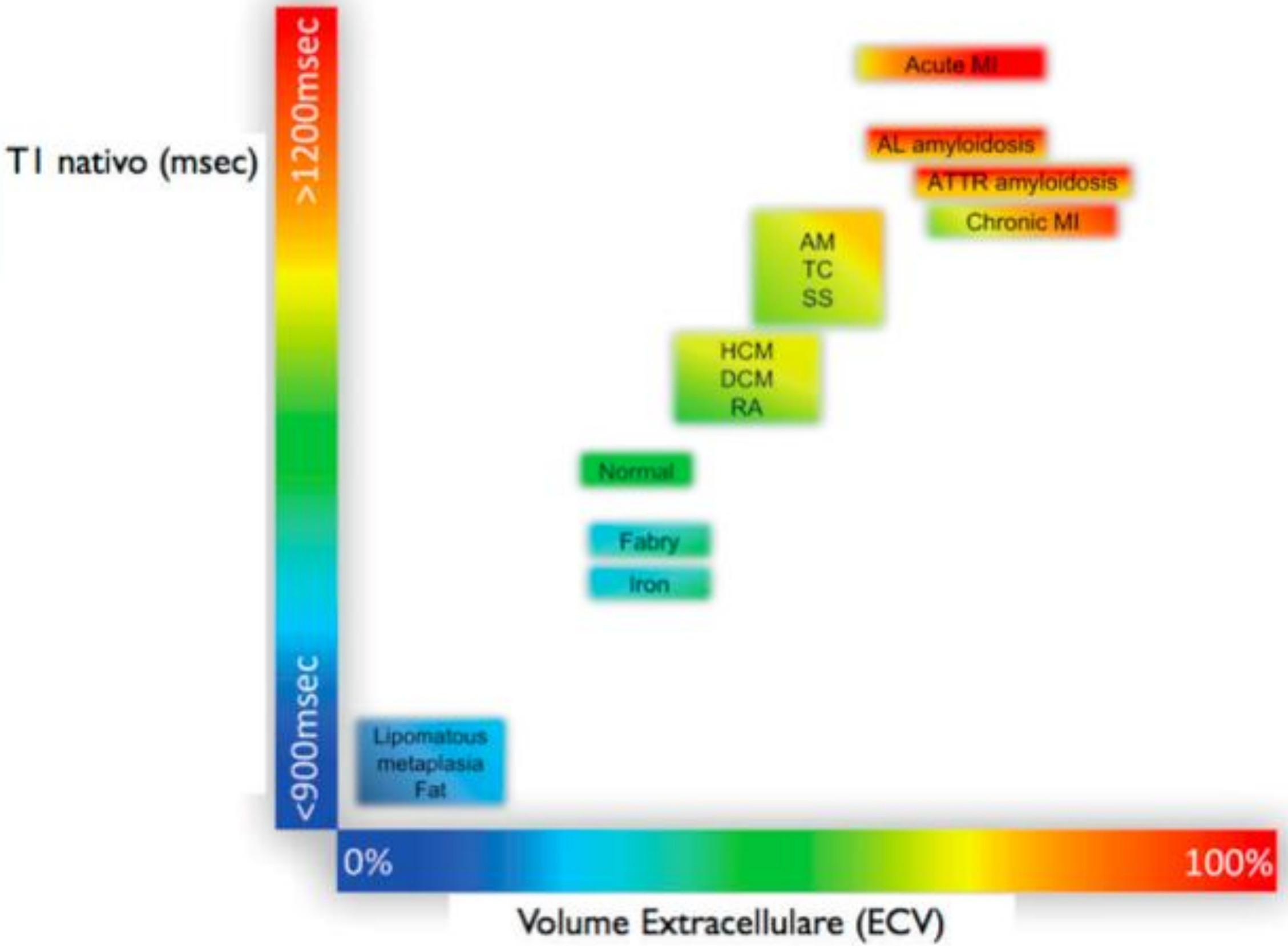
J Cardiovasc Magn Reson. 2016; 18: 89.
Sado et al. Circ Cardiovasc Imaging 2013;6:392–8.

3. Early treatment improves outcomes!

The effect of enzyme replacement therapy on clinical outcomes in male patients with Fabry disease: A systematic literature review by a European panel of experts

Molecular Genetics and Metabolism Reports 19 (2019) 100454

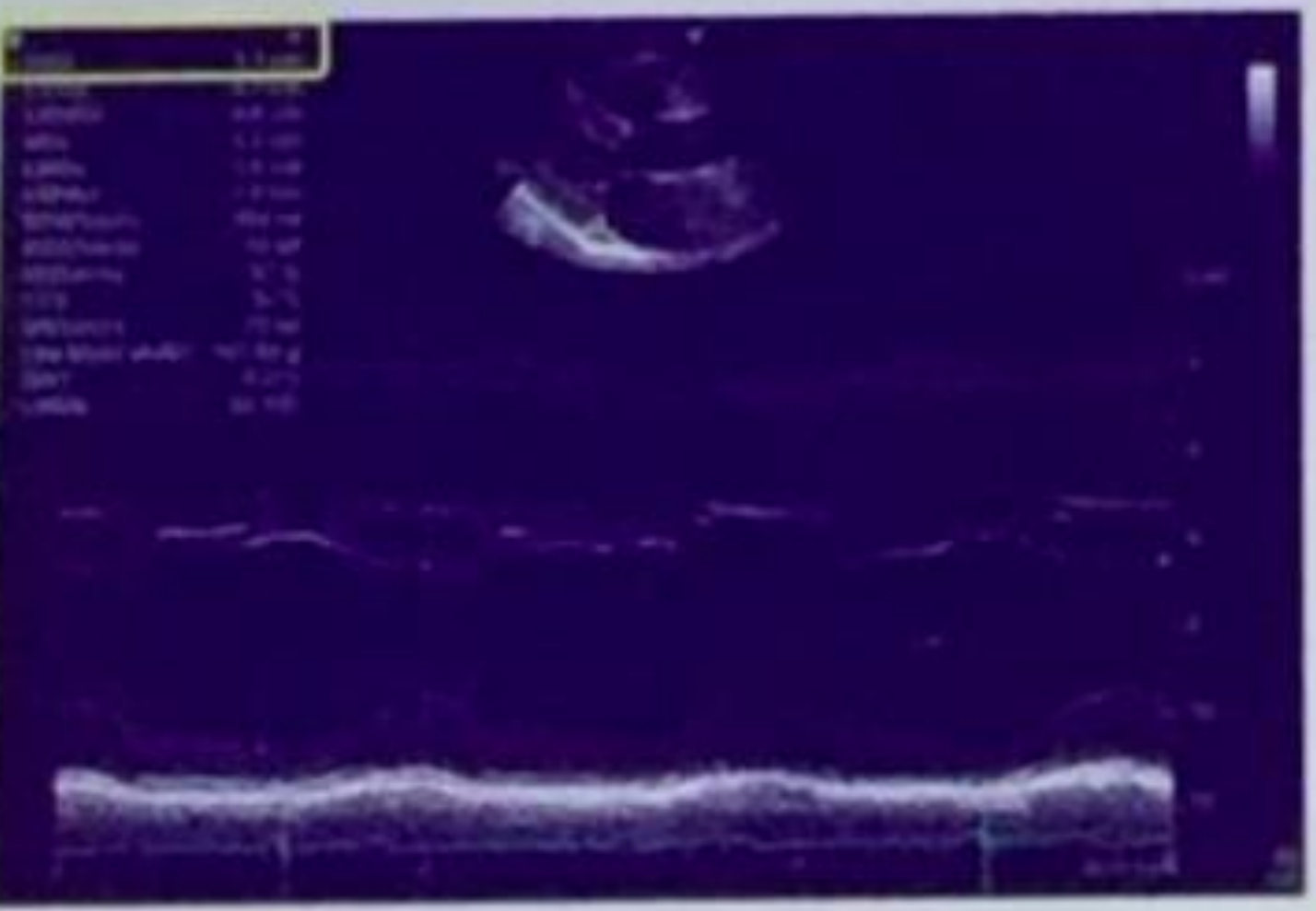
Mappaggio T1 e ECV nella pratica clinica



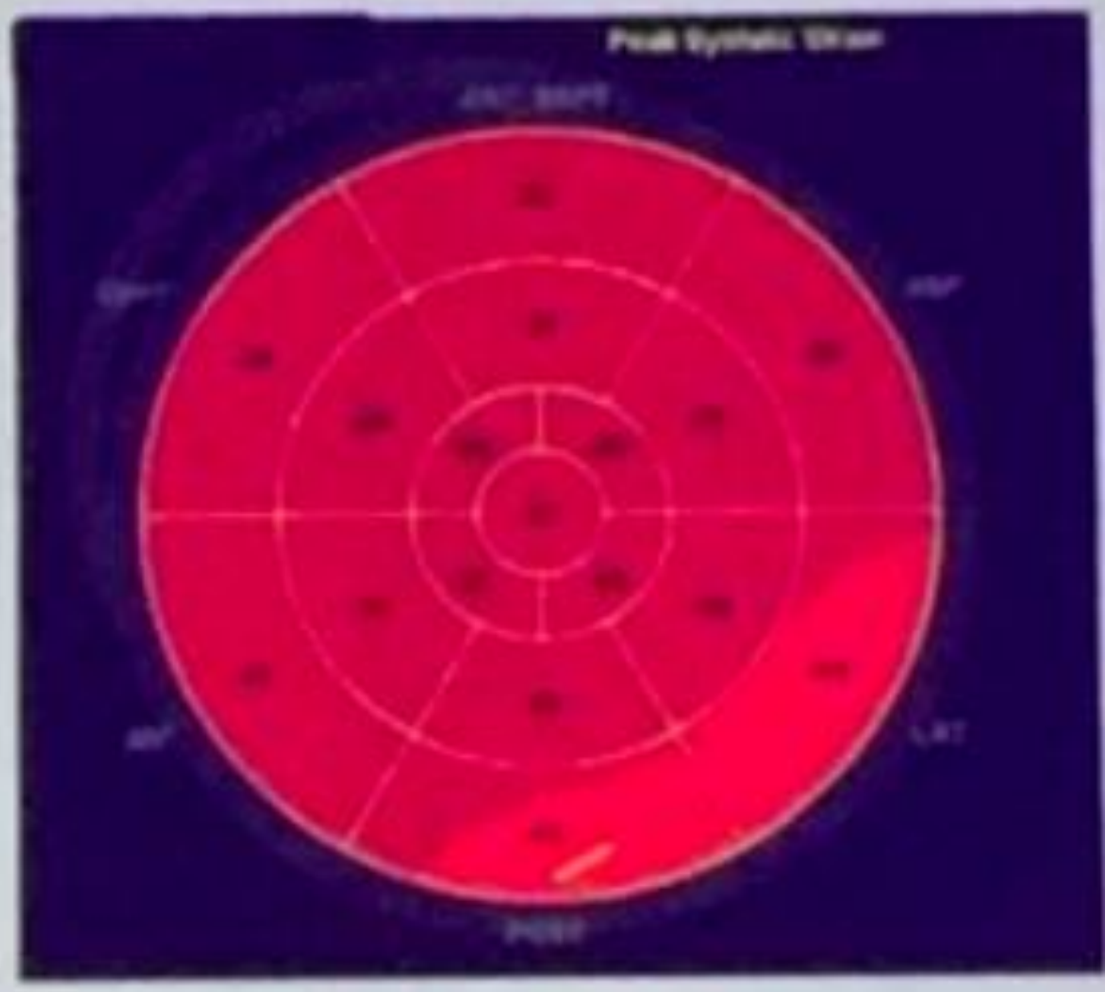
Courtesy of Dr. Imazio

AFD Disease: Typical early stage

FI, male, 45 years



EF = 58%



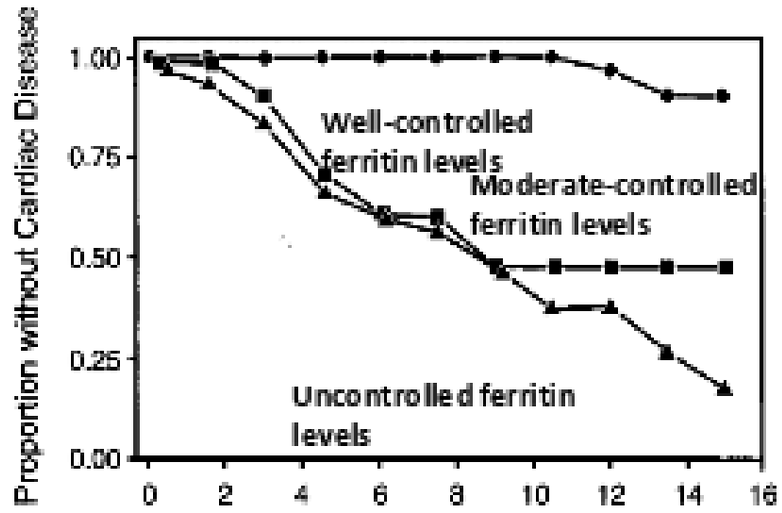
GLS = -22.2%

Initial septal hypertrophy and GLS pattern

Iron overload cardiomyopathy (IOC): key messages

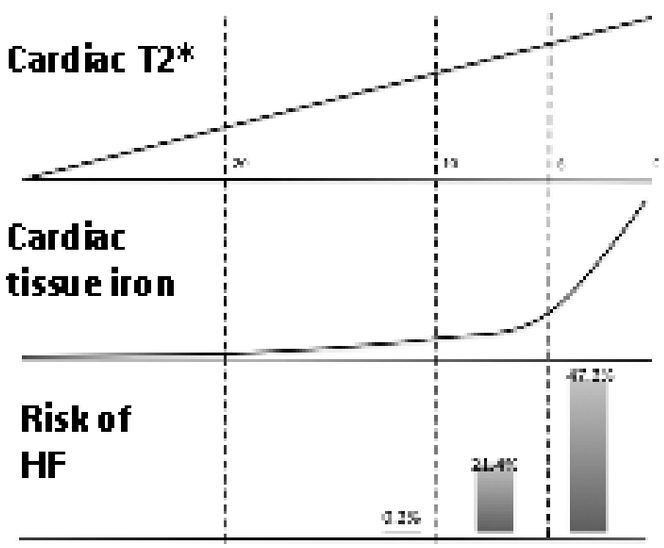
1. Epidemiology

- Constantly **increasing incidence** as an effect of improved mortality of patients with hereditary anemia and blood malignancies
- **Leading cause of death in patients receiving chronic blood transfusion therapy**
- Iron dose-dependent probability of HF-development

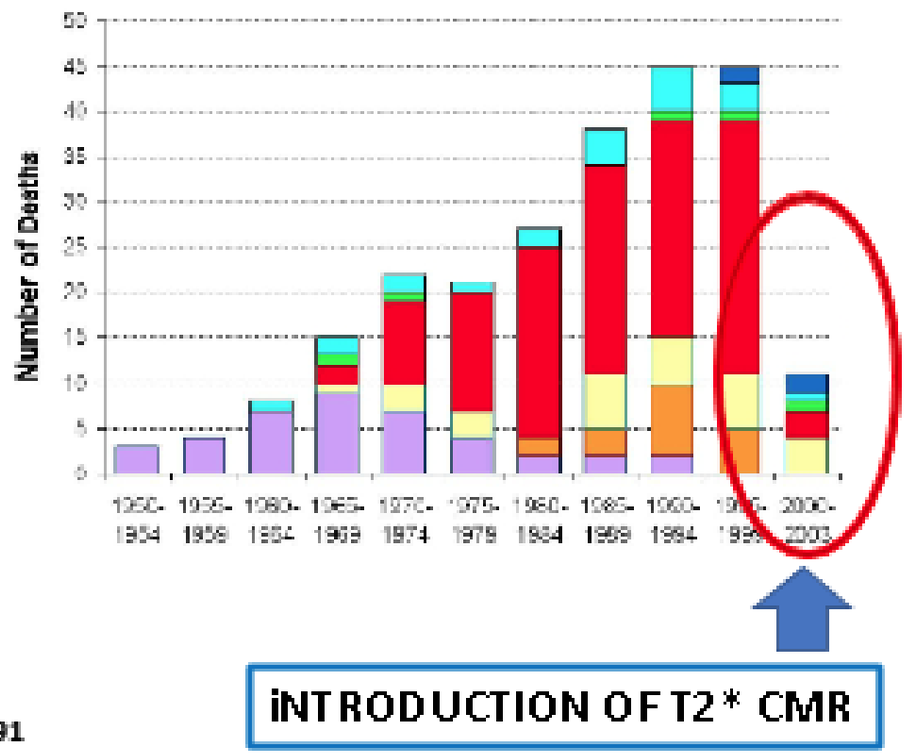


Olivieri et al. N Engl J Med 1994;331:574–8.

2. CMR-guided follow-up

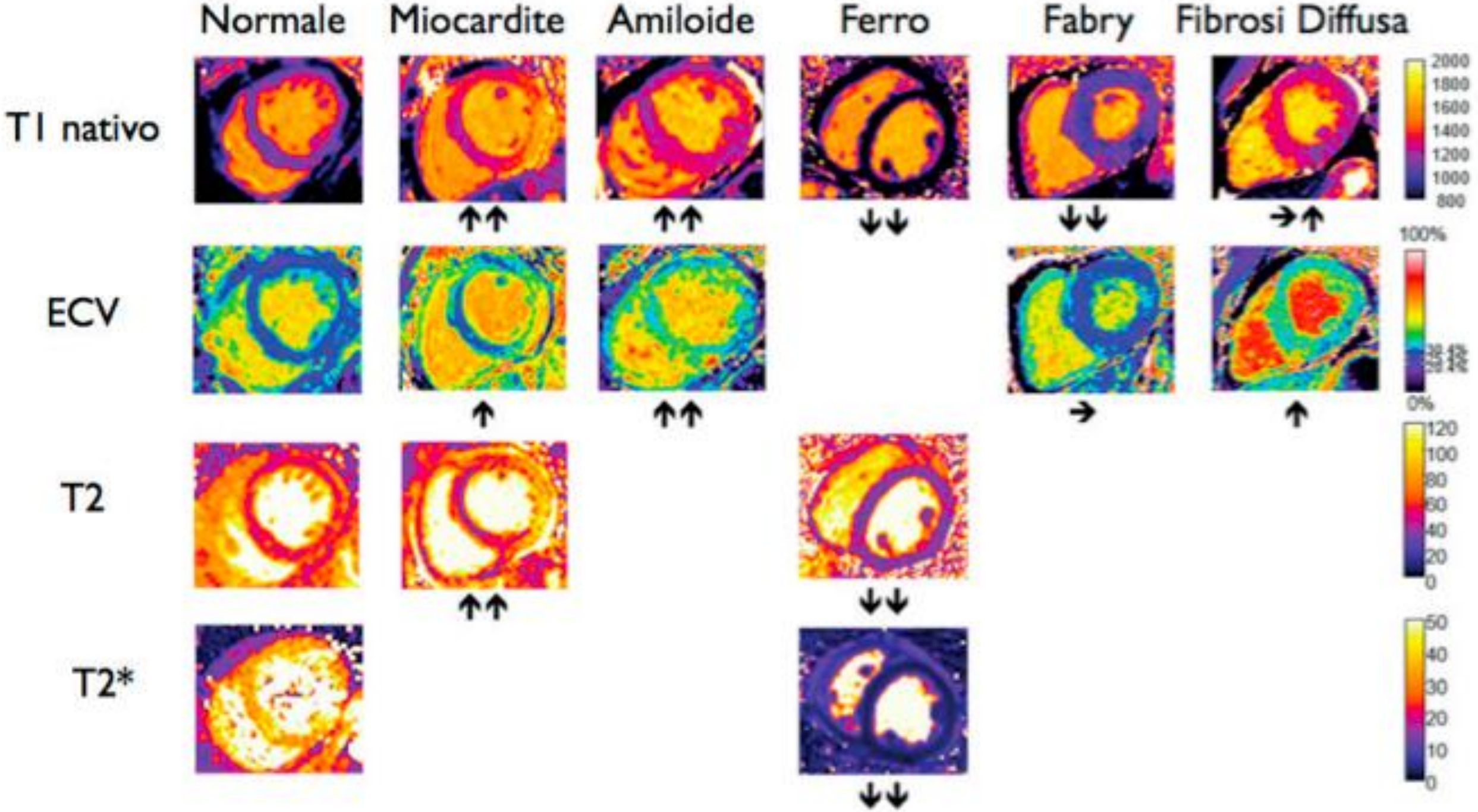


J Cardiovasc Magn Reson. 2008 Sep 25;10:42.
European Heart Journal – CI (2017) 18, 1090–1091



- **Cardiac ultrasound inadequate** to assess early cardiac structural changes and guide therapy
- **cardiac T2*** is the imaging of choice, **should be performed as early as possible and the effectiveness of iron chelation can be reliably guided by follow-up scans**

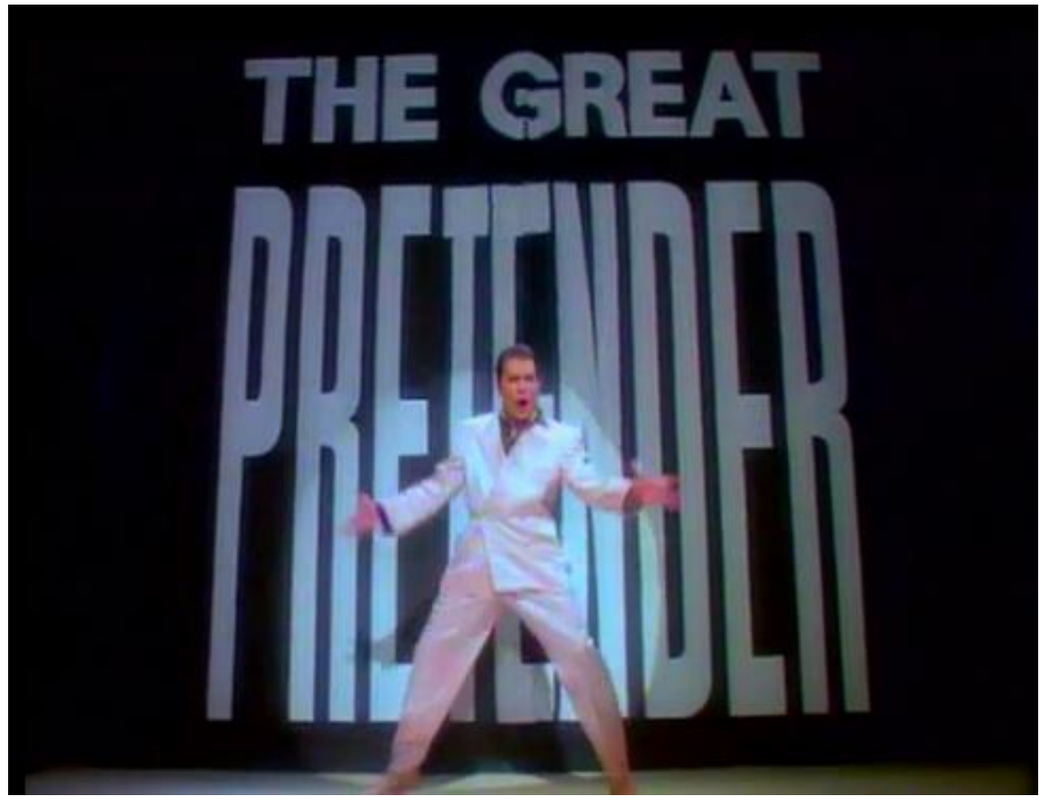
Mapping T1 e T2 e calcolo del volume extracellulare (ECV)



Courtesy of Dr. Imazio

ATTR: the great pretender

- Challenging
- Fascinating
- Mysterious
- Not as rare as supposed
- Relative easy to detect (when suspected!)
- Treatable !!



CARATTERISTICHE DELLE TRE FORME DI AMILOIDOSI SISTEMICA AD INTERESSAMENTO CARDIACO E DELLA CARDIOMIOPATIA IPERTROFICA

	ATTRm	AL	SSA	CMPI
Spessore parietale VS	Moderatamente aumentato (1.5-2.0 cm)	Lievemente aumentato (1.2-1.5 cm)	Severamente aumentato (1.8-12.2 cm)	Estremamente variabile (1.2-3.5 cm)
Ipertrofia VS	Tendenzialmente simmetrica	Tendenzialmente simmetrica	Tendenzialmente simmetrica	Variabile (asimmetrica, apicale, raramente simmetrica)
Frazione di eiezione VS	Lievemente ridotta	Normale/lievemente ridotta	Moderatamente ridotta	Normale o aumentata
Ipertrofia VD	Frequente	Frequente	Frequente	Possibile
Ispessimento del setto interatriale	Frequente	Frequente	Frequente	Assente
Disfunzione diastolica	Frequente	Frequente	Frequente	Frequente
Ispessimento delle valvole atrioventricolari	Frequente	Possibile	Frequente	Assente
Versamento pericardico	Frequente	Frequente	Frequente	Estremamente raro
Bassi voltaggi del QRS	<25% dei casi	Frequente	<25% dei casi	Estremamente raro
Captazione miocardica di 99mTc-DPD	Forte	Assente o debole	Forte	Assente
Valori di NT-proBNP e troponina cardiaca	Moderatamente aumentati	Severamente aumentati	Moderatamente aumentati	Moderatamente aumentati