

Διαγνωστική προσέγγιση του αρρώστου με πιθανή καρδιακή αμυλοείδωση

02/11/2011 12:08:32

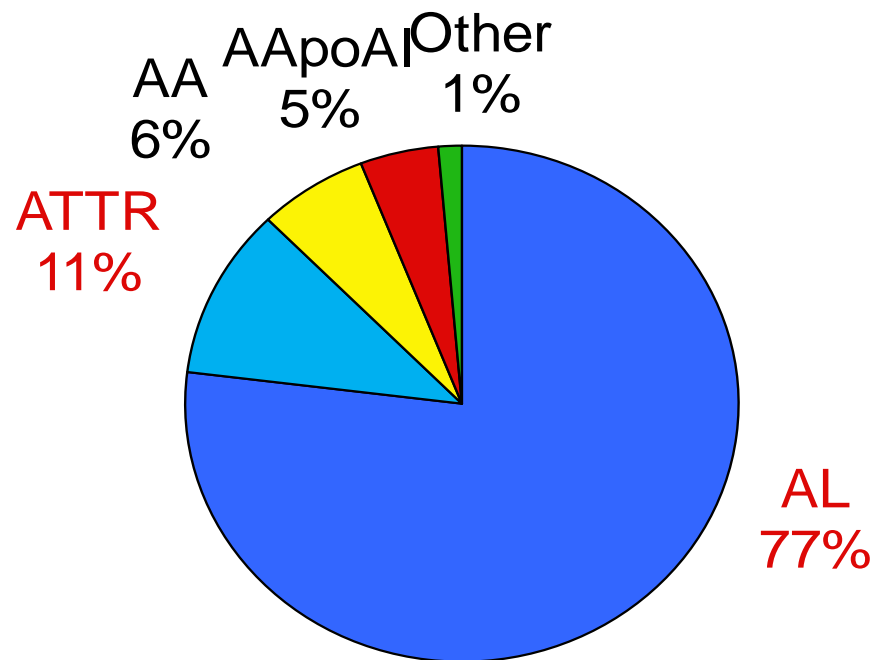


2:290 93 HR

Θεραπευτική Κλινική



Ιταλία: Συχνότητα των διαφόρων μορφών αμυλοείδωσης (στοιχεία σε 2338 ασθενείς από το κέντρο αμυλοείδωσης της Pavia)



1780 patients with AL amyloidosis
Incidence: 10 patients/million/year

Δεδομένα από Καθ. Κο G.Merlini



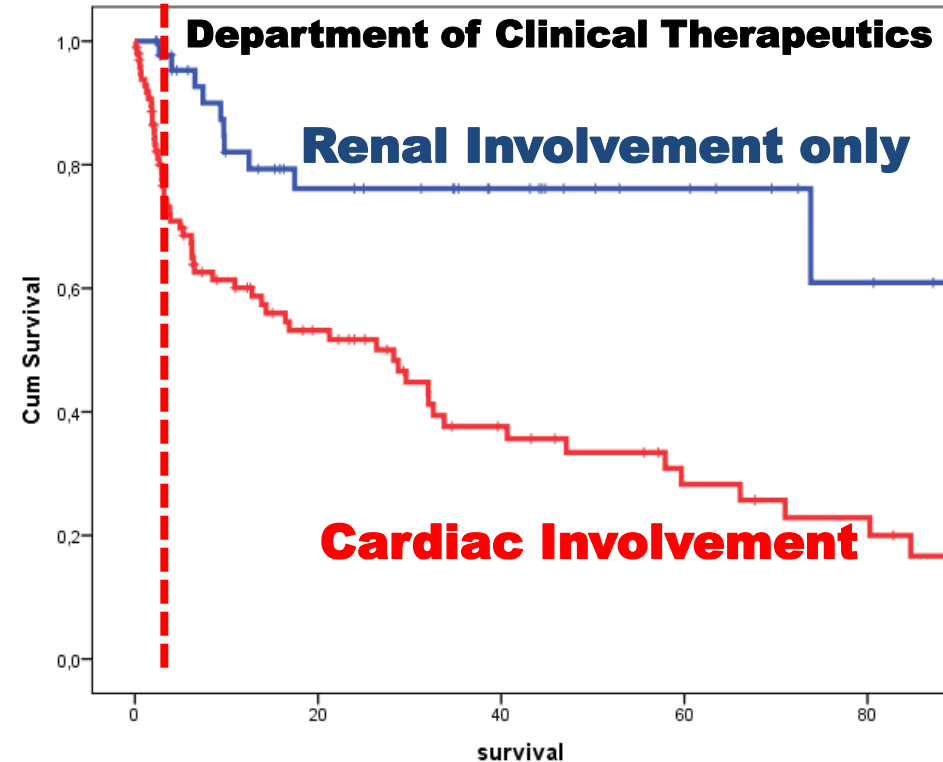
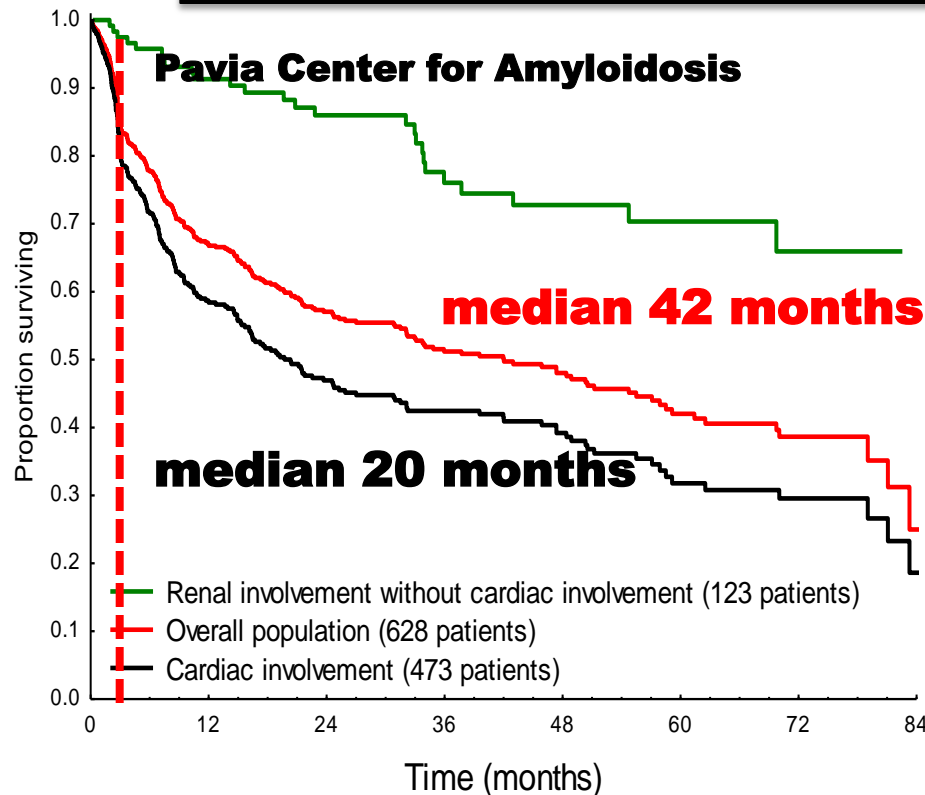
Κλινική παρουσίαση της πρωτοπαθούς αμυλοείδωσης

Γενικά συμπτώματα: Καταβολή, αδυναμία, απώλεια βάρους	60-90%
Οίδημα κάτω άκρων	55-65%
Δύσπνοια στην προσπάθεια	40-50%
Ορθοστατική υπόταση	27-30%
Δυσαισθησίες, παραισθησίες, σύνδρομο καρπιαίου σωλήνα	23-28%
Δυσγευσία	15-18%
Πορφύρα	8-15%
Μακρογλωσσία	10-15%
Διάρροια	8-15%
Αμυλοείδωμα (ογκόμορφη εντόπιση)	3-8%



Η προσβολή του μυοκαρδίου καθορίζει την πρόγνωση

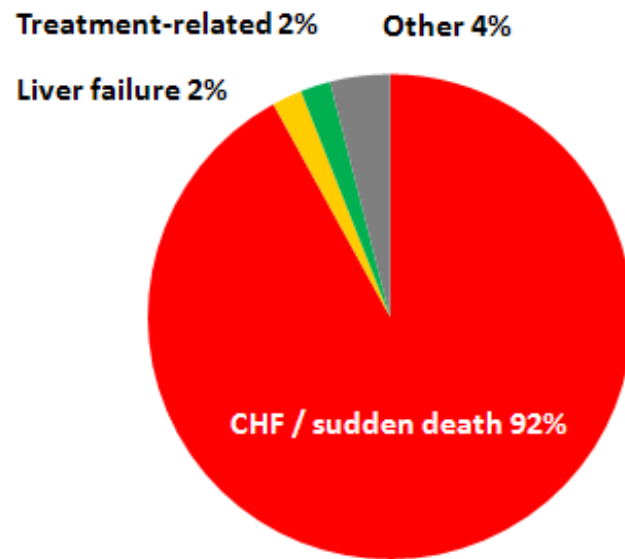
~20% των αρρώστων με AL και προσβολή της καρδιάς καταλήγουν στους πρώτους 3 μήνες



Η πρώιμη διάγνωση και έναρξη θεραπείας μπορεί να σώσει την ζωή αρρώστων με καρδιακή AL αμυλοείδωση

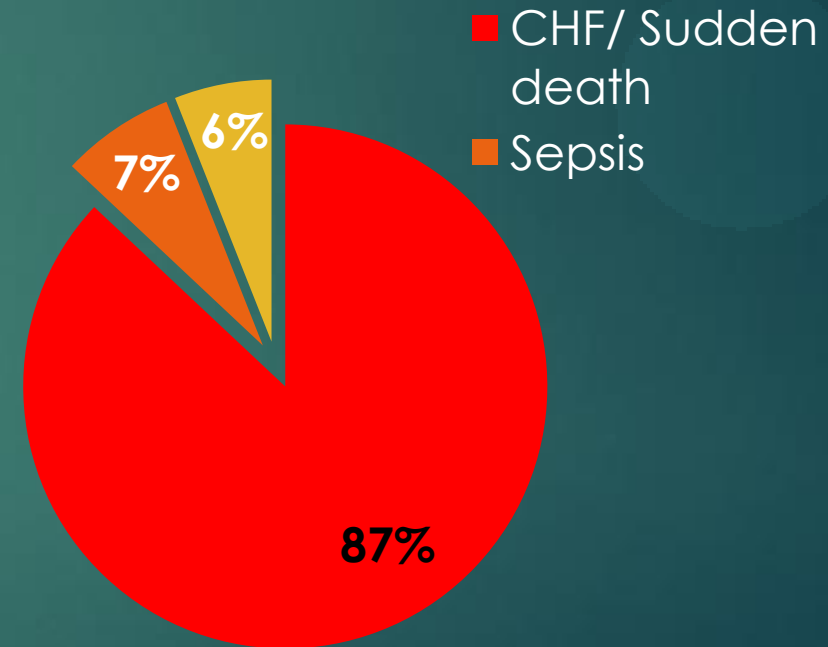
Πρόγνωση των αρρώστων με AL αμυλοείδωση

Causes of death in the first year after diagnosis in 169 patients with AL amyloidosis



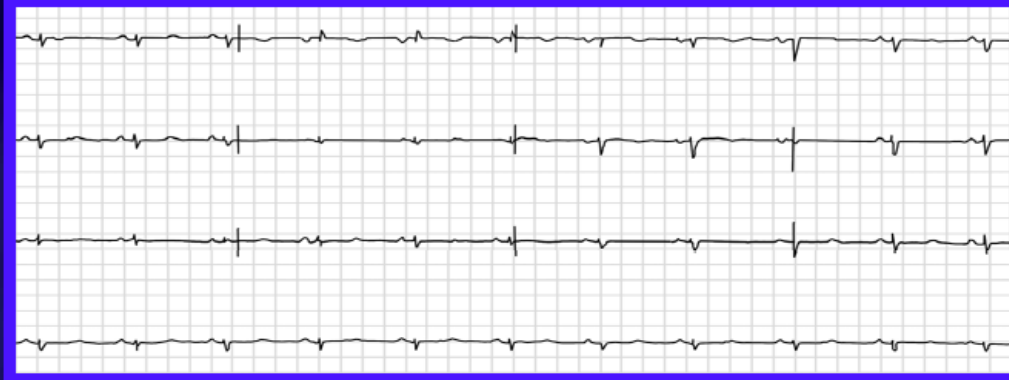
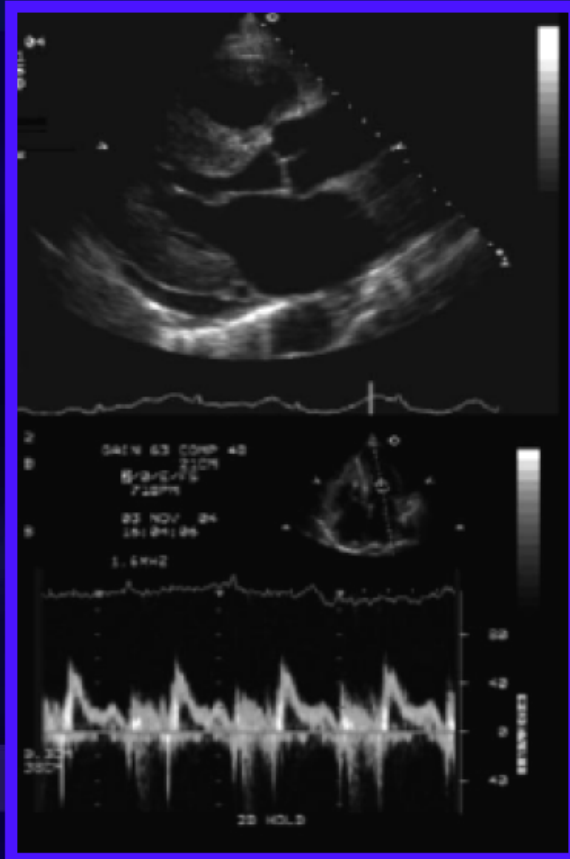
Courtesy of Prof Merlini

Cause of death N=132



Data: Department of Clinical Therapeutics 2017

Cardiac Amyloid

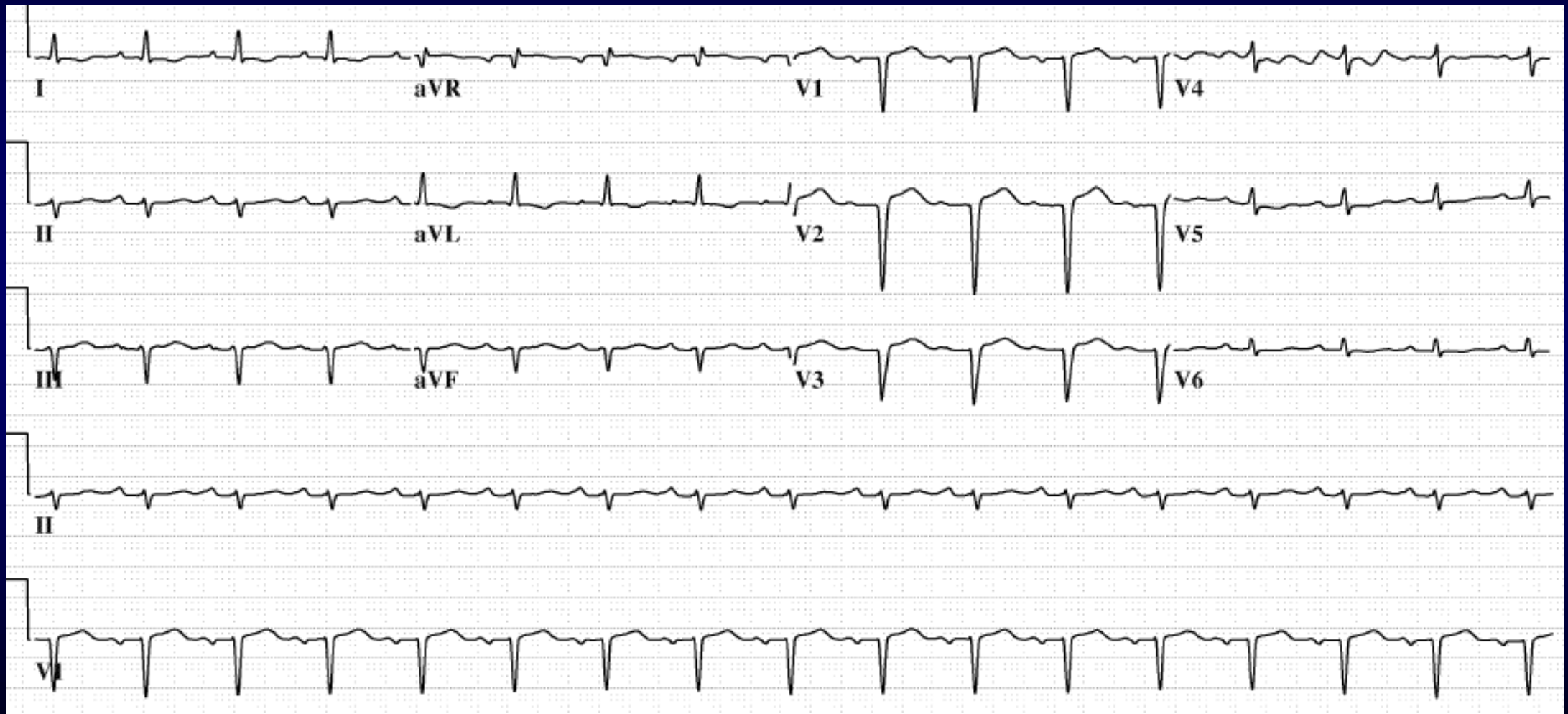


**In the proper clinical setting
these findings are as diagnostic
as myocardial biopsy**



ECG findings in cardiac amyloidosis

- ♥ **Low QRS voltage pattern**
- ♥ **Pseudoinfarction patterns**
- ♥ **Supraventricular arrhythmias (mainly atrial fibrillation)**
- ♥ **Atrioventricular or intra- and interventricular conduction defects**
- ♥ **Unusual axis deviations**
- ♥ **ECG may not demonstrate low-voltage complexes in individuals with ATTR**



15/04/2014 12:41:29

V

5

10



58
HR

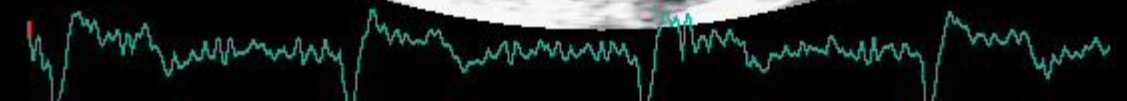
15/04/2014 12:53:19

V

5

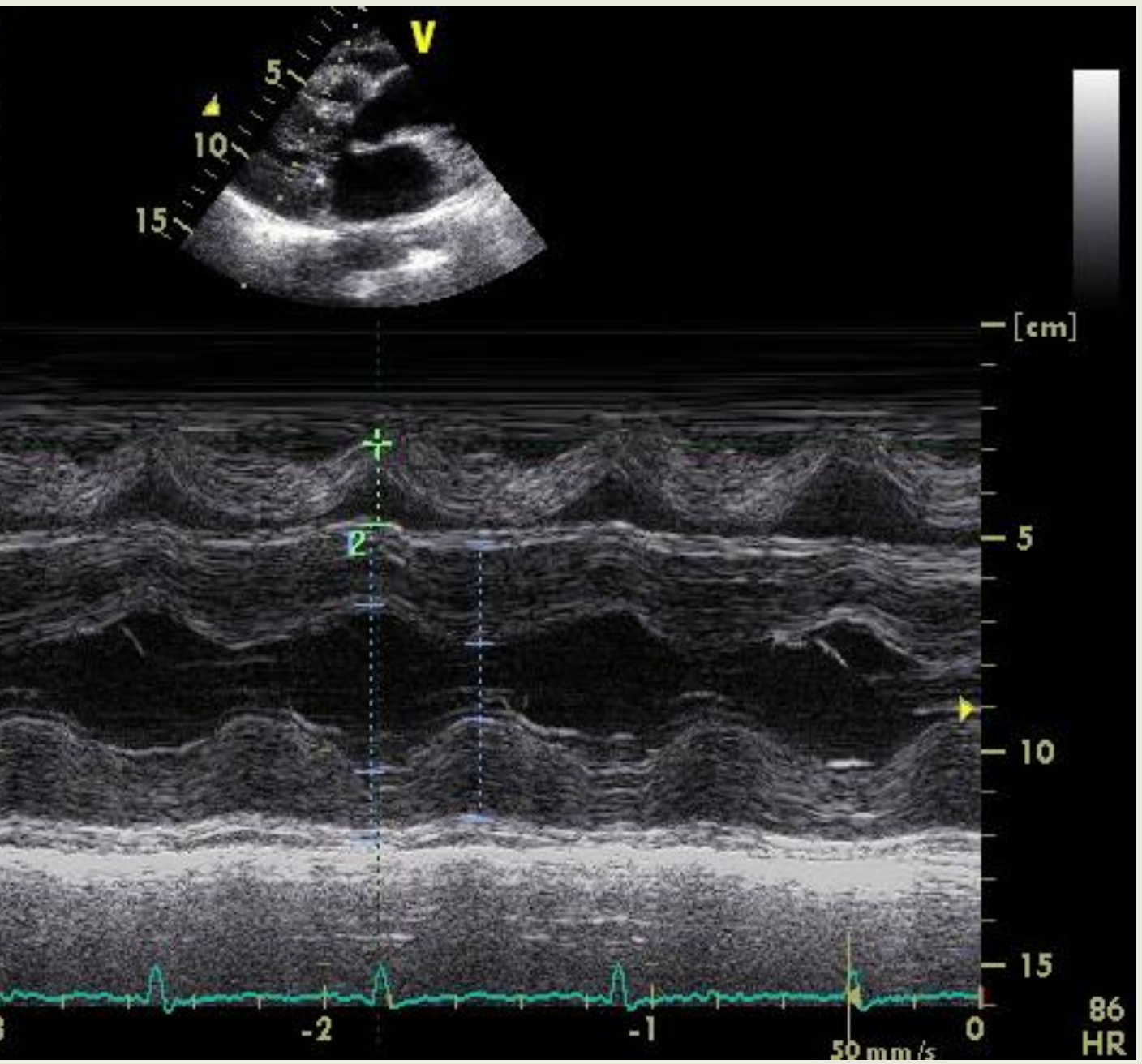
10

15

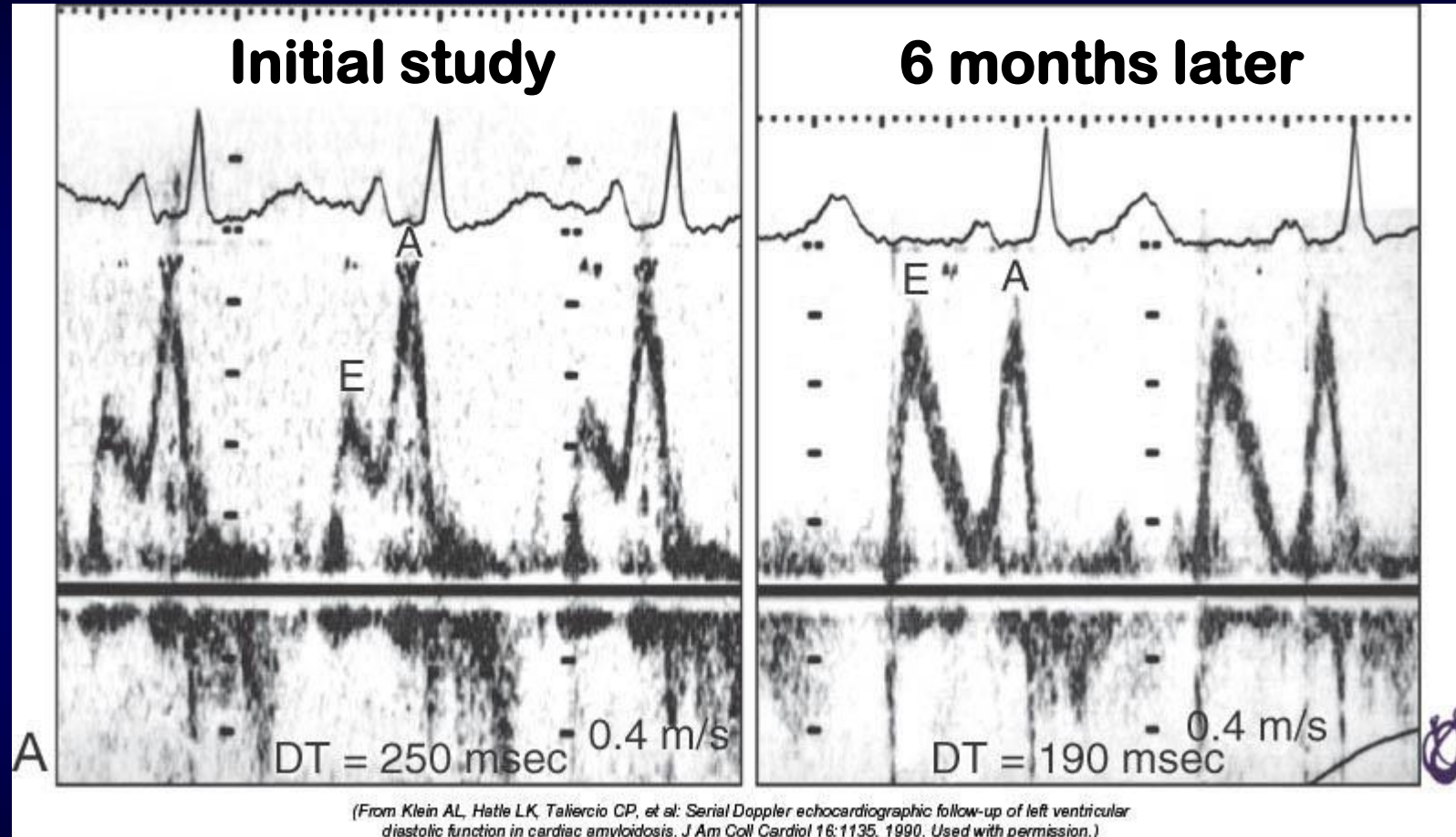


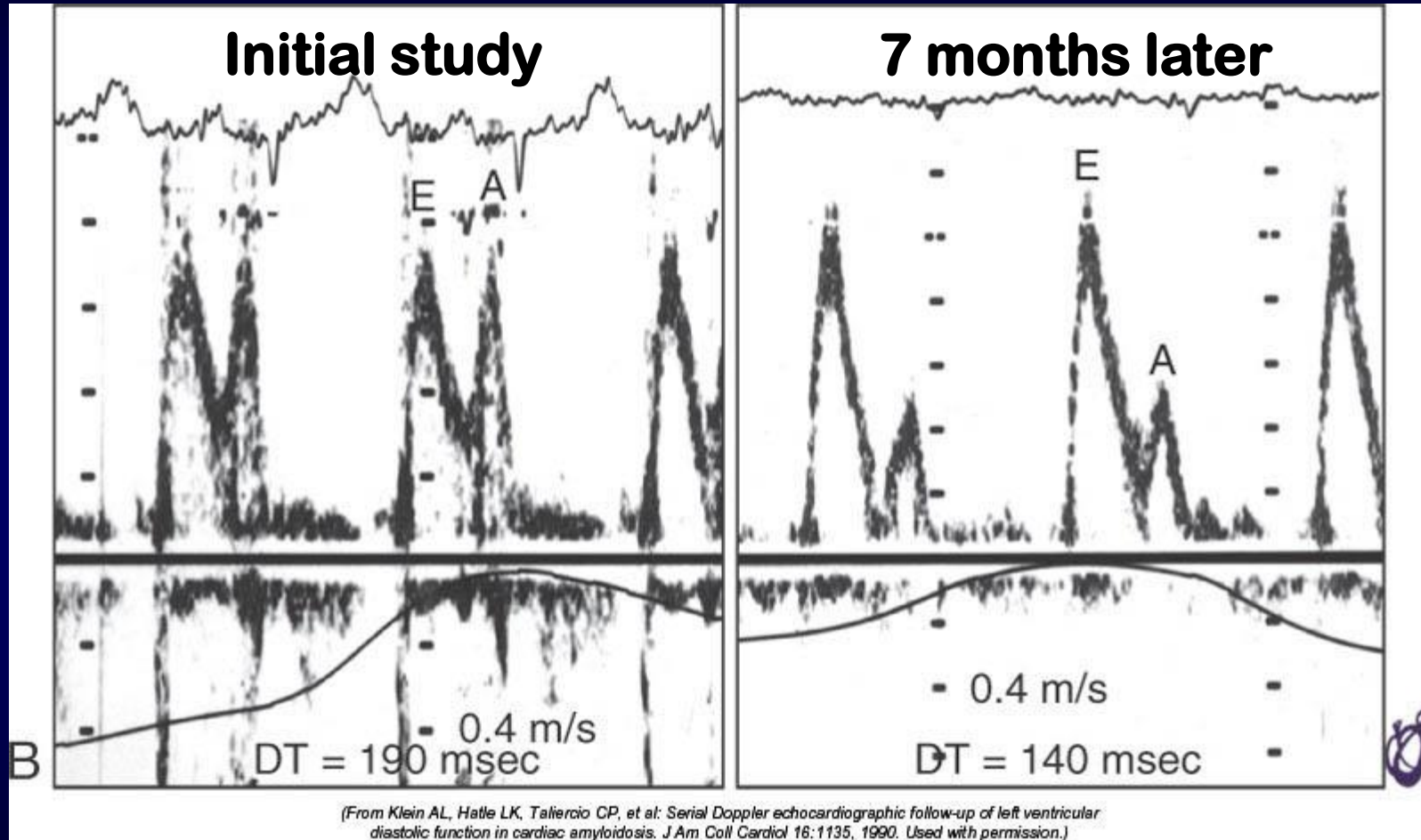
73
HR

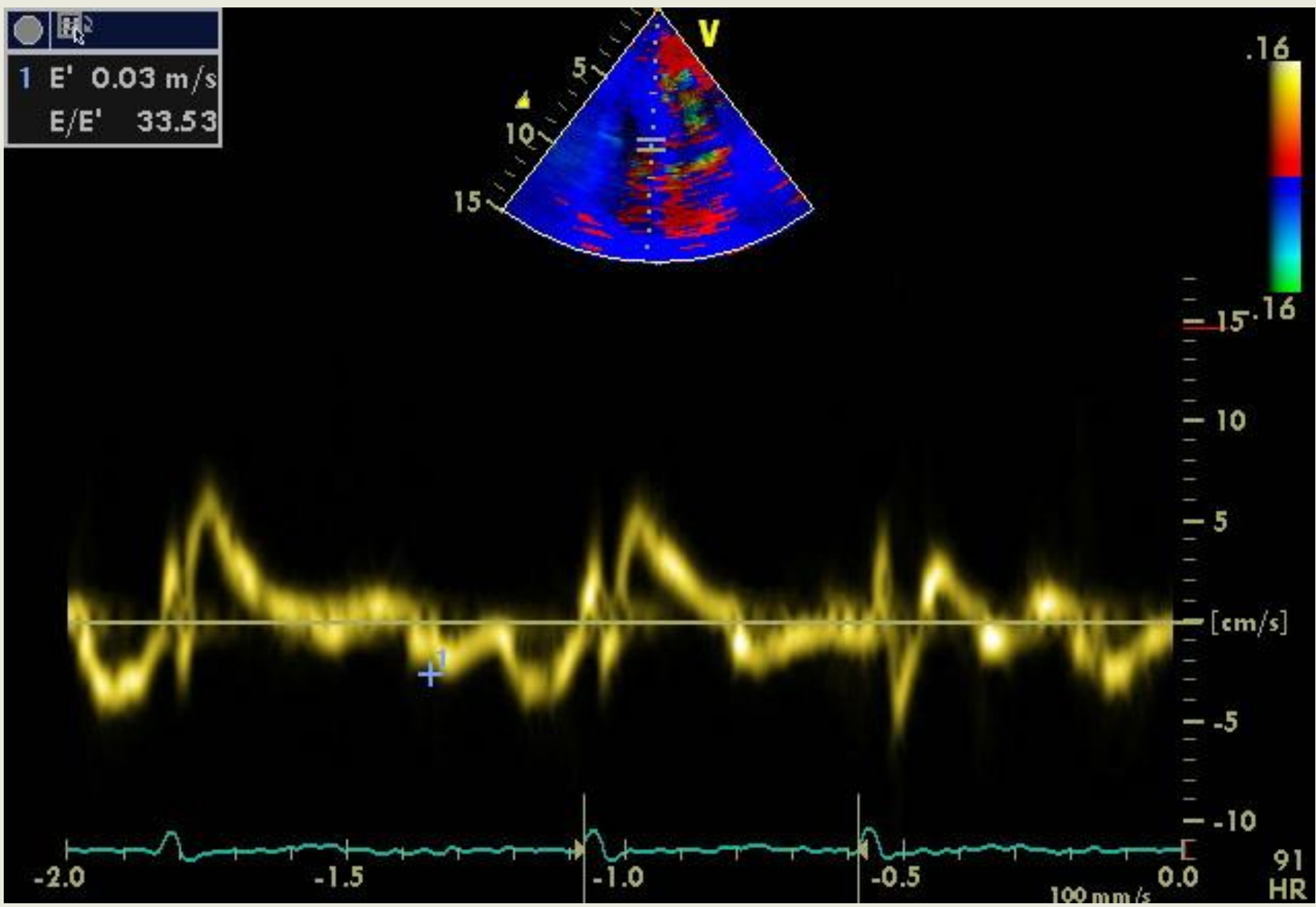
2	RVIDd	1.87 cm
1	IVSd	1.87 cm
	LVIDd	3.91 cm
	LVPWd	1.59 cm
	IVSs	2.38 cm
	LVIDs	1.76 cm
	LVPWs	2.38 cm
	EDV(Teich)	66.51 ml
	ESV(Teich)	9.16 ml
	EF(Teich)	86.23 %
	%FS	55.07 %
	SV(Teich)	57.35 ml



Diastolic dysfunction in cardiac amyloidosis

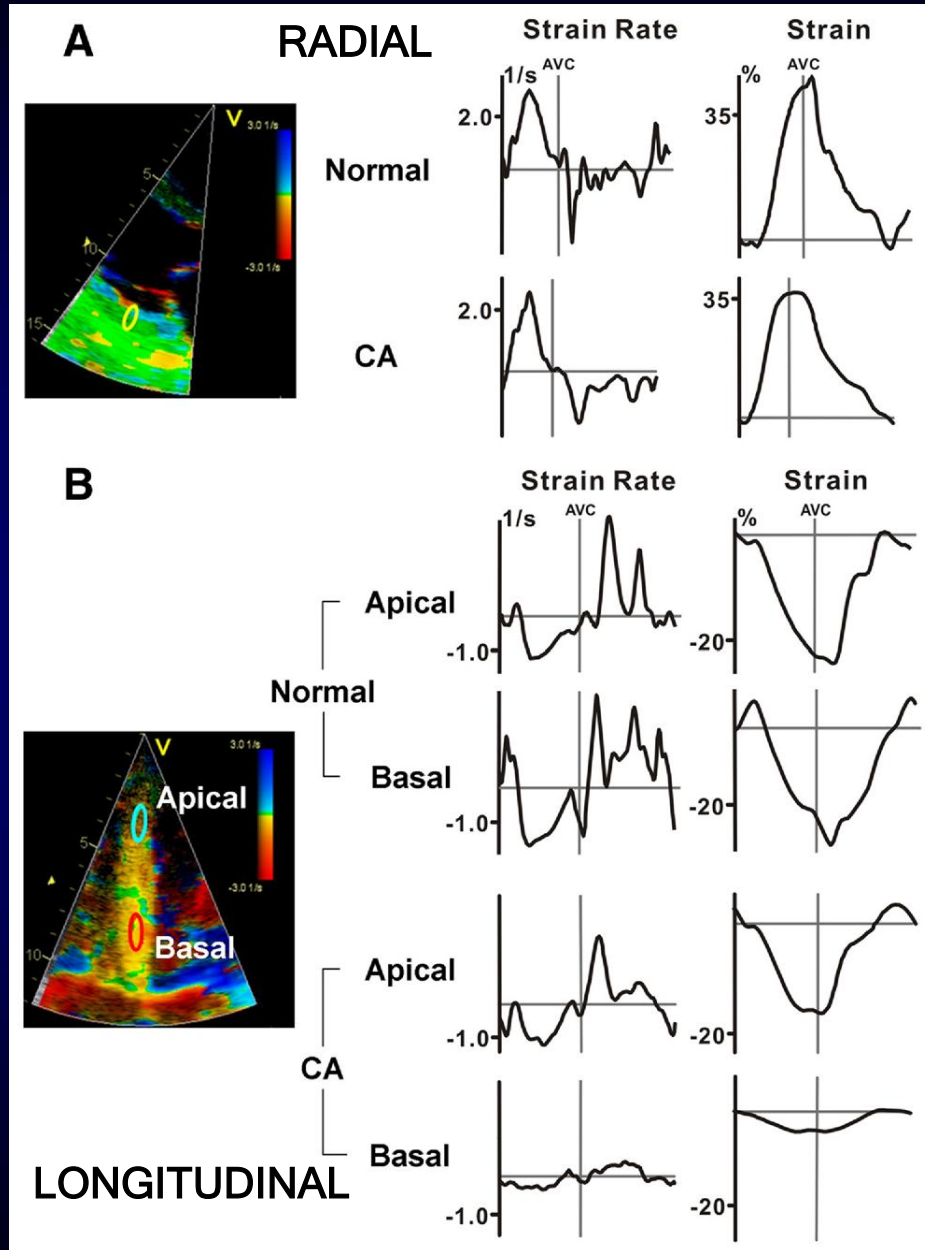






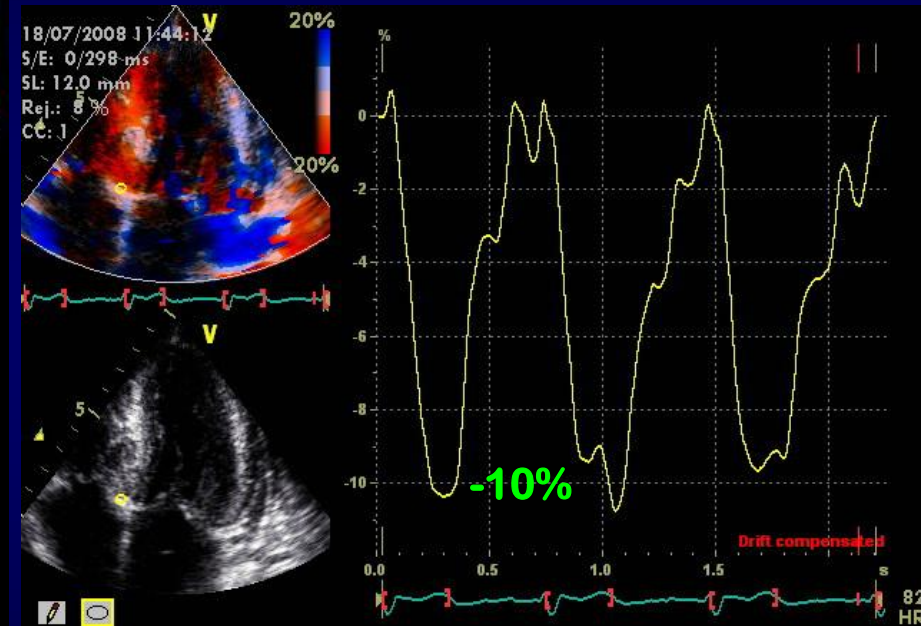
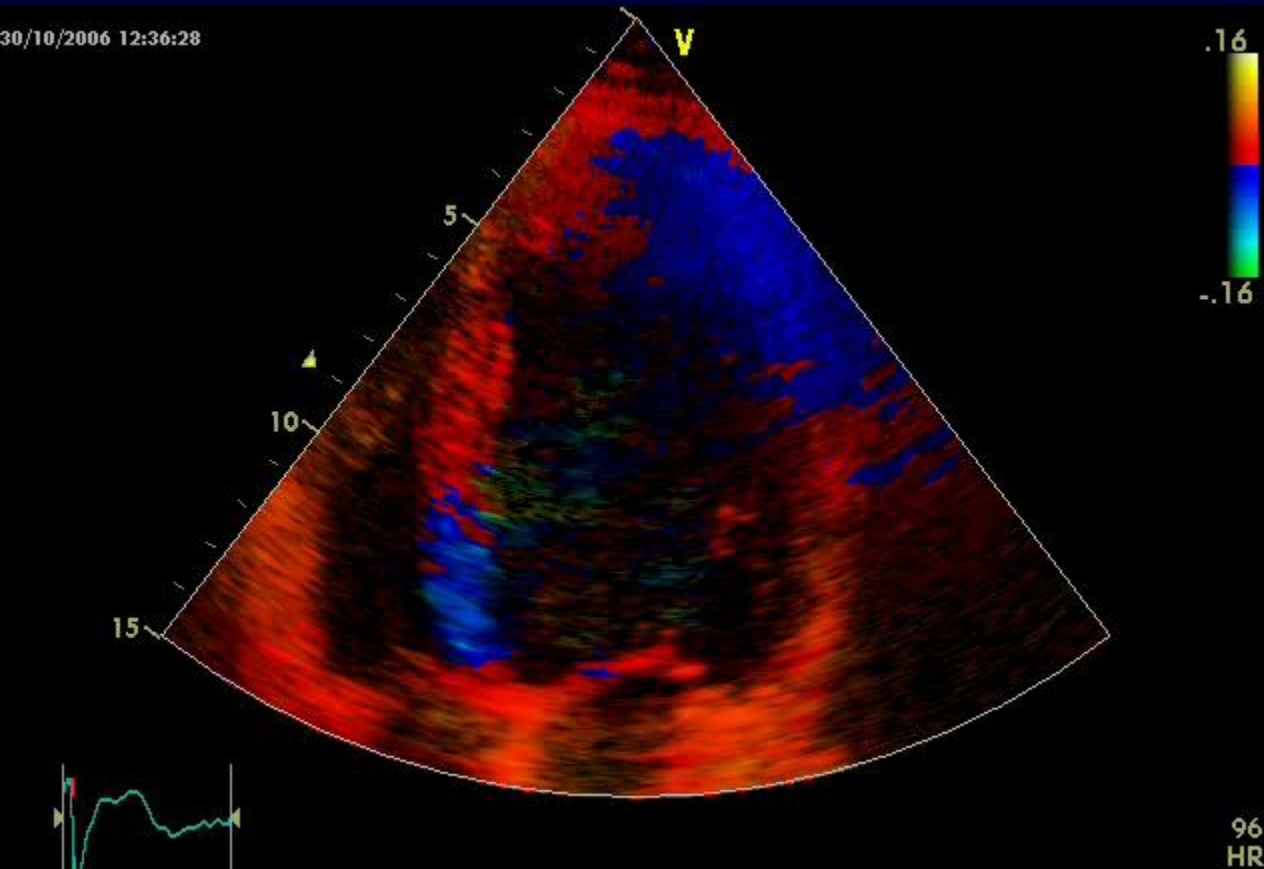


TDI derived strain and strain rate in cardiac amyloidosis

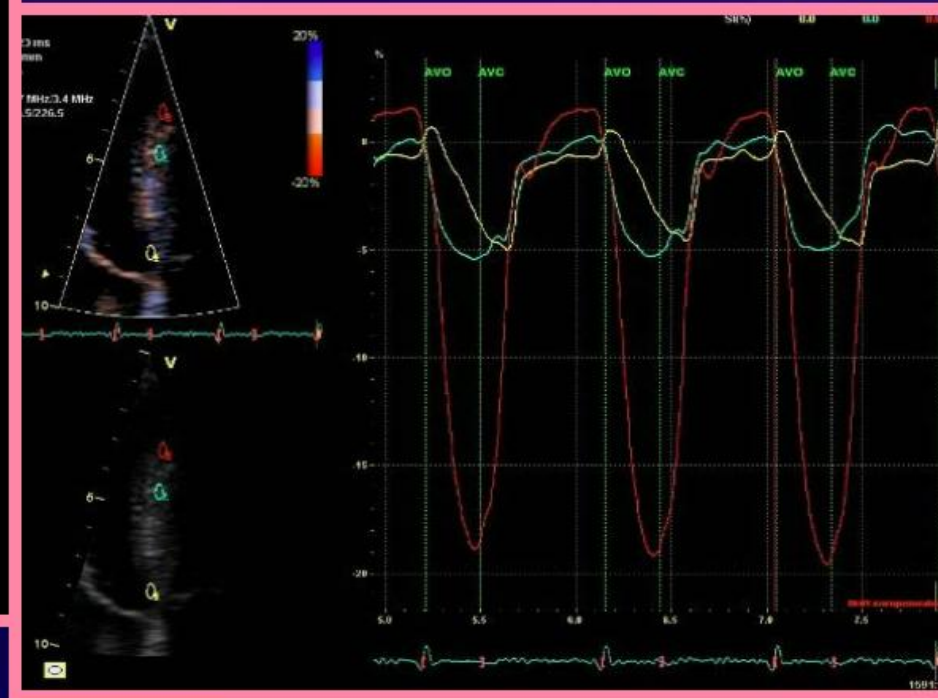




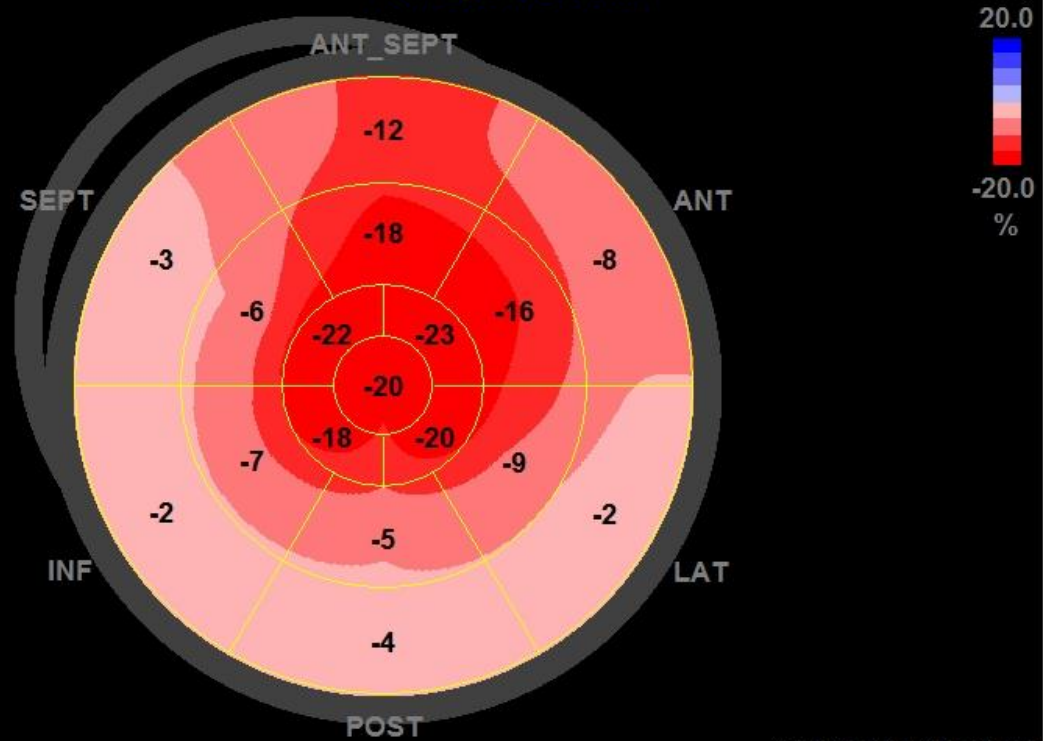
TDI derived strain in cardiac amyloidosis



Doppler Myocardial Imaging for Cardiac Amyloid

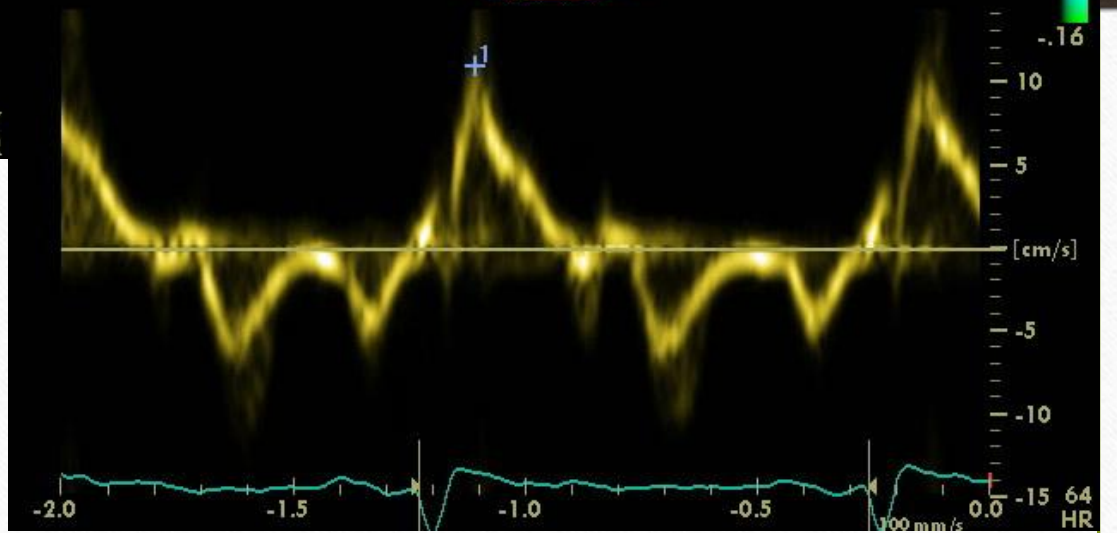
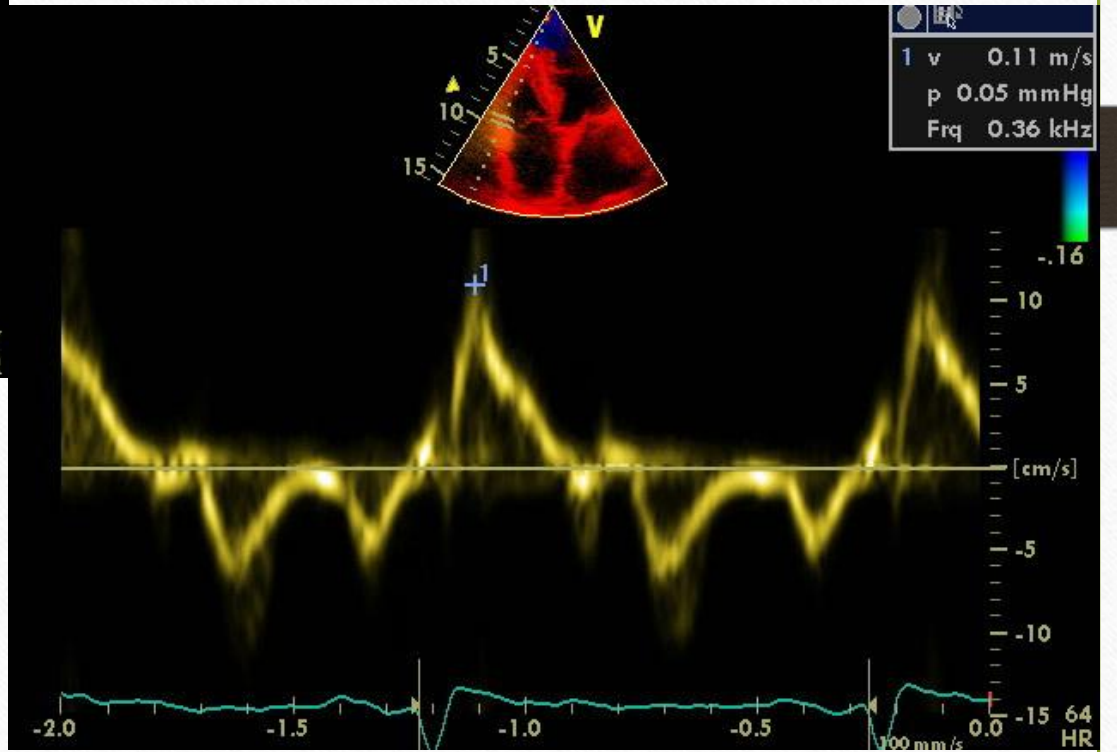
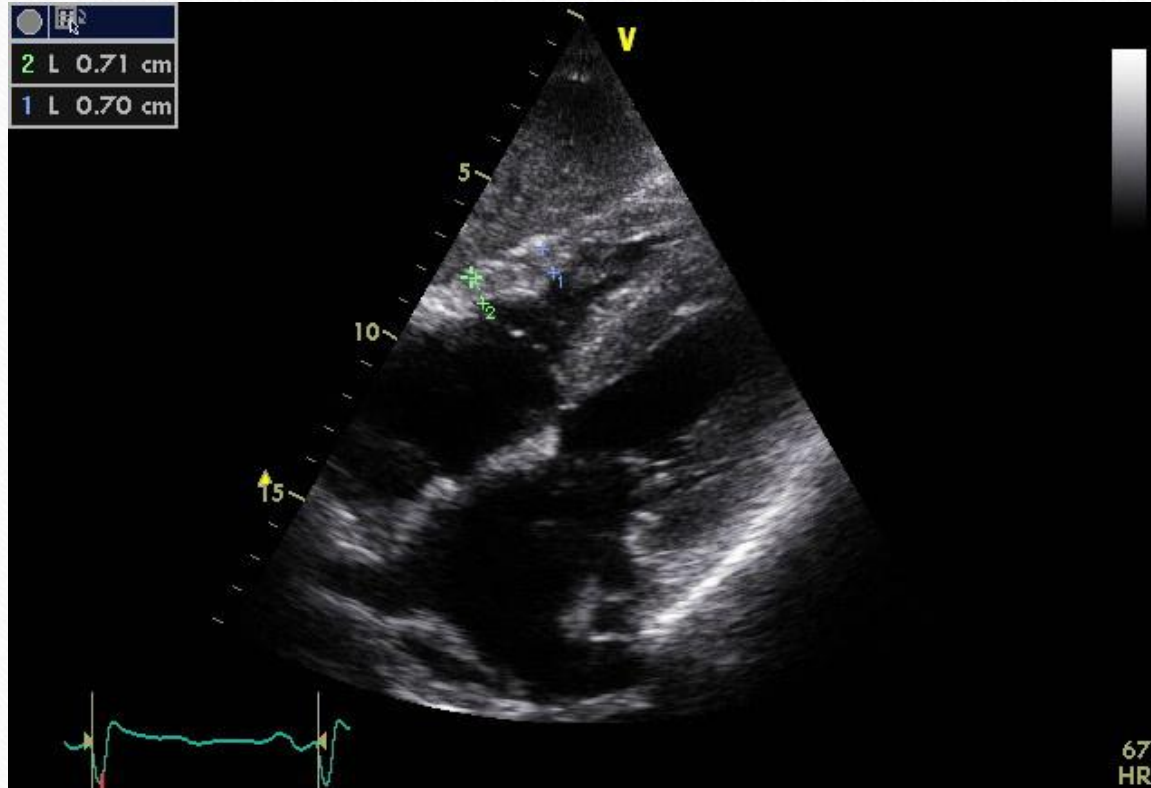


Peak Systolic Strain



09/01/2017-12:26:09

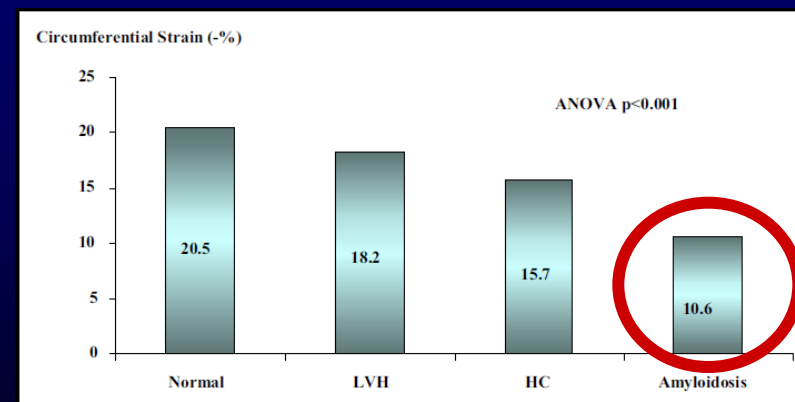
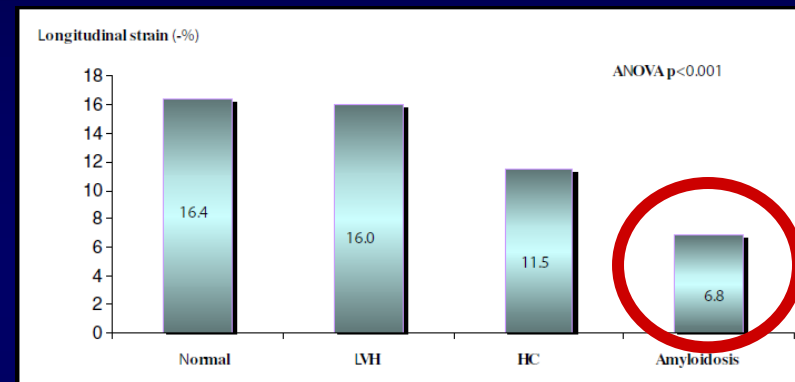
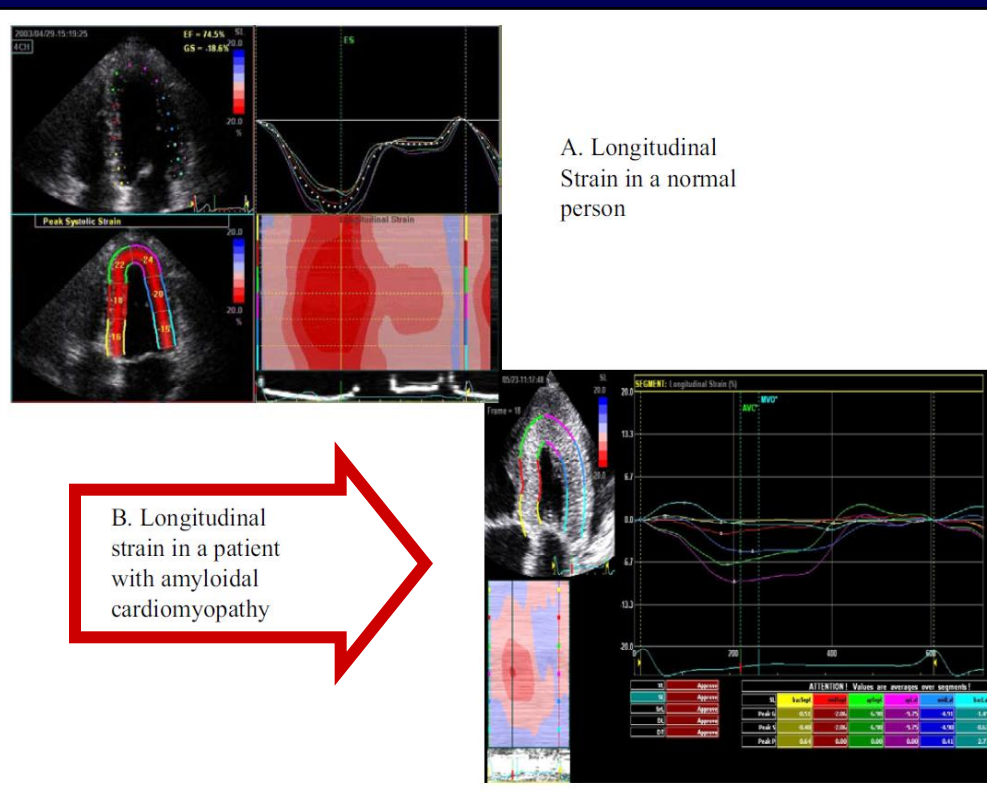
GLPS_LAX	-12.1 %	AVC_MAN	336 msec
GLPS_A4C	-11.5 %	HR_ApLAX	64.3 bpm
GLPS_A2C	-12.8 %		
GLPS_Avg	-12.1 %		

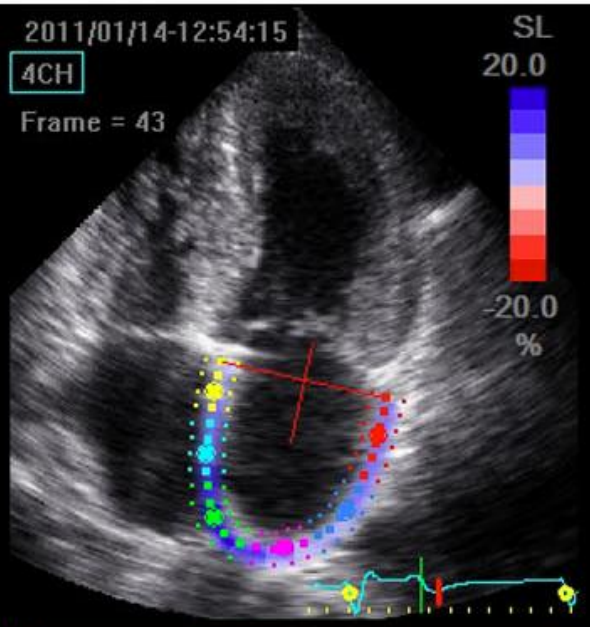
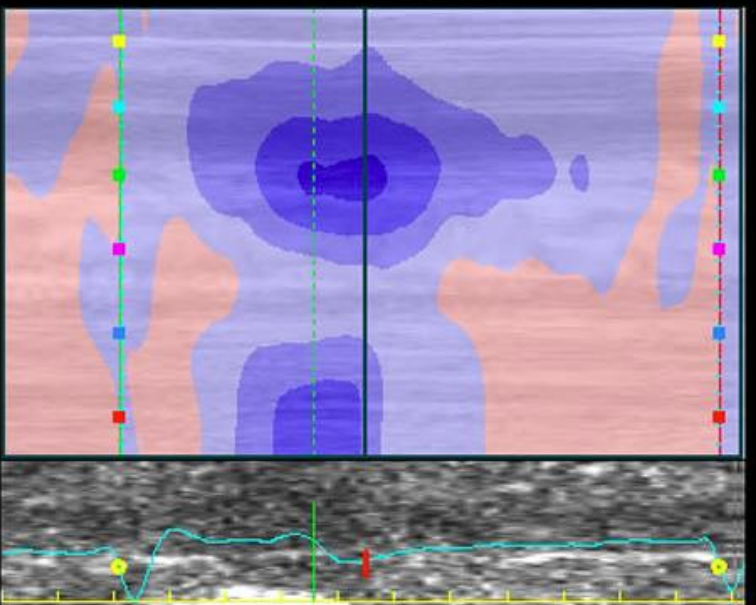
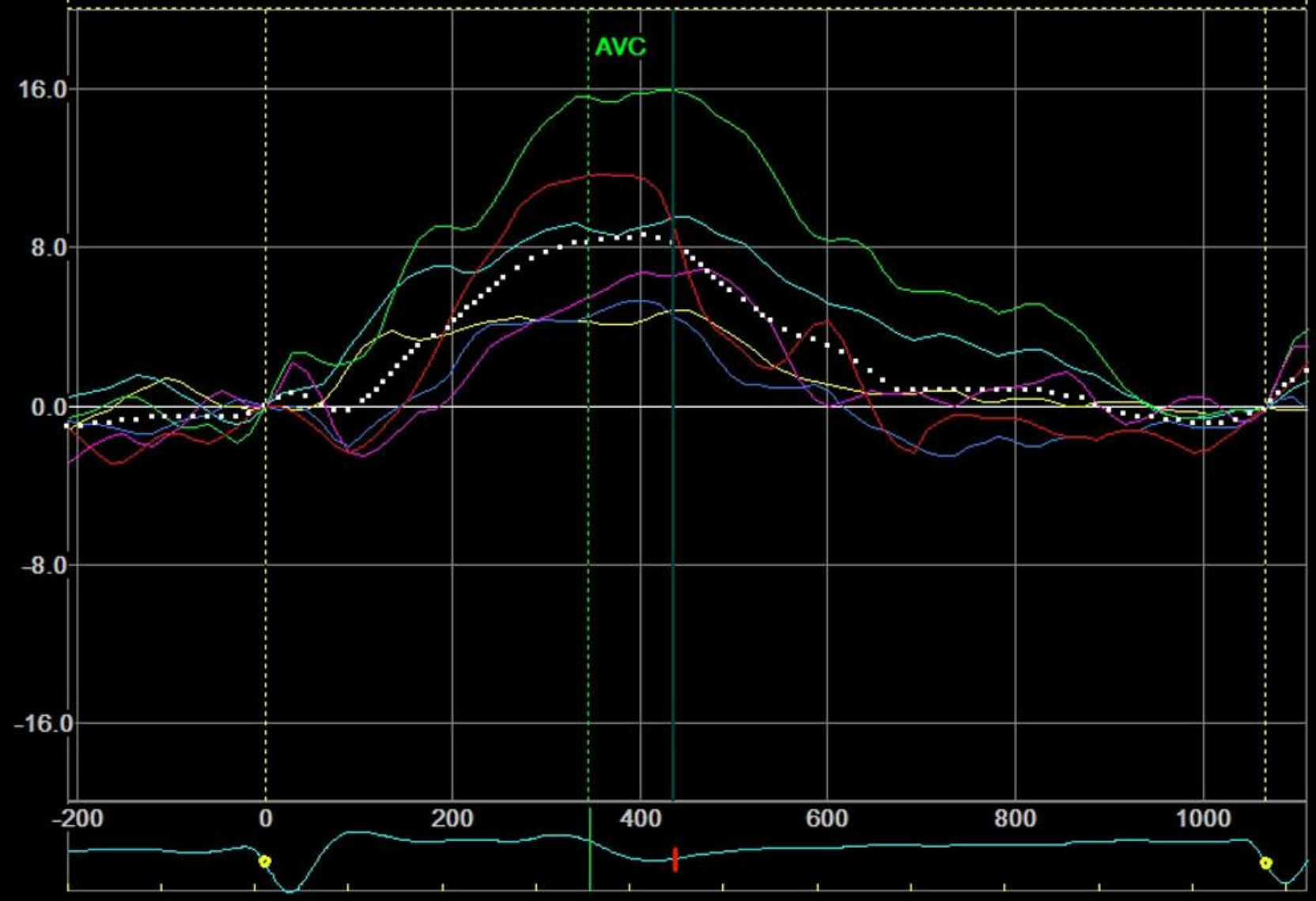




Differentiation of Hypertrophic Cardiomyopathy and Cardiac Amyloidosis from Other Causes of Ventricular Wall Thickening by Two-Dimensional Strain Imaging Echocardiography

Jing Ping Sun, MD^{a,*}, William J. Stewart, MD^b, Xing Sheng Yang, MD, PhD^a, Robert O. Donnell, MD^a, Angel R. Leon, MD^a, Joel M. Felner, MD^a, James D. Thomas, MD^b, and John D. Merlino, MD^a

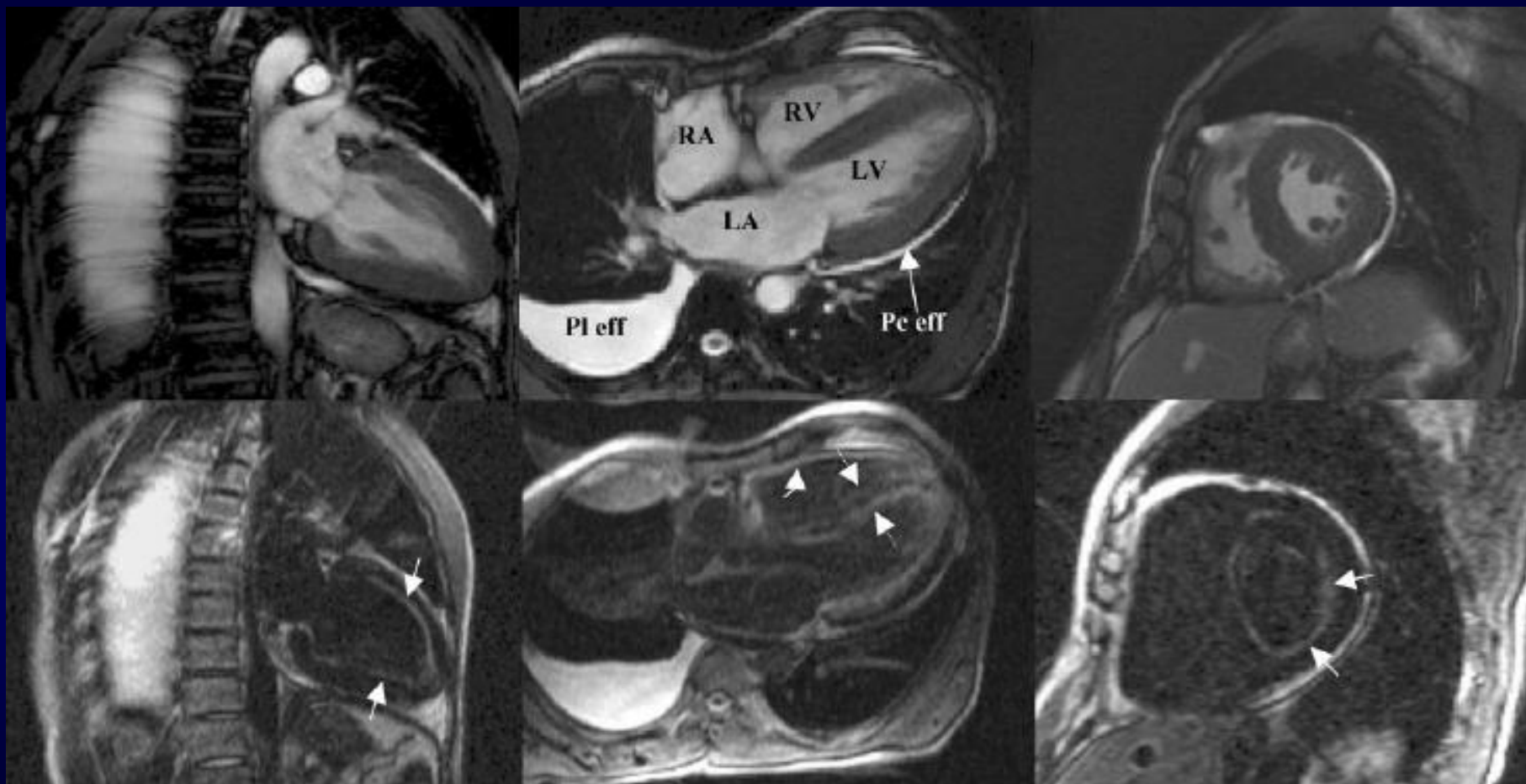


B**LOCAL: Longitudinal Strain (%) = -3.59****T=436 msec**



MRI in cardiac amyloidosis

Role of Gadolinium contrast-enhanced MRI



global subendocardial late enhancement

Maceira et al., Circulation 2005



Impact of cardiac stage

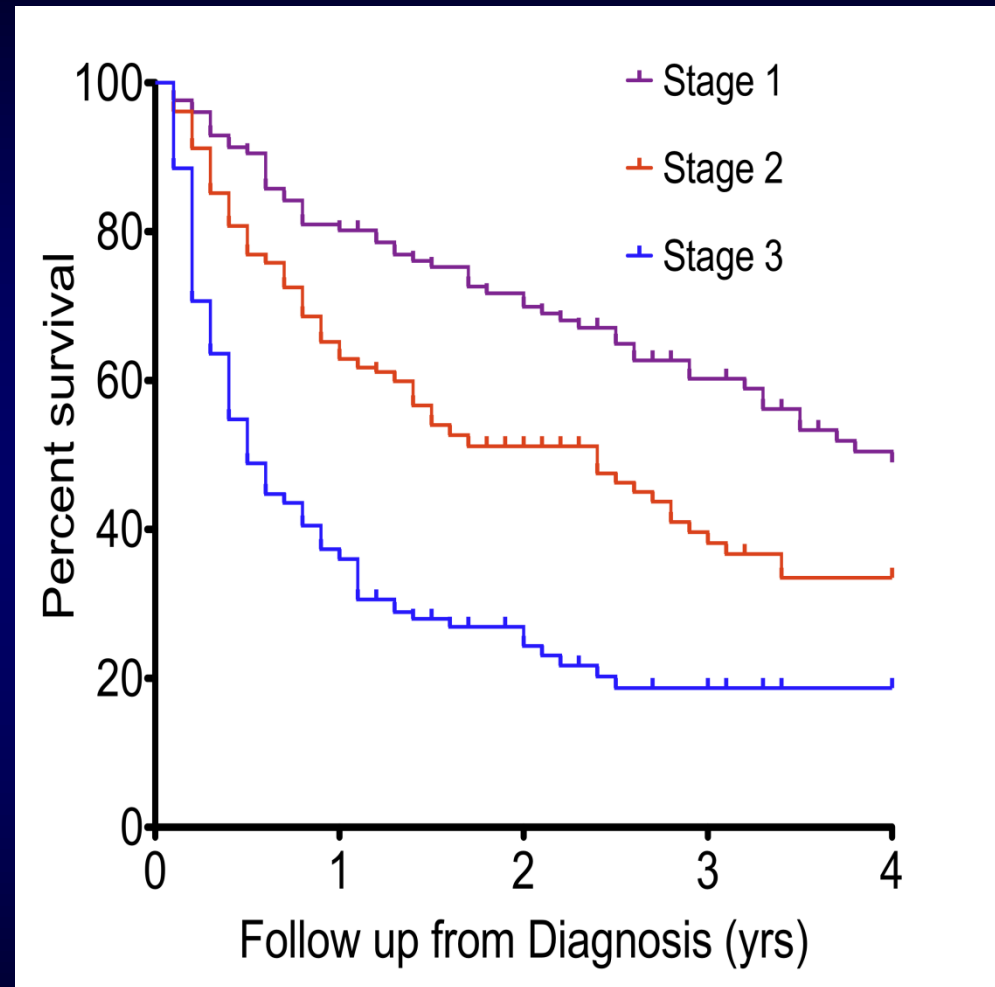
Troponin T < 0.035 ug/L

NT-proBNP < 332 ng/L

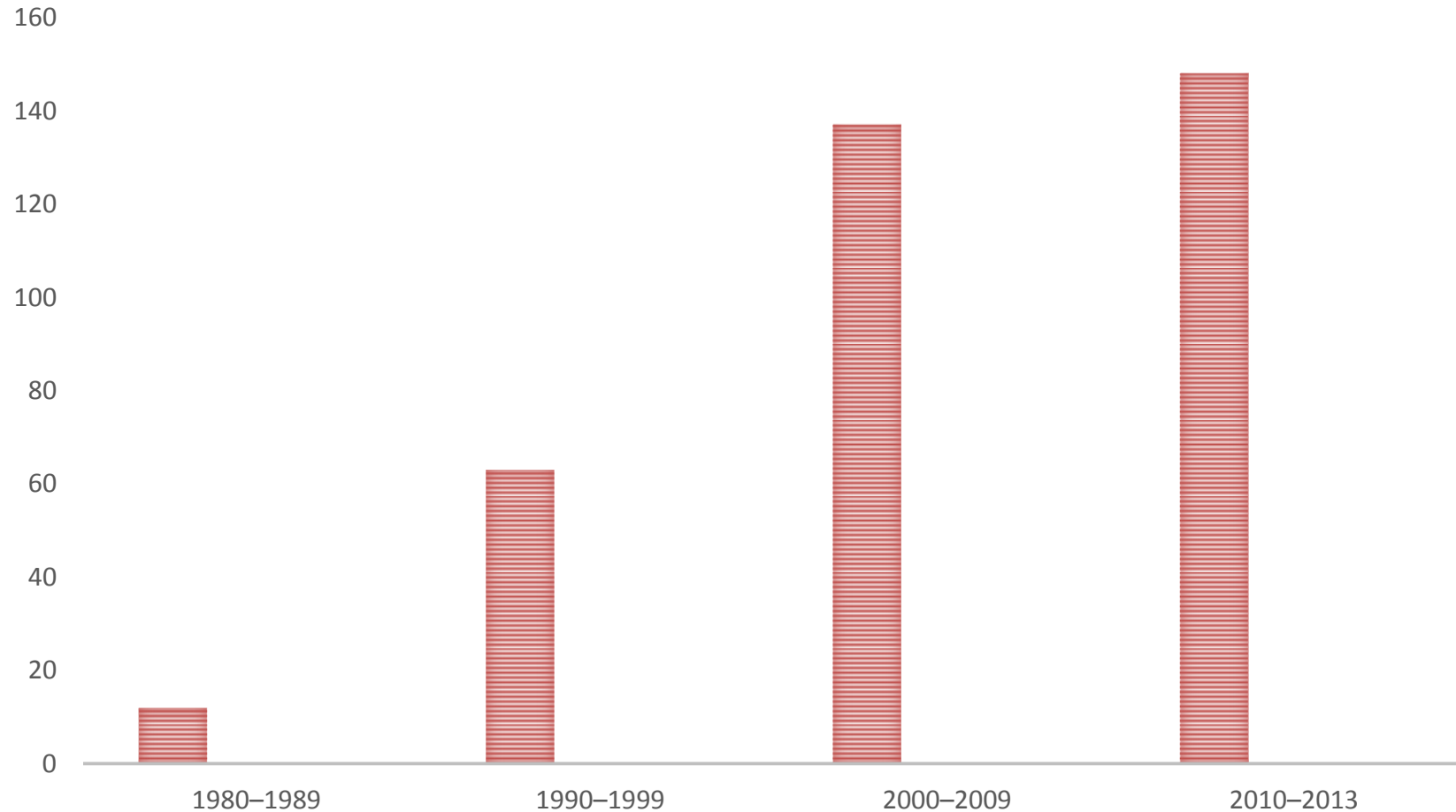
Stage 1: Neither high

Stage 2: Either high

Stage 3: Both high



Διάγνωση wtTTR καρδιακής αμυλοείδωσης: Mayo Clinic

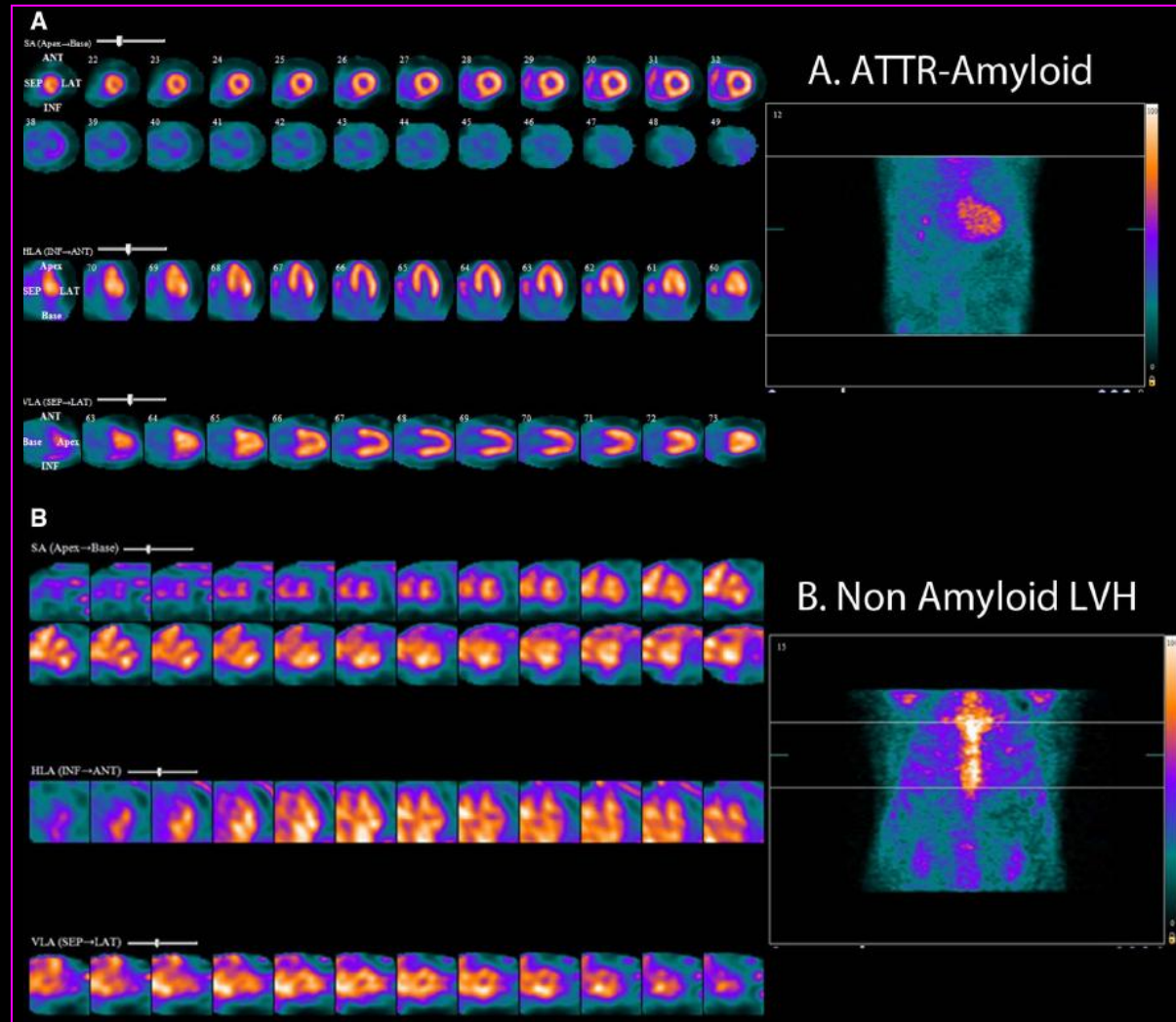




Radionuclide imaging of cardiac amyloidosis

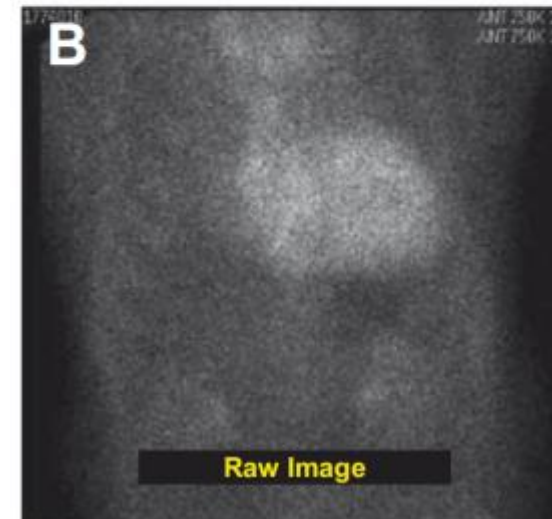
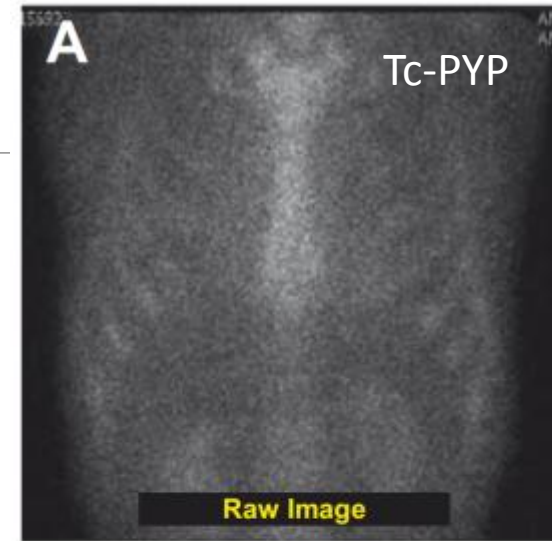
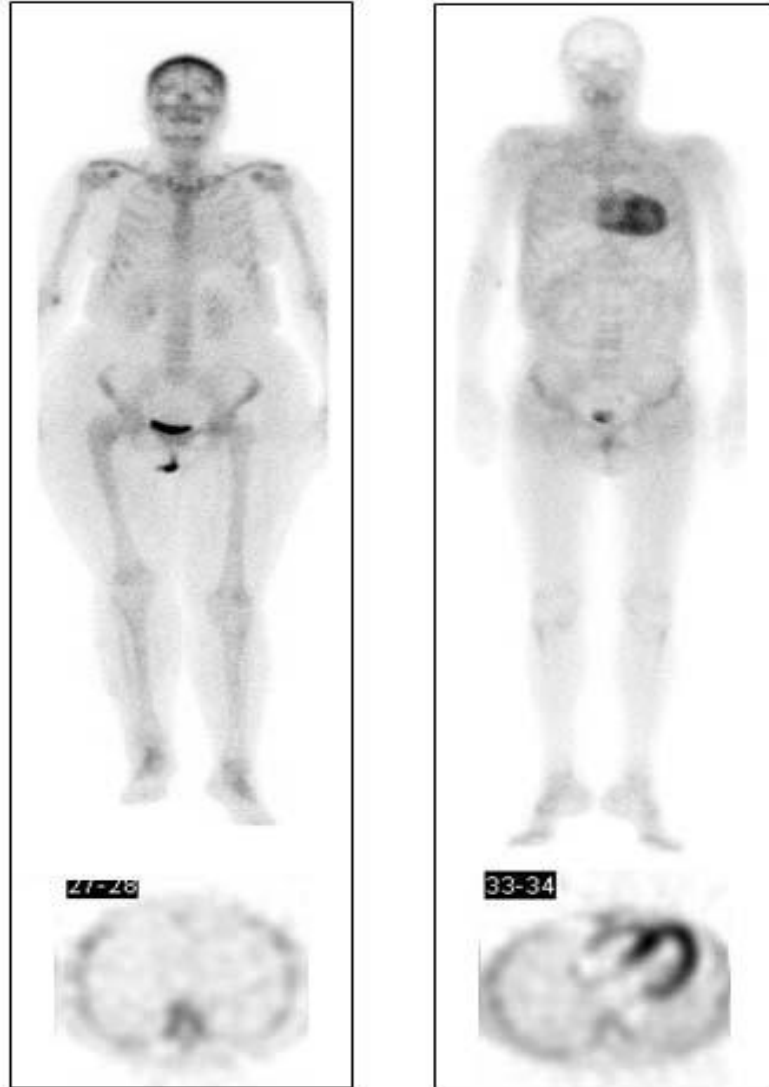
Tc-99m pyrophosphate single-photon emission computed tomography

Ευαισθησία 97%
Ειδικότητα ~100%
για ATTR έναντι AL καρδιακή αμυλοείδωση



Μη επεμβατική τυποποίηση: Σπινθηρογράφημα με Tc99^m-DPD ή Tc99^m-PYP για αμυλοείδωση από τρανσθυρετίνη

AL Tc-DPD ATTR



Aortic stenosis and transthyretin cardiac amyloidosis: the chicken or the egg?

