When the Left Ventricular Walls are Thick
An imaging perspective

Helen Thomson
Heart Centre at the Alfred

Disclosures

None

Case

43 year old architect

2/7 Hx of dry cough and SOB and decreased exercise tolerance

1/7 Hx of marked orthopnea

S/B A&E

More Hx

Smoker 1 pack/day

Binge drinker

Recreational drugs: MDMA, Ketamine, Cannabis, Heroin, Metamphetamines and ICE

Examination

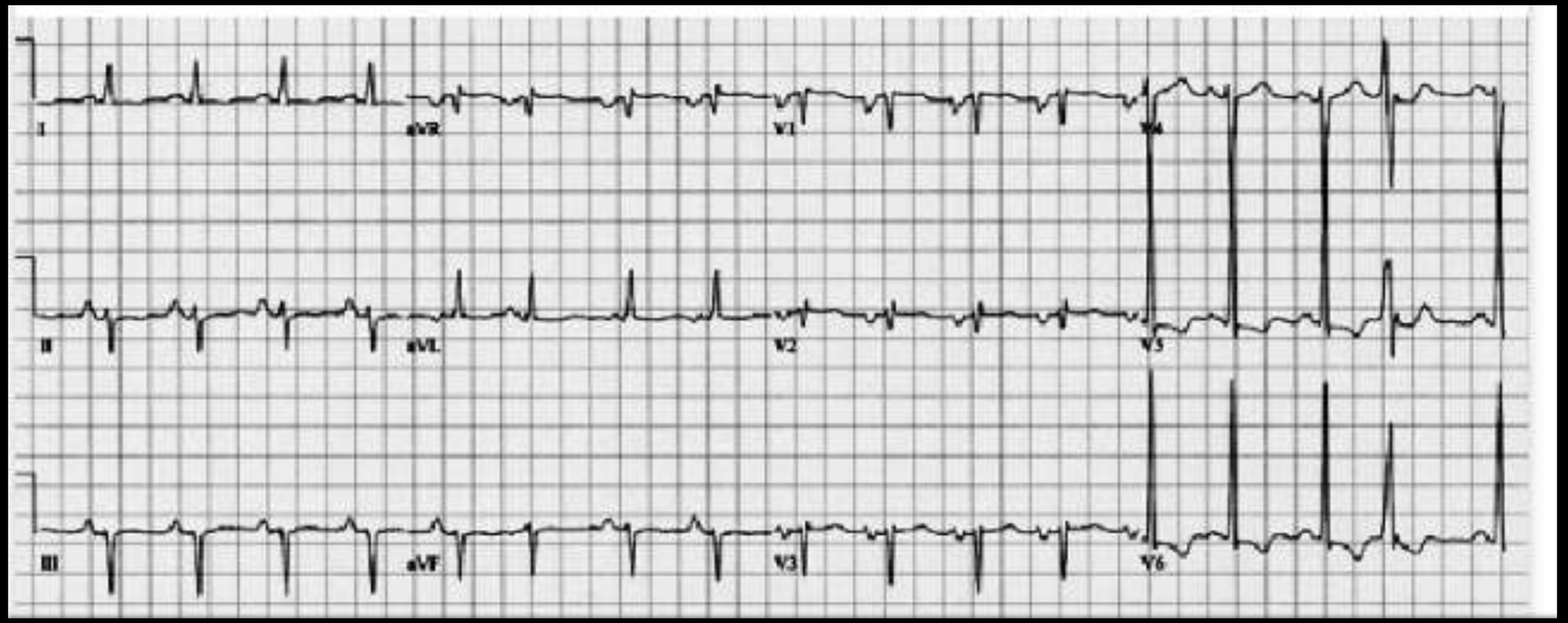
T=36.5 HR 98 BP=158/100

JVP=4cm

3/6 PSM apex to axilla

Bibasal creps

ECG



Investigations

Cr=145 ★

Troponin=908 ★

NT-ProBNP=3368 ★

Covid negative

CXR : cardiomegaly , APO ★

Case 1

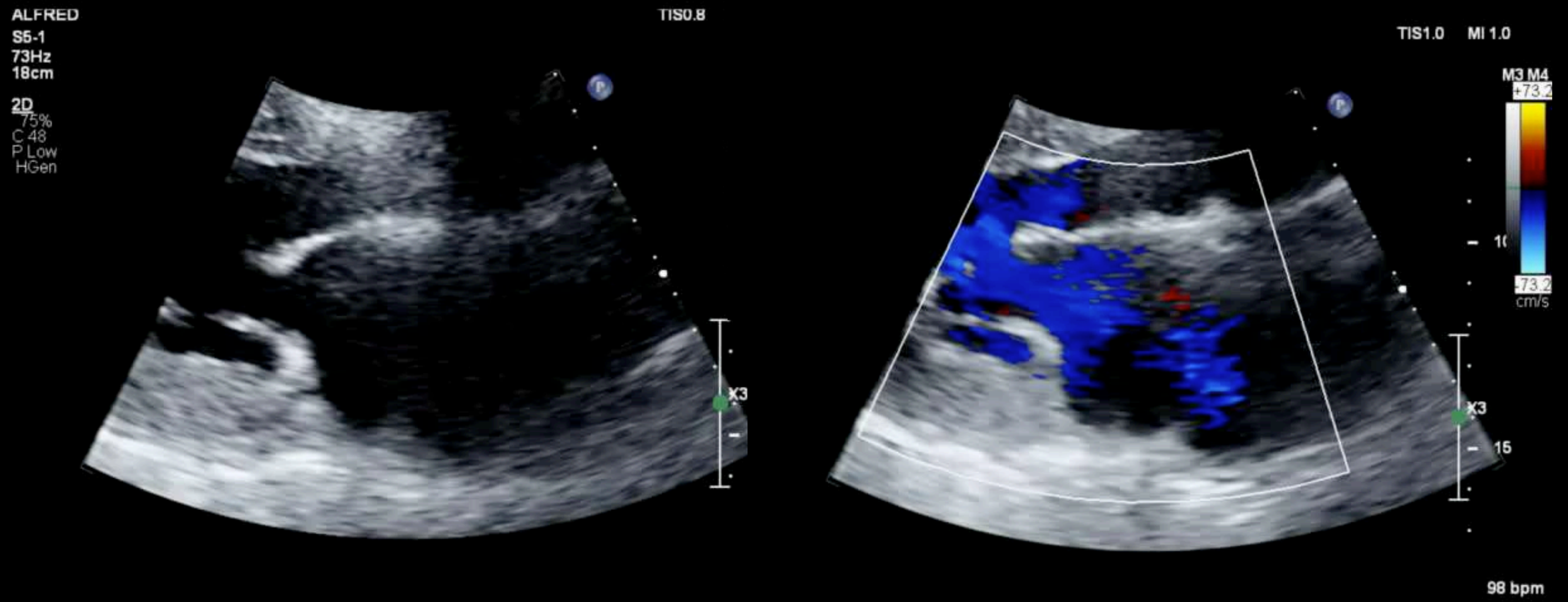
IVSd=2.0cm

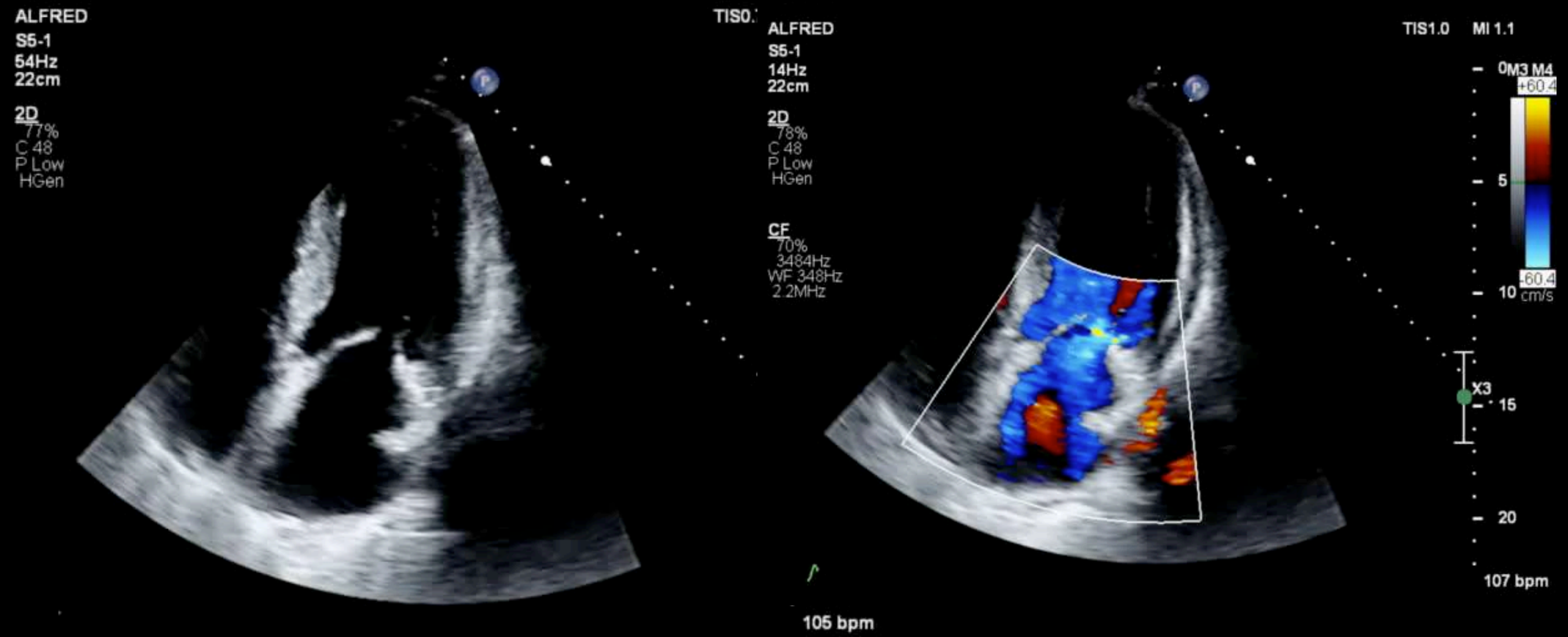
PWd=2.0cm

LVDd=4.8cm

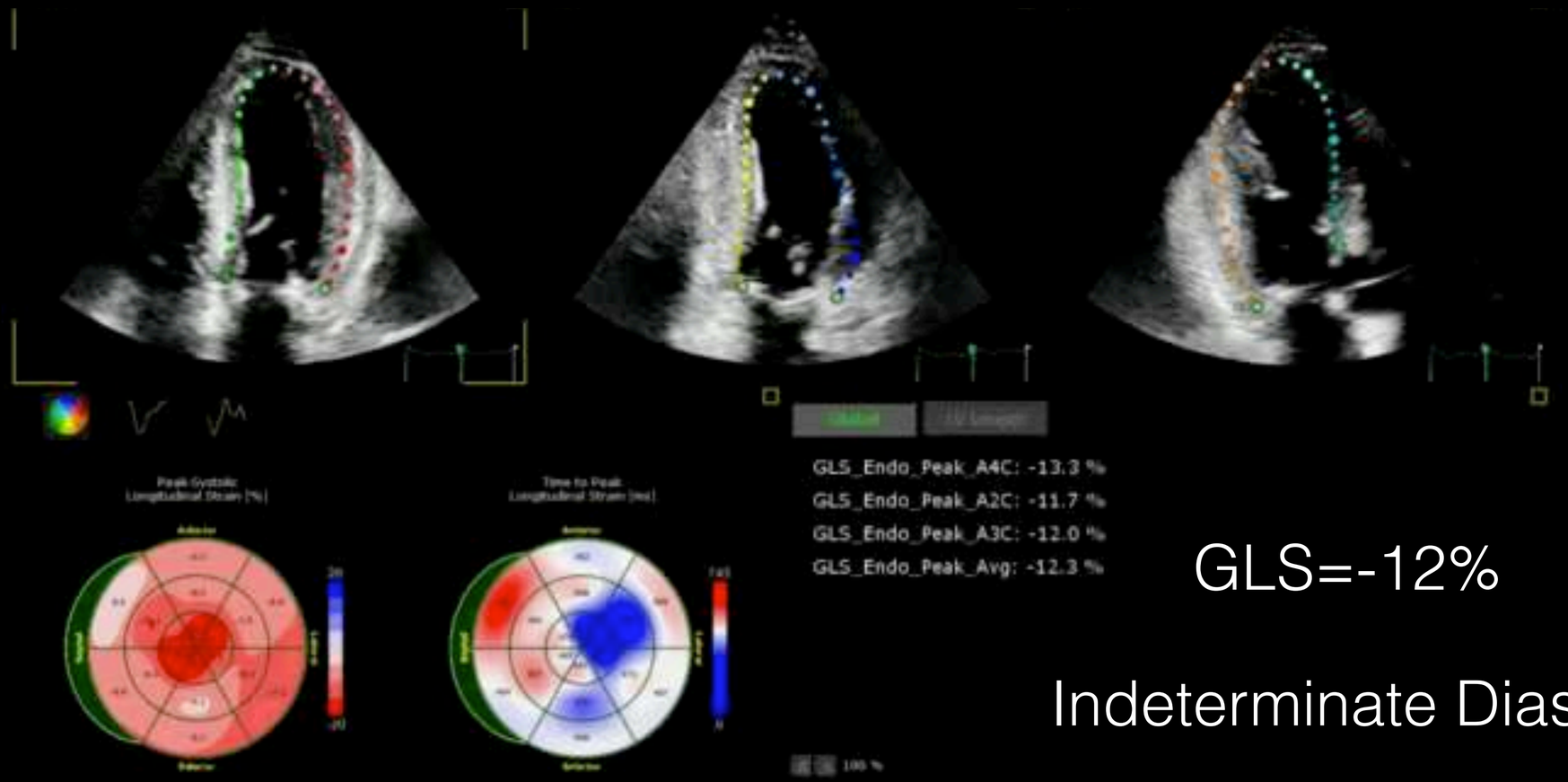
RWT=O.8

LVMi=265.8g/m_2


ALFRED
S5-1
73Hz
18cm
2D
75%
C 48
P Low
HGen
TIS0.8
X3
TIS1.0 MI 1.0
M3 M4
+73.2
-73.2
cm/s
X3
15
98 bpm


ALFRED
S5-1
54Hz
22cm
2D
77%
C 48
P Low
HGen
TIS0.
ALFRED
S5-1
14Hz
22cm
2D
78%
C 48
P Low
HGen
CF
70%
3484Hz
WF 348Hz
2.2MHz
105 bpm
TIS1.0
MI 1.1
0M3 M4
+60.4
5
-60.4
cm/s
10
X3
15
20
107 bpm

GLS



GLS=-12%

Indeterminate Diastology

TOE

Severe MR due to flail anterior leaflet and restricted posterior leaflet

Angiography

Normal

Questions

What is the cause of the increase in LV wall thickness?

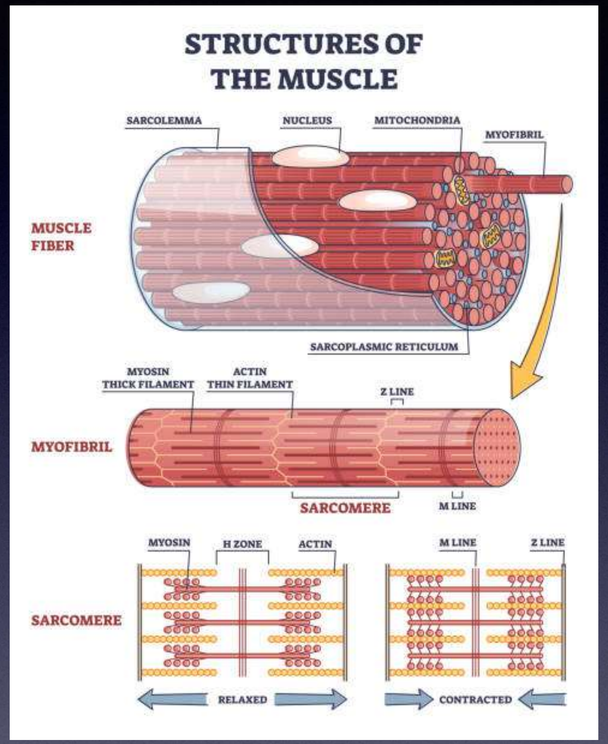
What is the cause of the mitral regurgitation?

What next?

Causes of Increased Left Ventricular Wall Thickness on Echo?

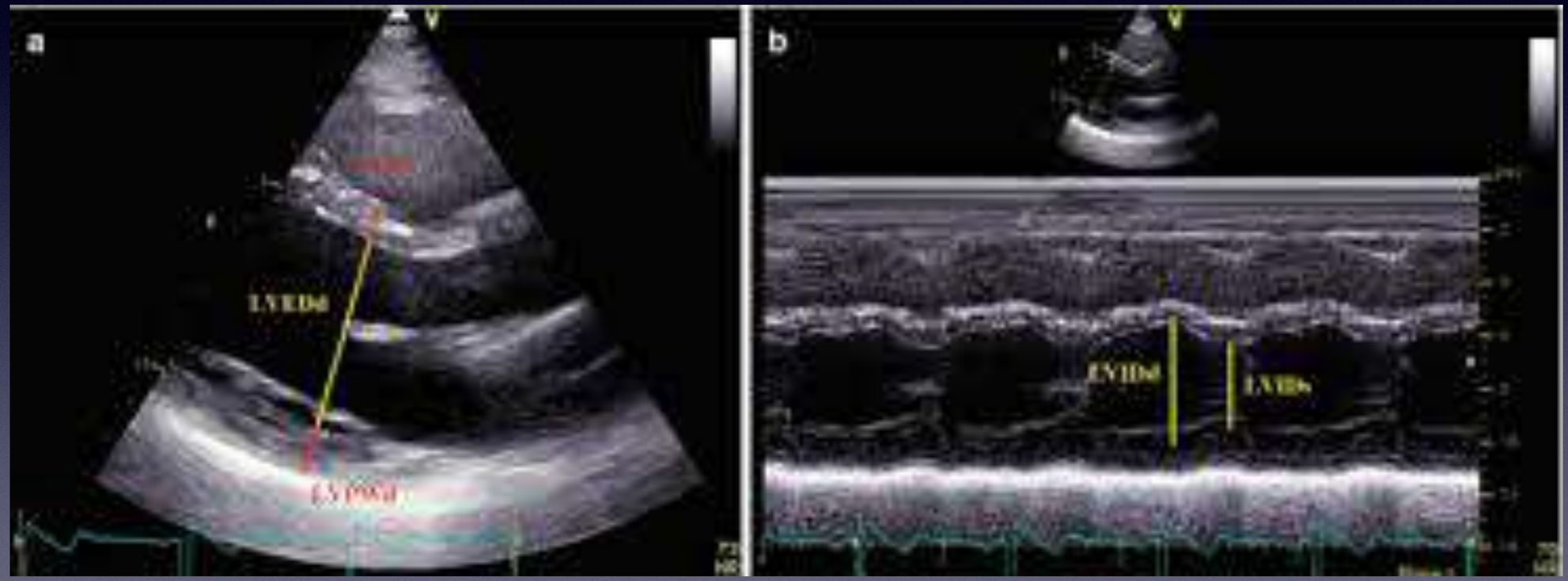
Most often due to left ventricular hypertrophy

LV Hypertrophy



Increase LVM due to increase in myocytes, sarcomeres and extracellular volume

Echo Assessment of LVH
Linear Methods



2D

M-mode

2D Linear Method

Biplane imaging assists with pependicularity

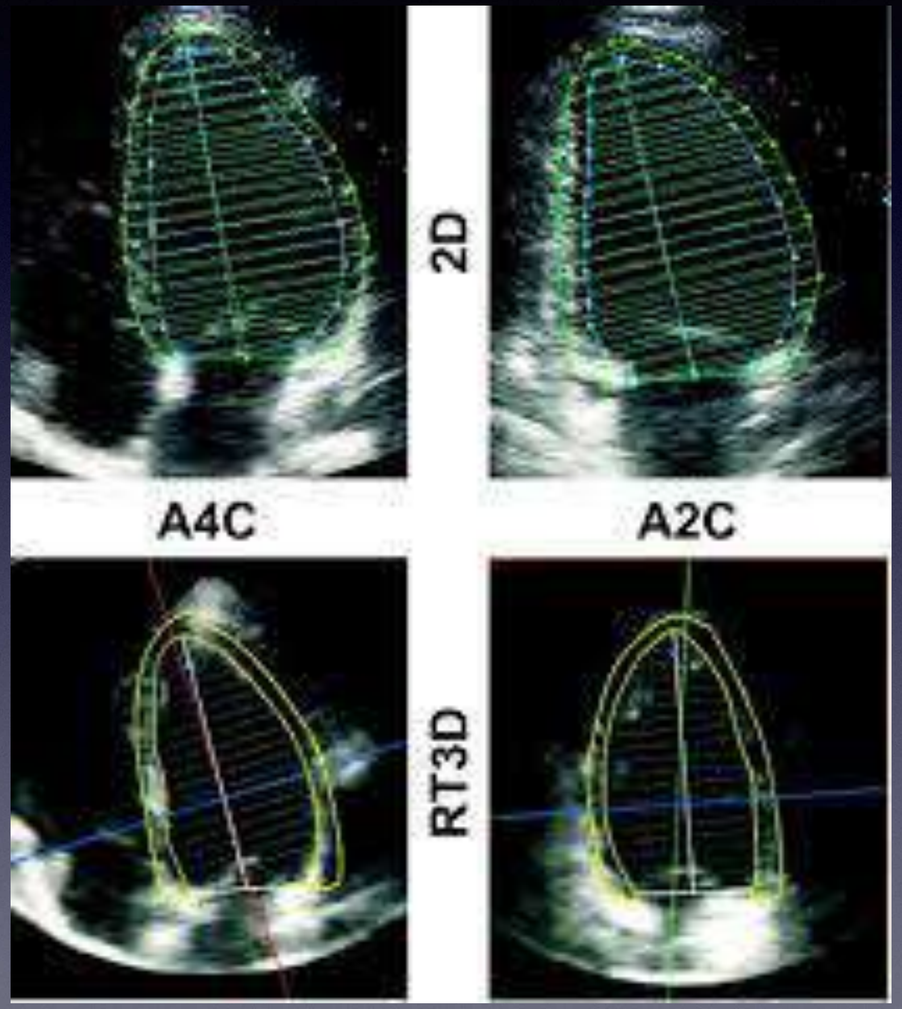
JASE Vol 32 issue 1 pages1-64 Jan 2019

Sigmoid Septum

Measurement is moved slightly apically beyond basal septal bulge.

JASE Vol 32 issue 1 pages1-64 Jan 2019

For the Non-Geometric Ventricle



ASE Formula for LVH

$$\text{LV mass (g)} = 0.8[1.04(\text{LVIDD} + \text{IVST} + \text{PWT})^3] - \text{LVIDD}^3) + 0.6$$

Where

LVIDD= left ventricular end diastolic dimension

IVST=interventricular septum thickness

PWT=posterior wall thickness

For the Geometric Ventricle

Definition of Abnormal LV mass

Male ≥ 115g/m2

Female ≥ 95g/m2

Recommendation for Cardiac Chamber Quantification by Echo in Adults.
An update from the ASE and European Association of CVI. Lang at al 2015

RWT

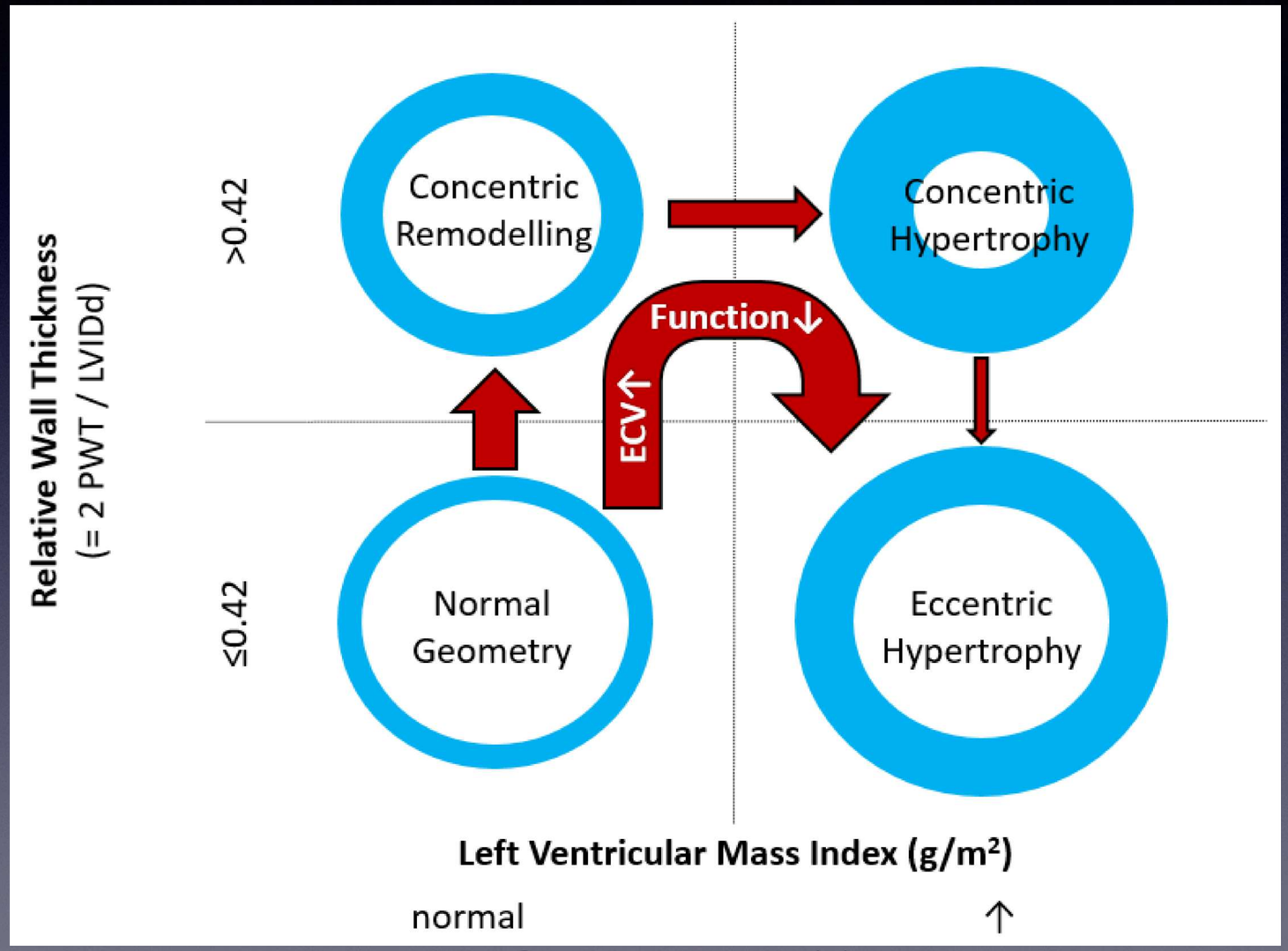
(IVSd+PWd)/LVEDd

or

(2PWd)/LVEDd

Where normal =<0.42

Classification of LV Geometry in HHD



Ismail TF J Clin Med 2023 12(9) 3122

Classification of LV Geometry in HHD

Normal	RWT≤0.42	LVM normal
LV remodelling	RWT>0.42	LVM normal
Concentric LVH	RWT>0.42	LVM increased
Eccentric LVH	RWT≤0.42	LVM increased

Types of LVH

Physiological: Pregnancy, Exercise training

Pathological: Pressure load: HTN, AS, Coarctation

Volume load: AR, MR

Genetic: HCM

Mimics of LVH: Amyloid , Fabry, Glycogen storage disorders

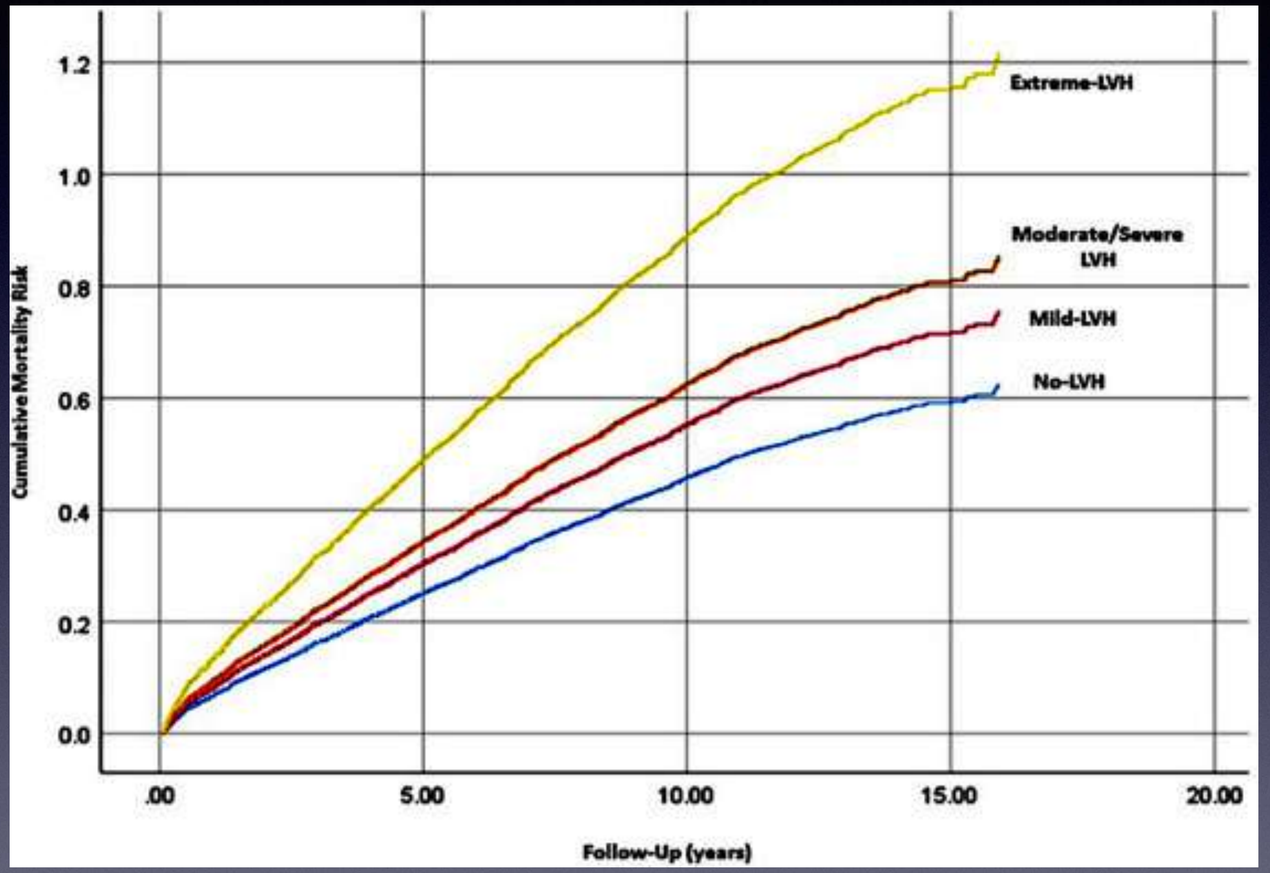
LVH due to HTN

LVH present in 15-20% of population

Prevalence of 19-48% of untreated hypertensives

Prevalence of 58-77% in high risk hypertensive patients

LVH & Mortality



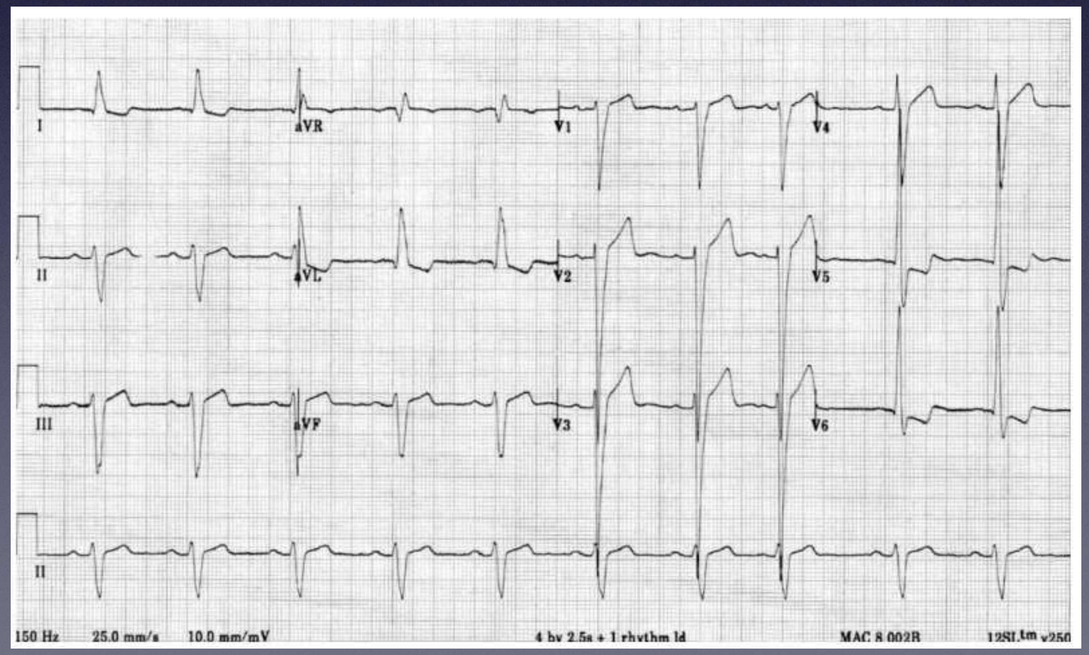
Papademetriou V et al . LVH & mortality risk in male veterans at high cardiovascular risk. Am J Cardiol 2020 125(6) 887

Clinical Features

Hx: Long standing Hx of HTN

ECG: High voltages & strain

Specific for LVH but not sensitive

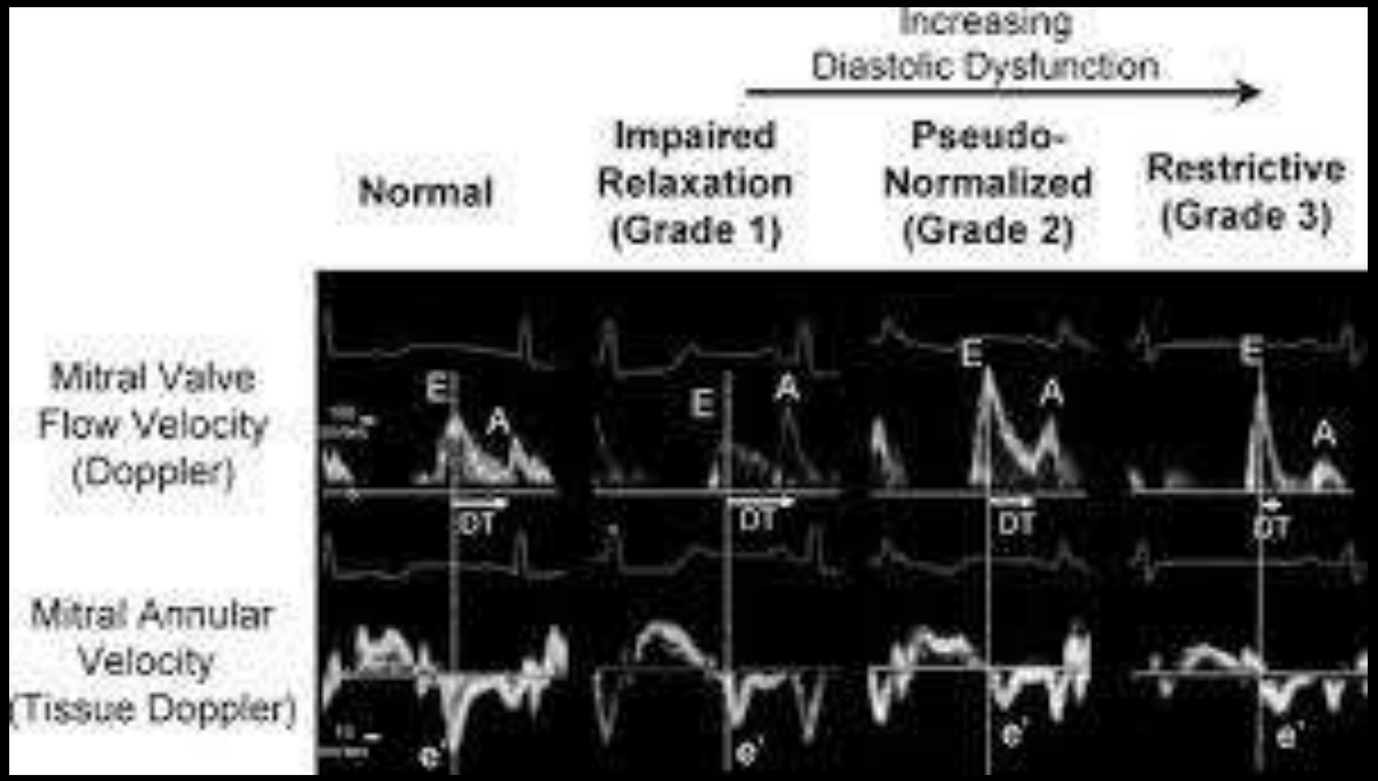


Echo Features in HHD

Wall thickness is usually ≤15mm (almost always <20mm)

LVH is usually concentric with no more than 2mm difference in wall thickness of different segments (with the exception of the basal septum in the elderly)

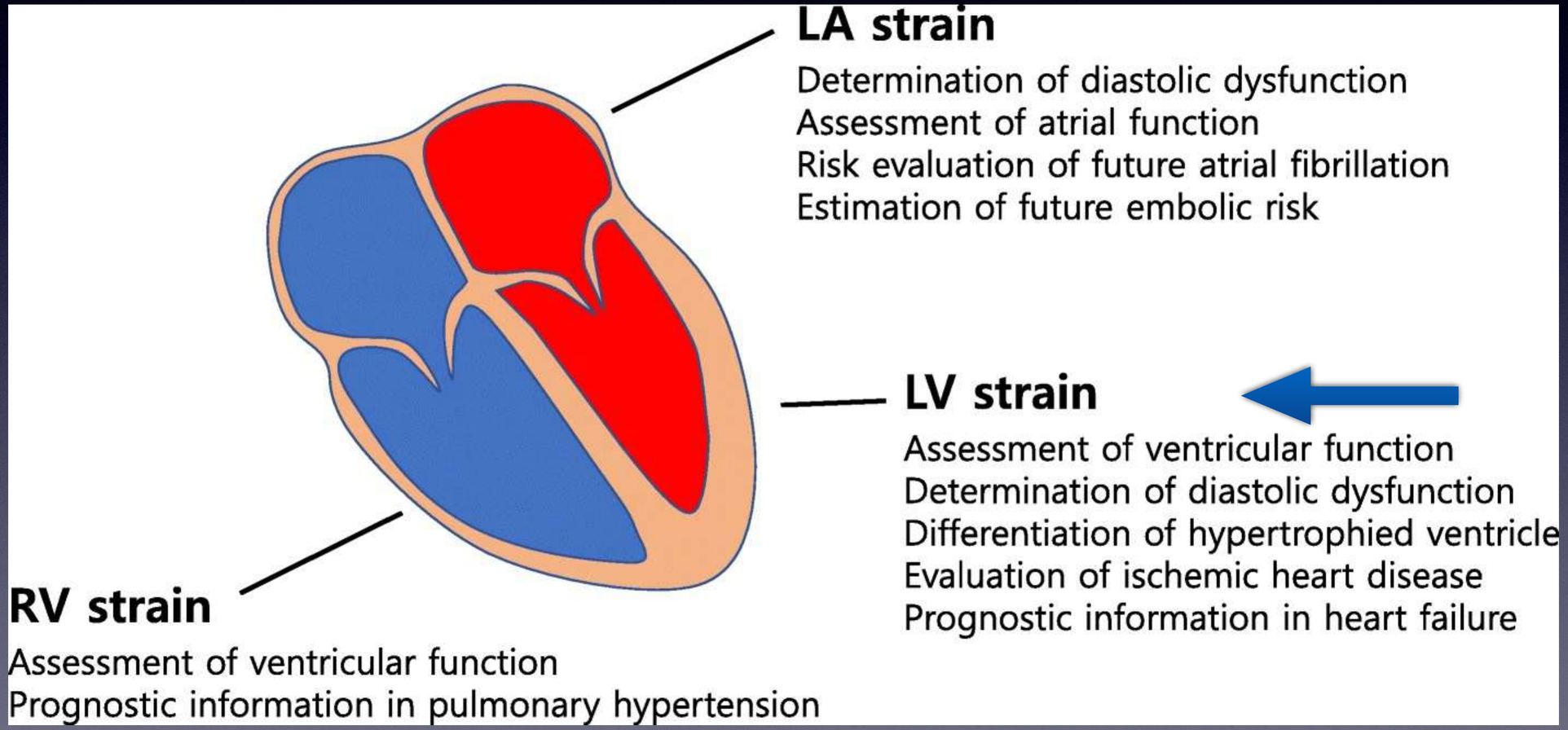
Diastolic Filling in HHD



+ Left atrial volume

+ PA systolic pressure

Role of Strain echo in HTN HD



Oh *JK and Park JH Role of Strain Echo in patients with hypertension. Clinical Hypertension Feb 2022*

Hypertensive Heart Disease

Clinical Suspicion

↓

ECG & Echo Findings

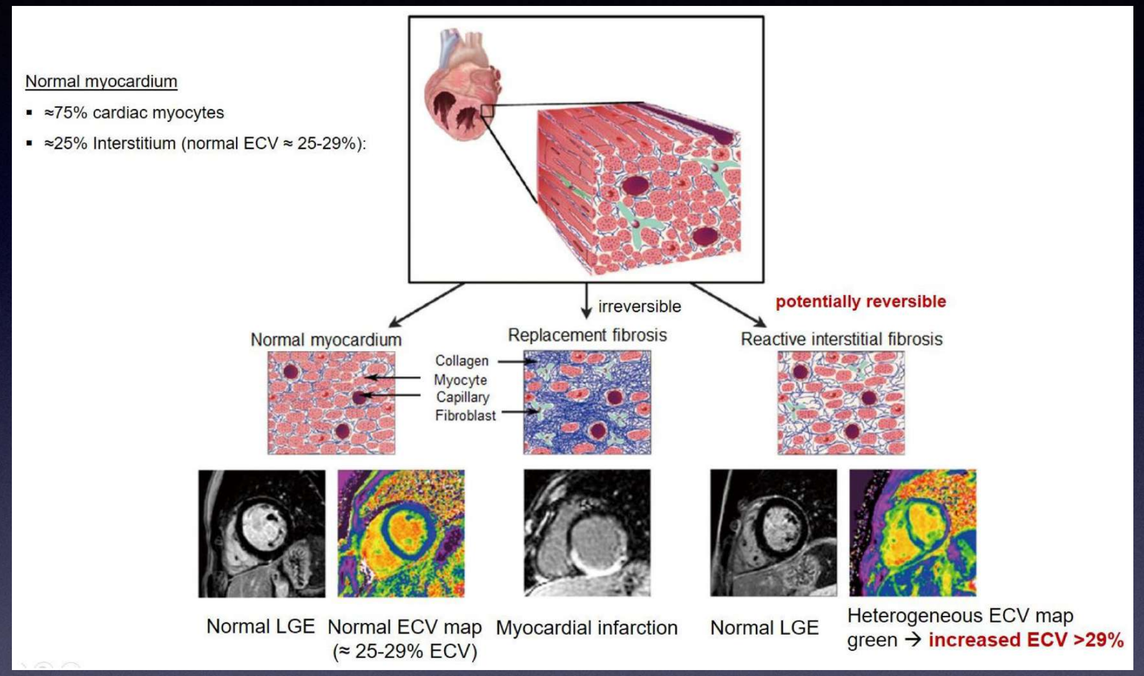
MRI usually not required

Increased LV wall thickness

Diffuse LGE in nonspecific pattern in up to 50% of cases

Normal or mildly increased ECV

MRI: Tissue characterisation with LGE and ECV in HHD



Diffuse LGE in non specific pattern in up to 50% of cases of LVH due to hypertension

Normal T1 and ECV normal or mildly increased

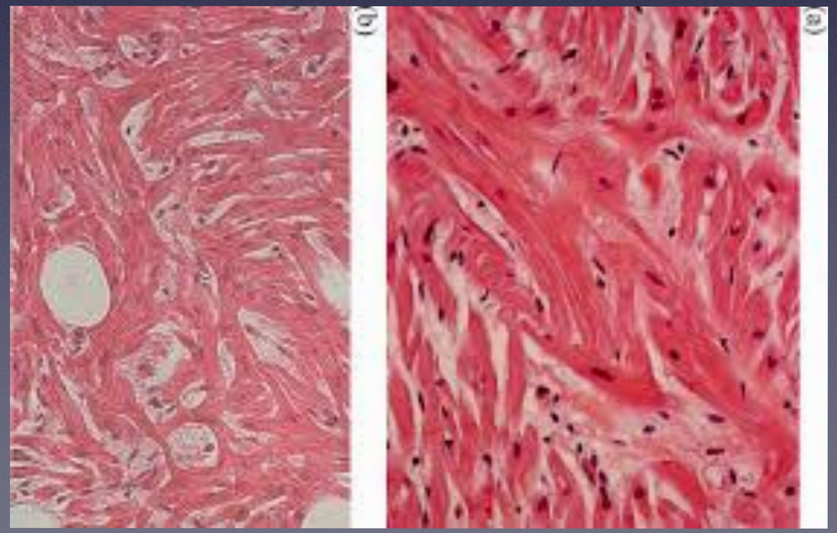
Ismail TF J Clin Med 2023 12(9) 3122

HCM

LVH in the absence of another explanation

1. in the presence of an identified genetic mutation causing a sarcomere (or sarcomere-related) variant

2. or when genetic etiology remains unresolved (40% of cases)

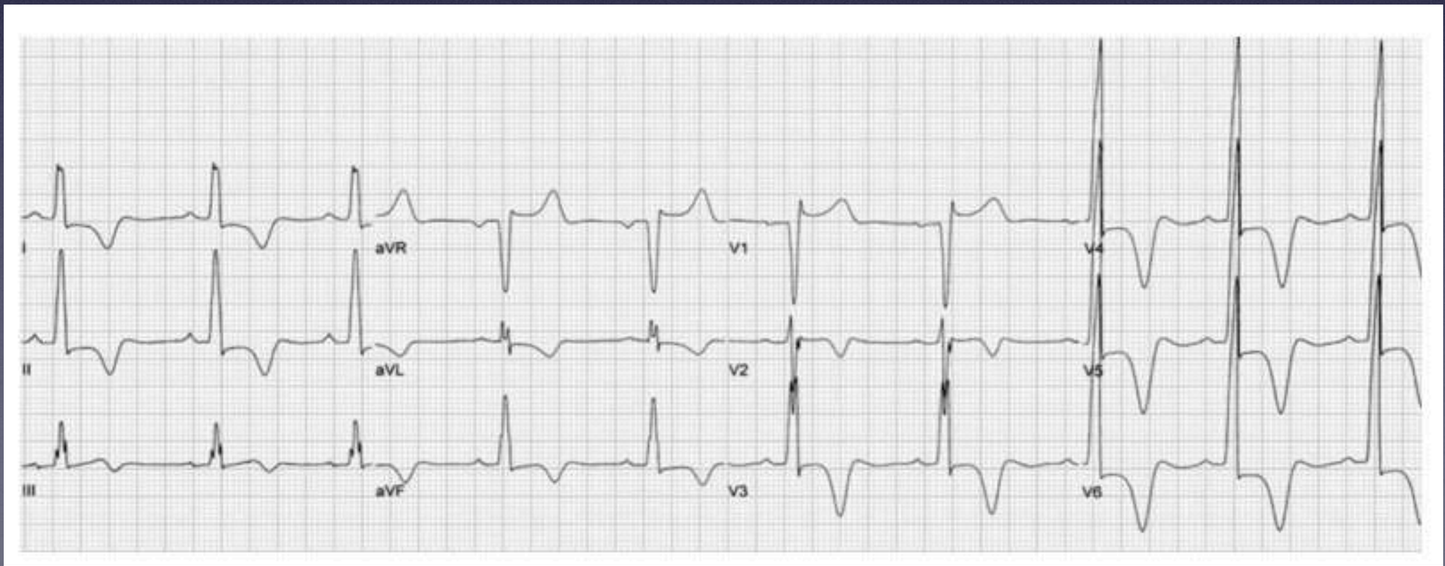


Clinical Features

Hx: Sx of SOB, CP, syncope, palpitations

Positive Family Hx,

ECG: High voltages, Negative T waves in apical variant



Echo based Diagnostic Criteria for Adults

Unexplained hypertrophy of non-dilated left ventricle

LVSD≥15mm

LVSD≥13mm (if family Hx)

Usually widespread but 10% have hypertrophy in only one or two segments

Features of LVH in HCM

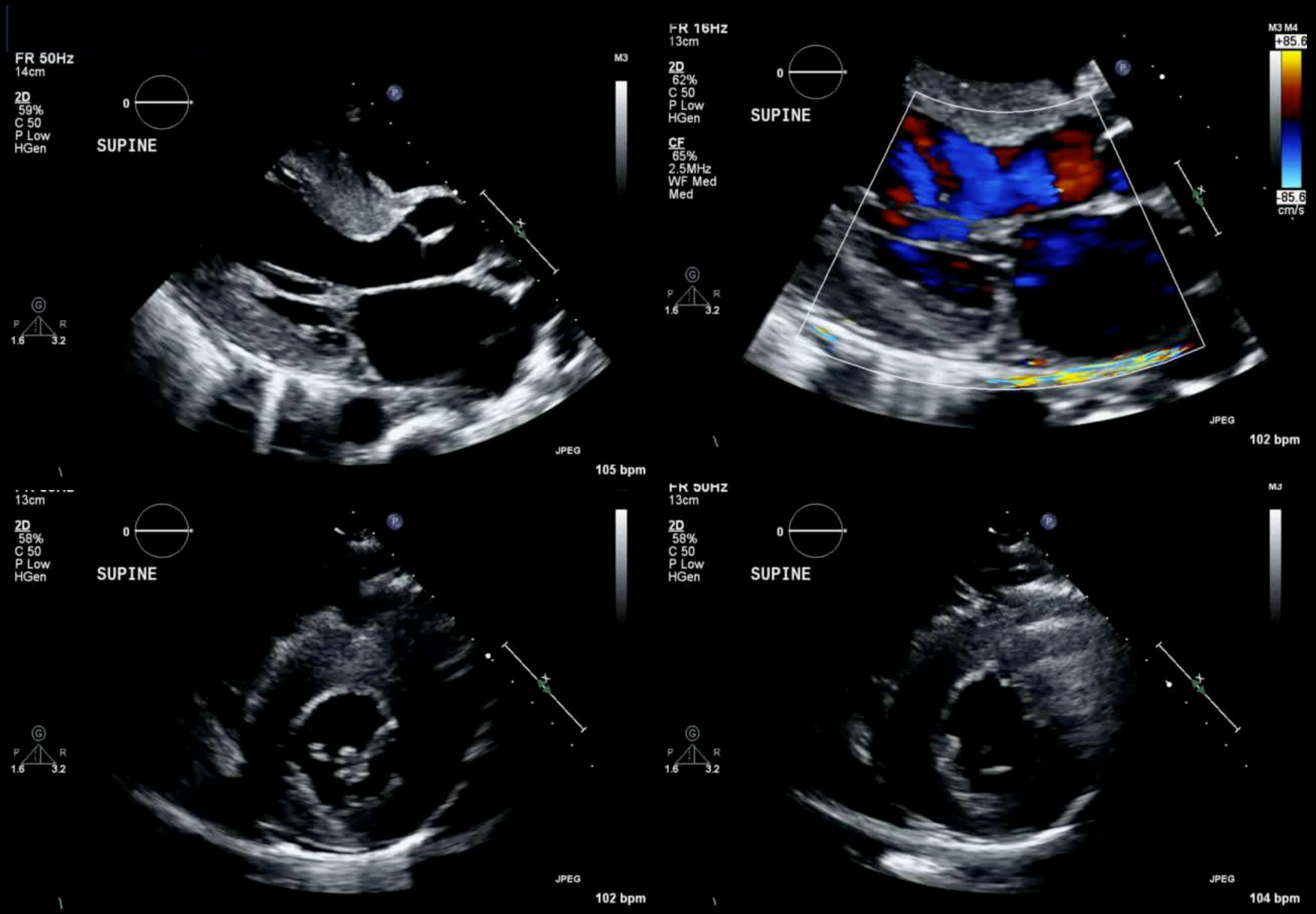
Distribution of LVH: Often asymmetric but not always

Other Features

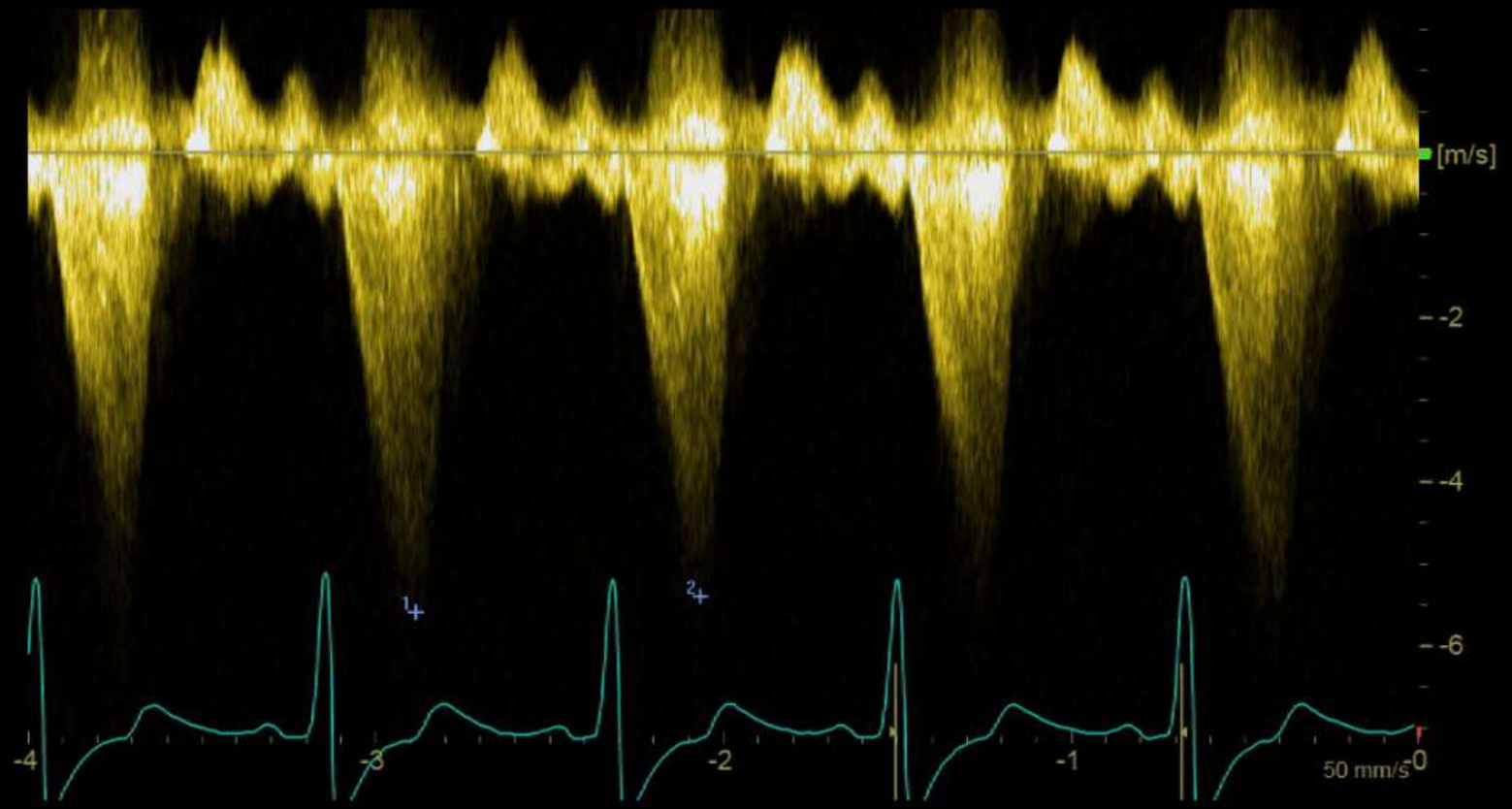
LVOT

Abnormalities of mitral valve

HCM


FR 50Hz
14cm
2D
59%
C 50
P Low
HGen
SUPINE
M3
JPEG
105 bpm
FR 16Hz
13cm
2D
62%
C 50
P Low
HGen
CF
65%
2.5MHz
WF Med
Med
SUPINE
M3 M4
+85.6
-85.6
cm/s
JPEG
102 bpm
13cm
2D
58%
C 50
P Low
HGen
SUPINE
JPEG
102 bpm
FR 50Hz
13cm
2D
58%
C 50
P Low
HGen
SUPINE
M3
JPEG
104 bpm

LVOT gradient



Role of CMR

For confirmation of diagnosis

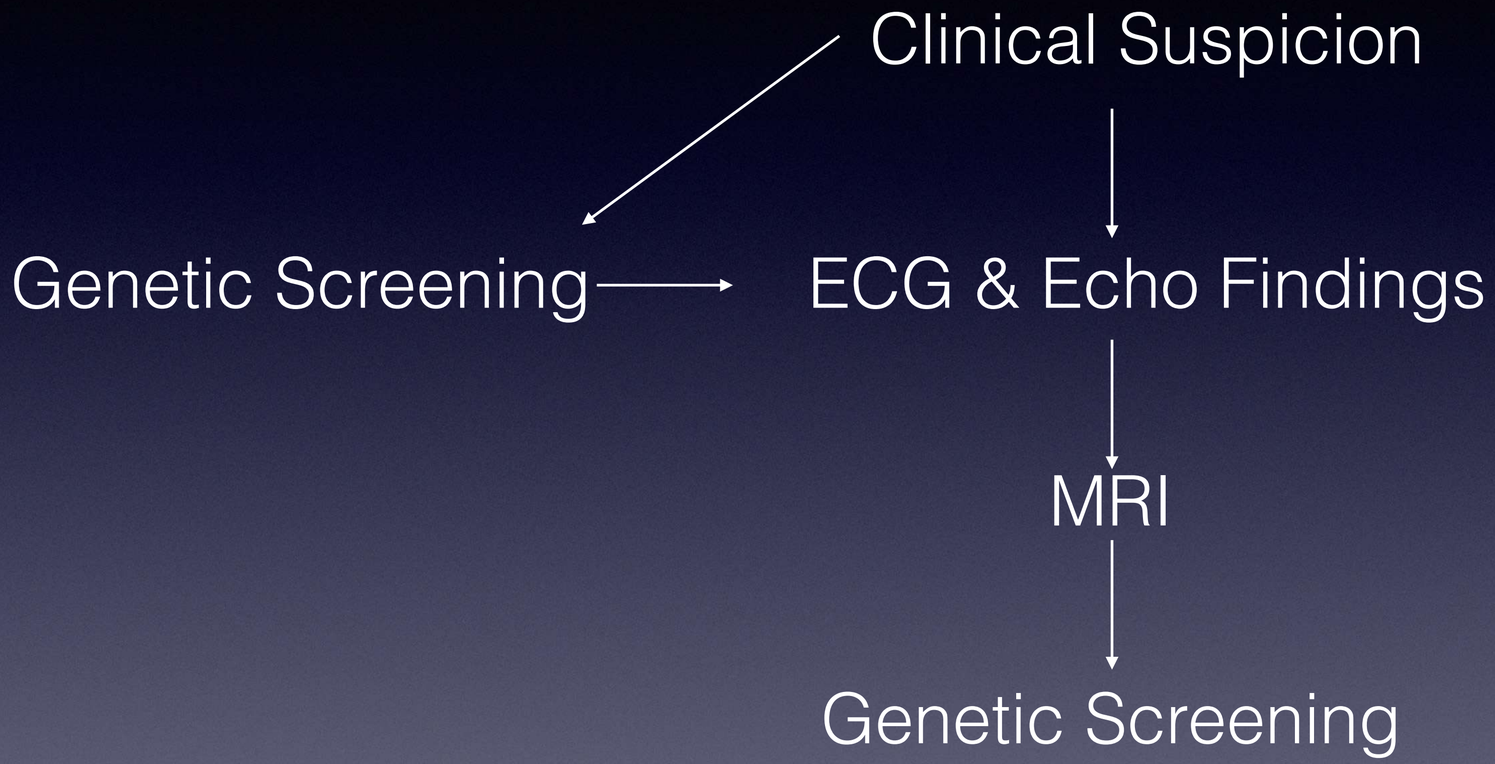
For risk factor stratification.

a. Degree and distribution of hypertrophy.

b. Scar with late gadolinium enhancement

c. Apical aneurysm

HCM


Clinical Suspicion
Genetic Screening
ECG & Echo Findings
MRI
Genetic Screening

Mimics of LVH:Thick LV Walls

Infiltration: Amyloid, Fabry, Danons,

Oedema: Rejection in OHTx, Myocarditis, Sarcoid

Cardiac Amyloid

Abnormality of folding of amyloid protein so the protein forms insoluble beta pleating which forms rigid fibrils

These fibrils deposit in extracellular tissue of the heart and other organs

Cardiac Amyloid

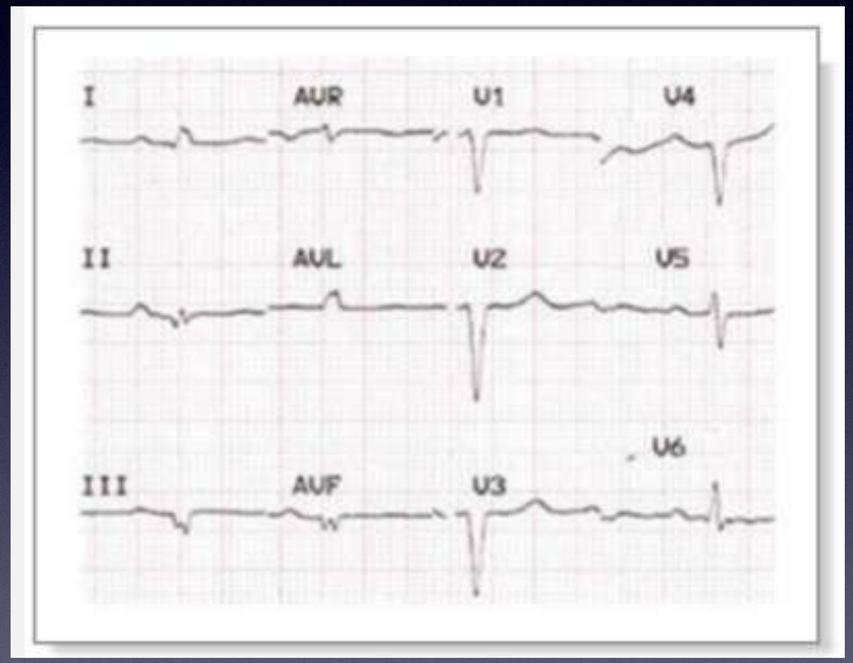
ALL Amyloid: Plasma cell dyscrasia- Abnormal light chains associated with immunoglobulin

Familial Amyloid: Liver produces abnormal transthyretin protein

Wild type Amyloid: Liver produces abnormal transthyretin protein

Cardiac Amyloid

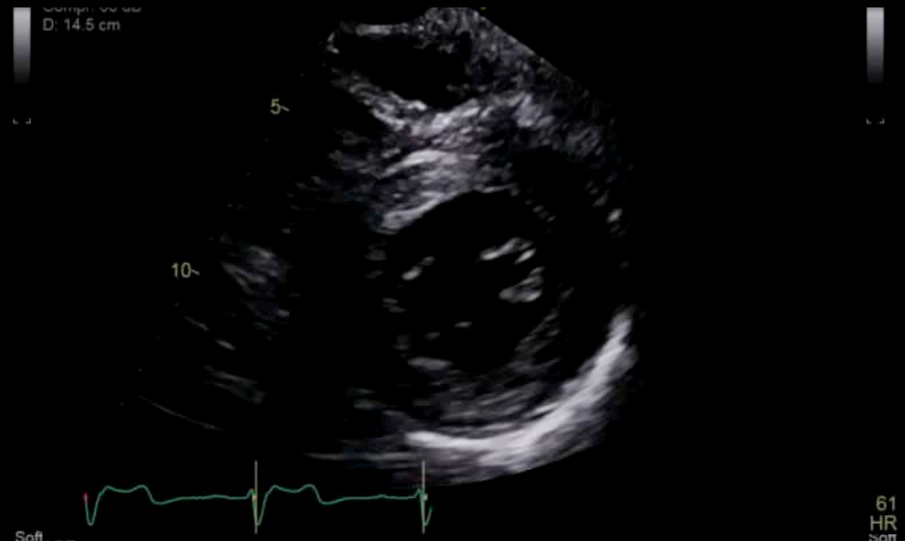
ECG

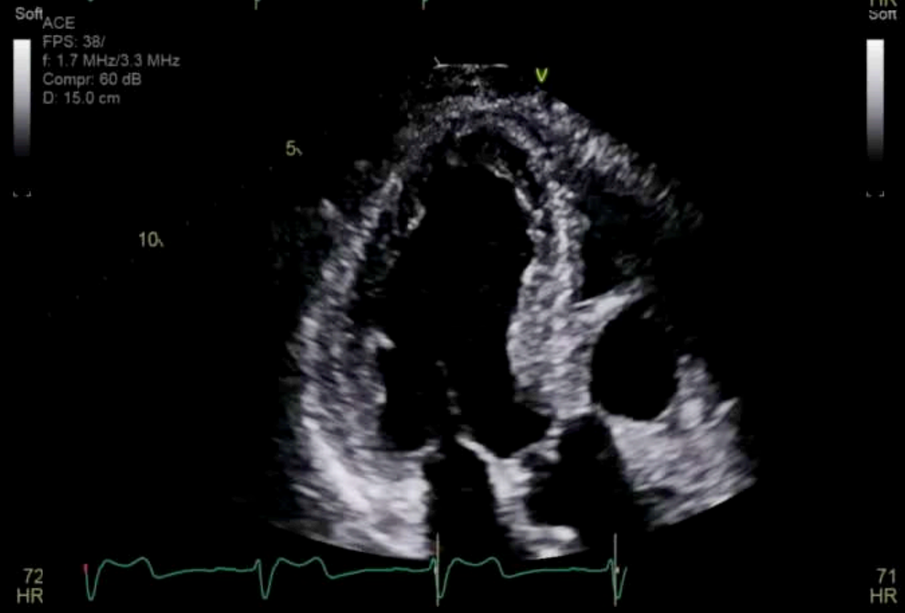


BNP elevated

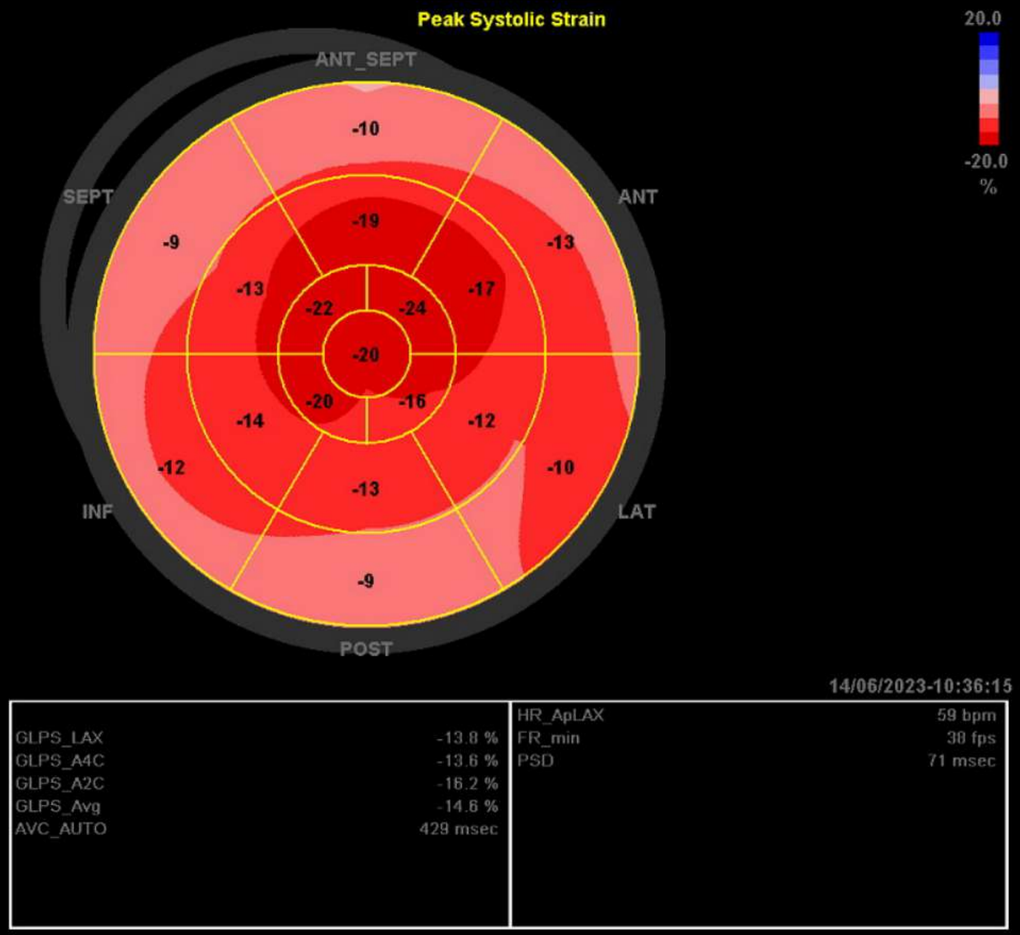
Troponin elevated

Cardiac Amyloid





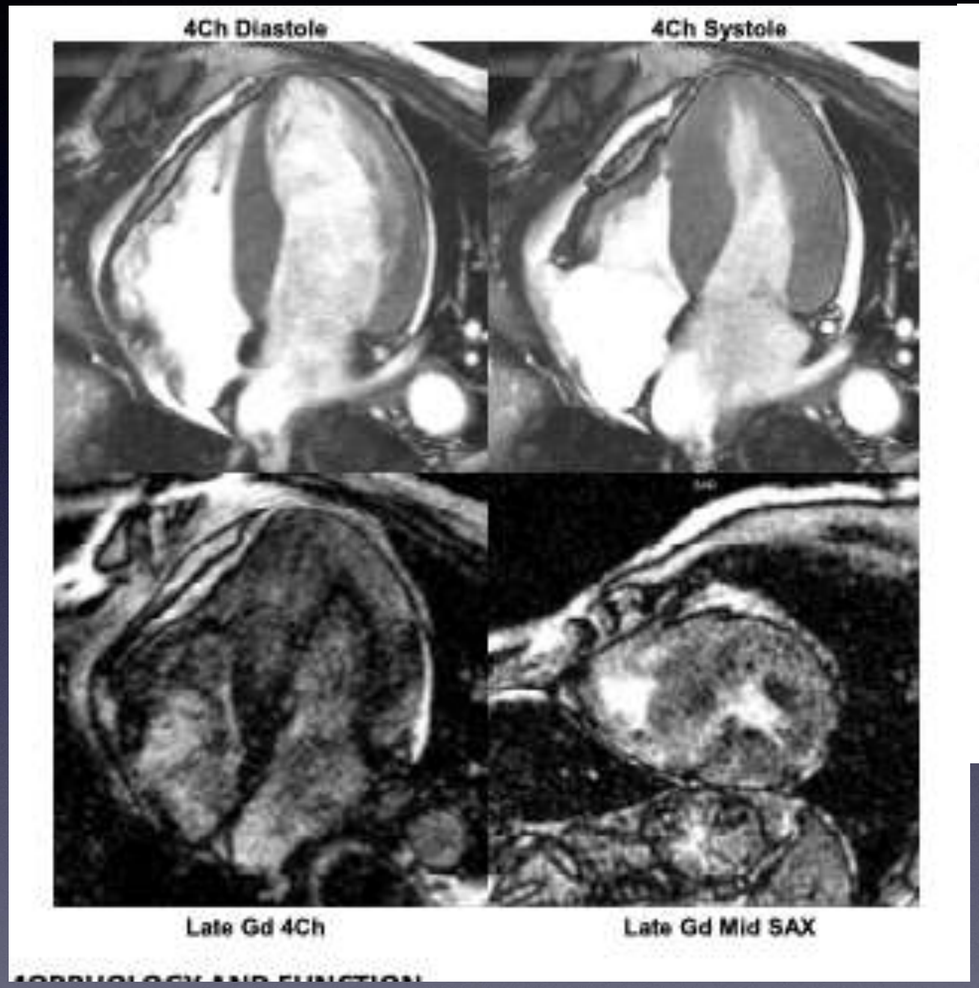
GLS in Cardiac Amyloid



Decreased

Apical Sparing

MRI in Cardiac Amyloid



LV MORPHOLOGY AND FUNCTION

Left ventricle mildly dilated with markedly elevated mass in the setting of moderate to severe LV wall thickening, measuring 22mm at maximal point (septum). Preserved systolic function (LVEF 62%). Right ventricle mildly dilated with normal systolic function. Normal global STIR ratio (no evidence of myocardial oedema). Prolonged native T1 time (1334ms) consistent with amyloidosis. Small circumferential pericardial effusion.

VALVES AND ATRIA

Aortic valve leaflets not well seen but appear restricted with flow acceleration in the aorta, suggestive of aortic stenosis. Mitral and tricuspid valves appear structurally normal. Tricuspid regurgitation seen but not quantified (appears mild). Mild left and moderate right atrial enlargement. Thickened interatrial septum, most consistent with lipomatous hypertrophy.

POST CONTRAST IMAGING

Myocardium is nulled at short T1 inversion time with diffuse myocardial enhancement consistent with infiltration. Diffuse patchy late gadolinium enhancement (LGE).

CONCLUSION

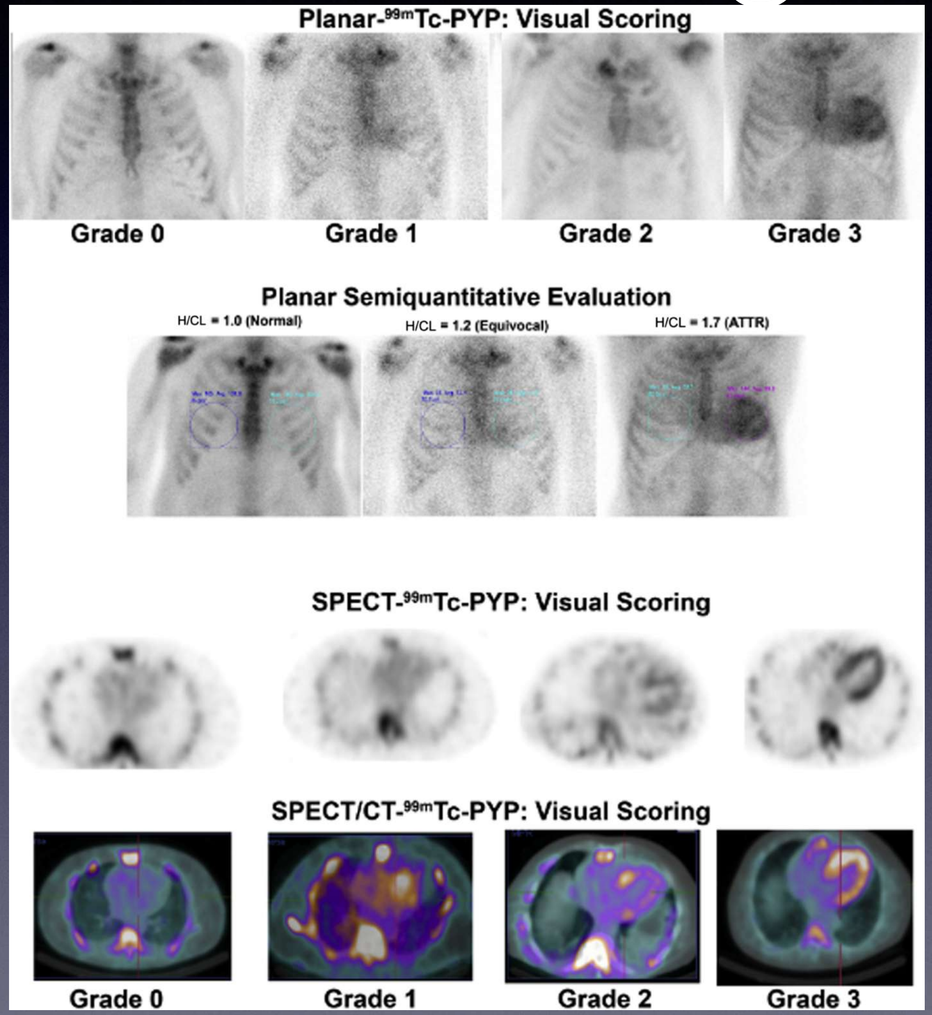
CMR features consistent with a diagnosis of advanced cardiac amyloidosis.

Marked increase in LVM

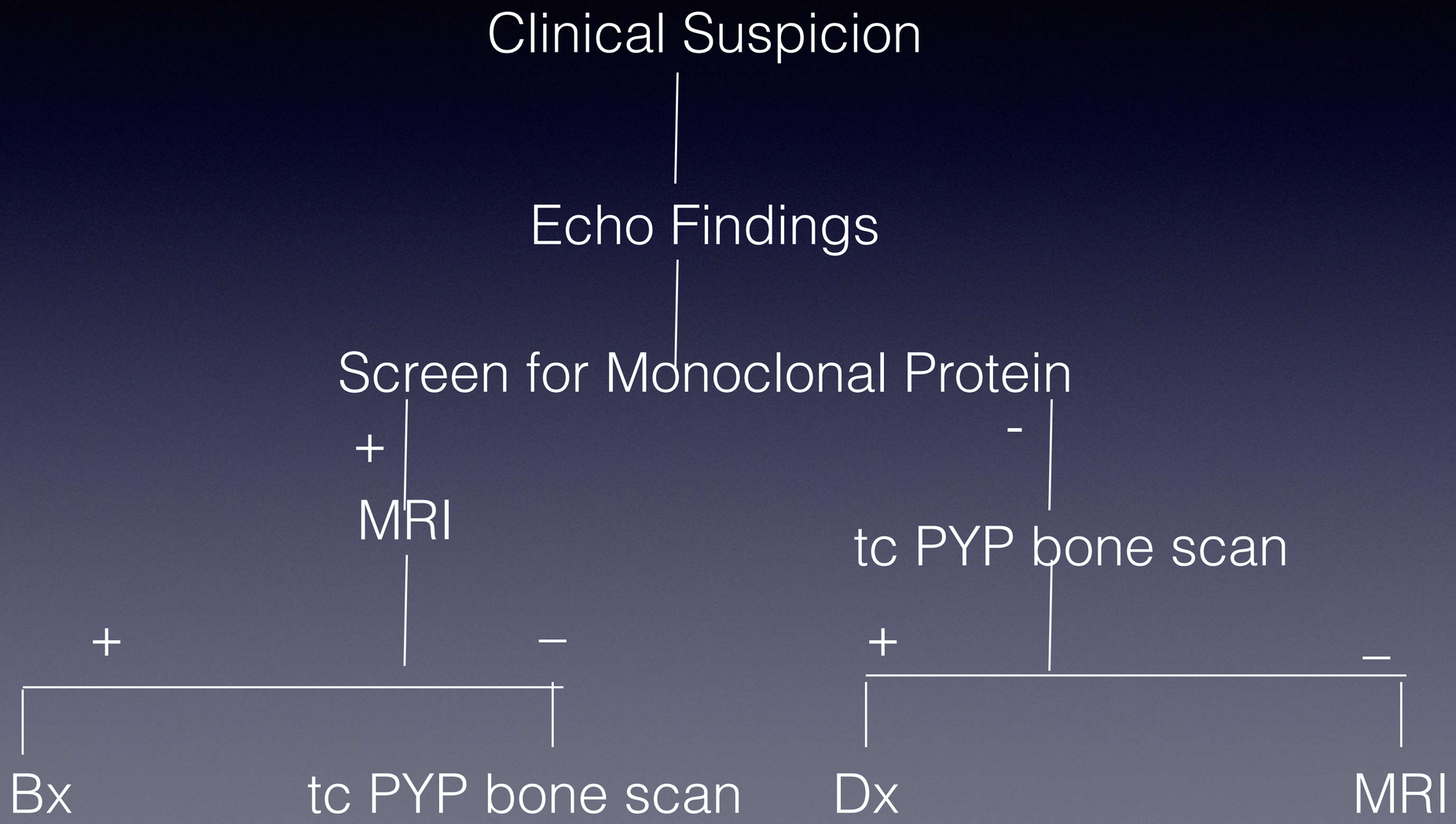
Diffuse patchy gadolinium enhancement

Myocardium nulled at short T1 inversion time c/w infiltration

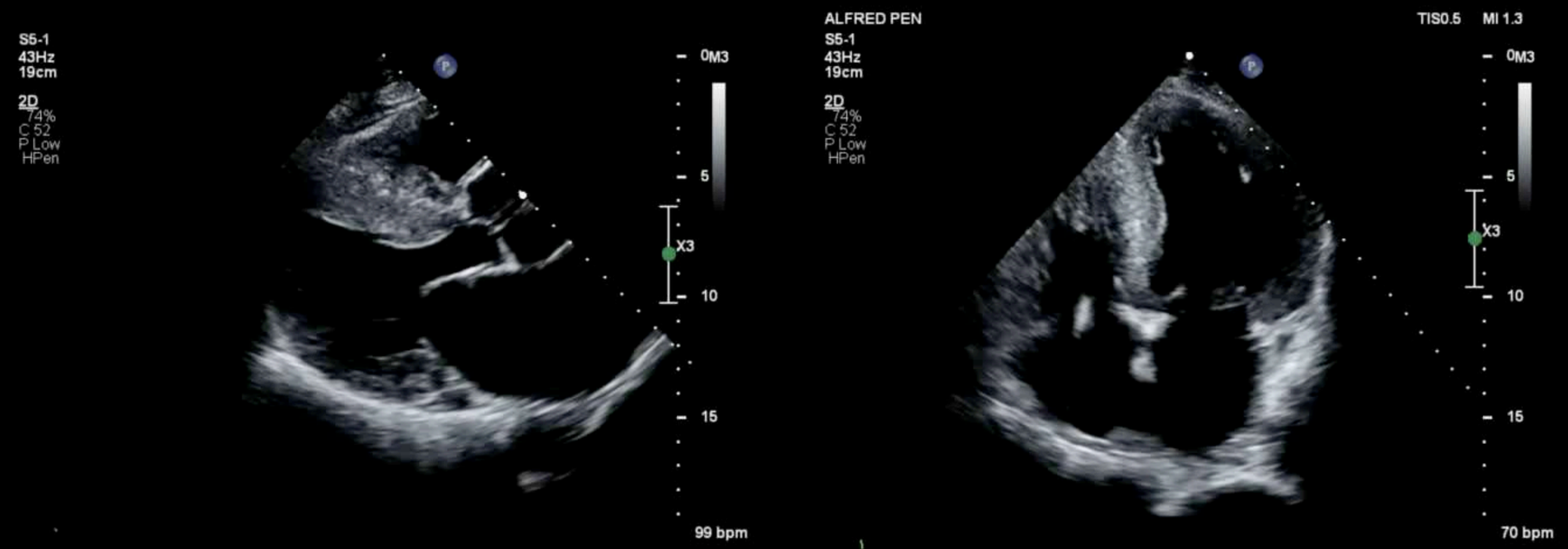
Tc PYP Imaging



Cardiac Amyloidosis

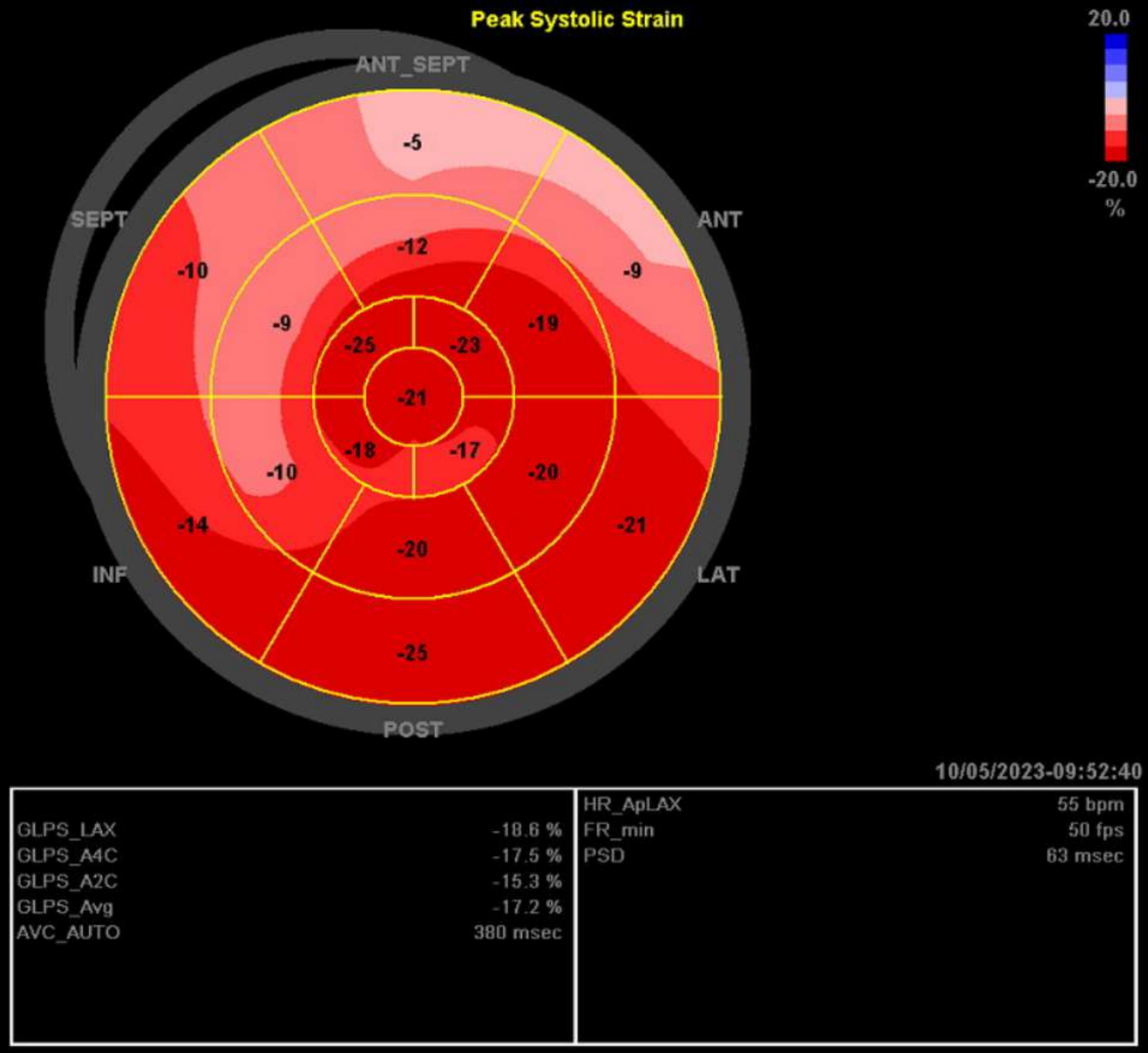


Fabry Disease



X-linked lysosomal storage disease
Deficiency in Alpha Galactosidase A

TREATABLE



Peripheral neuropathy

Cerebrovascular Accident

Cornea Verticillata

Renal failure

Angiokeratoma

Anhidrosis

Back to our case

Why the increase in LV wall thickness?

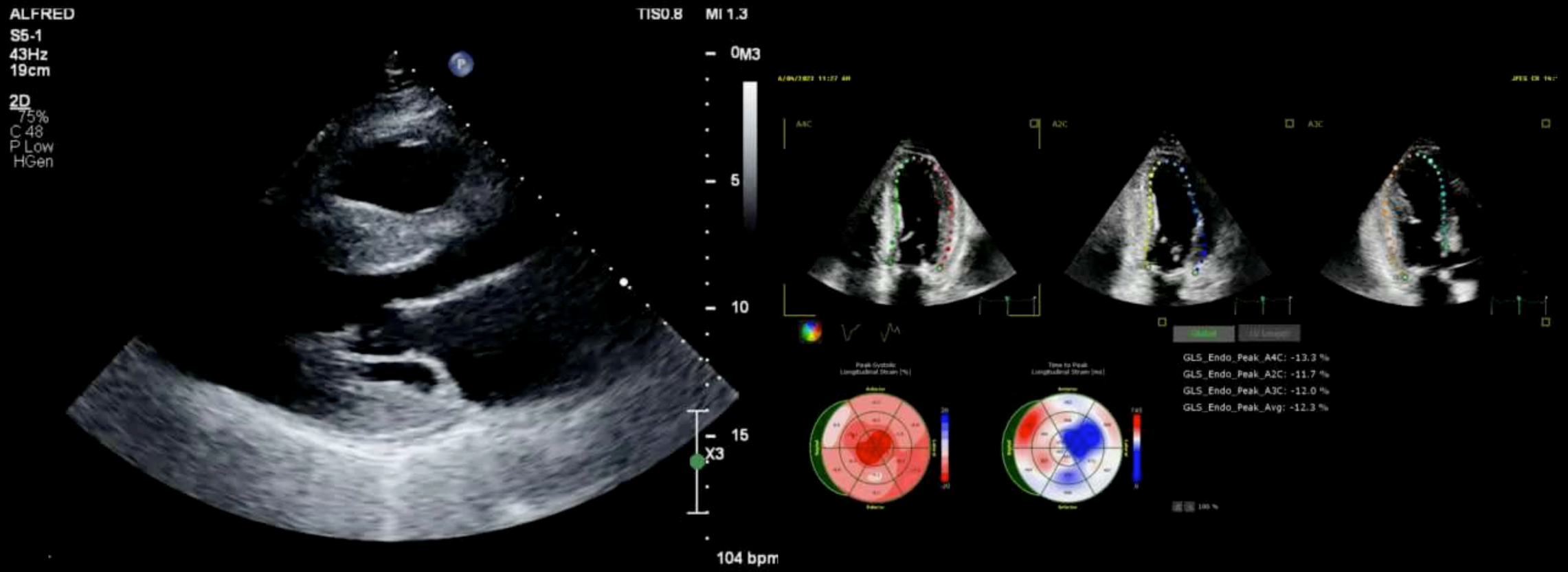
Past Hx of Dx of HTN in 2018 but did not commence treatment

Significant HTN in hospital

ECG: LVH on voltage criteria

?HHD

Echo



Severe LVH (? out of range for HHD)

Apical sparing of GLS

MRI

MEASUREMENTS

LEFT VENTRICLE SAX STACKS

LVEDV	237	ml
LVEDV Index	109	ml/m^2
LVESV	123	ml
LV Stroke Vol.	114	ml
LV EF	48	%
LV Mass	298	gm
LV Mass Index	137	gm/m^2

Normal LV Values

	Men	Women
Volume	156+/-21	128+/-21
Vol/BSA	80+/-9	75+/-9
Mass	146+/-20	108+/-18
Mass/BSA	74+/-9	63+/-8

RIGHT VENTRICLE

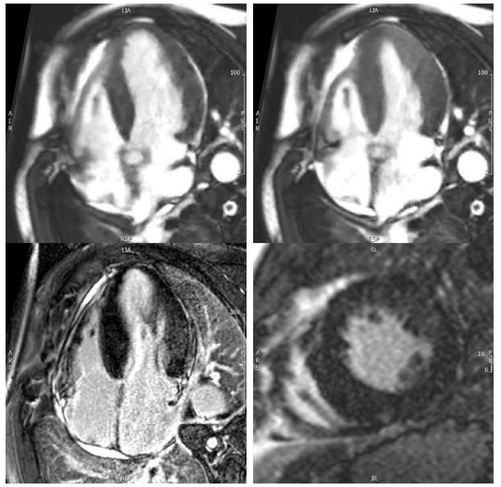
RV Dimension		mm
RVEDV Index		
RVESV Index		
RV Stroke Vol		
RVEF		%

ATRIA

LA Area		cm^2
LAESVi	31	ml/m^2
RA Area	22	cm^2

MYOCARDIAL OEDEMA & RELATIVE ENHANCE.

STIR Average	2.20	(N < 2.2)
STIR Regional		
ERE Average		(N < 4.0)
Abs Myo enh		(N < 45%)



Alfred Radiology & Heart Centre Report

LV MORPHOLOGY AND FUNCTION

Left ventricle normal size with mild systolic dysfunction (LVEF 48%). Severe concentric hypertrophy (up to 19mm). Hypokinesis of the basal to mid inferior and inferolateral walls. Right ventricle normal size with mild to moderate systolic dysfunction. Consolidation noted in the right upper lobe, right lower lobe and a smaller amount in left upper lobe.

VALVES AND ATRIA

Both atria normal size. Mitral regurgitation noted but not formally quantified - appears severe and posteriorly directed. The appearances are consistent with the known flail anterior leaflet and restricted posterior leaflet.

POST CONTRAST IMAGING

The myocardium is well nulled at TI 220. There is non-specific patchy myocardial late gadolinium enhancement at the inferior RV septal insertion point with some impression of more diffuse hazy LGE in the mid lateral wall.

CONCLUSION

Severe concentric LV hypertrophy with mild overall LV systolic dysfunction. Mild to moderate RV systolic dysfunction. Severe mitral regurgitation, consistent with known flail anterior leaflet on TOE. Some non-specific hazy late gadolinium enhancement. Appearances consistent with severe hypertensive heart disease (note history of amphetamines).

Severe LVH
Mild LV dysfunction
Hazy non-specific LGE
C/W HHD

Other Ix

Angiogram NAD

-ve PEP

-ve urine for light chains

Normal levels of Alpha Galactosidase

Negative screen for secondary causes of HTN

Surgery

Ruptured chords to anterior leaflet

Restriction of posterior leaflet and thickening of subvalvular apparatus.

MV repair with ring and artificial chords

Summary

LVH due to HTN

Severe MR? Role for metamphetamine affecting posterior mitral leaflet and subvalvular apparatus

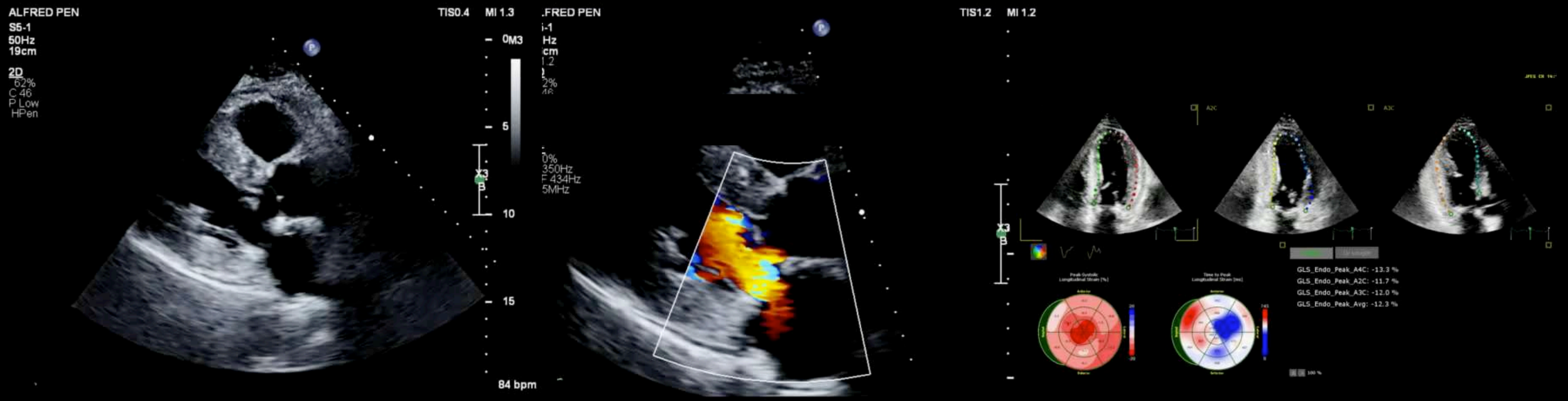
Mx

Sx: MV repair

Lifestyle changes

Tight BP control

18 months later



IVS=17mm

PW=16mm

LVEDD=17mm

LVMi=268.5g/m_2

Conclusion

Spectrum of diseases causing a thick left ventricle

Appropriate weighting of clinical data, ECG and Echo vital

Role of Cardiac MRI when LVH is disproportionate or asymmetric for tissue characterisation

Biopsy, enzyme testing and genetics may be required if infiltration or HCM is suspected.