CARDIO X CLINIC

MAYO CLINIC

MULTIMODAL IMAGING IN HYPERTROPHIC CARDIOMYOPATHY

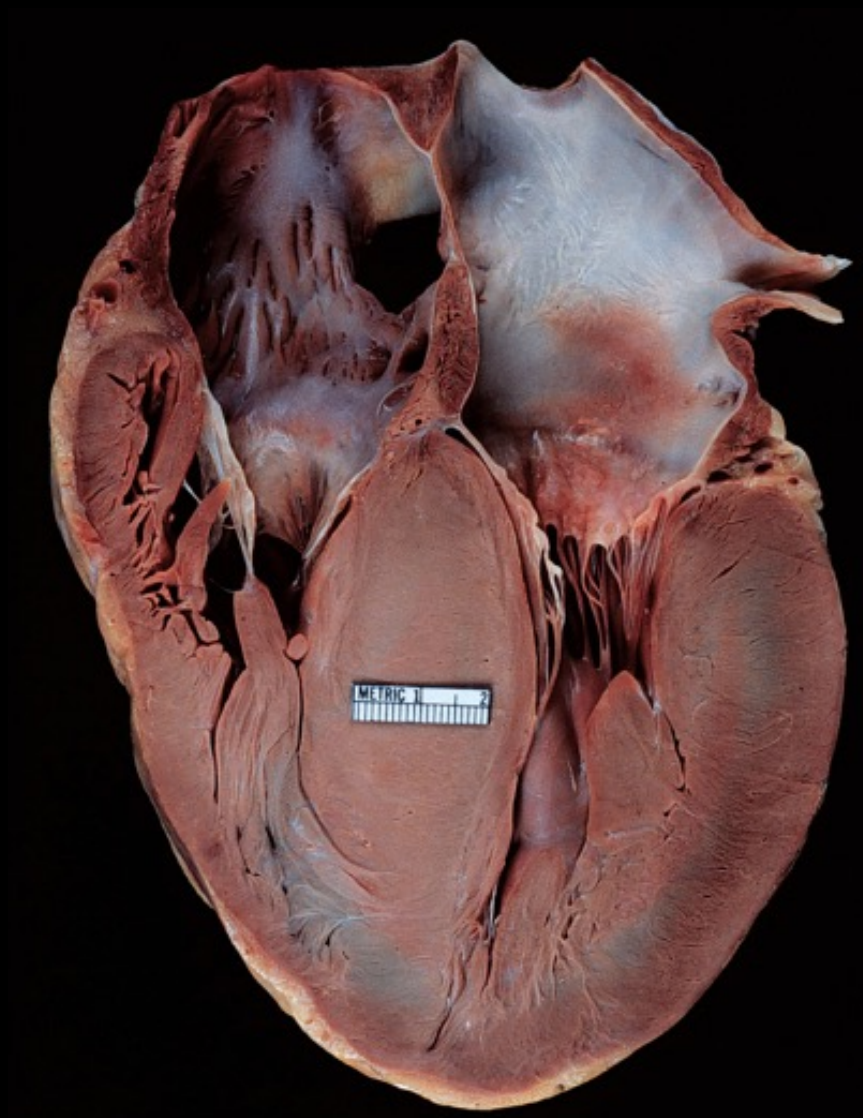
NOT SO RARE BUT NOT SO EASY

BENJAMIN ESSAYAGH, MD, FESC, FACC
CARDIO X CLINIC, Cannes, FR / MAYO CLINIC, Rochester, MN

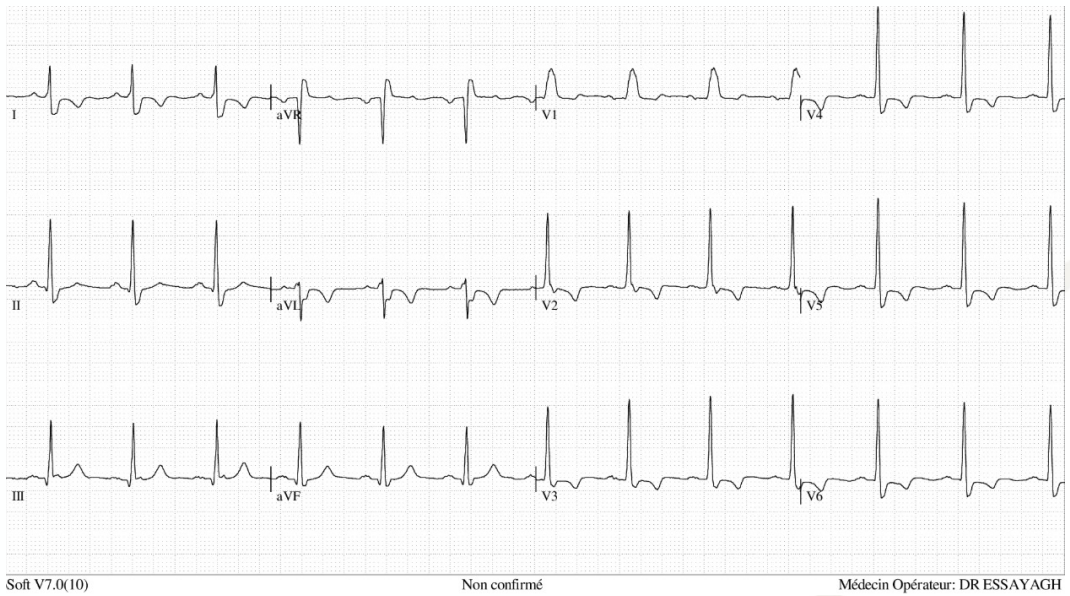
NO DISCLOSURES

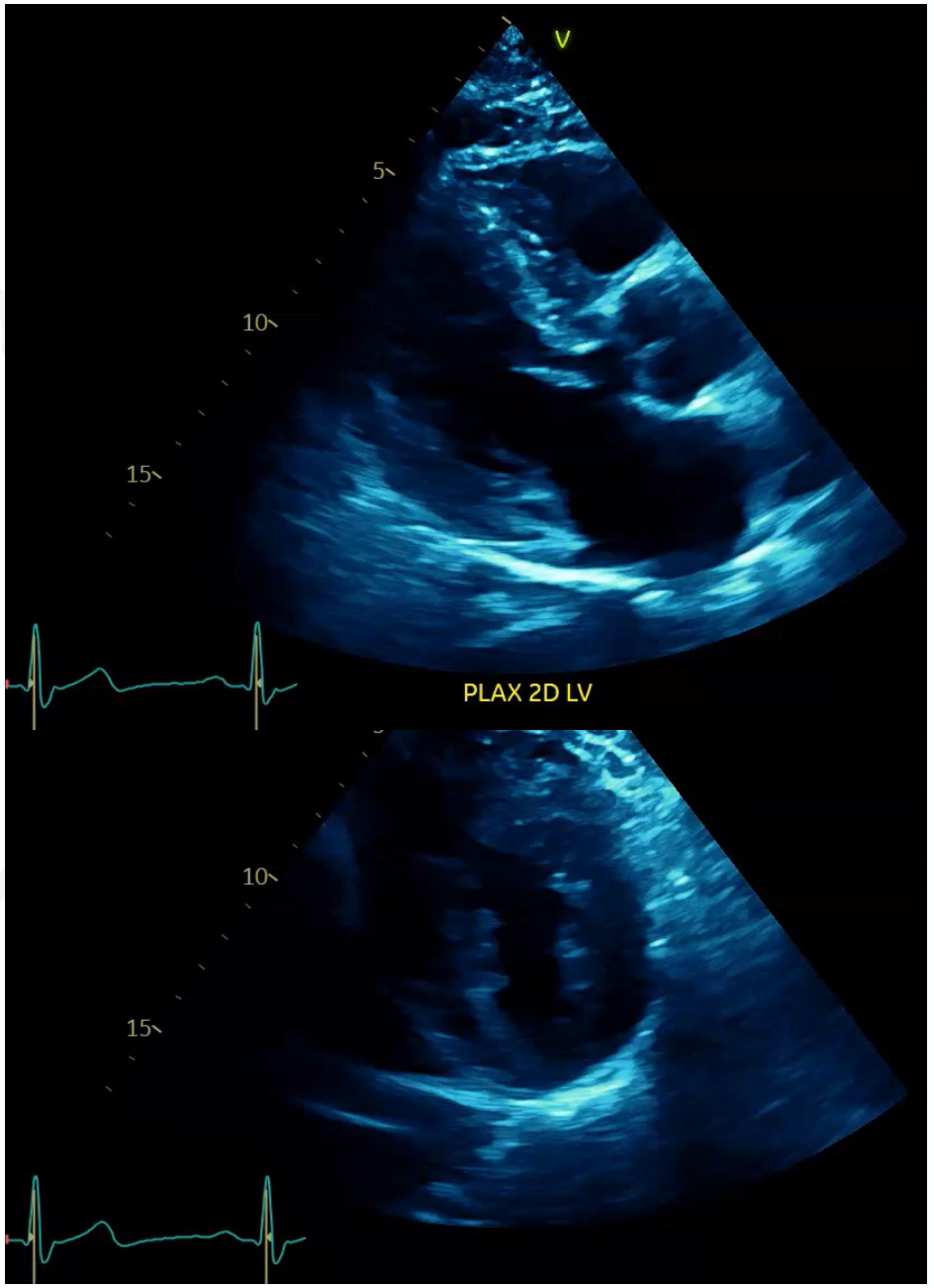
 | 0000001-3

HCM



Courtesy of Dr. W. D. Edwards, Mayo Clinic, Rochester, MN





/ 58yo man, smoker +++

/ dyspnea NYHA III

/ RCA 30%, normal cardiac scintigraphy

/ 3/6 aortic systolic murmur, no edema

/ 63% EF EDLVD 36mm EDLVV 68ml/m², IVSd 17mm, NTproBNP 862ng/mL

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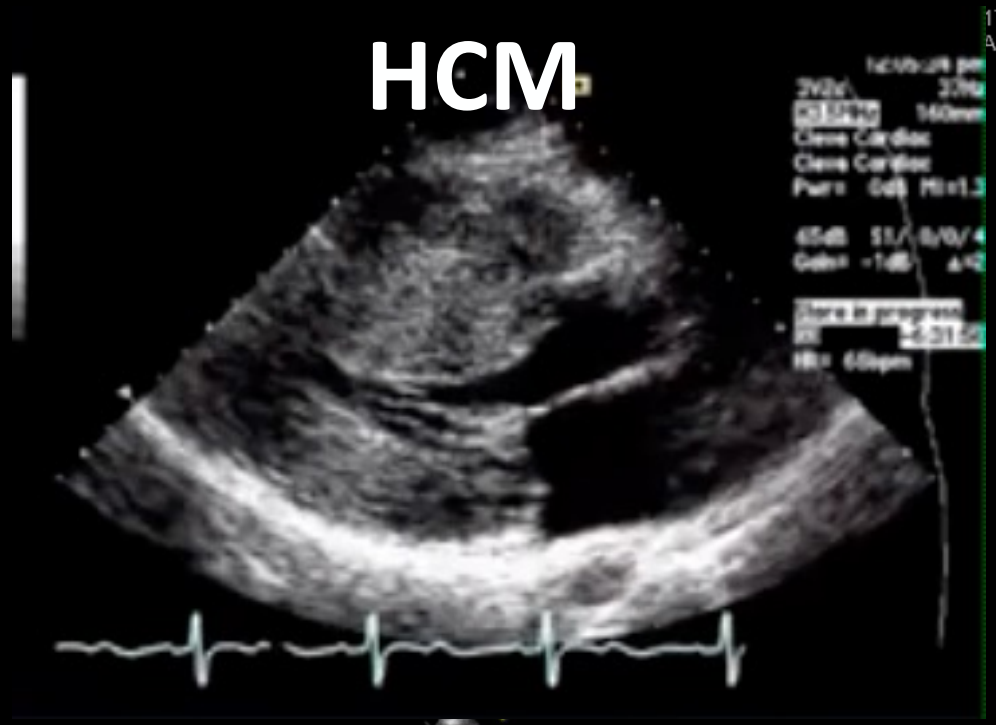
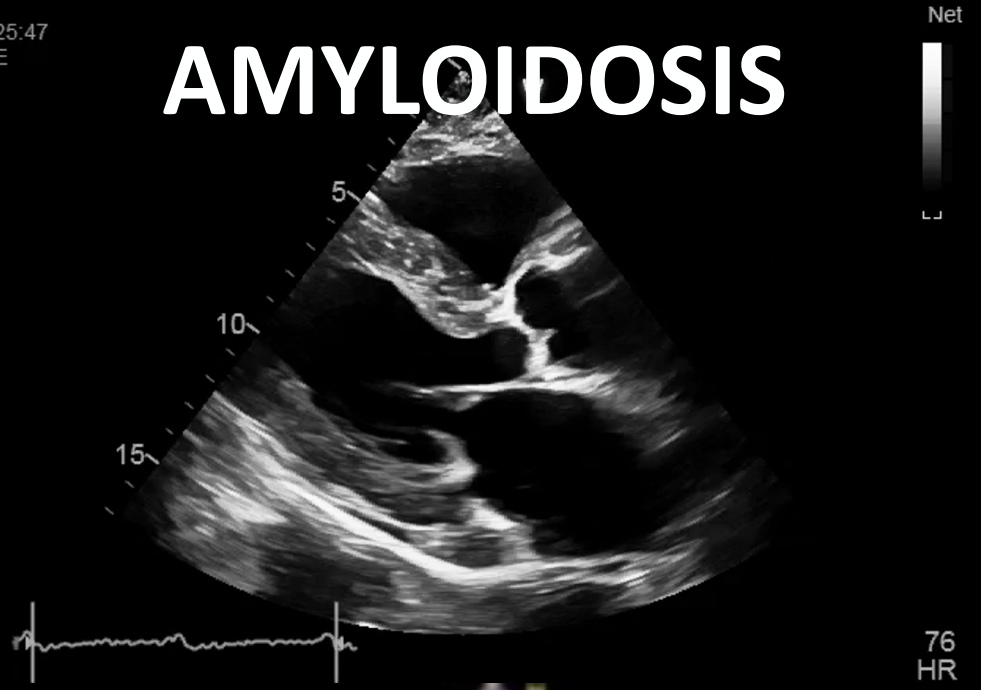
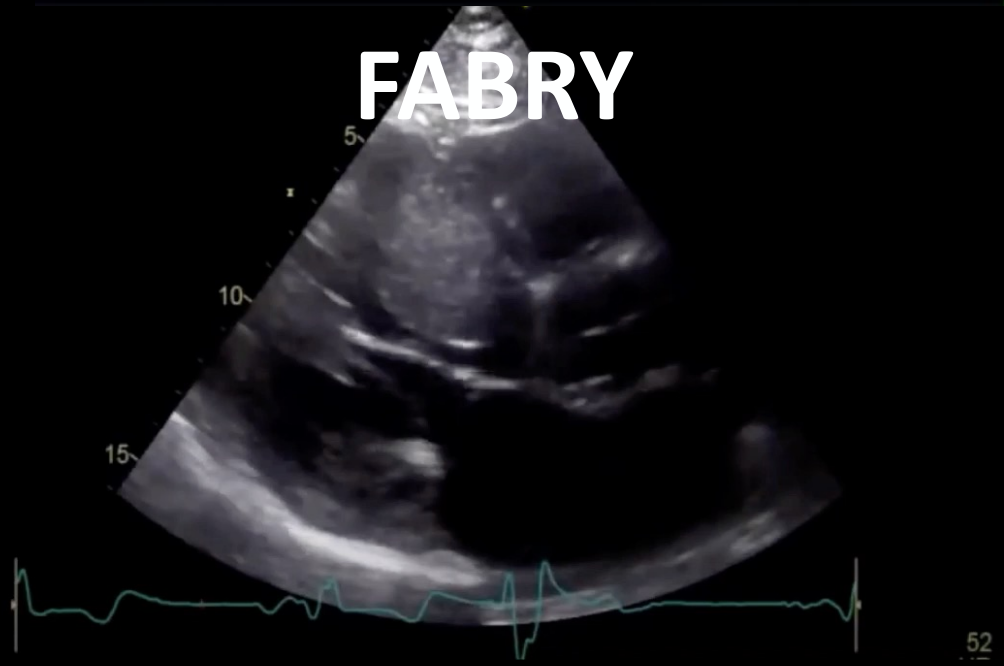
HCM



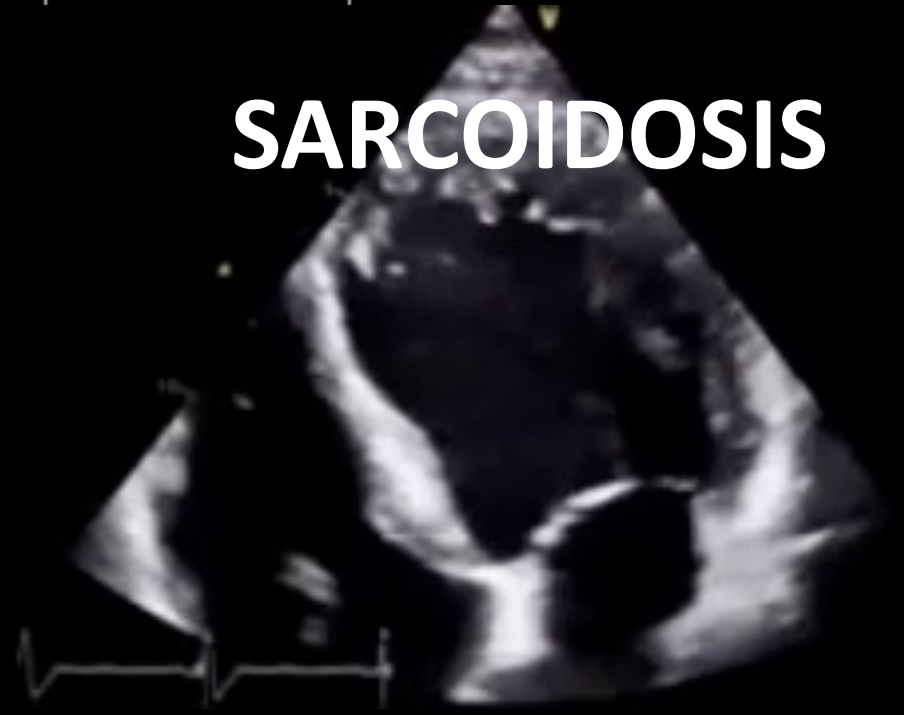
AMYLOIDOSIS



FABRY



SARCOIDOSIS



CARDIO X CLINIC

MAYO CLINIC

ACCA - HCM

WHAT IS AN HYPERTROPHIC CARDIOMYOPATHY ?

CARDIO X CLINIC

MAYO CLINIC

DIAGNOSTIC CRITERIA

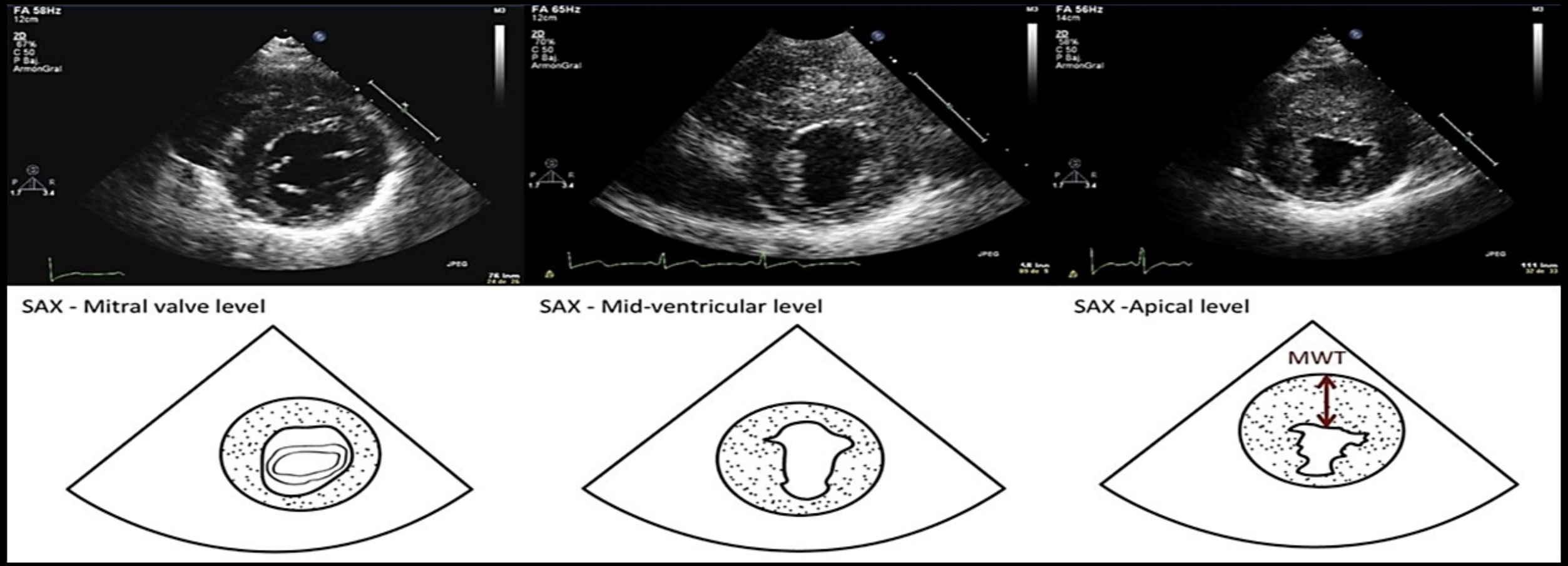
ADULTS: LV wall thickness ≥ 15mm in any myocardial segment

RELATIVES: LV wall thickness ≥ 13mm

CHILDREN: LV wall thickness > 2SD (2 z-score)

LEFT VENTRICULAR HYPERTROPHY IN HCM

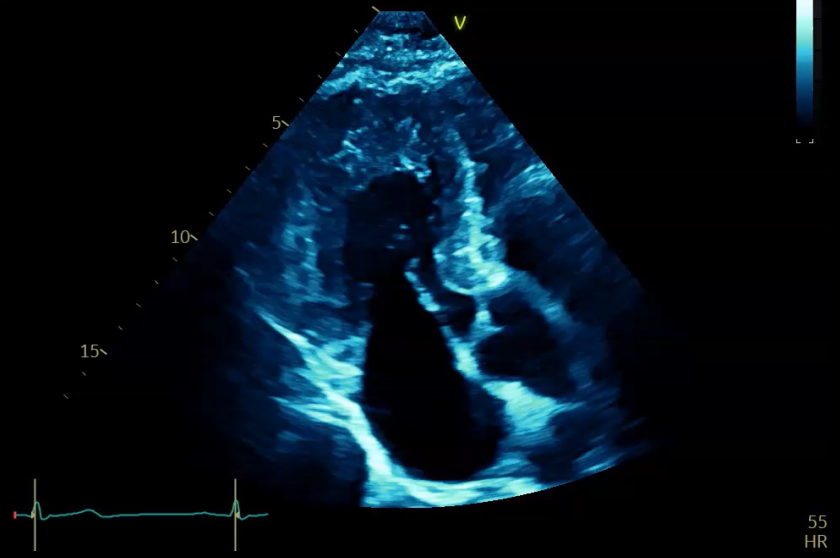
Assess MWT at different levels



Dominguez, et al. *Heart.* 2018.

TYPICAL FINDINGS IN SEVERE IN HCM

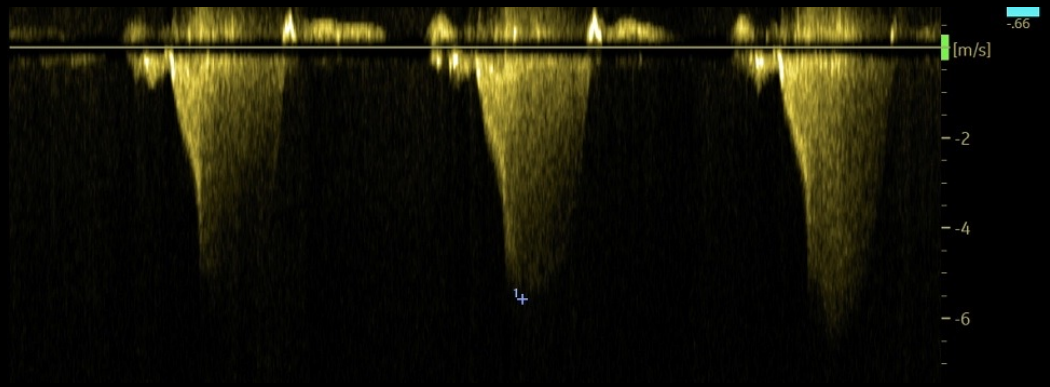
Systolic anterior motion (SAM) of the mitral valve



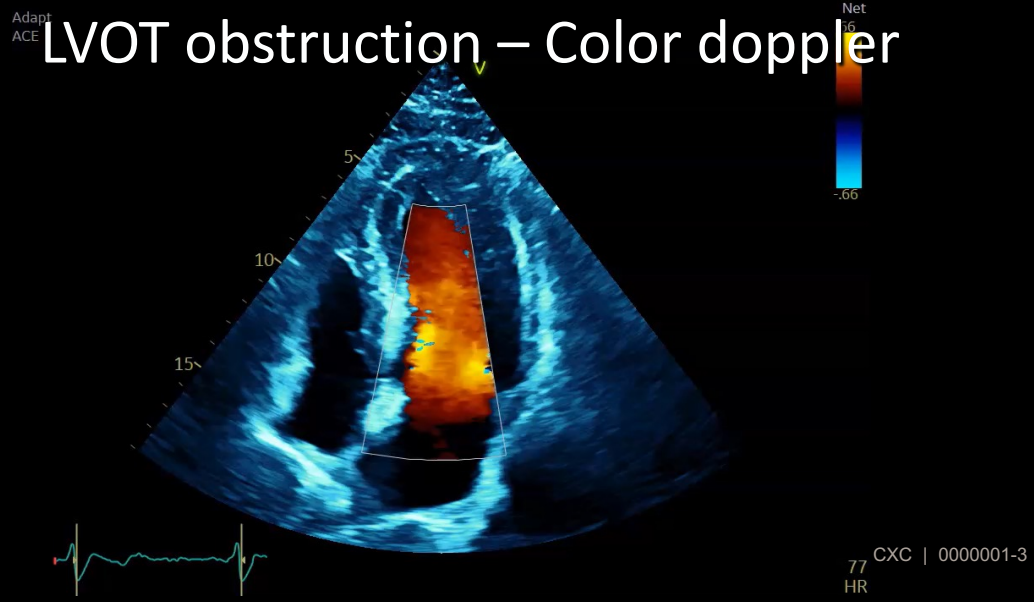
Mitral regurgitation

LVOT obstruction – CW doppler

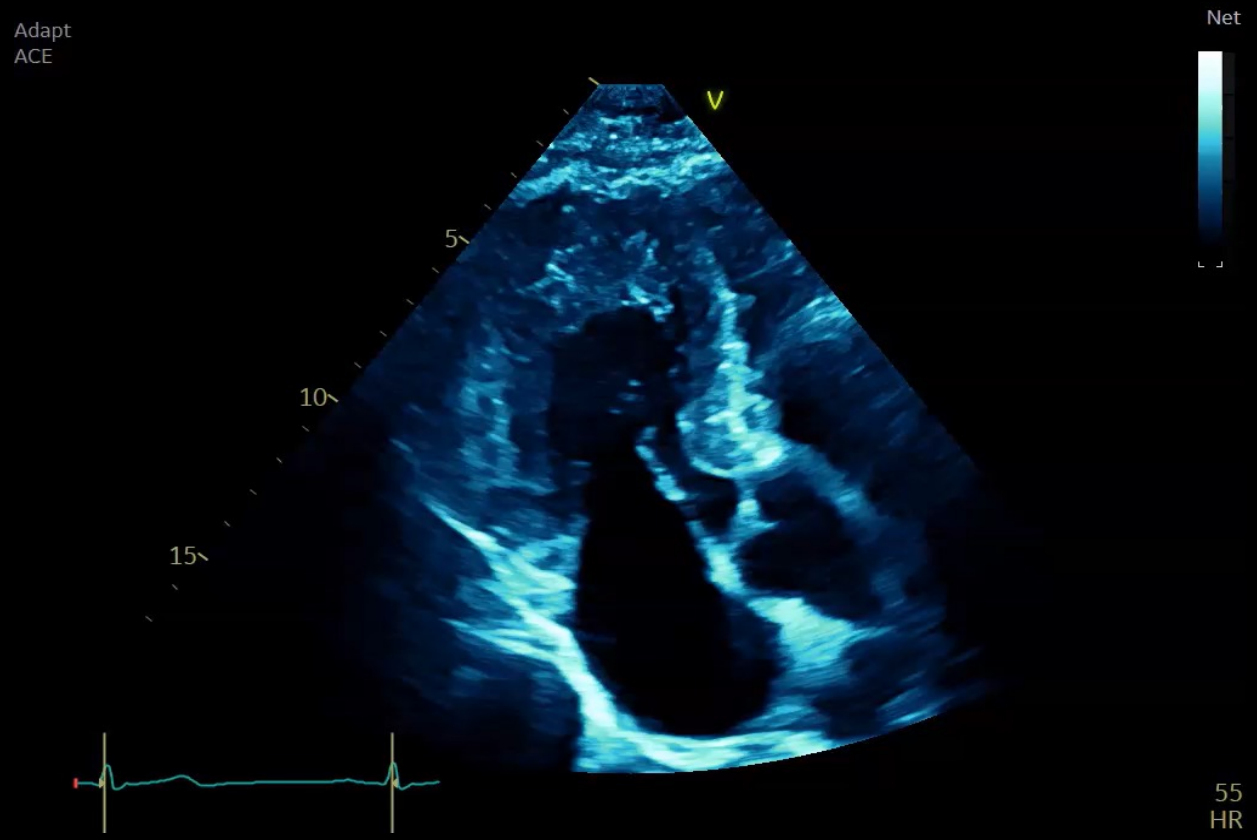
Typical dagger-shaped envelope



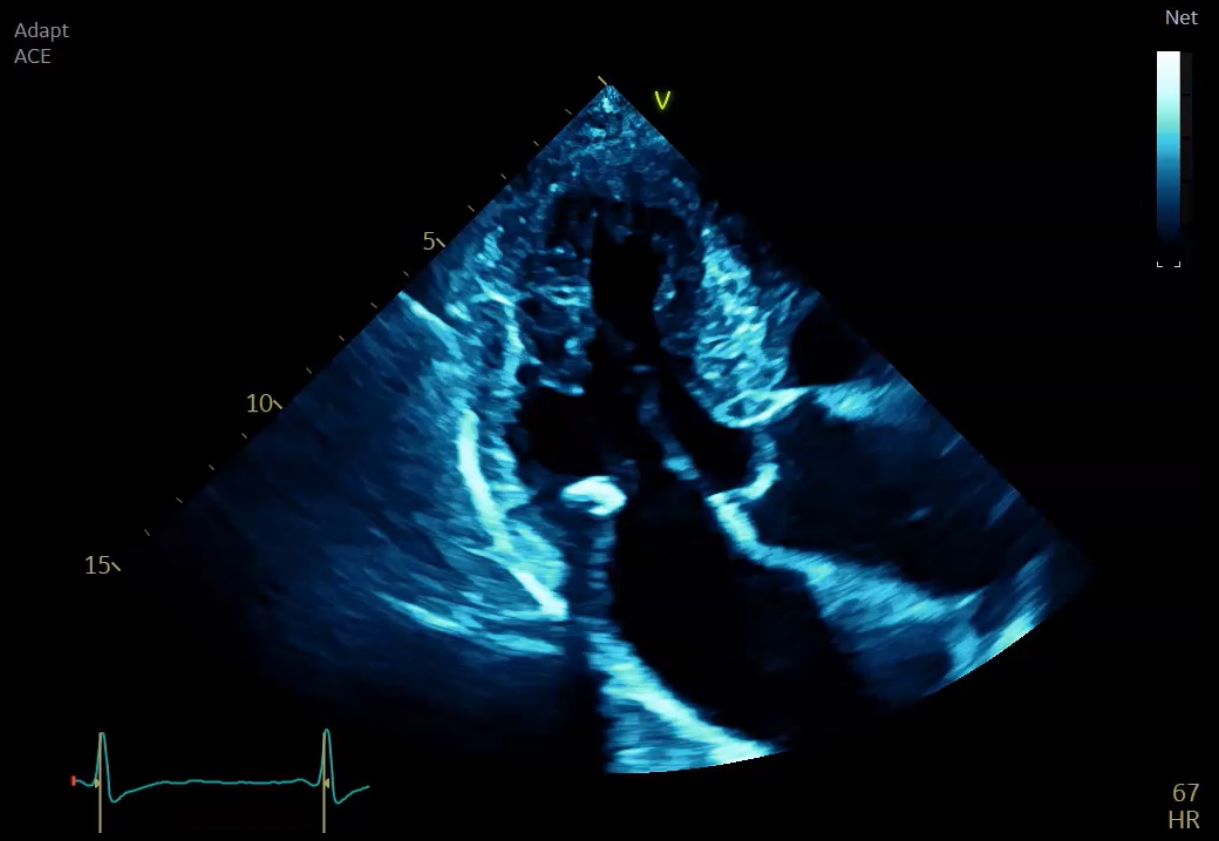
LVOT obstruction – Color doppler



TYPICAL FINDINGS IN SEVERE IN HCM



Septal HCM
17mm
SAM



Anterior and septal HCM
23mm
No SAM

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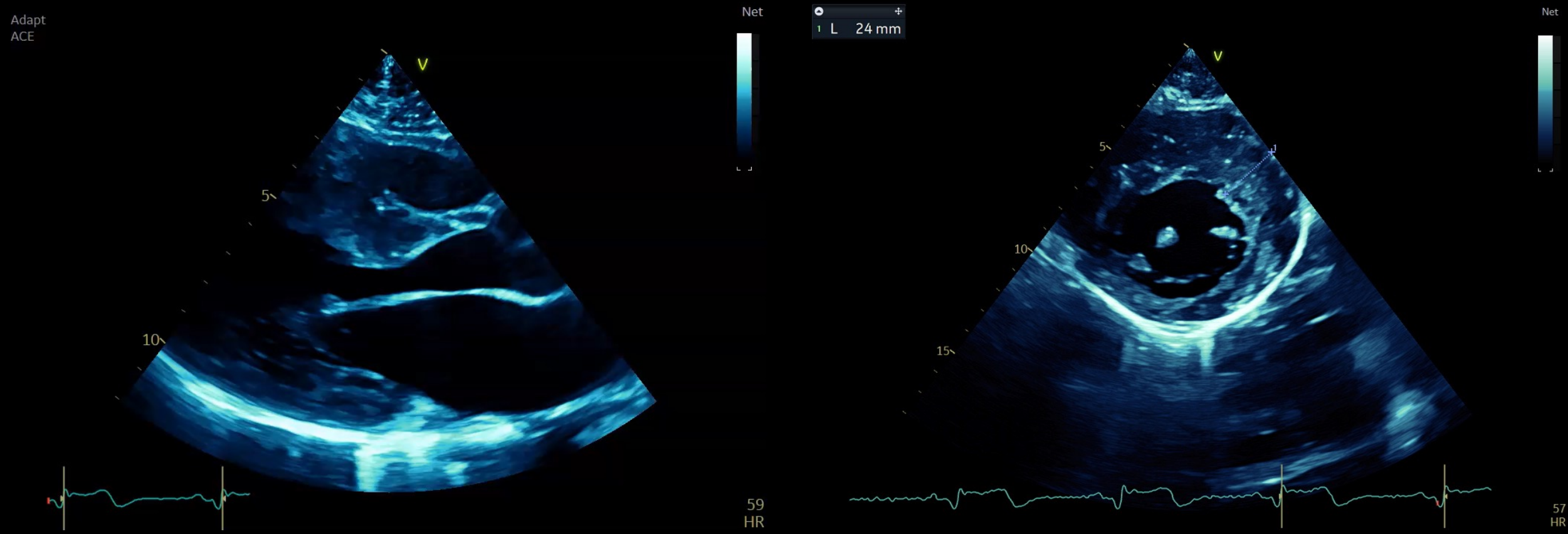
TYPICAL FINDINGS IN SEVERE IN HCM



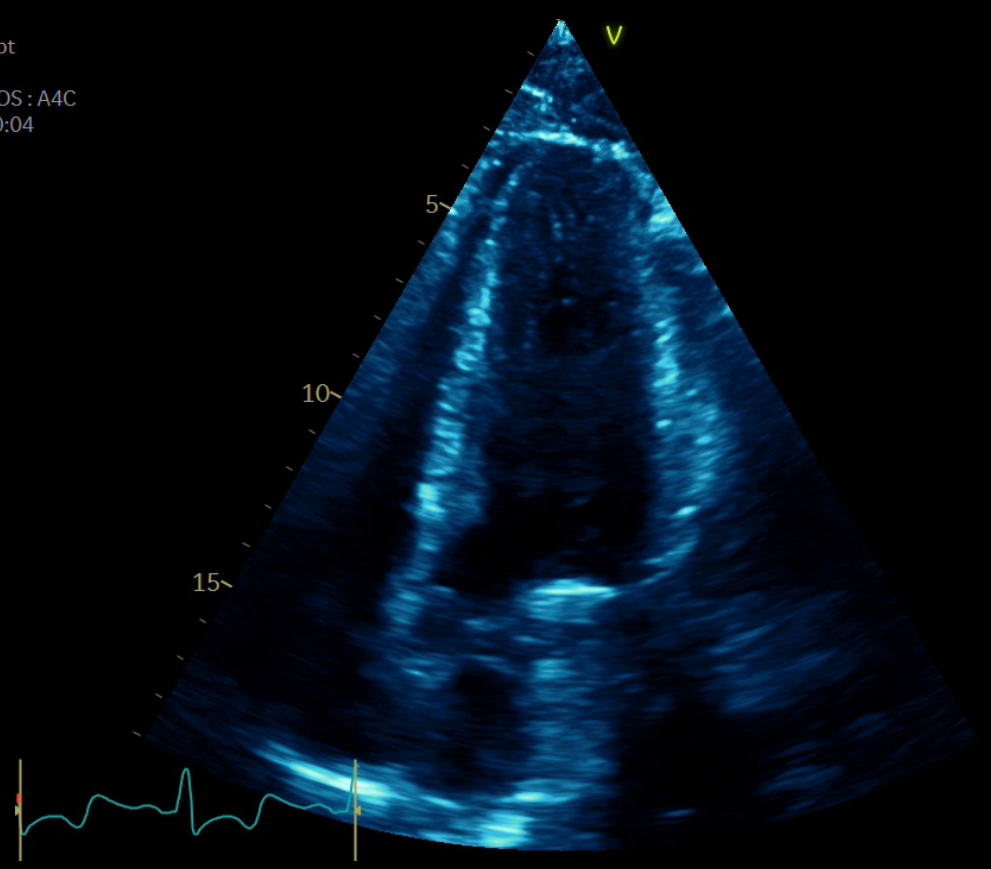
Anterior HCM

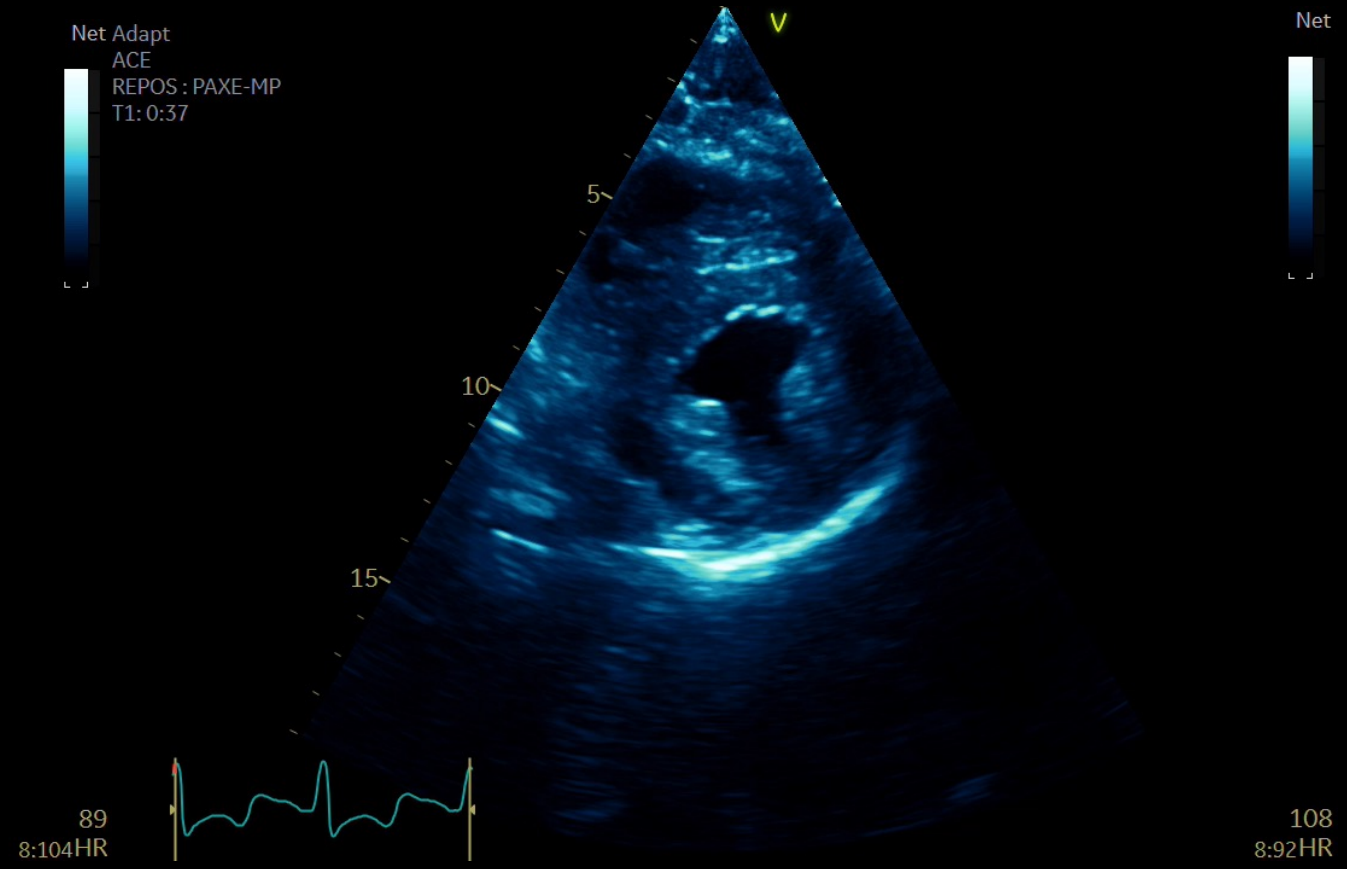
24mm

TNNI3+

Courtesy of Dr. J. LAÏK, CANNES, FR

TYPICAL FINDINGS IN SEVERE IN HCM





Apical HCM
21mm

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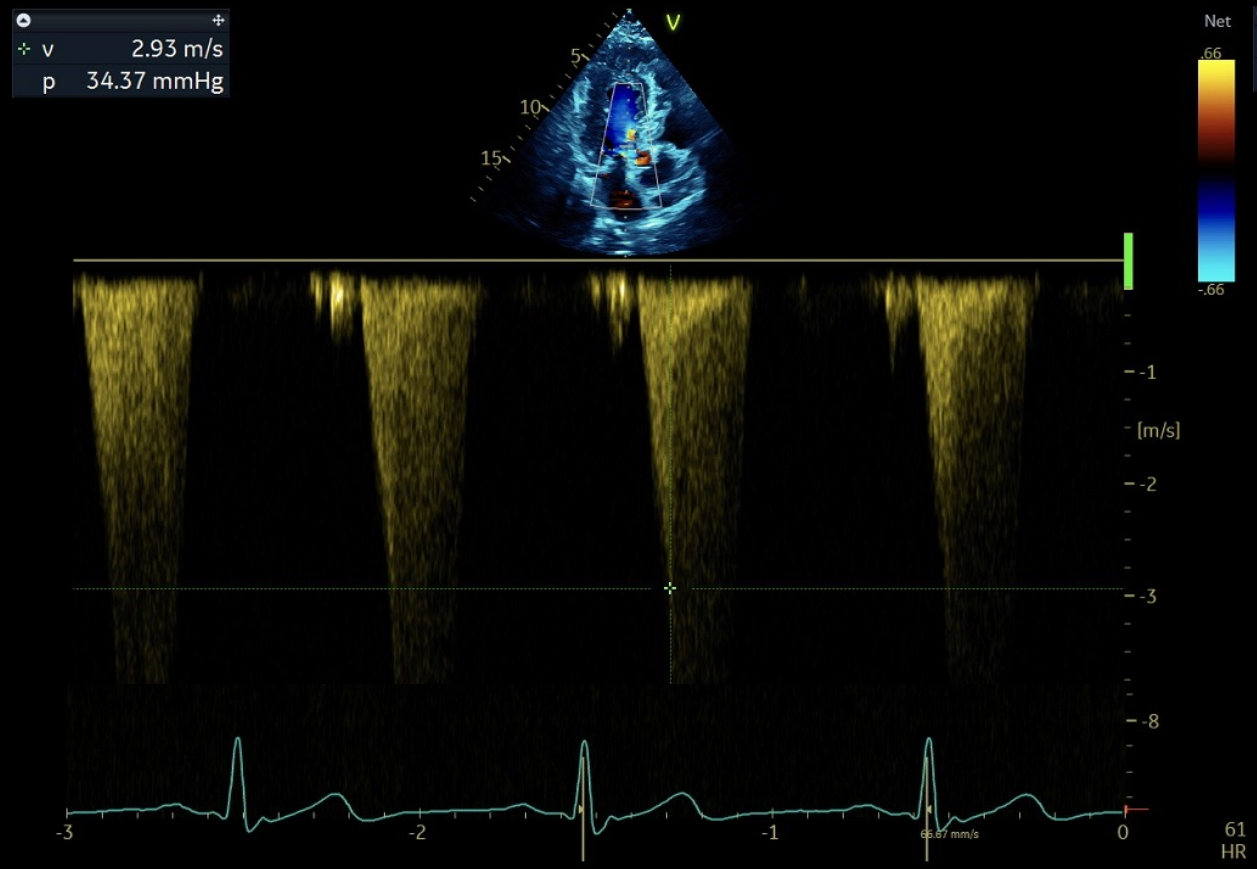
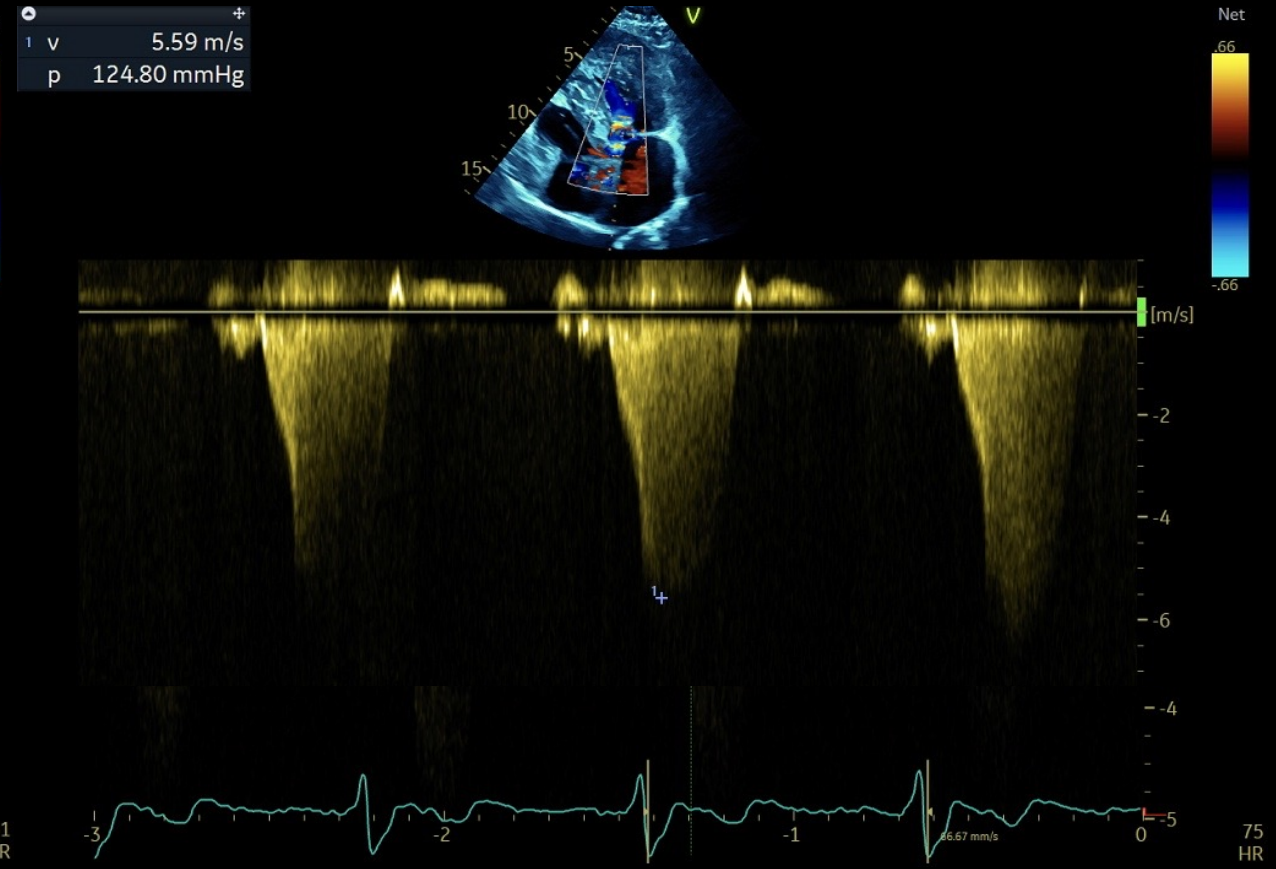
SQUATS IN HCM



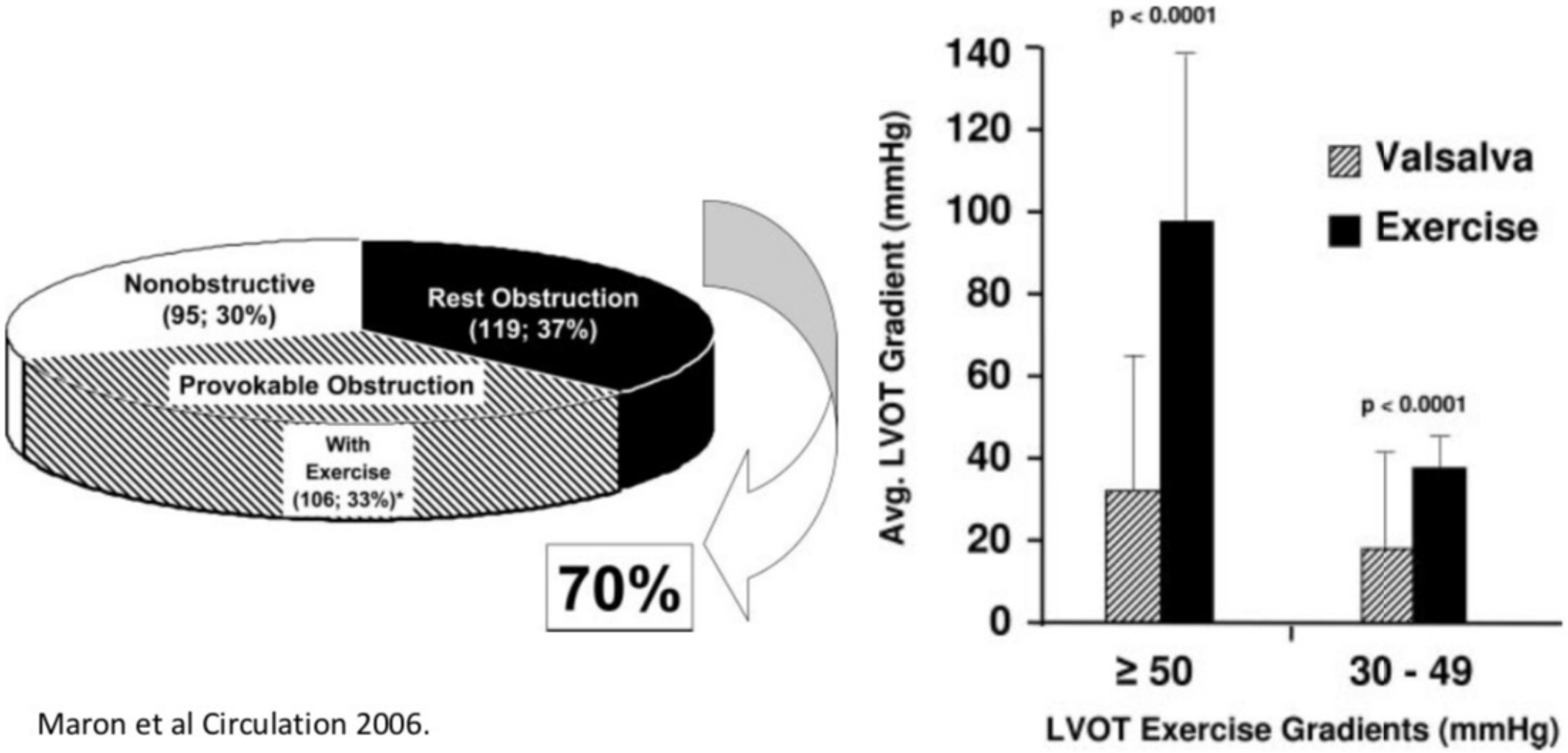
V= 3 m/s
Gmax 35 mmHg



V= > 5 m/s
Gmax >100 mmHg

CARDIO X CLINIC
MAYO CLINIC

PROVOCABLE LVOTO IN HCM

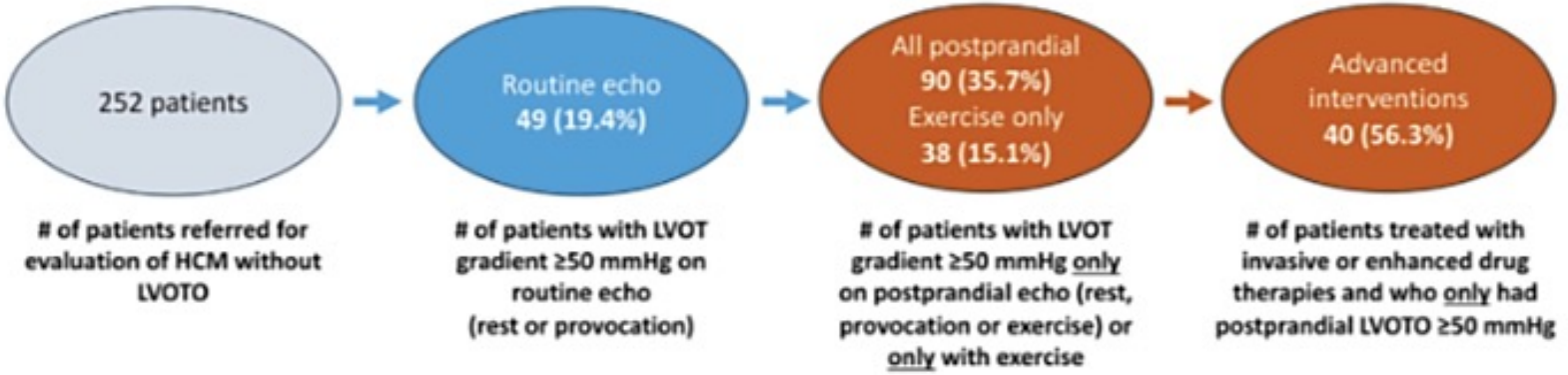


Maron et al Circulation 2006.

CARDIO X CLINIC MAYO CLINIC

Unmasking Obstruction in Hypertrophic Cardiomyopathy With Postprandial Resting and Treadmill Stress Echocardiography

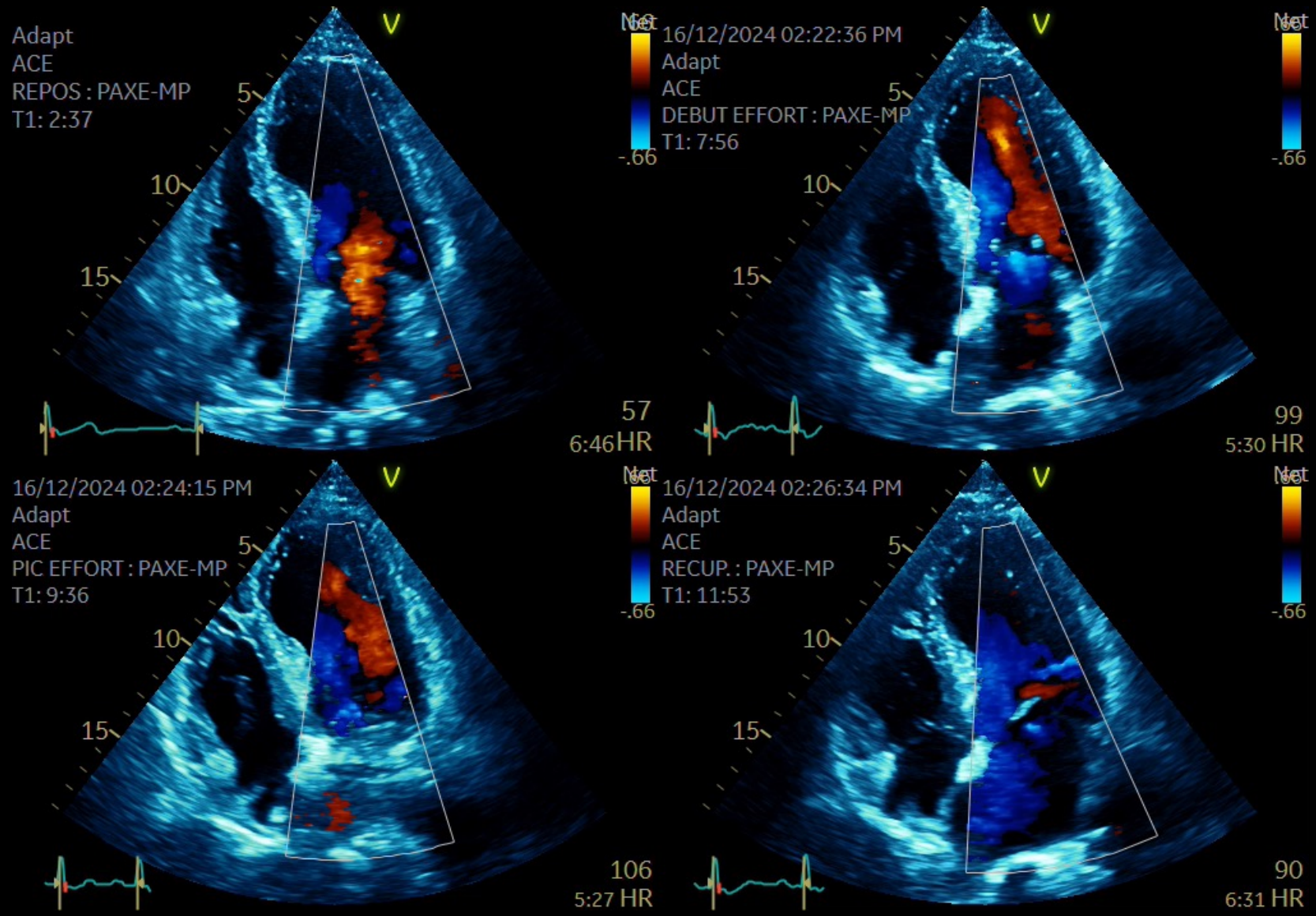
Daniele Massera, MD, MSc, Clarine Long, MD, Yuhe Xia, MS, Les James, MD, MPH, Elizabeth Adlestein, BA, Isabel C. Alvarez, BS, MPH, Woon Y. Wu, FNP, Maria C. Reuter, AGACNP, Milla Arabadjian, PhD, Eugene A. Grossi, MD, Muhamed Saric, MD, PhD, and Mark V. Sherrid, MD, *New York and Mineola, New York*



Journal of the American Society of Echocardiography
■ 2024

CARDIO X CLINIC
MAYO CLINIC

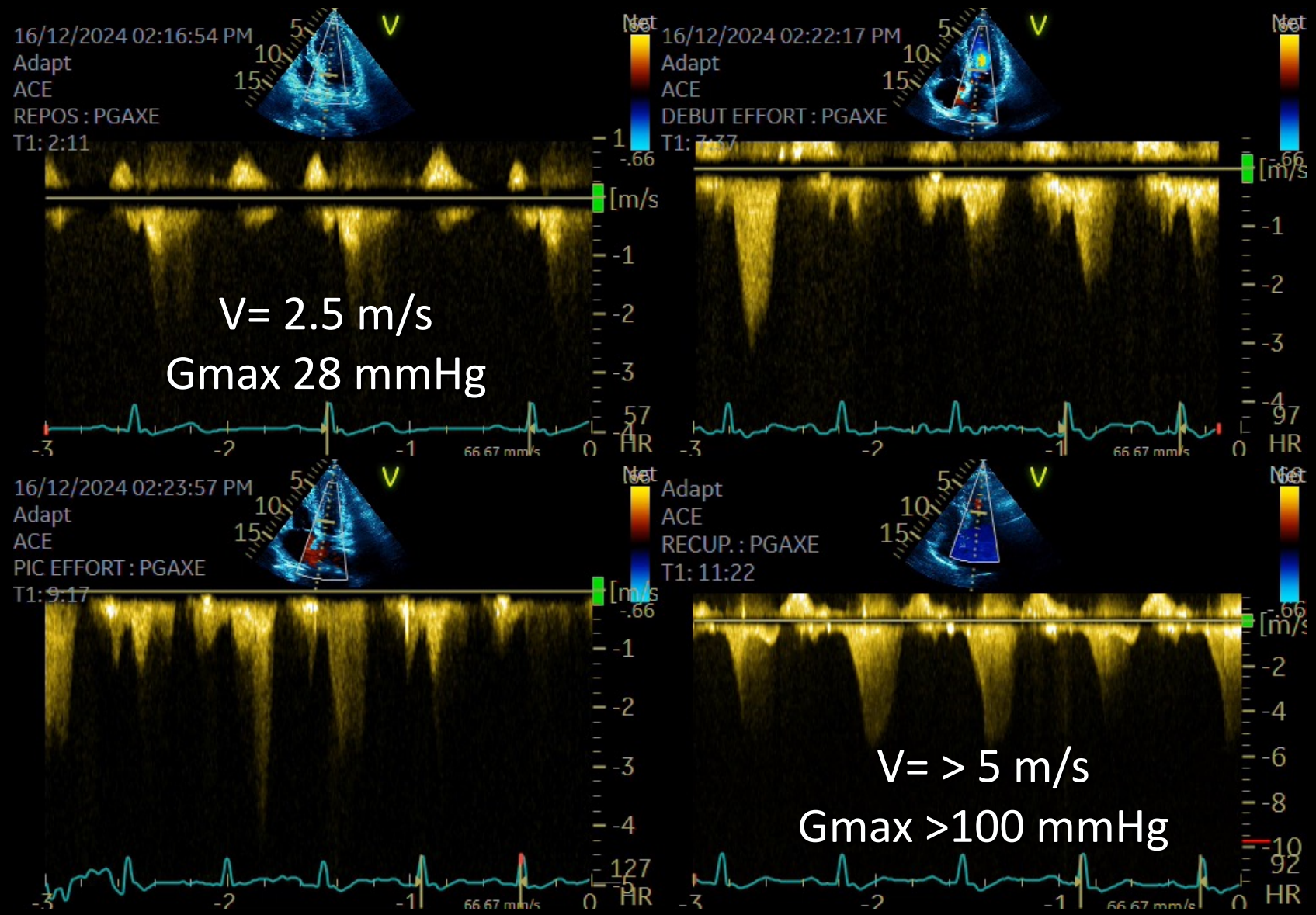
EXERCISE ECHOCARDIOGRAPHY IN HCM



CARDIO X CLINIC

MAYO CLINIC

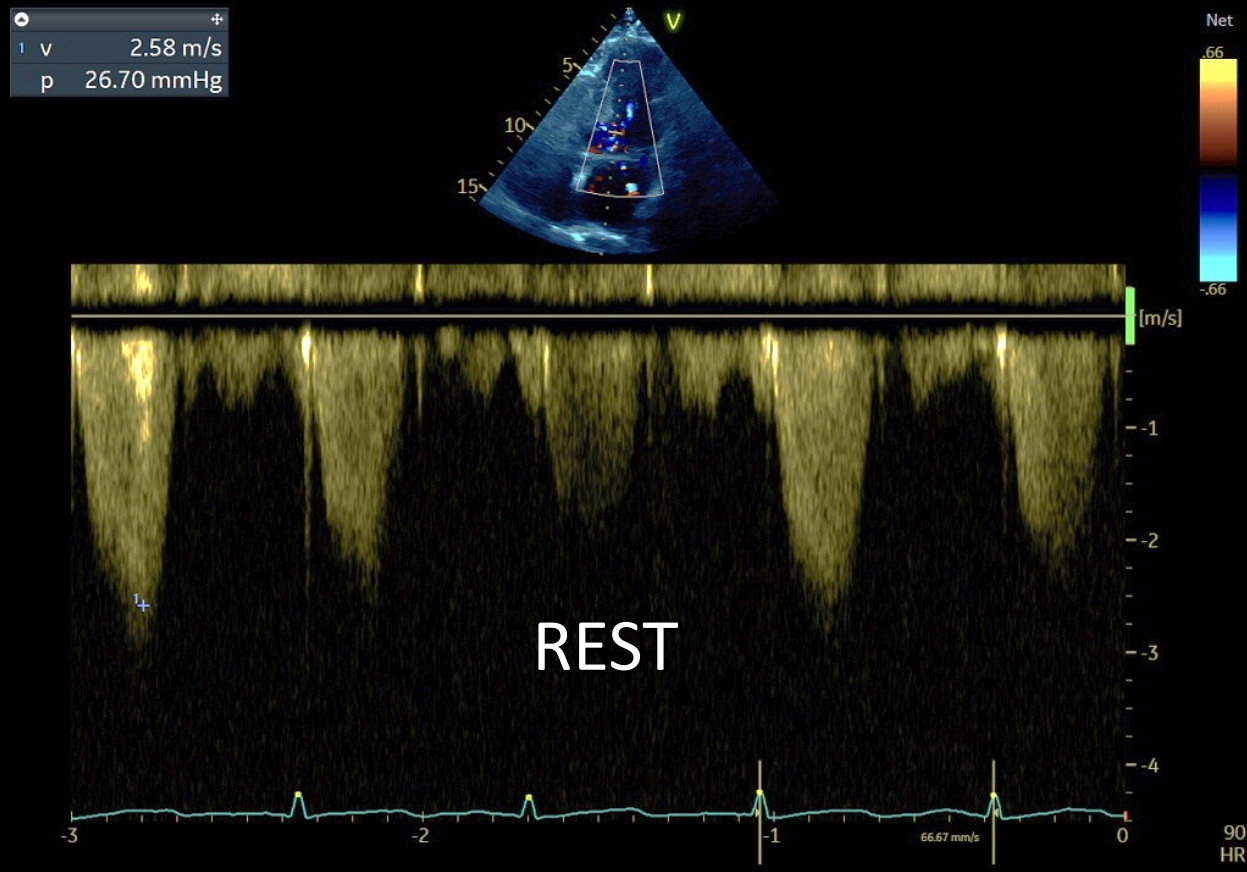
EXERCISE ECHOCARDIOGRAPHY IN HCM



CARDIO X CLINIC

MAYO CLINIC

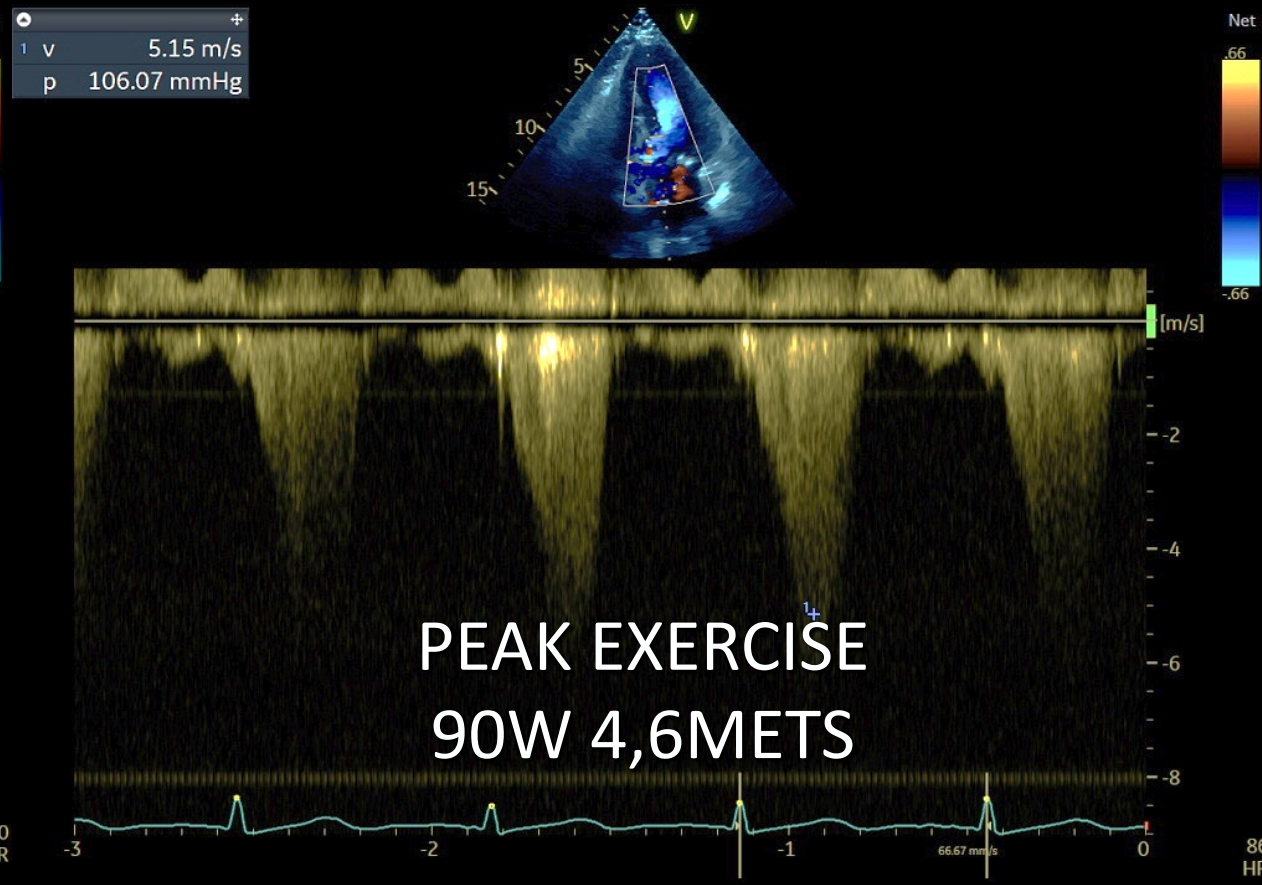
EXERCISE ECHOCARDIOGRAPHY IN HCM



REST

V= 2.6 m/s

Gmax 27 mmHg



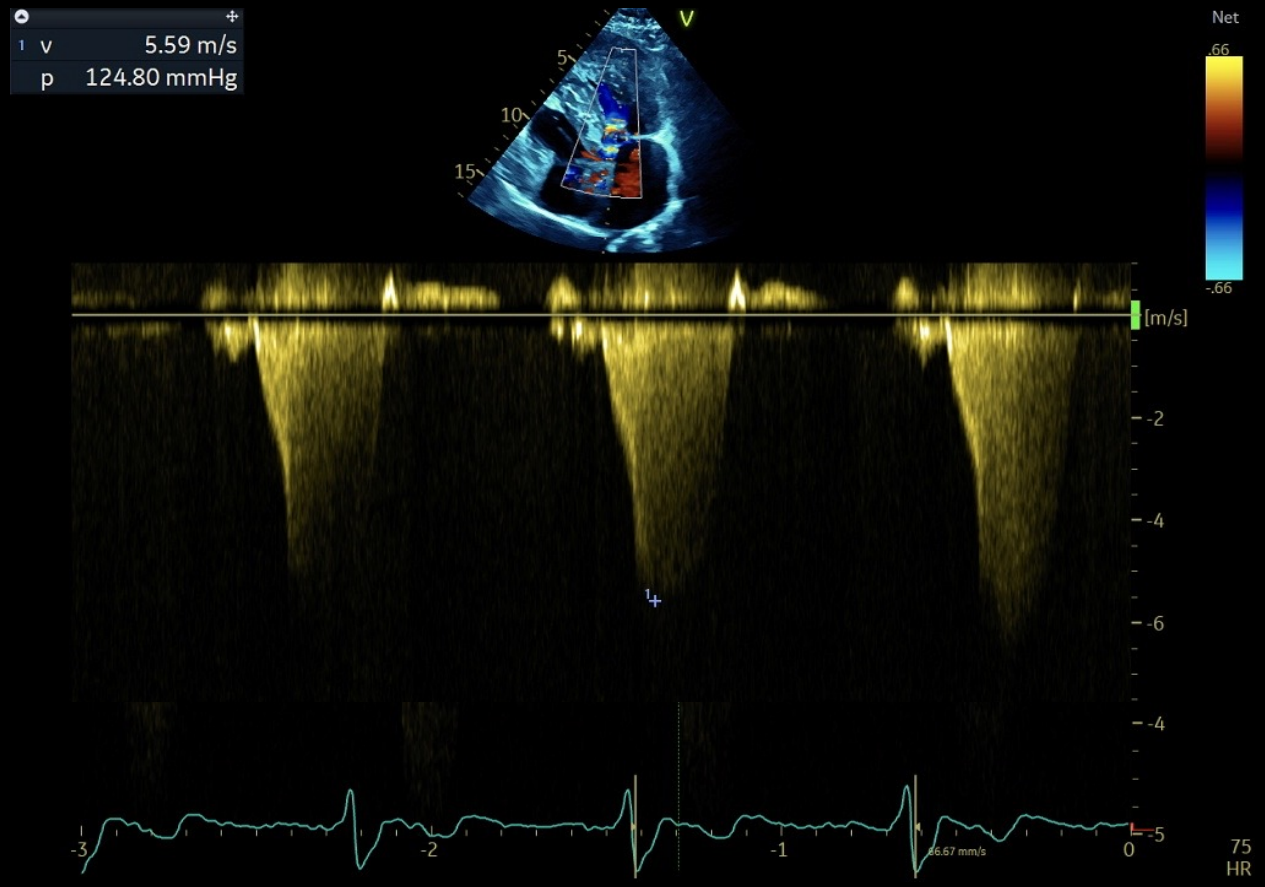
PEAK EXERCISE
90W 4,6METS

V= > 5 m/s

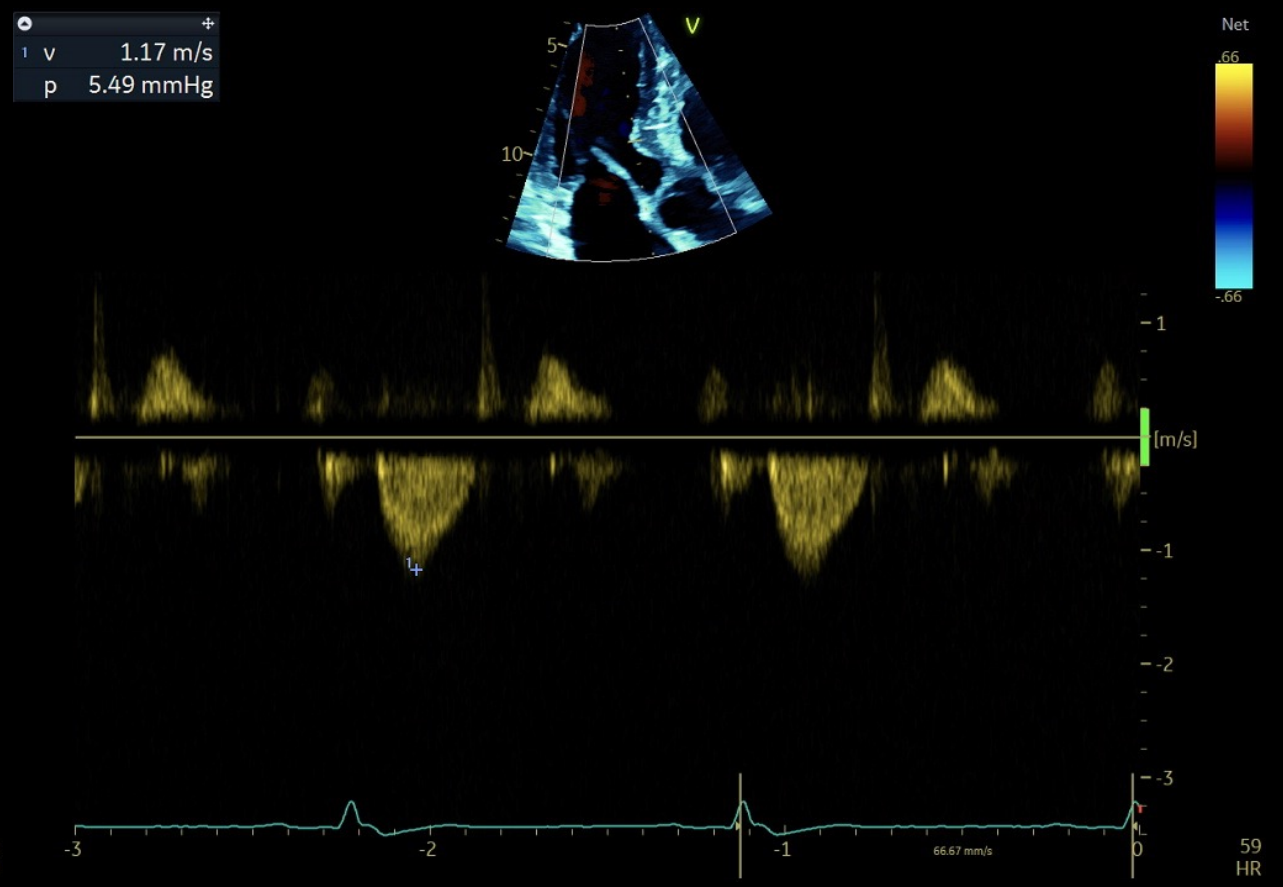
Gmax >100 mmHg

MAYO CLINIC
CARDIO X CLINIC

Courtesy of Dr. J. LAÏK, CANNES, FR



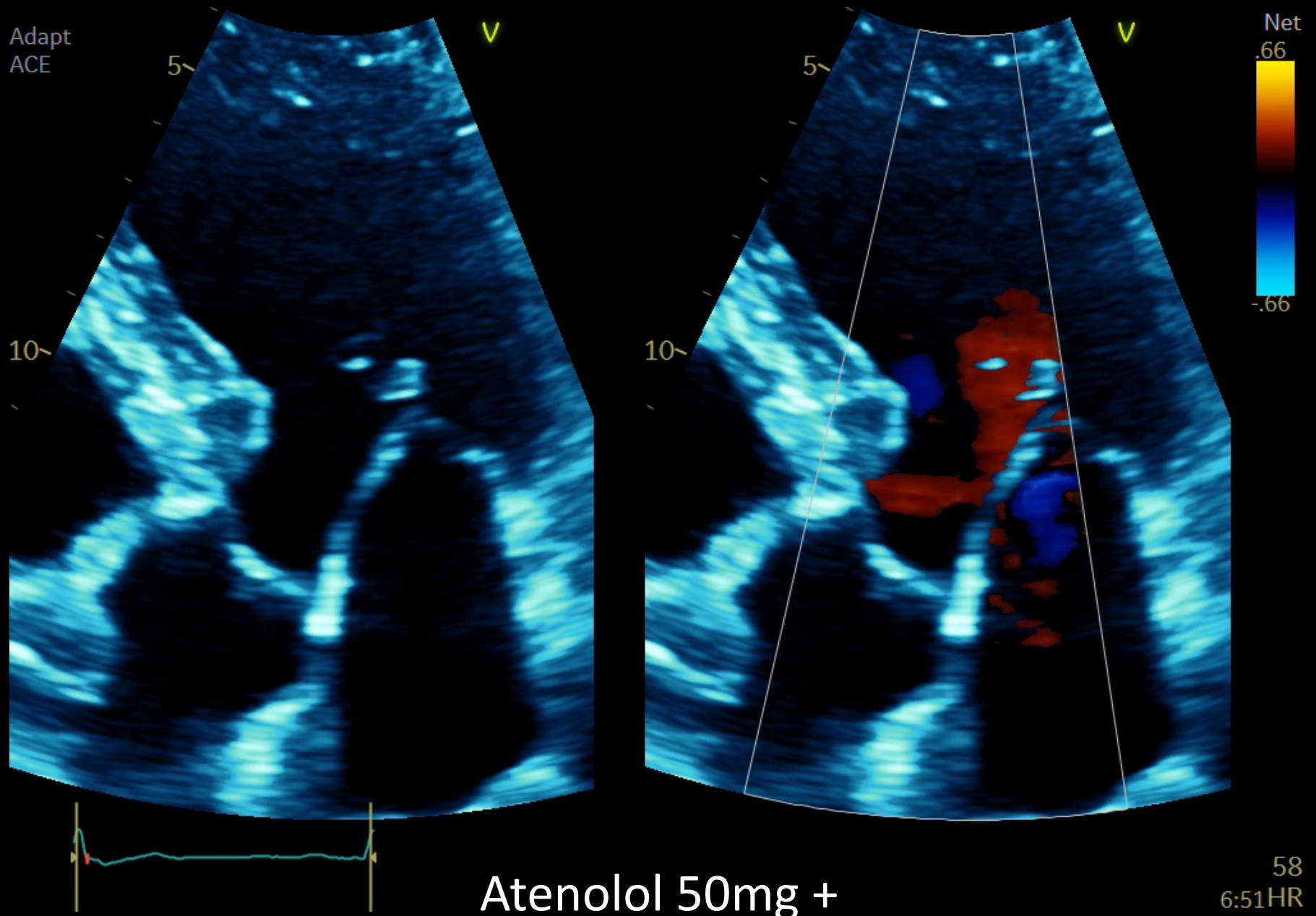
Atenolol 50mg



Atenolol 50mg +
Mavacamten 5mg

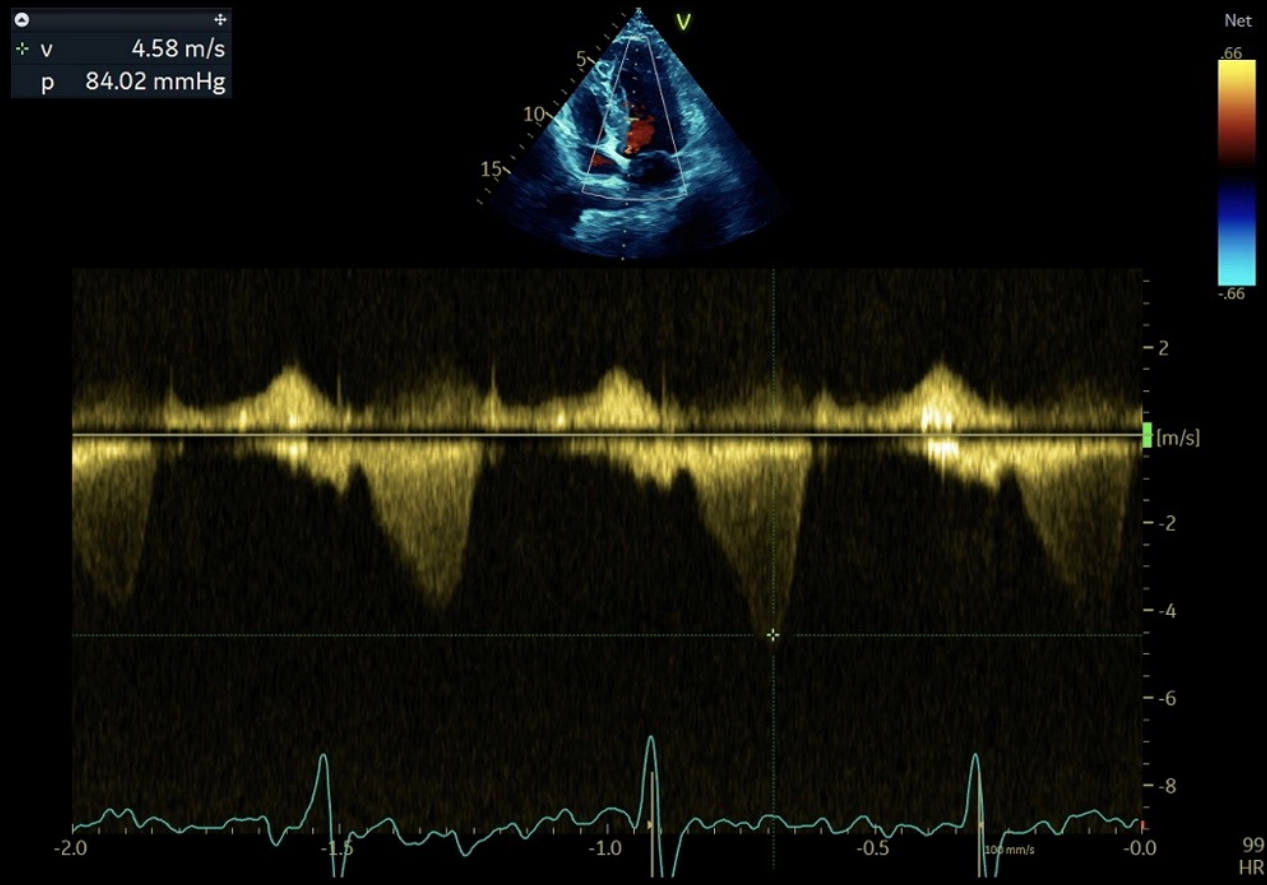
CARDIO X CLINIC

MAYO CLINIC

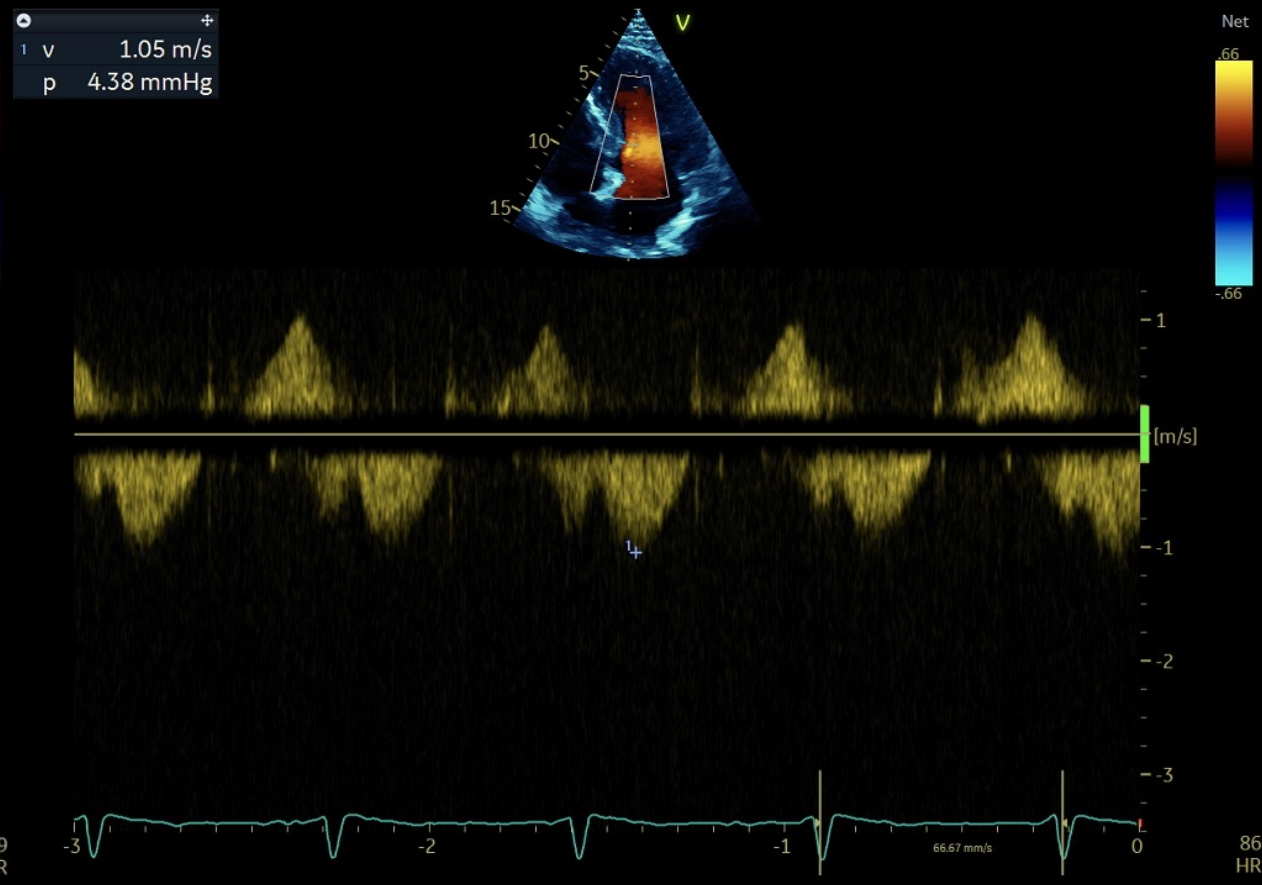


Atenolol 50mg +
Mavacamten 10mg

CARDIO X CLINIC
MAYO CLINIC



Corgard 40mg



Corgard 40mg +
Mavacamten 10mg

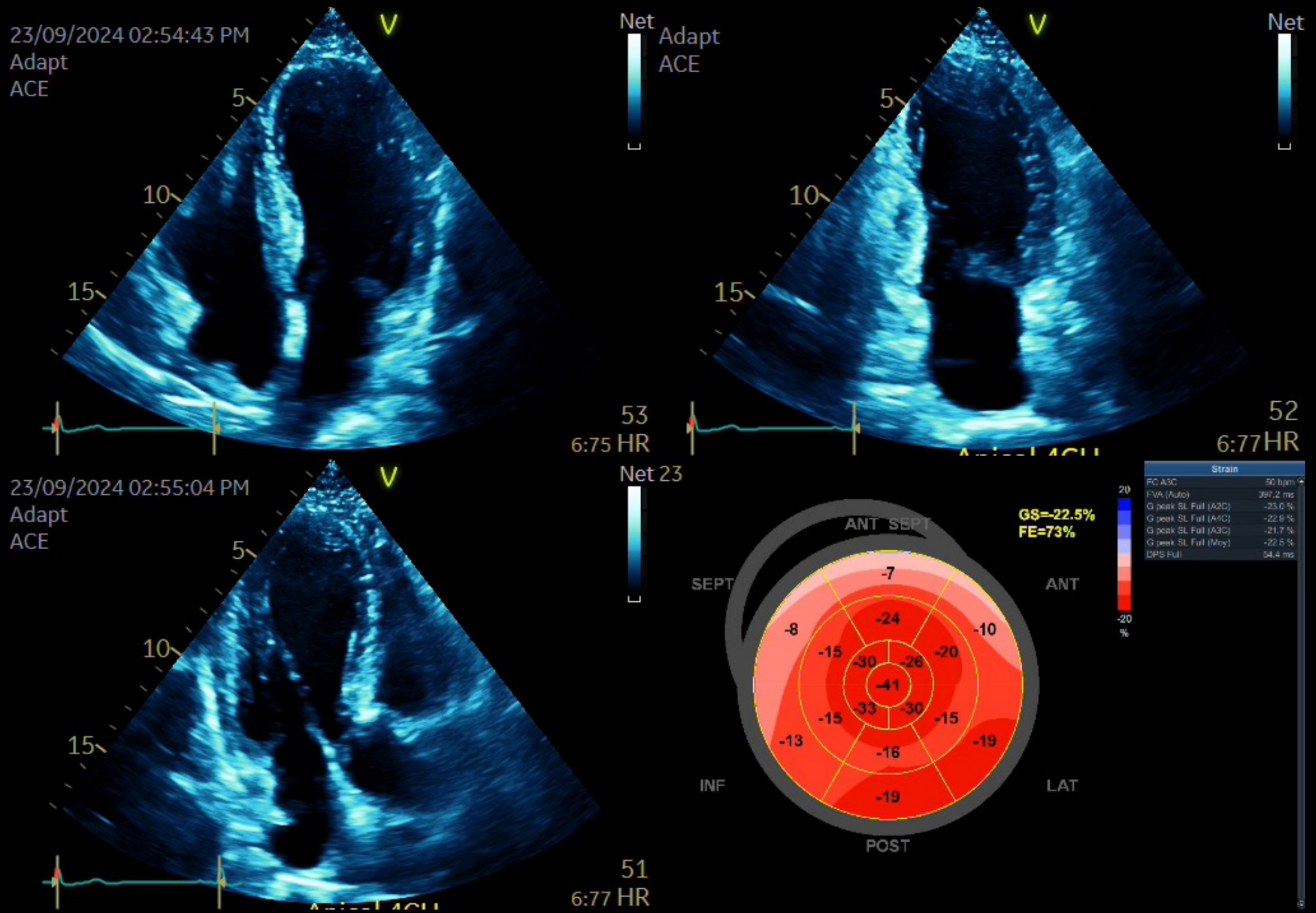
CARDIO X CLINIC
MAYO CLINIC

ACCA - HCM

HOW STRAIN ECHO HELPS IN HCM ?

CARDIO X CLINIC

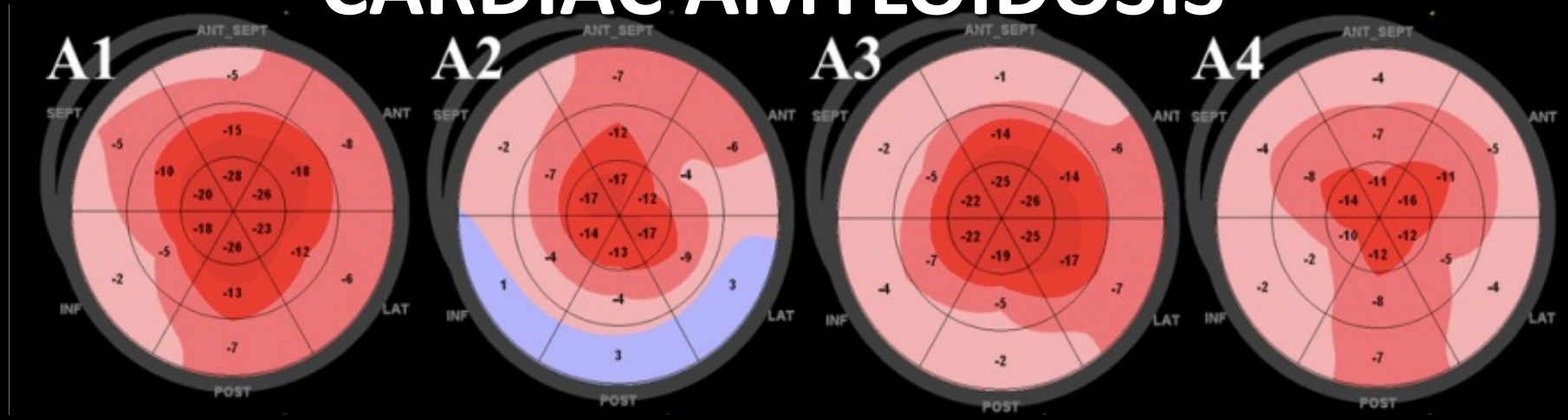
MAYO CLINIC



Courtesy of Dr. J. LAÏK, CANNES, FR

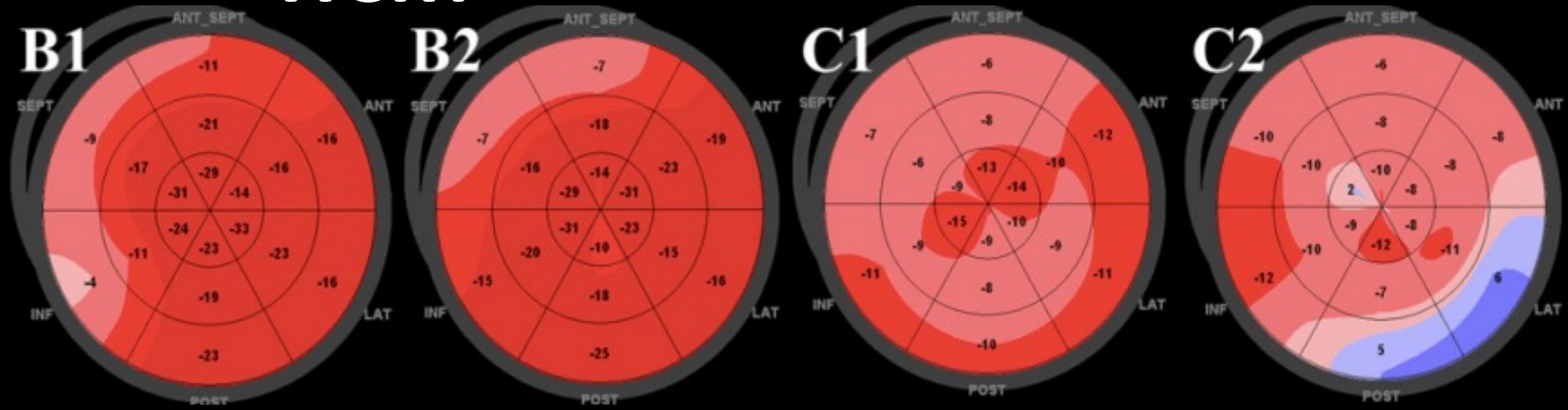
DIFFERENTIAL DIAGNOSIS

CARDIAC AMYLOIDOSIS



HCM

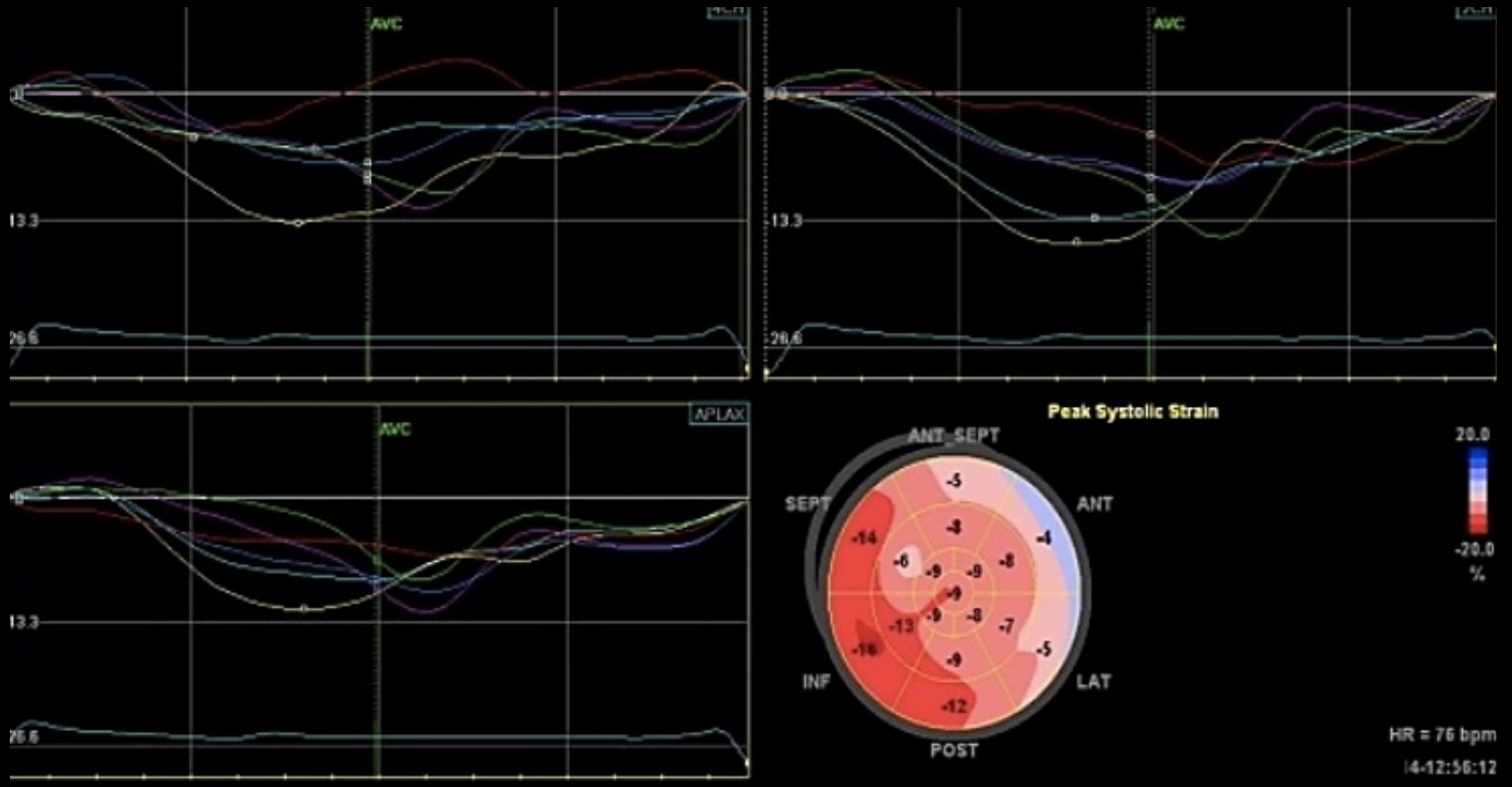
AS



CARDIO X CLINIC

MAYO CLINIC

HCM FAMILY MEMBERS GEN+ PHEN-



35y man
3 SCD family

EF = 65%
GLS = -9%

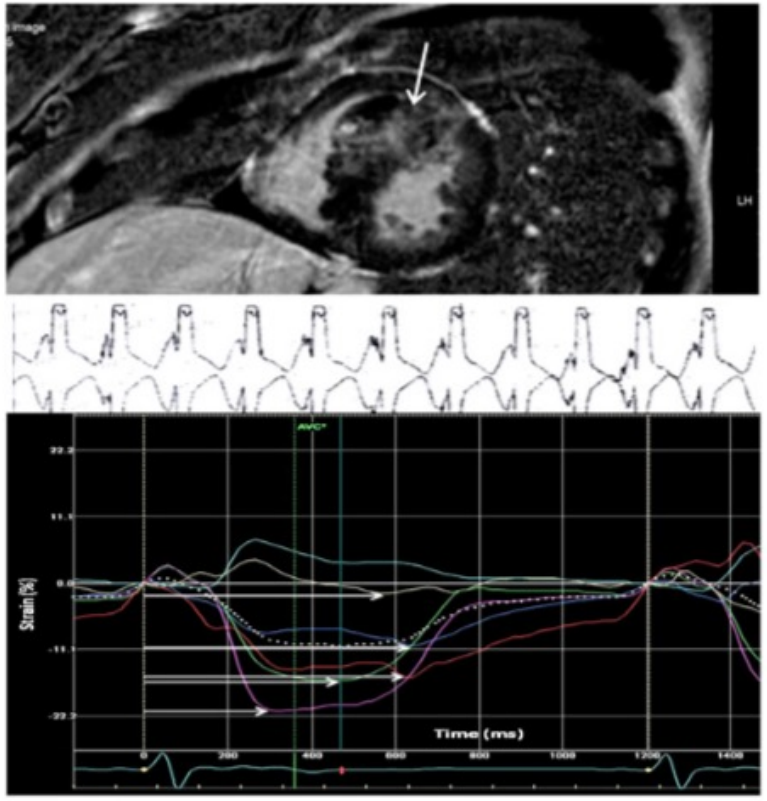
STRAIN IS ABNORMAL

CARDIO X CLINIC
MAYO CLINIC

STRAIN IS LINKED TO VA AND FIBROSIS

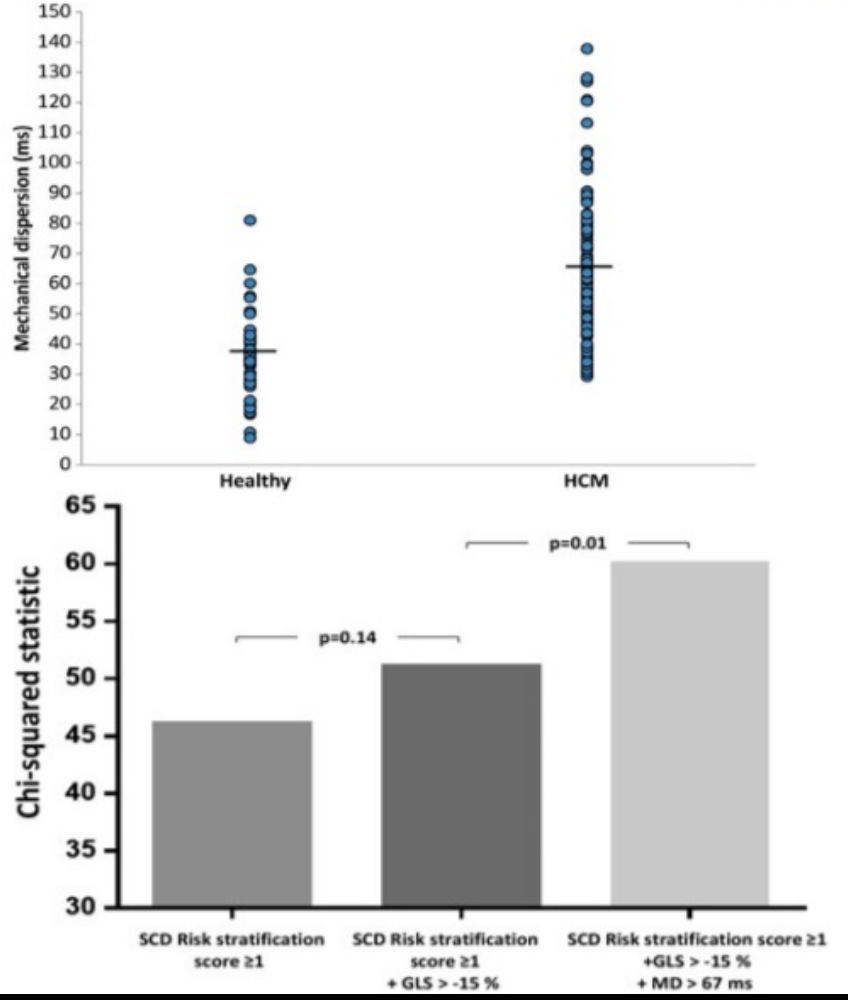
Strain echocardiography is related to fibrosis and ventricular arrhythmias in hypertrophic cardiomyopathy

Trine F. Haland[1,2,3,4], Vibeke M. Almaas[1,2,3], Nina E. Hasselberg[1,2,3,4], Jørg Saberniak[1,2,3,4], Ida S. Leren[1,2,3,4], Einar Hopp[2,3,5], Thor Edvardsen[1,2,3,4], and Kristina H. Haugaa[1,2,3,4]*



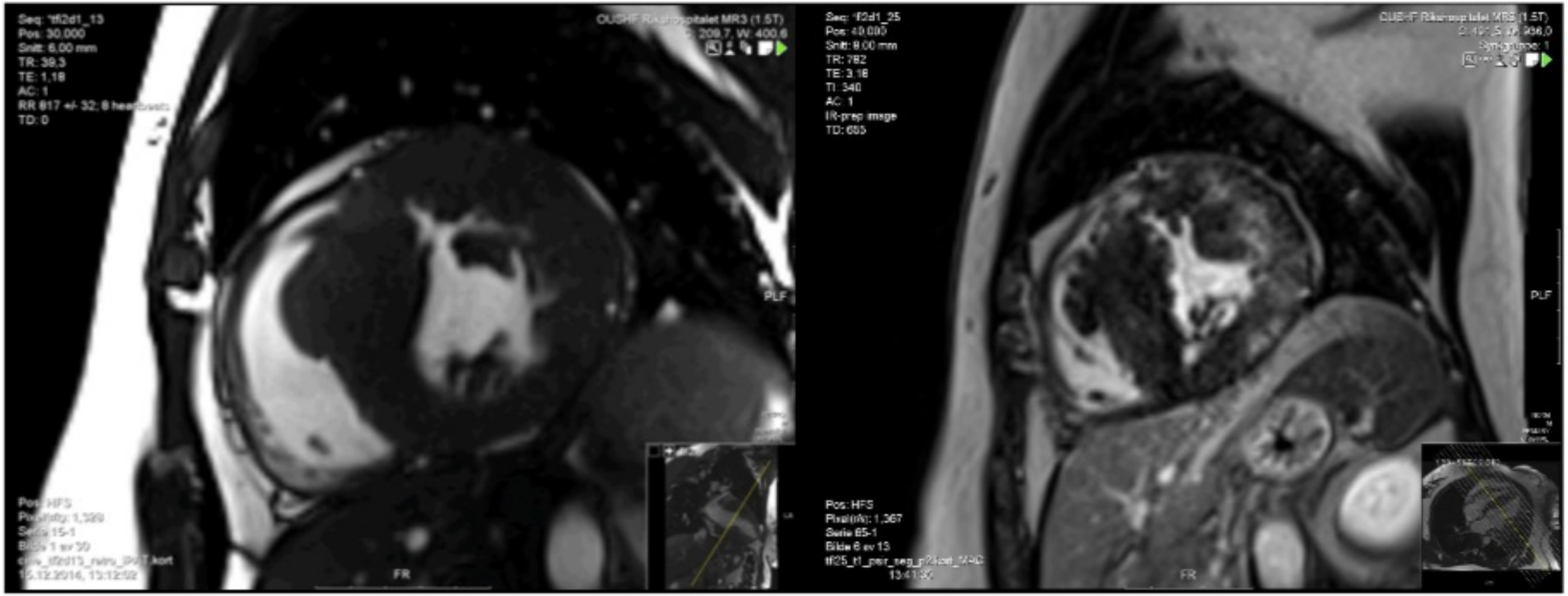
European Heart Journal – Cardiovascular Imaging
doi:10.1093/ehjci/jew005

2016

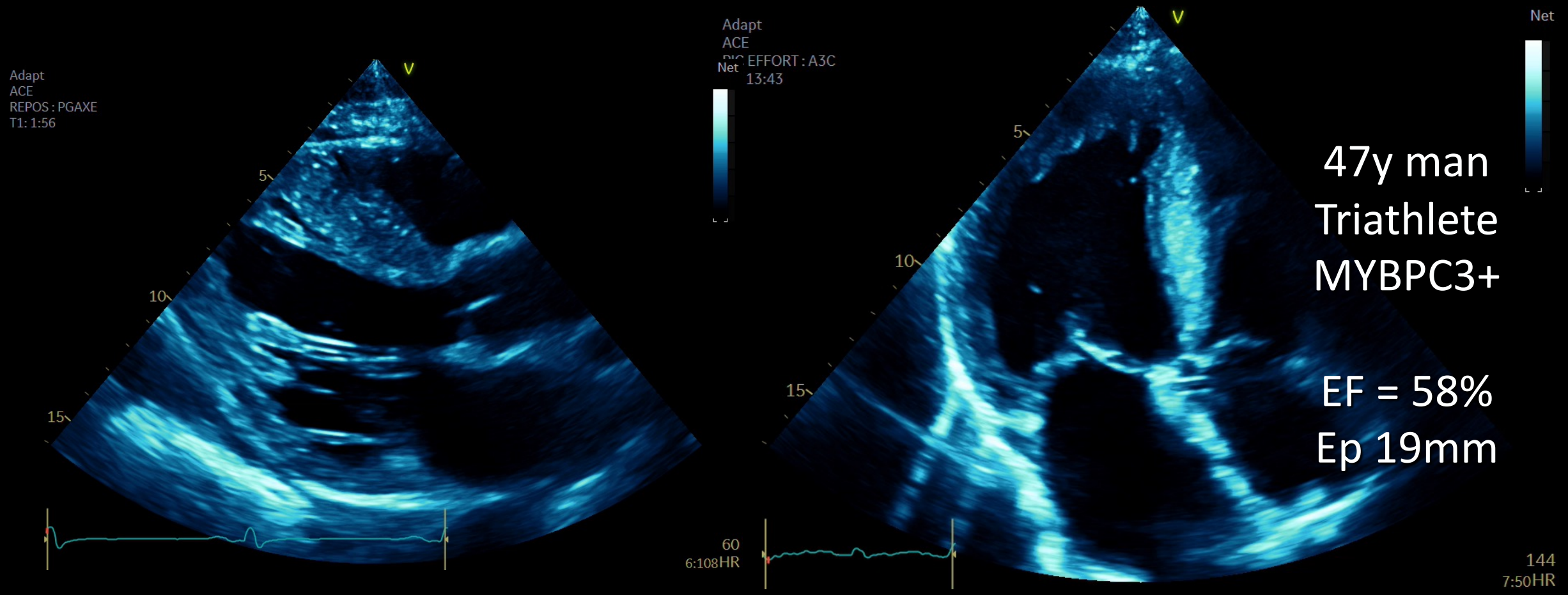


CMR IN HCM

35 y man, 3 SCD family



GENETIC TEST IN HCM



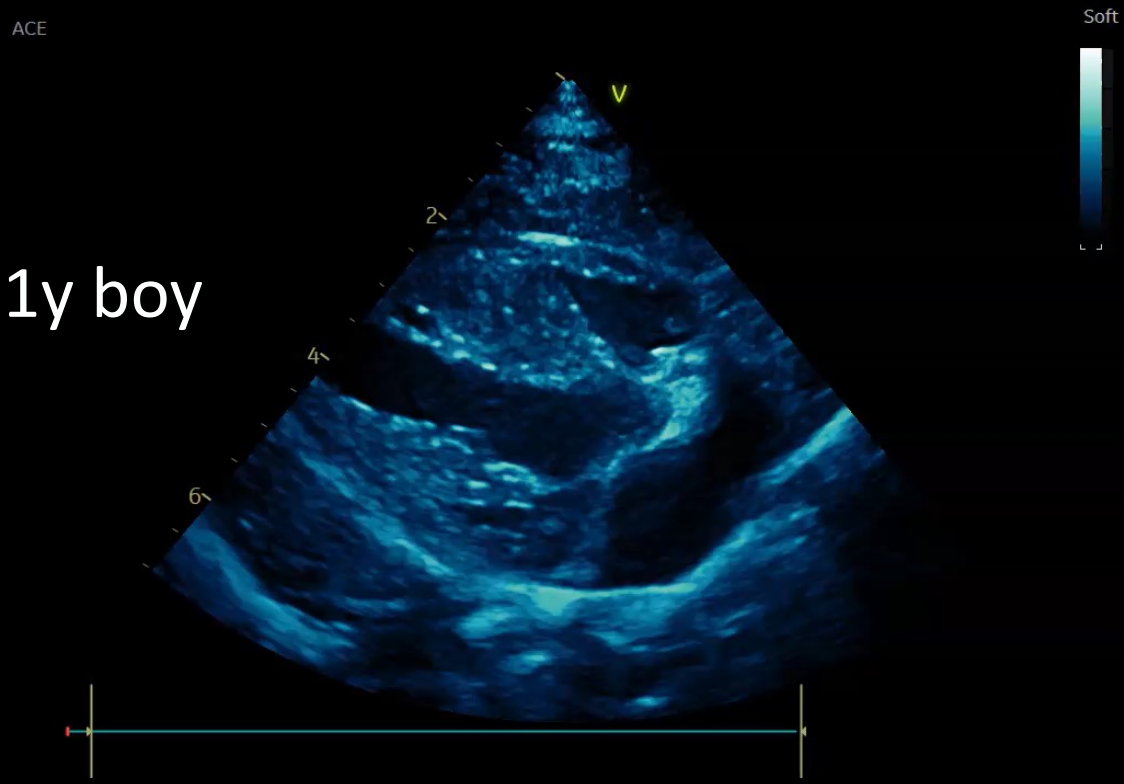
47y man
Triathlete
MYBPC3+

EF = 58%
Ep 19mm

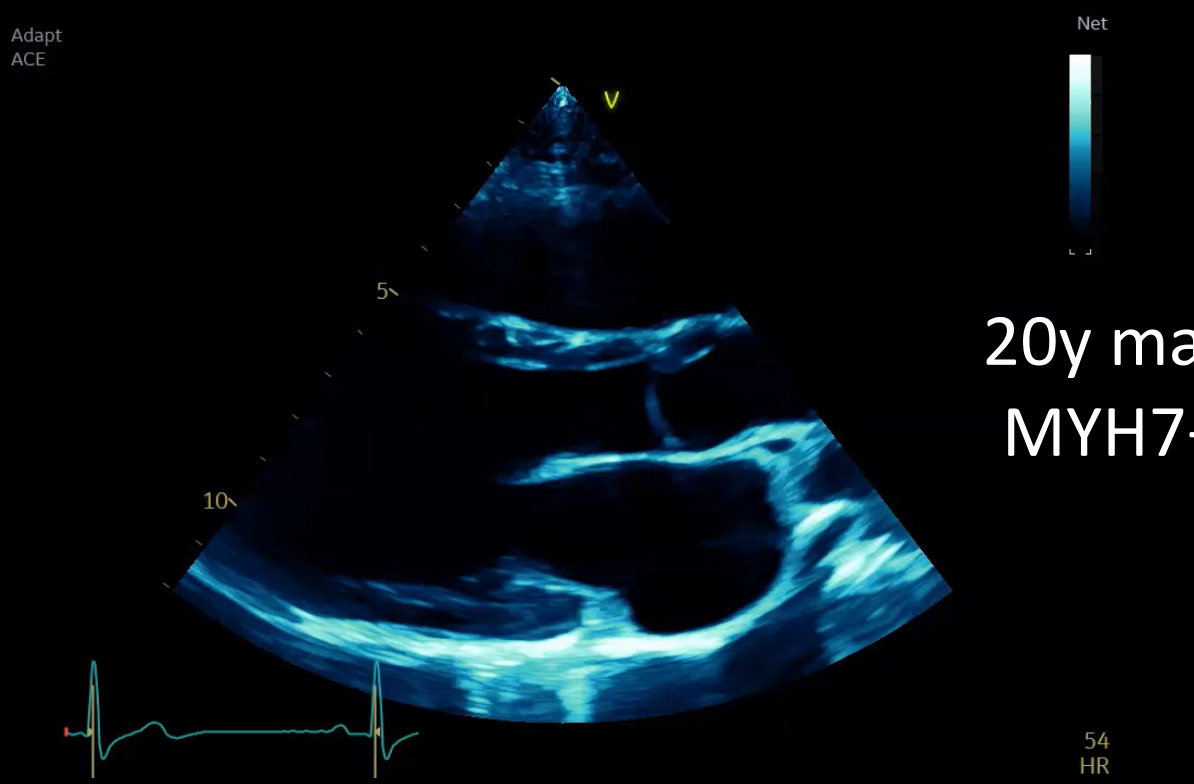
MAYO CLINIC
CARDIO X CLINIC

GENETIC TEST HAVE LIMITS

Genotype negative HCM



Genotype positive phenotype negative HCM



40% of HCM patients do not carry known pathogenic mutations

Courtesy of Dr. J. LAÏK, CANNES, FR

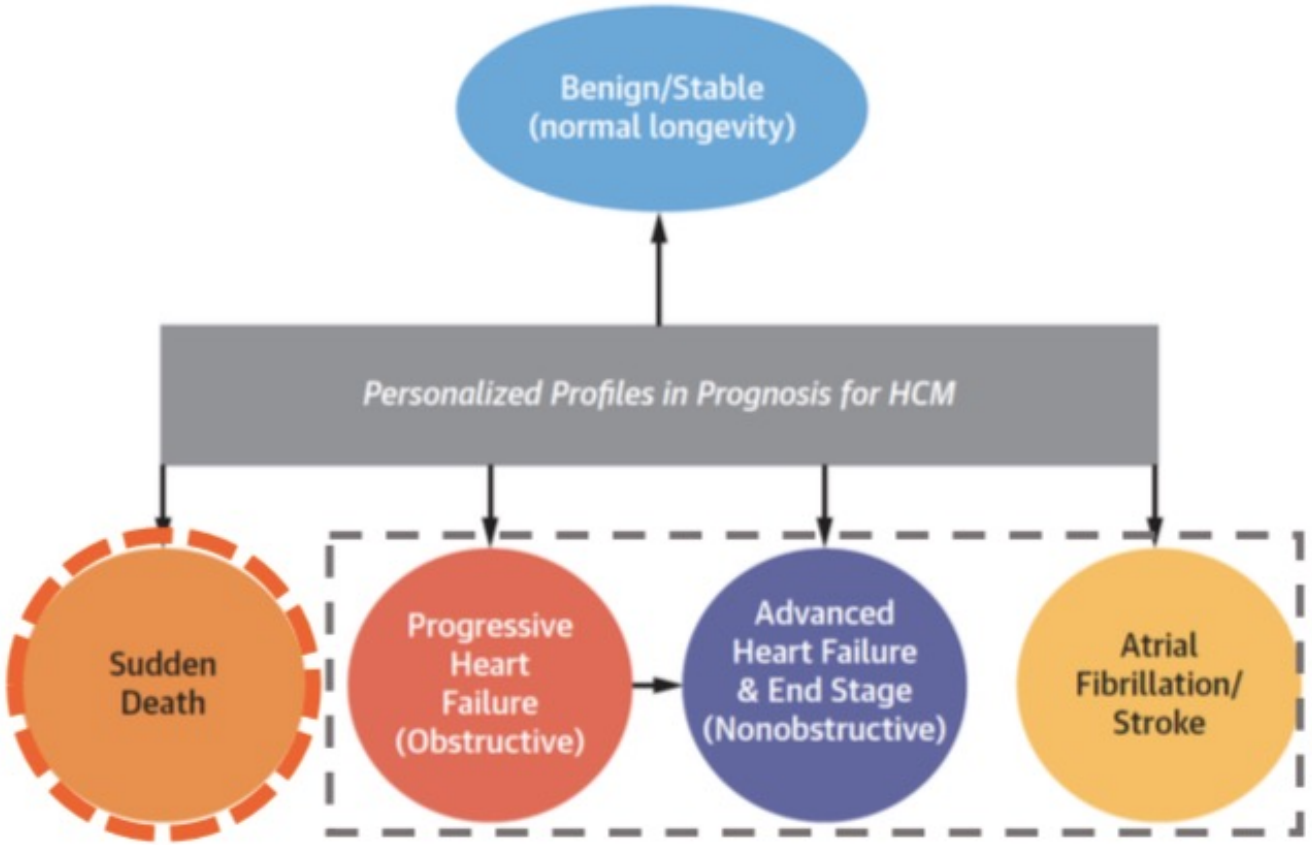
ACCA - HCM

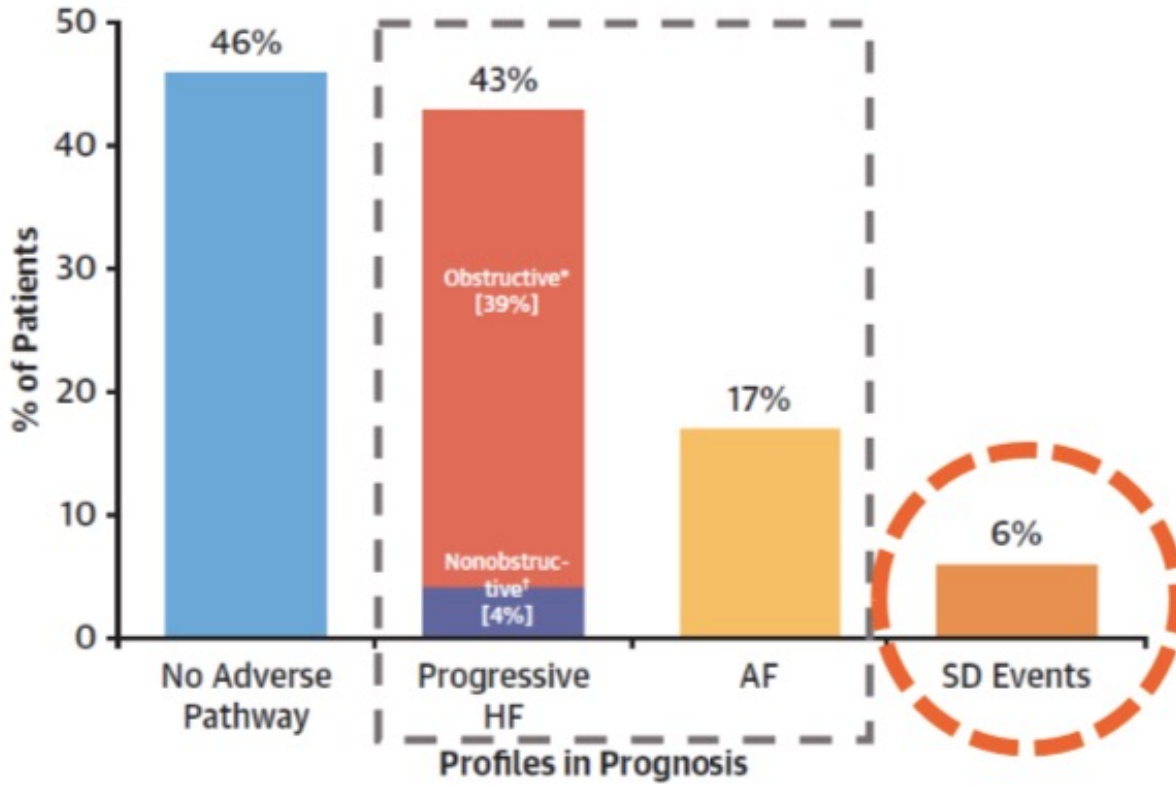
WHO DO WE ASSESS THE RISK ?

CARDIO X CLINIC

MAYO CLINIC

PROGNOSTIC PROFILS IN HCM





Maron BJ et al. J Am Coll Cardiol 2022

LOOK AT THE LEFT ATRIUM

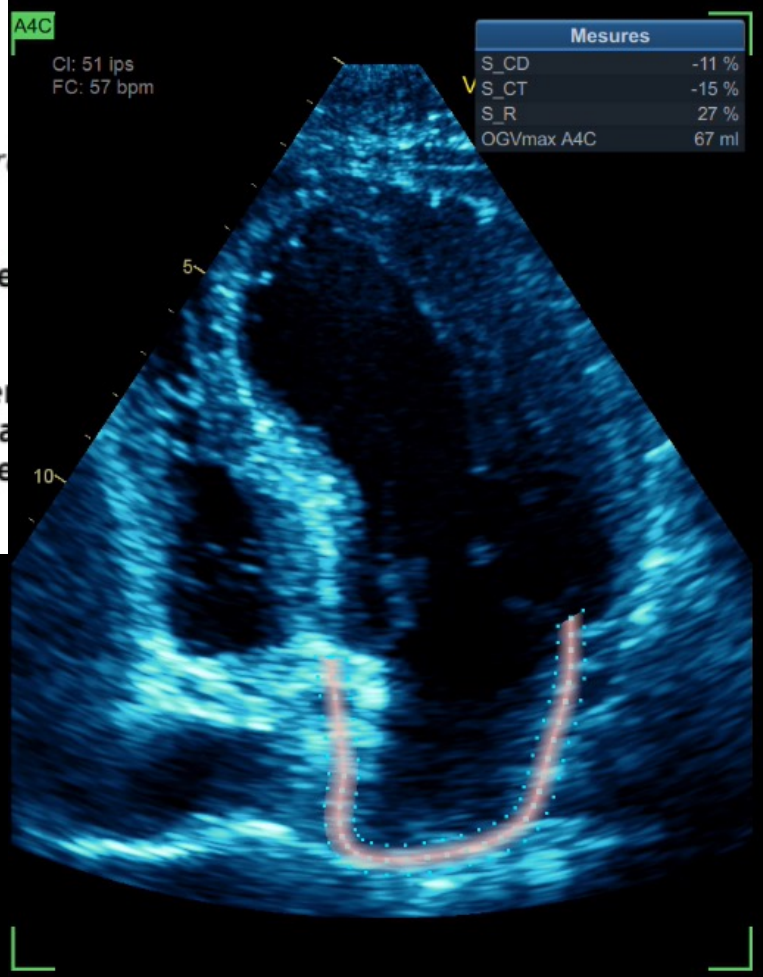
CLINICAL RESEARCH

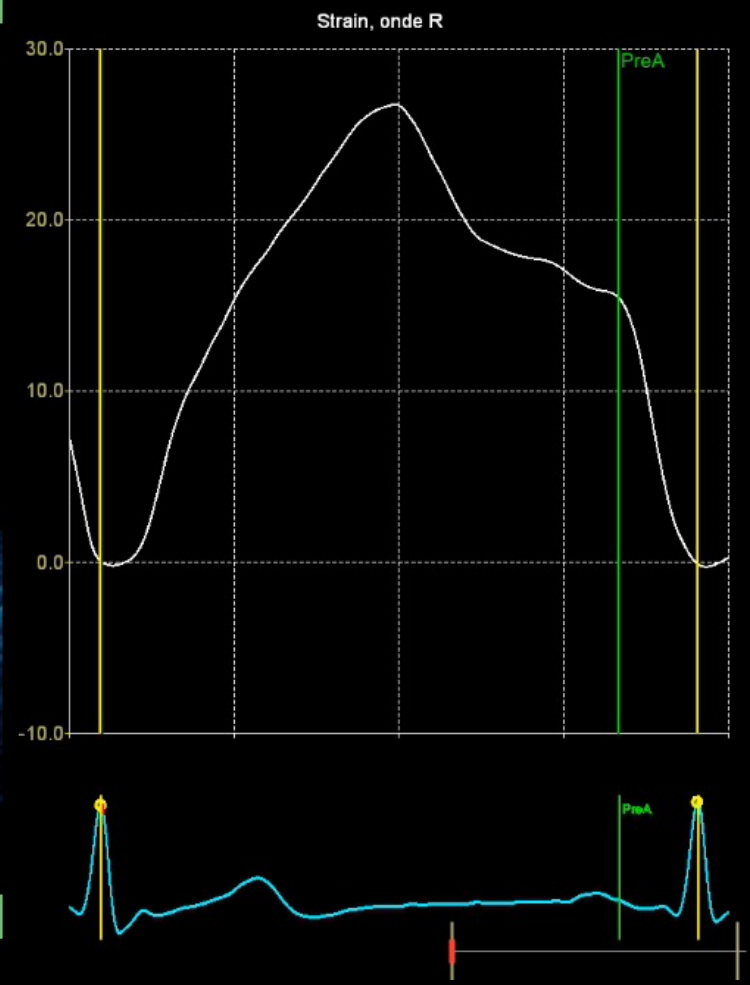
Left atrial dysfunction as marker of poor outcome in patients with hypertrophic cardiomyopathy

Valeur pronostique de la dysfonction atriale gauche dans la car hypertrophique

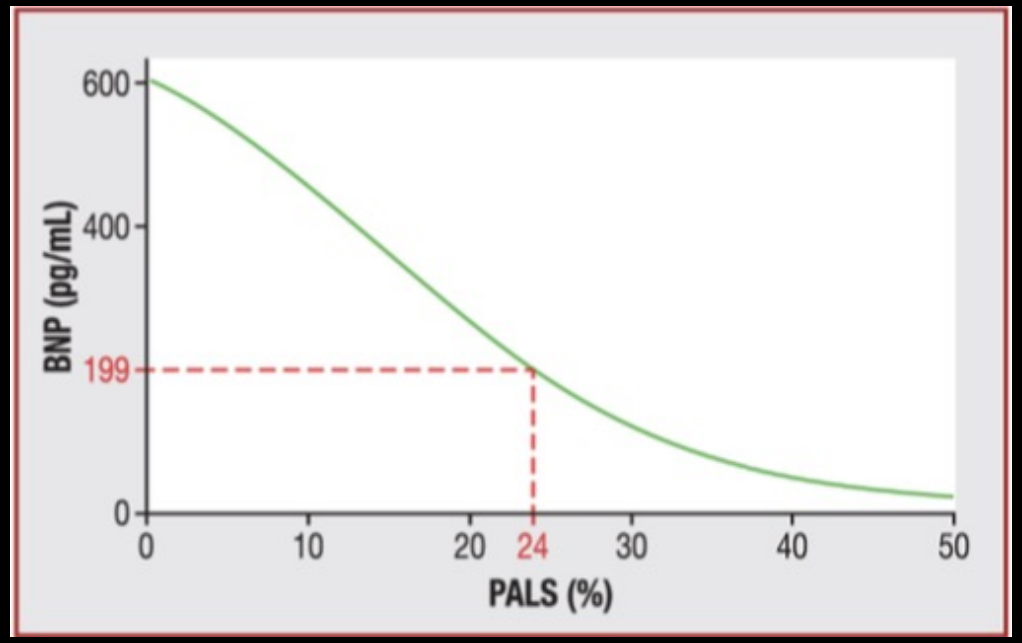
Benjamin Essayagh[a,g], Noémie Resseguie
Nicolas Michel[a], Anne-Claire Casalta[a],
Sébastien Renard[a], Valeria Donghi[a],
Andreina Carbone[a], Chiara Piazzai[a], Pie
Franck Levy[c], Hélène Martel[a], Hilla Géra
Jean-François Avierinos[a], Karine N'Guye
Gilbert Habib[a,f,*]

LA SR 27%





LOOK AT THE LEFT ATRIUM



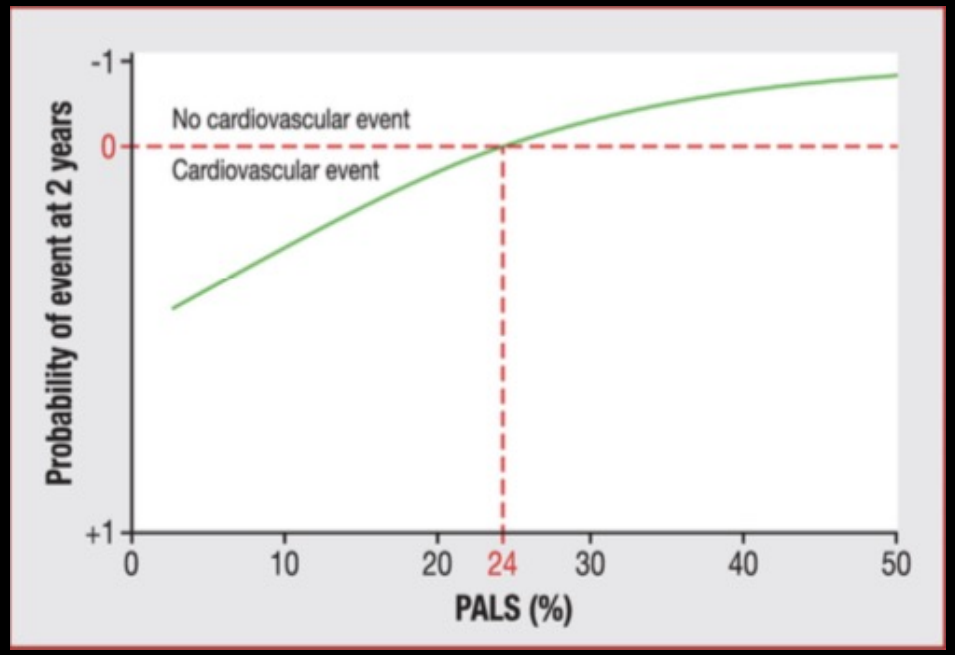
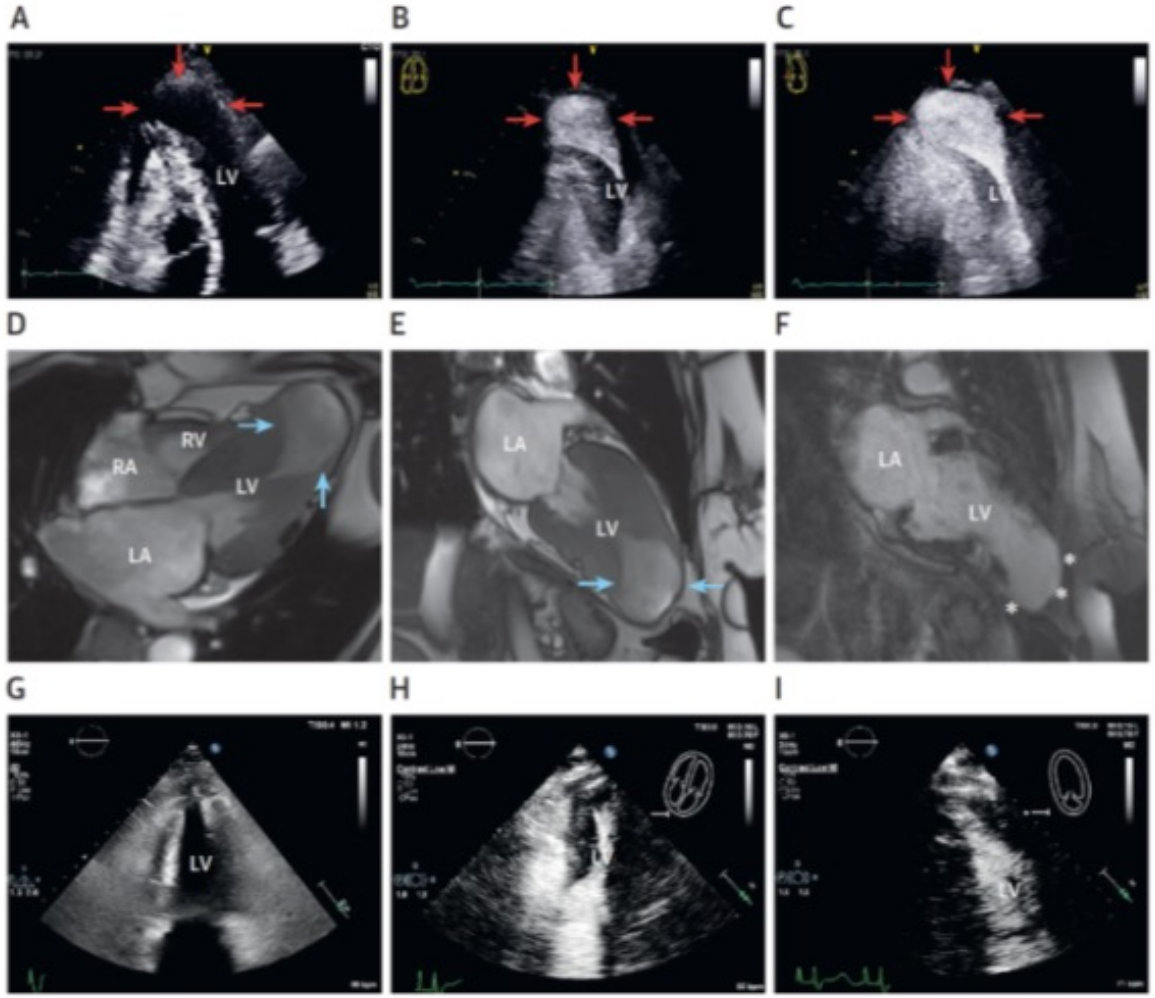


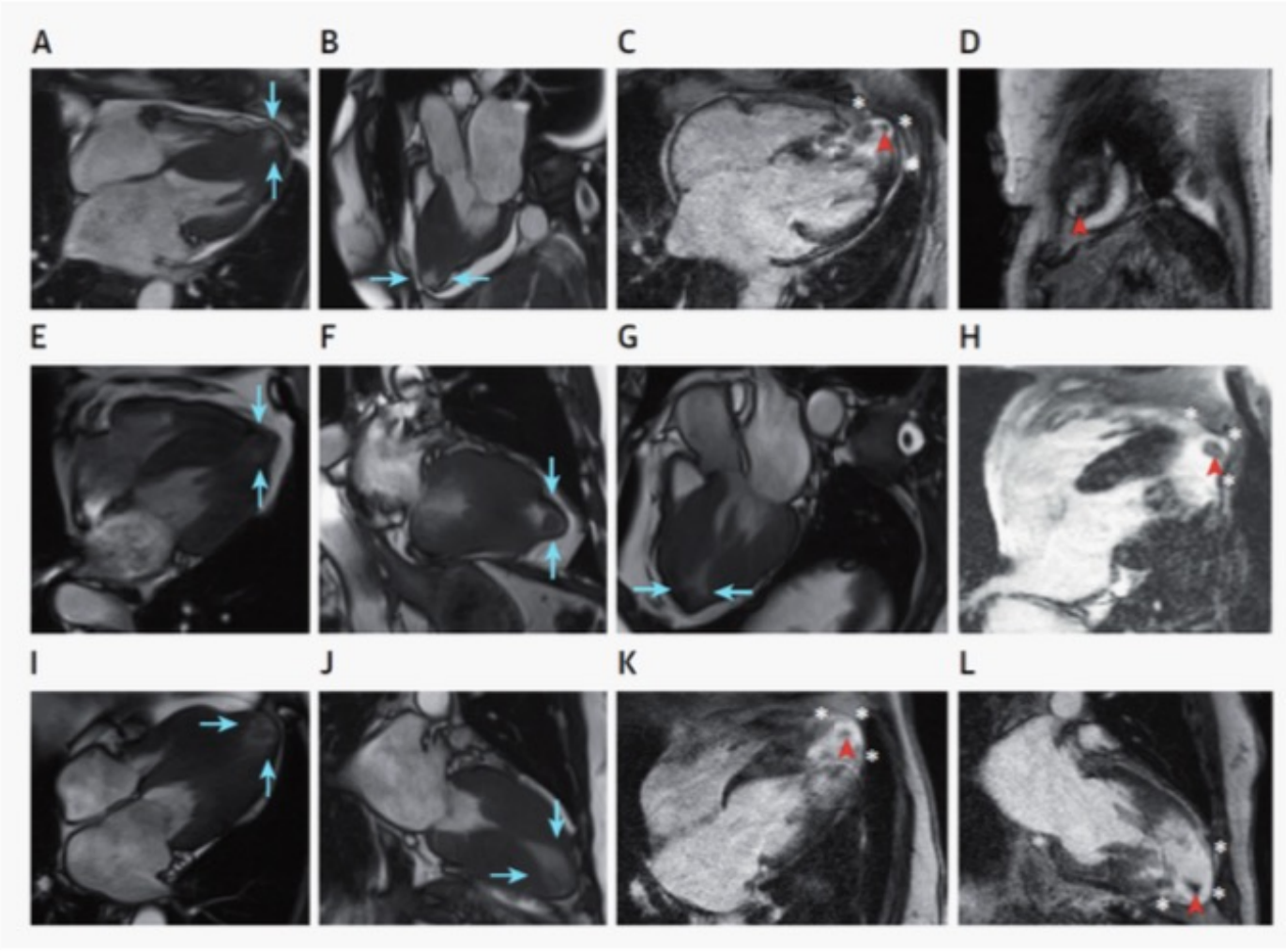
Table 3 Multivariable predictors of hypertrophic cardiomyopathy-related events and mortality.

	Univariate analysis		Multivariable analysis[a]	
	HR (95% CI)	*P*	HR (95% CI)	*P*
Echocardiographic variables				
Maximal LV thickness	1.12 (1.09–1.13)	<0.0001	1.13 (1.00–2.28)	0.04
LV global longitudinal strain	1.24 (1.12–1.37)	<0.0001	1.06 (0.88–1.28)	0.5
MR	2.10 (1.18–3.74)	0.02	1.45 (0.40–5.26)	0.6
PALS	0.94 (0.92–0.97)	<0.0001	0.86 (0.79–0.94)	0.0008

CARDIO X CLINIC MAYO CLINIC

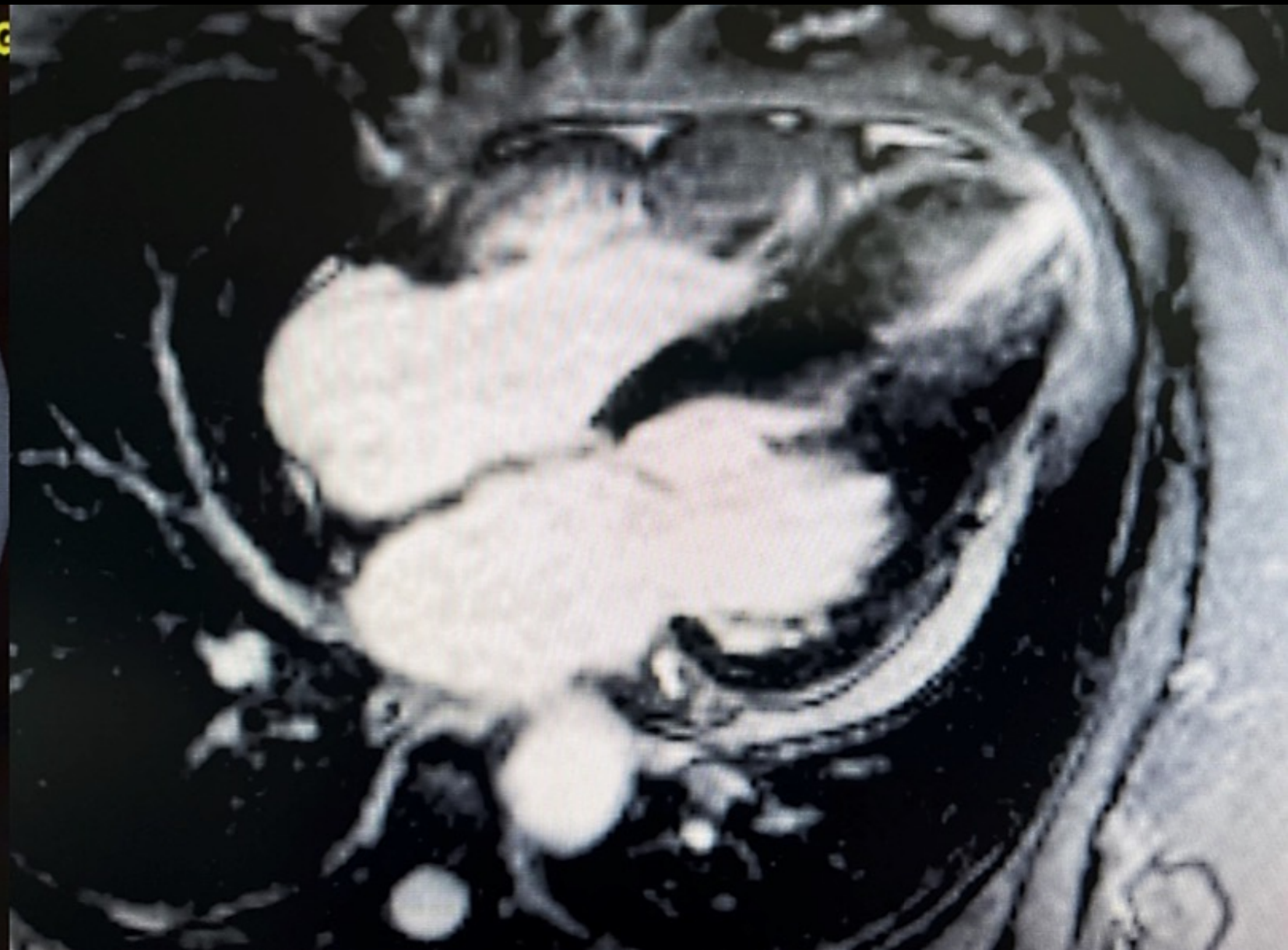
APICAL ANEURYSM IN HCM





Lee DZJ et al. Jacc Img 2022

APICAL ANEURYSM IN HCM



SCD RISK STRATIFICATION

HCM Risk-SCD Calculator

Age	**Years**	*Age at evaluation*
Maximum LV wall thickness	**mm**	*Transthoracic Echocardiographic measurement*
Left atrial size	**mm**	*Left atrial diameter determined by M-Mode or 2D echocardiography in the parasternal long axis plane at time of evaluation*
Max LVOT gradient	**mmHg**	*The maximum LV outflow gradient determined at rest and with Valsalva provocation (irrespective of concurrent medical treatment) using pulsed and continuous wave Doppler from the apical three and five chamber views. Peak outflow tract gradients should be determined using the modified Bernouilli equation: Gradient= $4V^2$, where V is the peak aortic outflow velocity*
Family History of SCD	○ No ○ Yes	*History of sudden cardiac death in 1 or more first degree relatives under 40 years of age or SCD in a first degree relative with confirmed HCM at any age (post or ante-mortem diagnosis).*
Non-sustained VT	○ No ○ Yes	*3 consecutive ventricular beats at a rate of 120 beats per minute and <30s in duration on Holter monitoring (minimum duration 24 hours) at or prior to evaluation.*
Unexplained syncope	○ No ○ Yes	*History of unexplained syncope at or prior to evaluation.*

Risk of SCD at 5 years (%):	
ESC recommendation:	

Reset

2014 ESC Guidelines on Diagnosis and Management of Hypertrophic Cardiomyopathy (Eur Heart J 2014 – doi:10.1093/eurheartj/ehu284)

O'Mahony C et al Eur Heart J (2014) 35 (30): 2010-2020

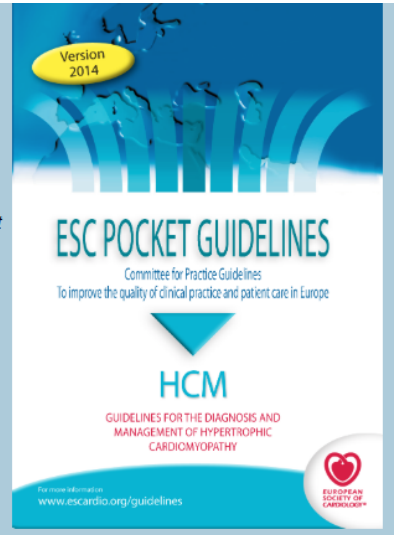
HCM Risk-SCD should not be used in:

- ***Paediatric patients (<16 years)***
- ***Elite/competitive athletes***
- ***HCM associated with metabolic diseases (e.g. Anderson-Fabry disease), and syndromes (e.g. Noonan syndrome).***
- ***Patients with a previous history of aborted SCD or sustained ventricular arrhythmia who should be treated with an ICD for secondary prevention.***

Caution should be exercised when assessing the SCD in patients following invasive reduction in left ventricular outflow tract obstruction with myectomy or alcohol septal ablation.

Pending further studies, HCM-RISK should be used cautiously in patients with a maximum left ventricular wall thickness ≥35 mm.

HCM = hypertrophic cardiomyopathy; LV = left ventricular; LVOT = left ventricular outflow tract; NSVT = non-sustained ventricular tachycardia; SCD = sudden cardiac death; VT = ventricular tachycardia



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SCD RISK STRATIFICATION

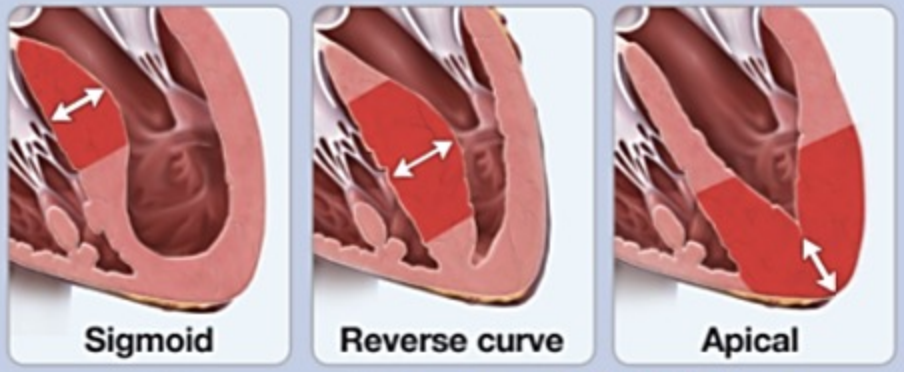
Goals of Echocardiographic Assessment in Hypertrophic Cardiomyopathy (HCM)

Establish diagnosis & determine pattern of hypertrophy

Clinical diagnosis should be suspected with imaging evidence of a maximal end-diastolic wall thickness of >15 mm anywhere in the left ventricle, absent another cause of hypertrophy in adults

Differentiate sigmoid septum (with ovoid cavity) versus reverse curve (with crescent cavity) versus apical hypertrophic phenotypes

Massive left ventricular hypertrophy >30 mm in any left ventricular segment is a risk factor for sudden cardiac death (SCD)



Evaluate global myocardial function

Systolic dysfunction defined as LVEF <50%

Strain abnormalities correlate with increased wall thickness & delayed gadolinium enhancement by MRI

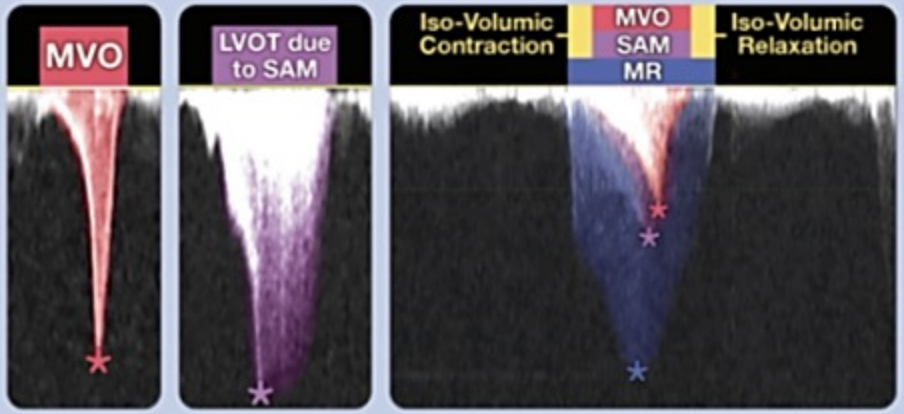
Establish presence & severity of LVOT obstruction

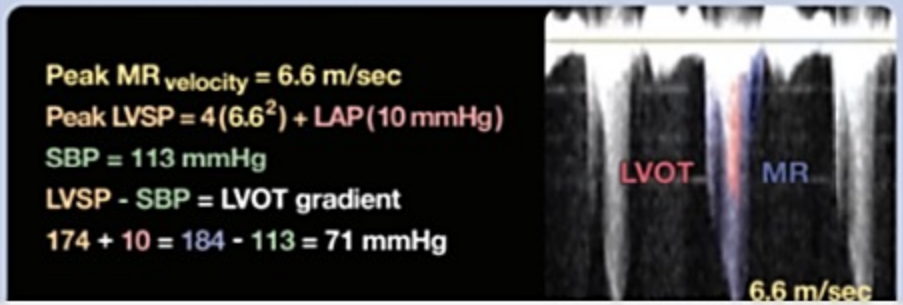
Peak LVOT gradient of ≥50 mmHg at rest or with provocation or exercise indicates obstruction

Differentiate SAM-mediated LVOT obstruction from mid-ventricular obstruction (MVO; "dagger" shaped)

Caution with contamination of LVOT signal with MR. MR velocity is higher & signal is of longer duration (spanning isovolumic contraction & relaxation) vs LVOT signal. MR contour may be incomplete if Doppler signal not optimally aligned

Estimated LVOT gradient from MR signal calculated as: LV Pressure - Systolic BP, where





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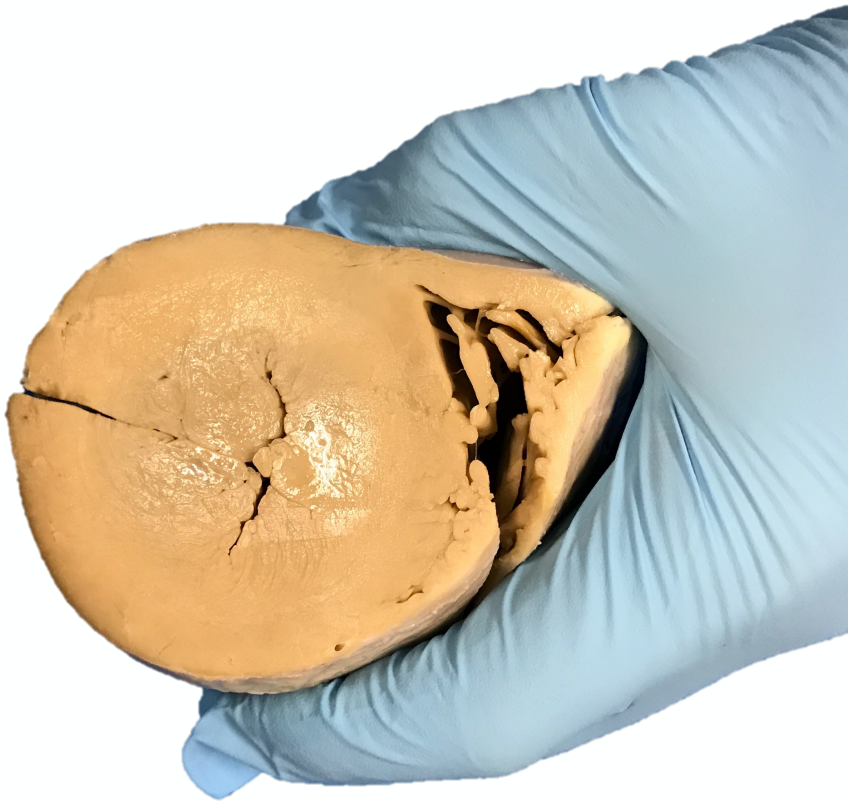

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