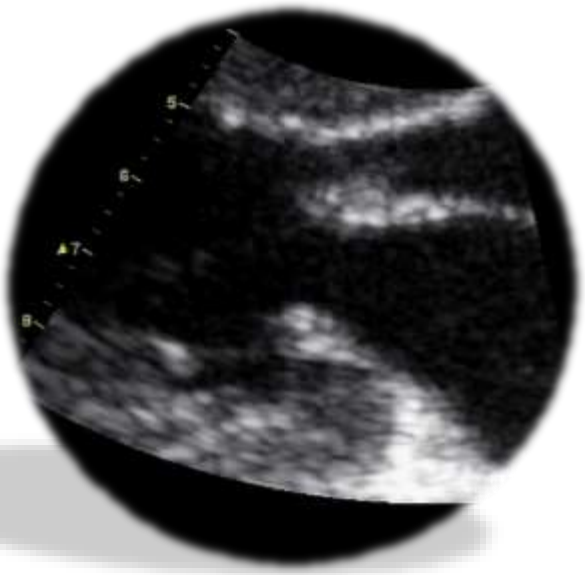
STRASBOURG 2024

CMH
Le cœur haltéré

Modérateurs : Marion CHATOT – Pierre LEDDET

GANG

STRASBOURG 2024



OPEN BAR SUR LE SEPTUM

Olivier HUTTIN

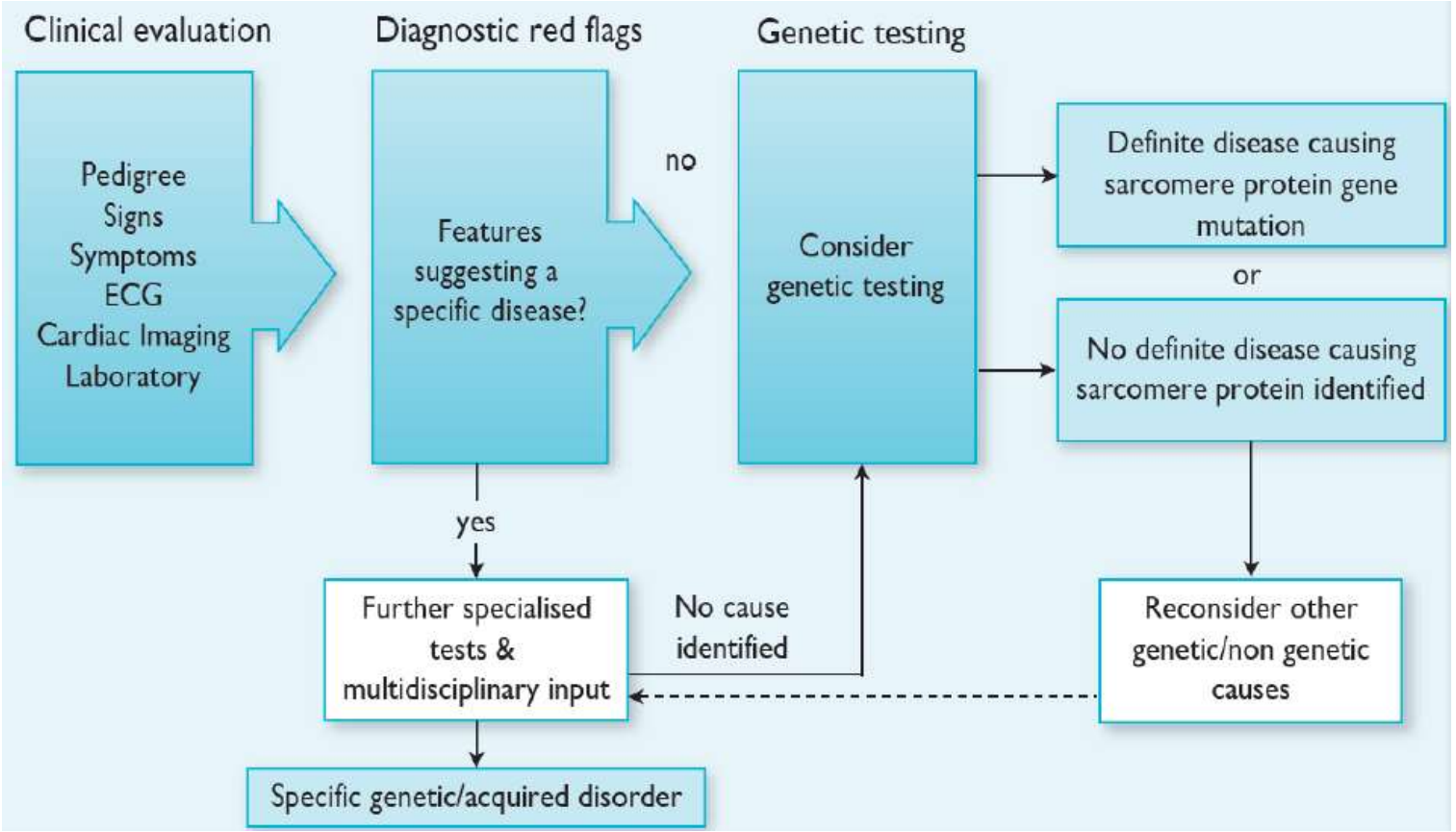
“SUB” PHENOTYPE PHENOCOPIES

Sarcomeric *Non Sarcomeric*

HCOM **NOCM** **APICAL ANEURYSM** **wTTR CA** **AL CA**

Hypertrophic Septum > Diagnosis red flags

CLINICAL CHARACTERISTICS



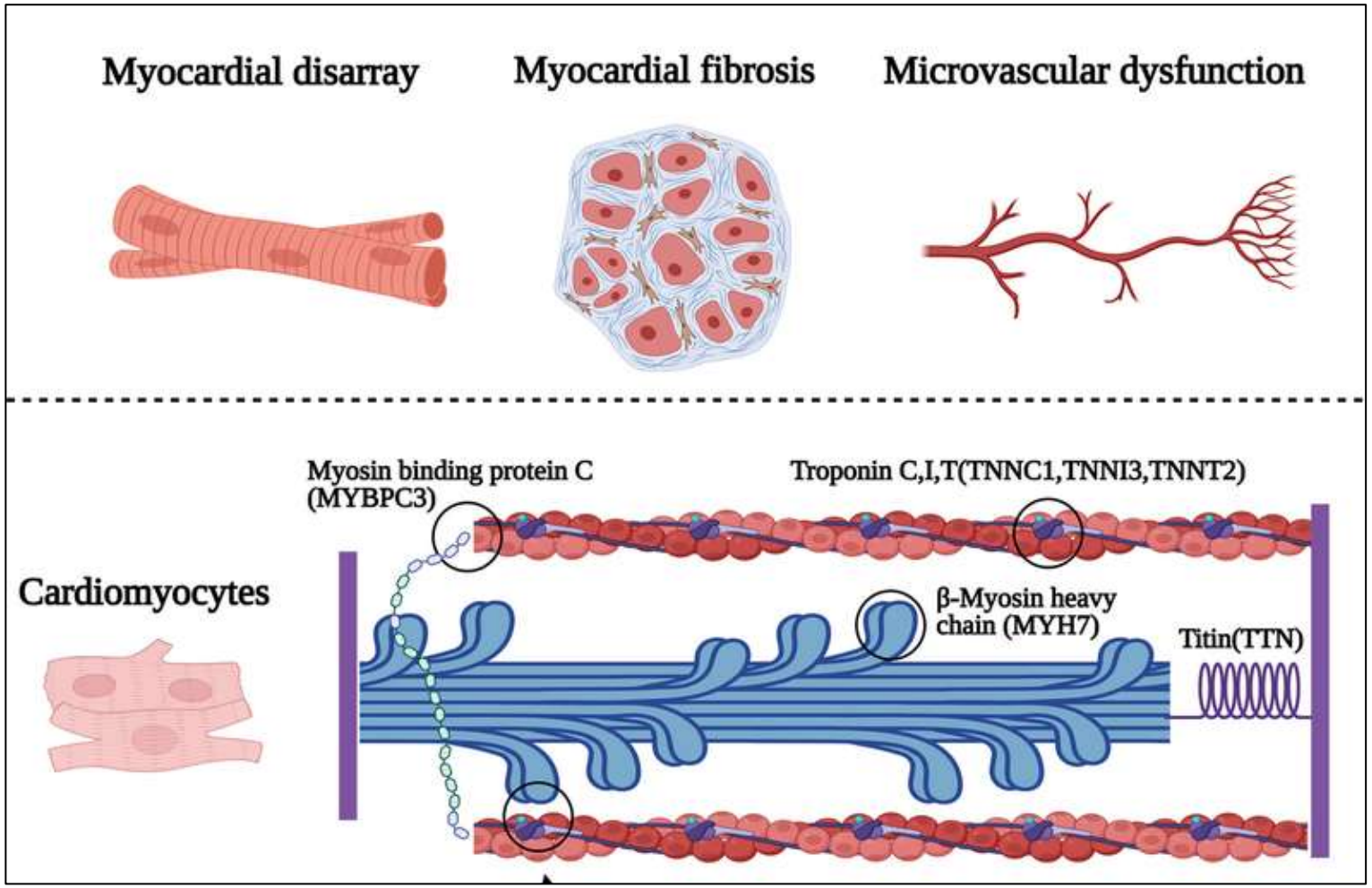
CIRCULATING BIOMARKERS

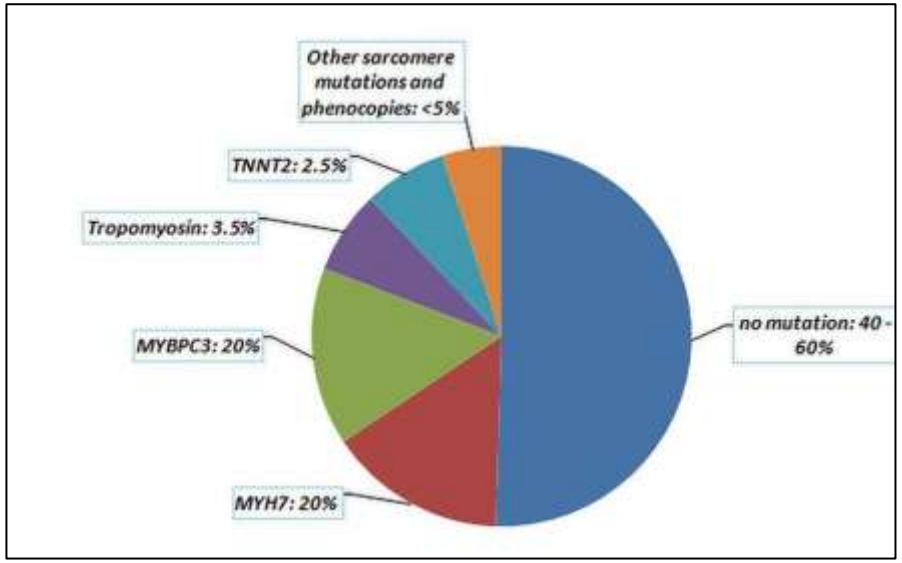
MULTIMODALITY CARDIAC IMAGING

GENETIC TESTING

Elliott PM Eur Heart J. 2014 Oct 14;35(39):2733-79.

Hypertrophic cardiomyopathy

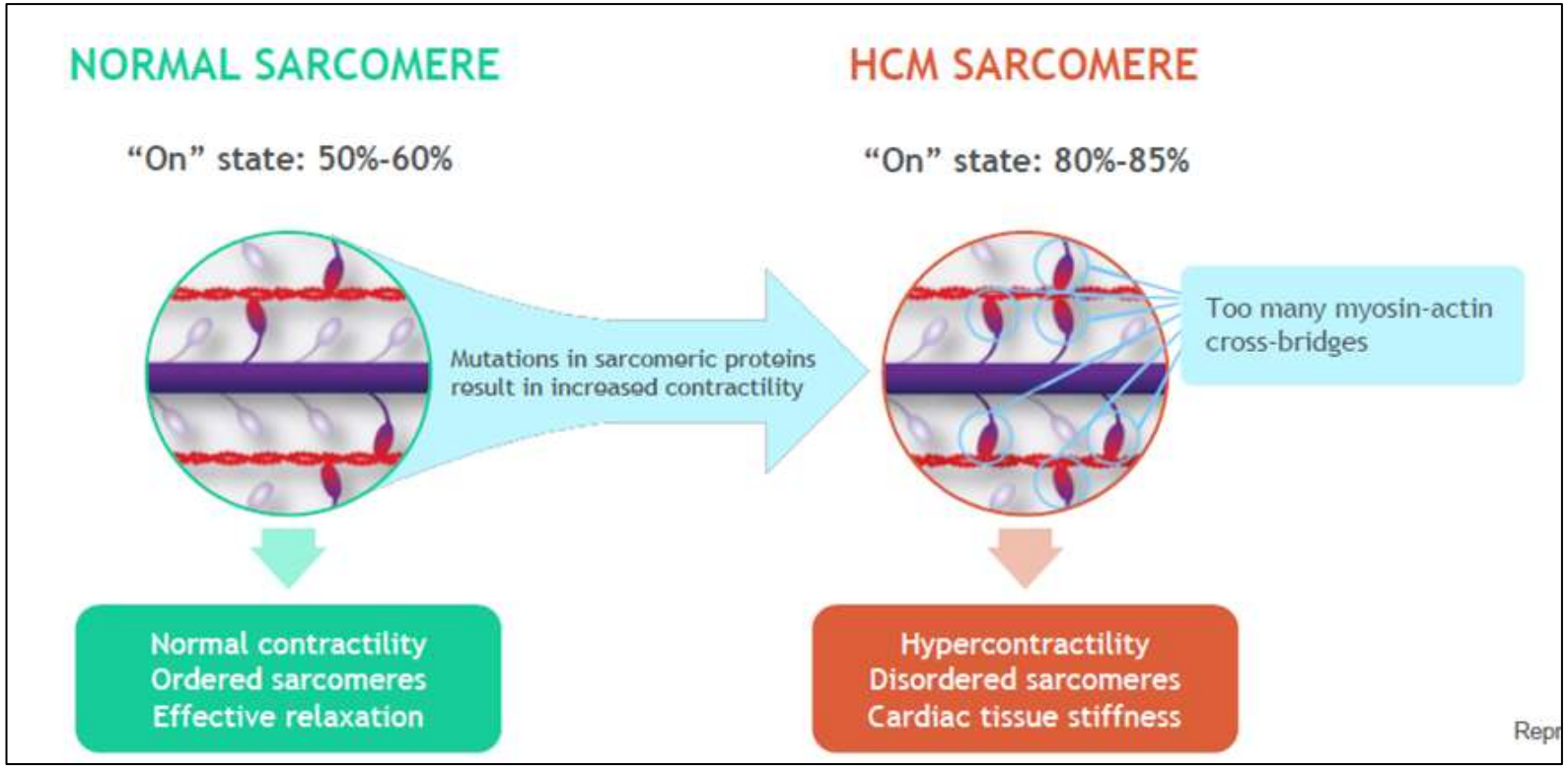




1. Septal hypertrophy
2. LV obstruction
3. Myocardial fibrosis

Wall Thickness

Obstruction



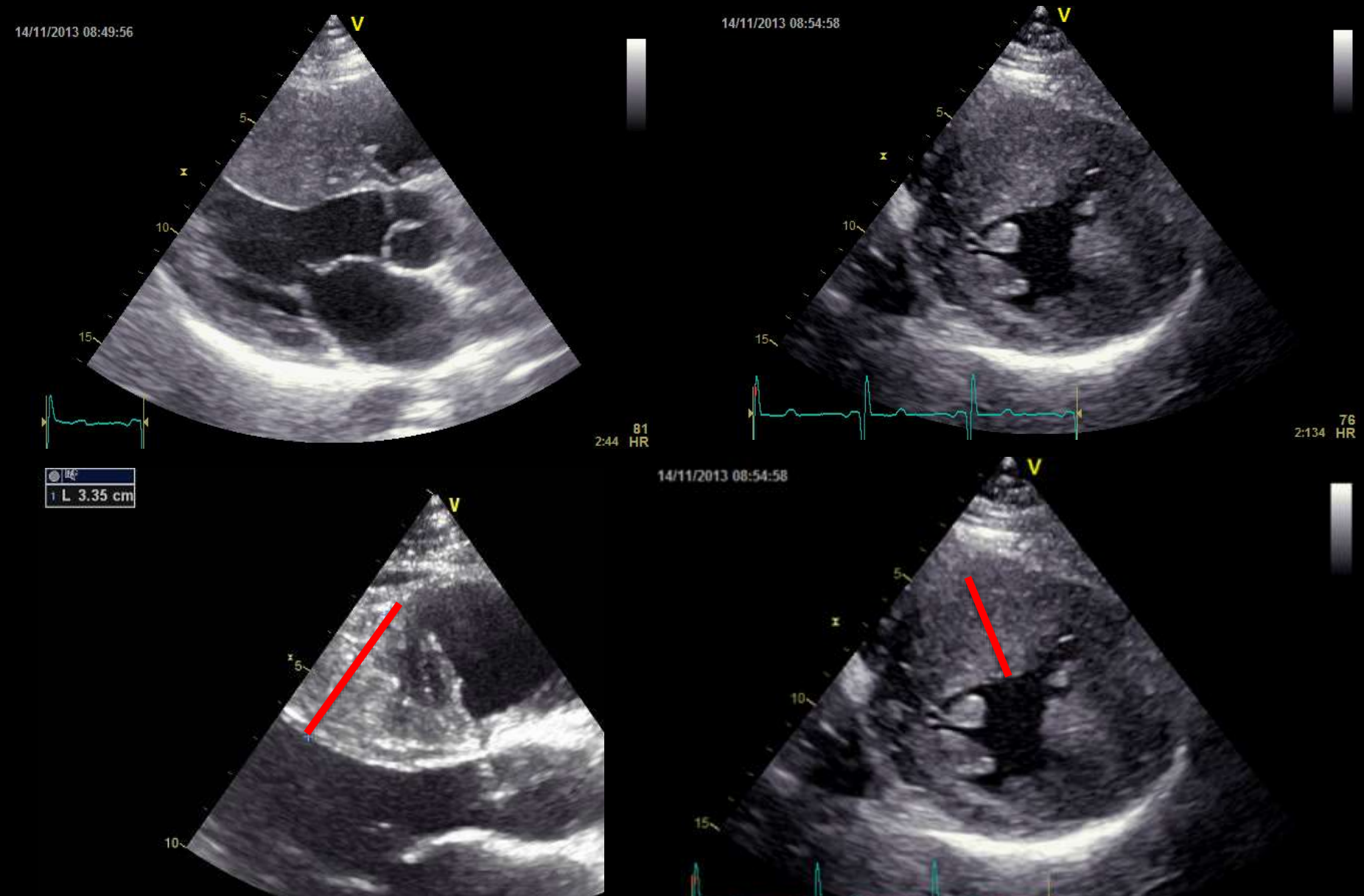
Wall thickness

- Involved segments and maximal thickness (consider contrast echo); septal:posterior wall ratio
- Consider asymmetric septal hypertrophy and septal morphology, concentric, midventricular, and apical variants
- Consider RV hypertrophy

Left ventricular outflow tract obstruction

- Mechanism, provocable vs fixed obstruction
- Level of obstruction (consider midventricular obstruction)
- Presence and severity at rest and under provocative manoeuvres: Valsalva, standing (obstructive, provocable obstructive, or non-obstructive HCM)

Septum ≠ Maximal Wall thickness

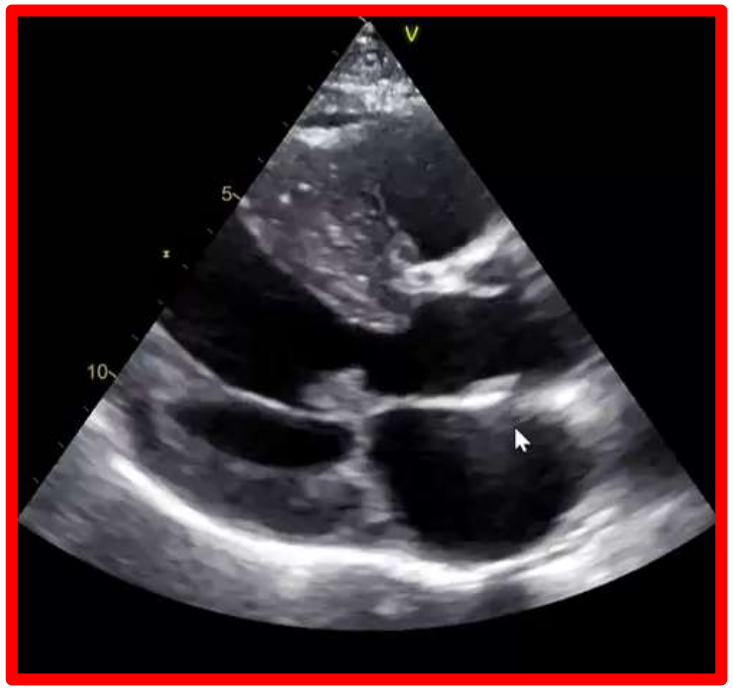


Septum How to ?

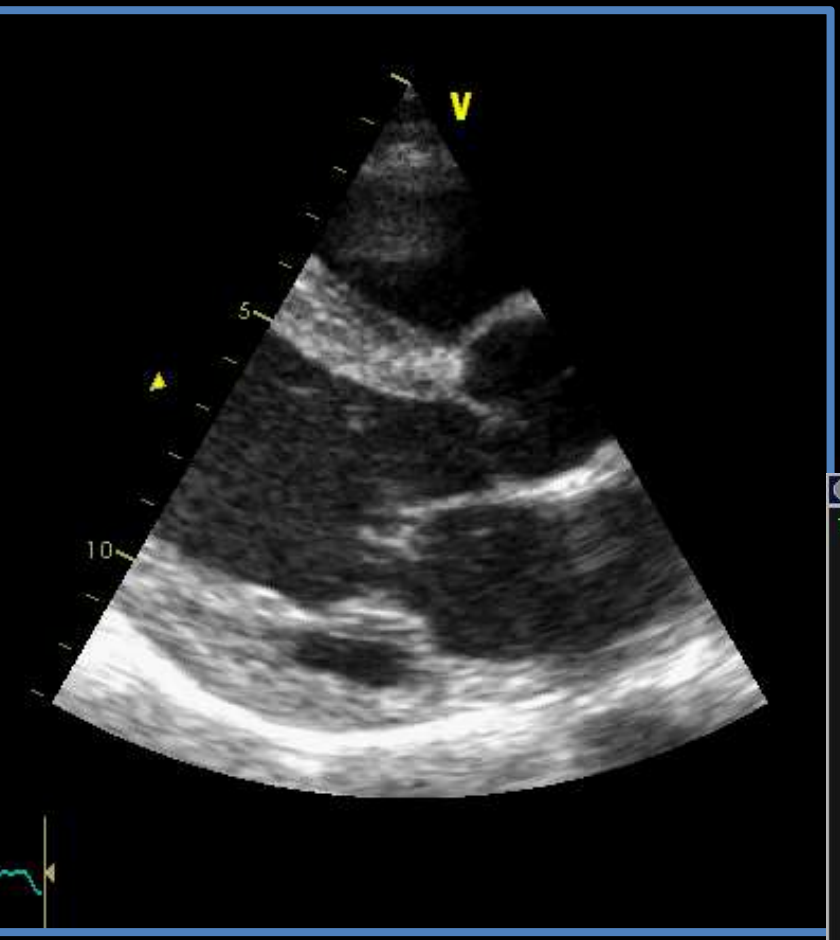
1. Parasternal view
2. Zoom
3. Diastoly

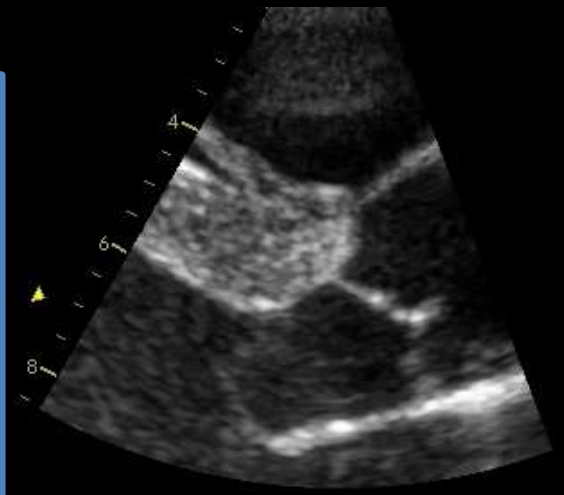
4. Confirmation in Short axis

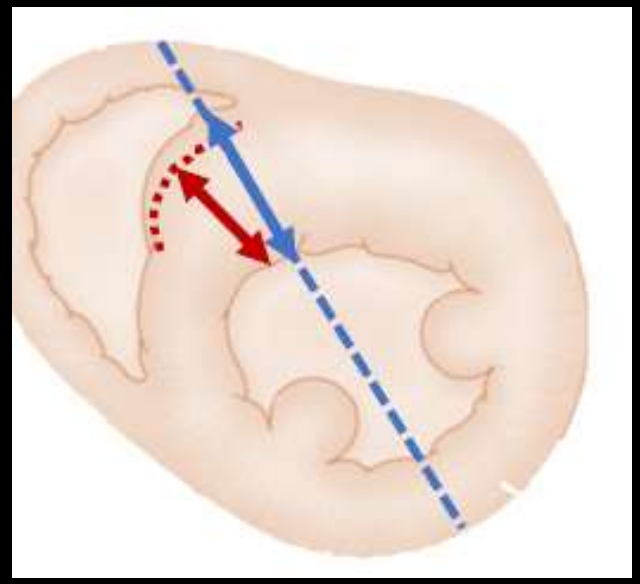
5. 16 segments measurements

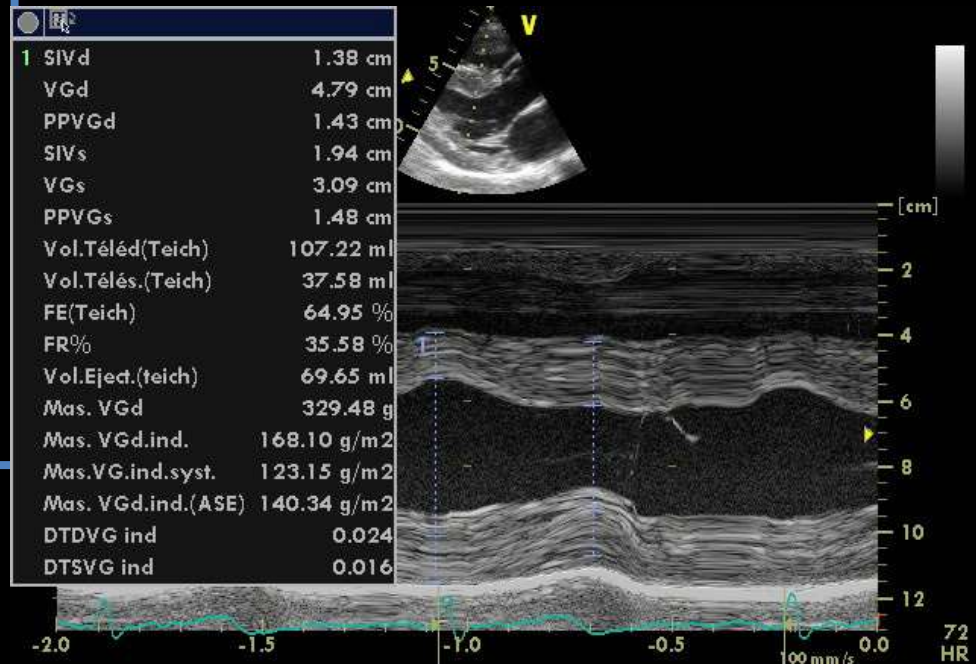


SEPTUM: Tip and tricks



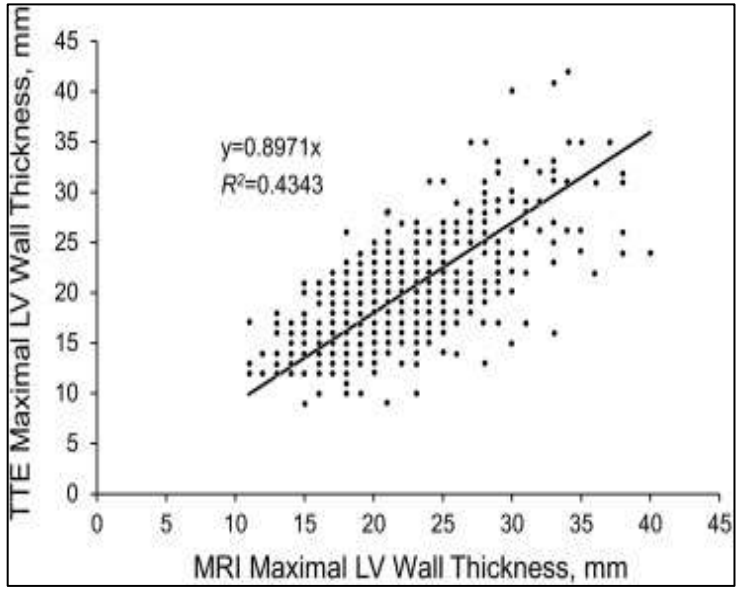




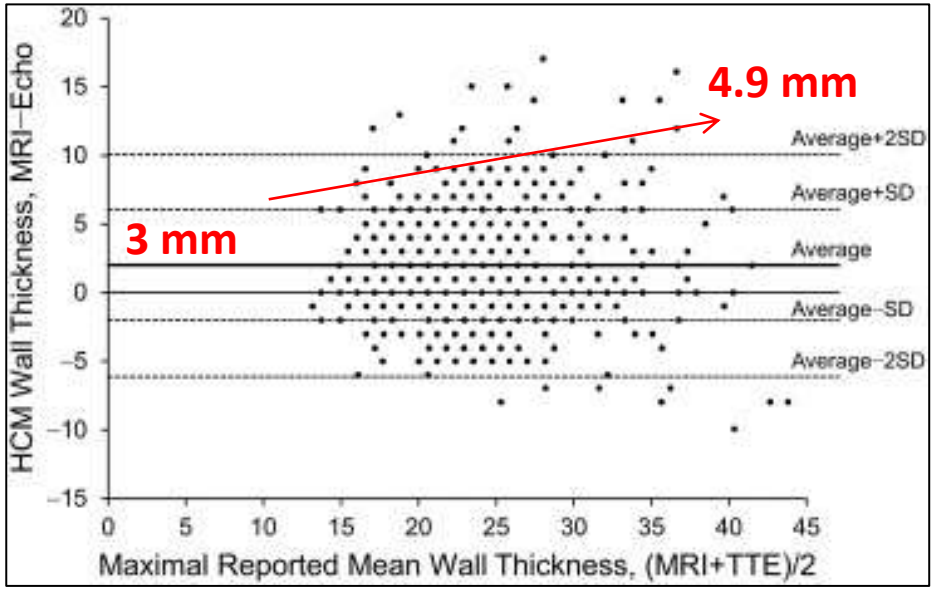


According to cardiac imaging

CMR vs TTE

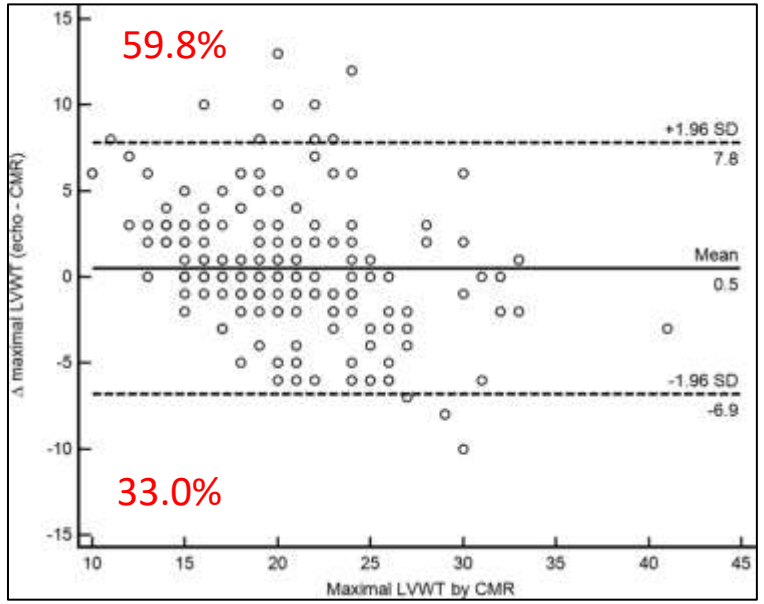


CMR > TTE



TTE>CMR

- *RV/LV trabeculations*
- *obliquity planes*



CMR>TTE

- erroneous myocardial delineation
- failure to visualize maximal LVH

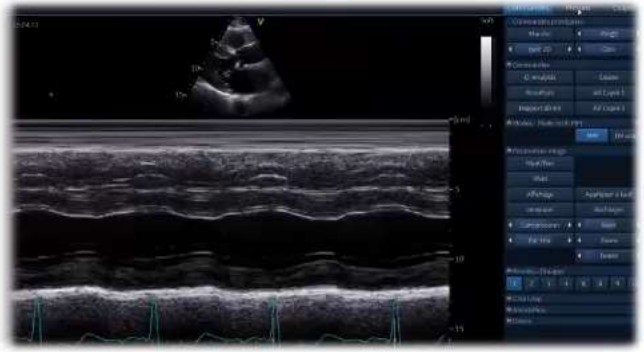
John P. Bois The American Journal of Cardiology, Volume 119, Issue 4, 2017, 643–650

Waseem Hindieh, Circ Cardiovasc Imaging. 2017

Implement : M-Mode / 2D / 3D / Multiplan / Auto

2D LV measure

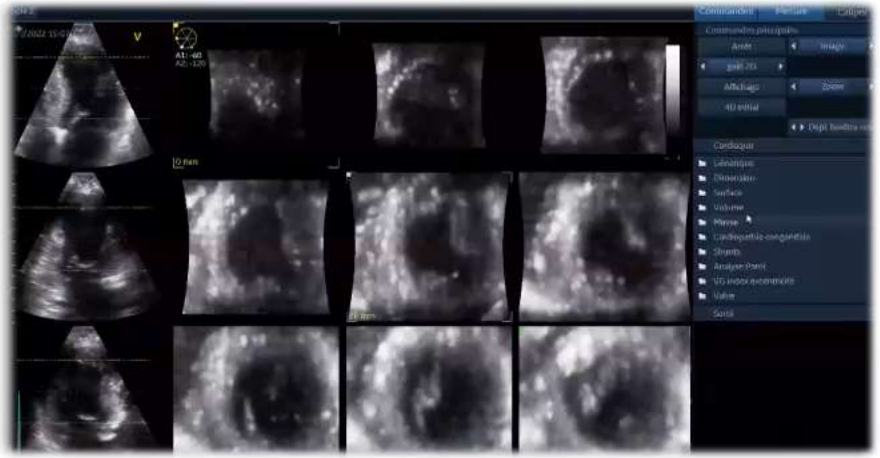
M-mode LV measure



Anatomic M Mode

Multiplan LV measure

3D LV mass

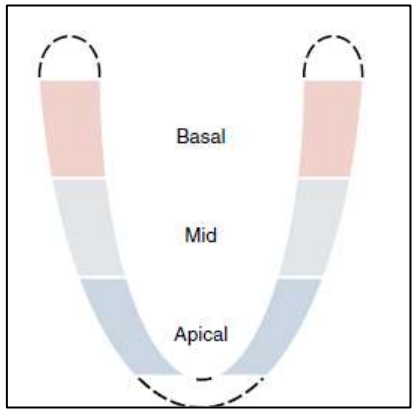


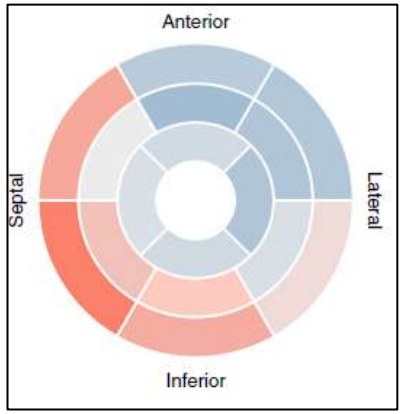
Artificial intelligency

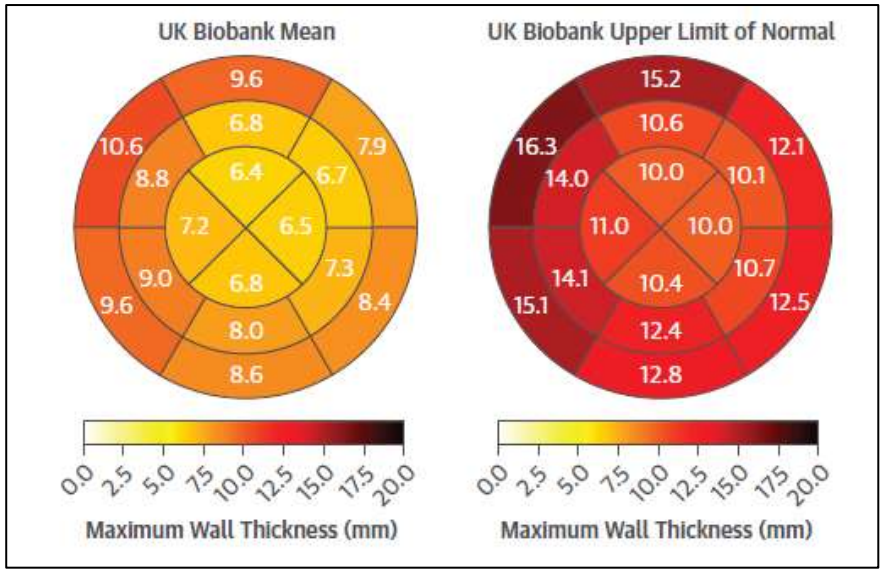
2DLV Auto measure

Normal Wall Thickness ?

- > 2 DS compared to mean value for age and weight
- LV wall thickness
  - > 15mm
  - > 13mm (familly history)
  - Less in case of apical HCM





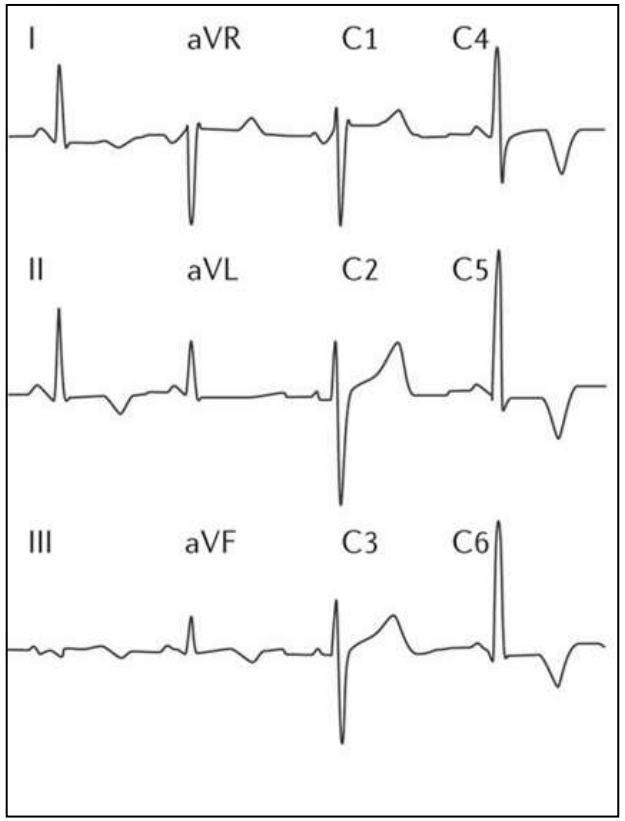


- AHA-ACC: Wall thickness ≥ 30 mm = SCD risk
- ESC: Wall thickness is included in the 5-year SCD risk predictor

Hughes RK, et al. J Am Coll Cardiol Img. 2023

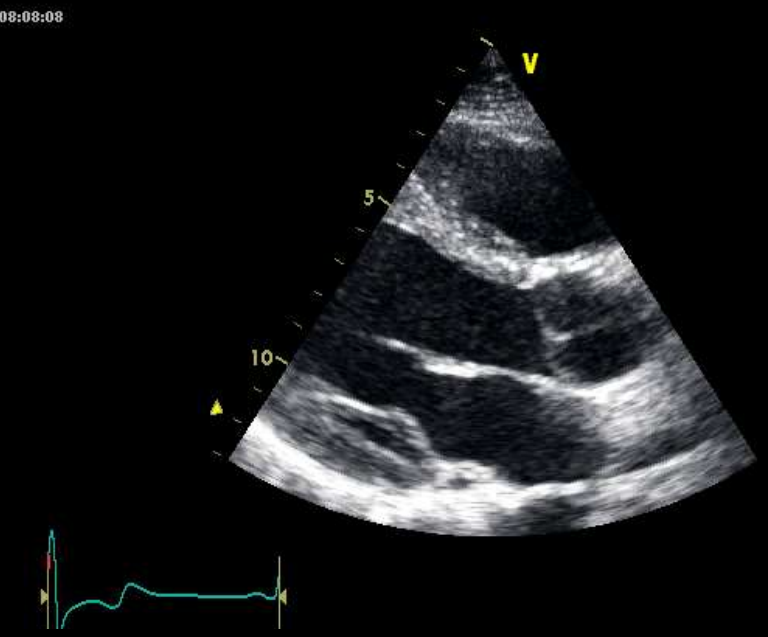
HCM without SEPTAL HYPERTROPHY

APICAL - ANTERO BASAL FORM

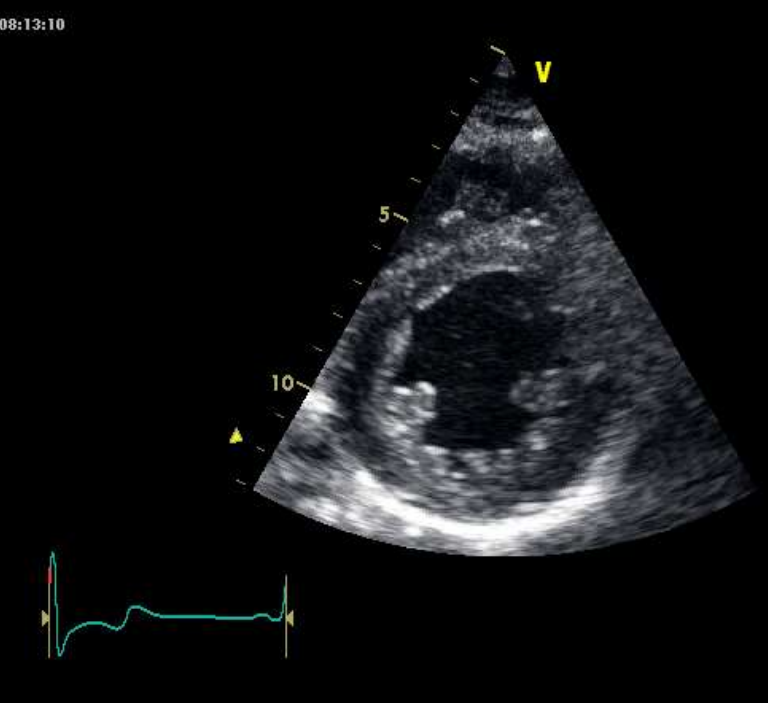


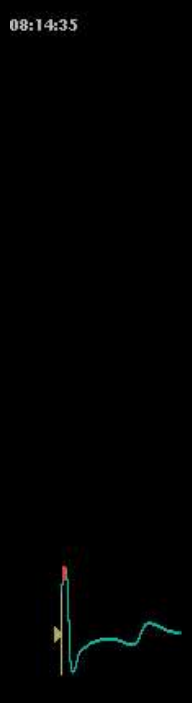
Normal Echo
Septum 10.5mm

Sen-Chowdhry, S. et al. Nat Rev Cardiol. 2016

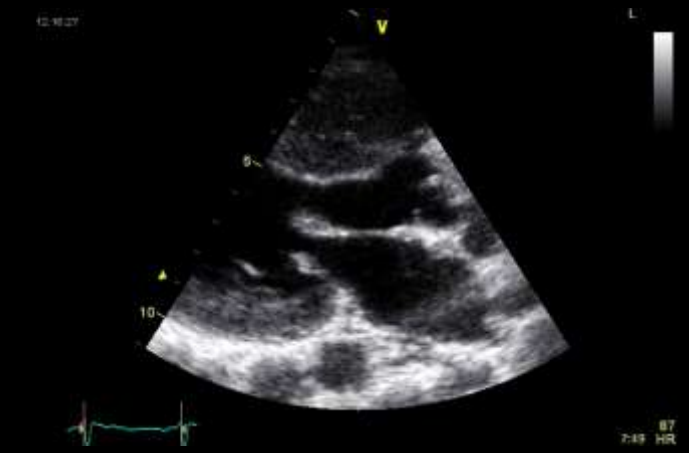

08:08:08
V
5
10

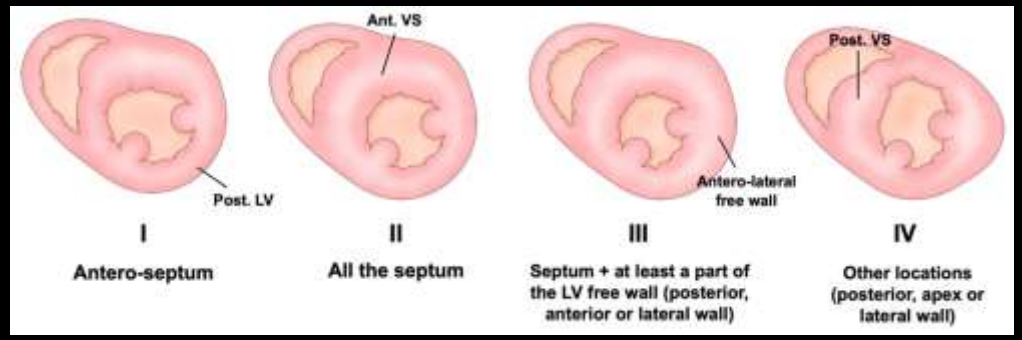

08:11:01

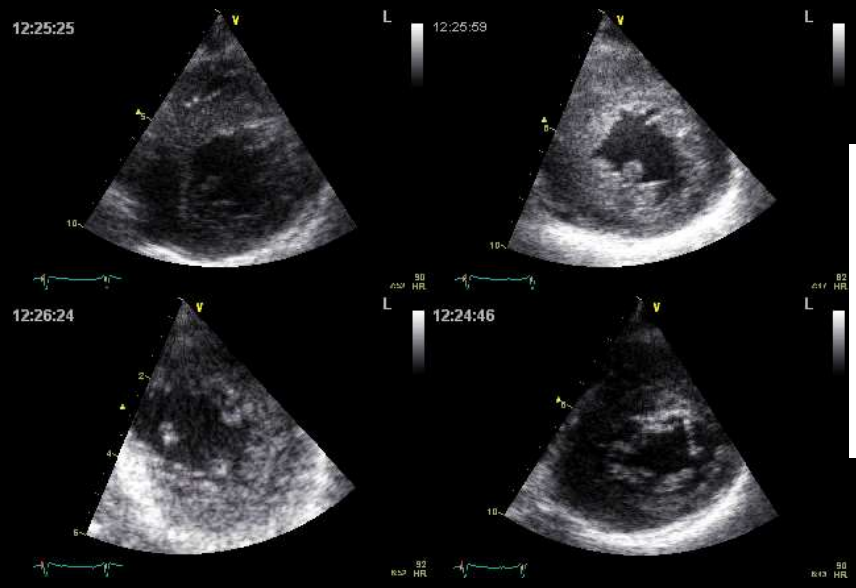

08:13:10
V
5
10

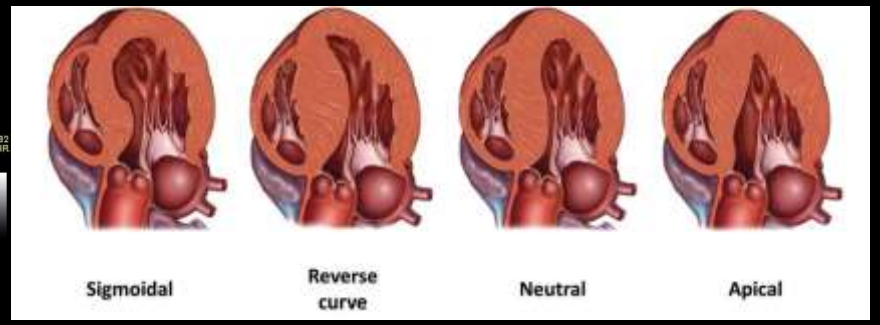

08:14:35

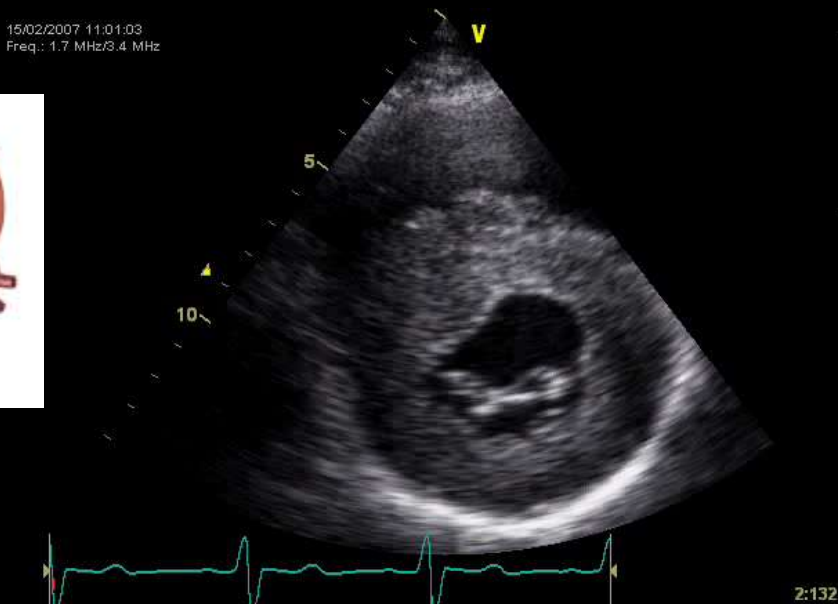
Septal / diffuse / apical phenotype









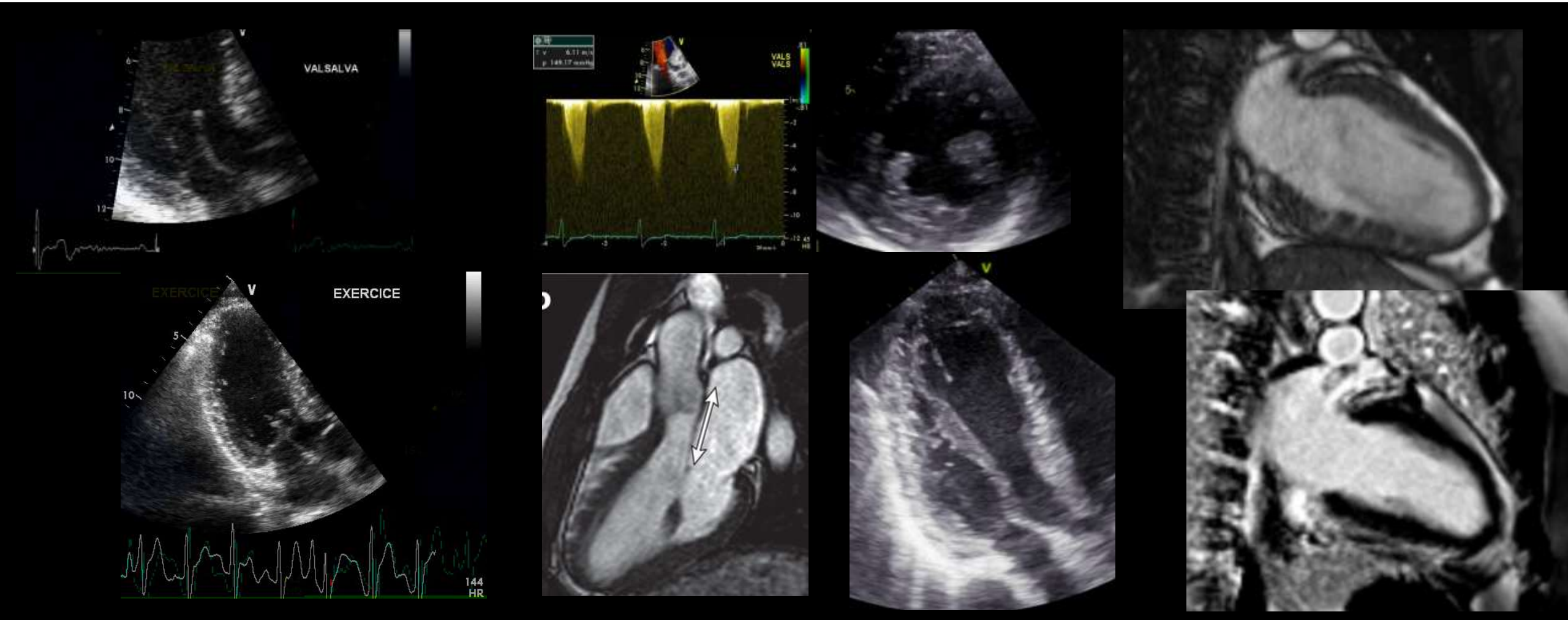


HCM without LVH

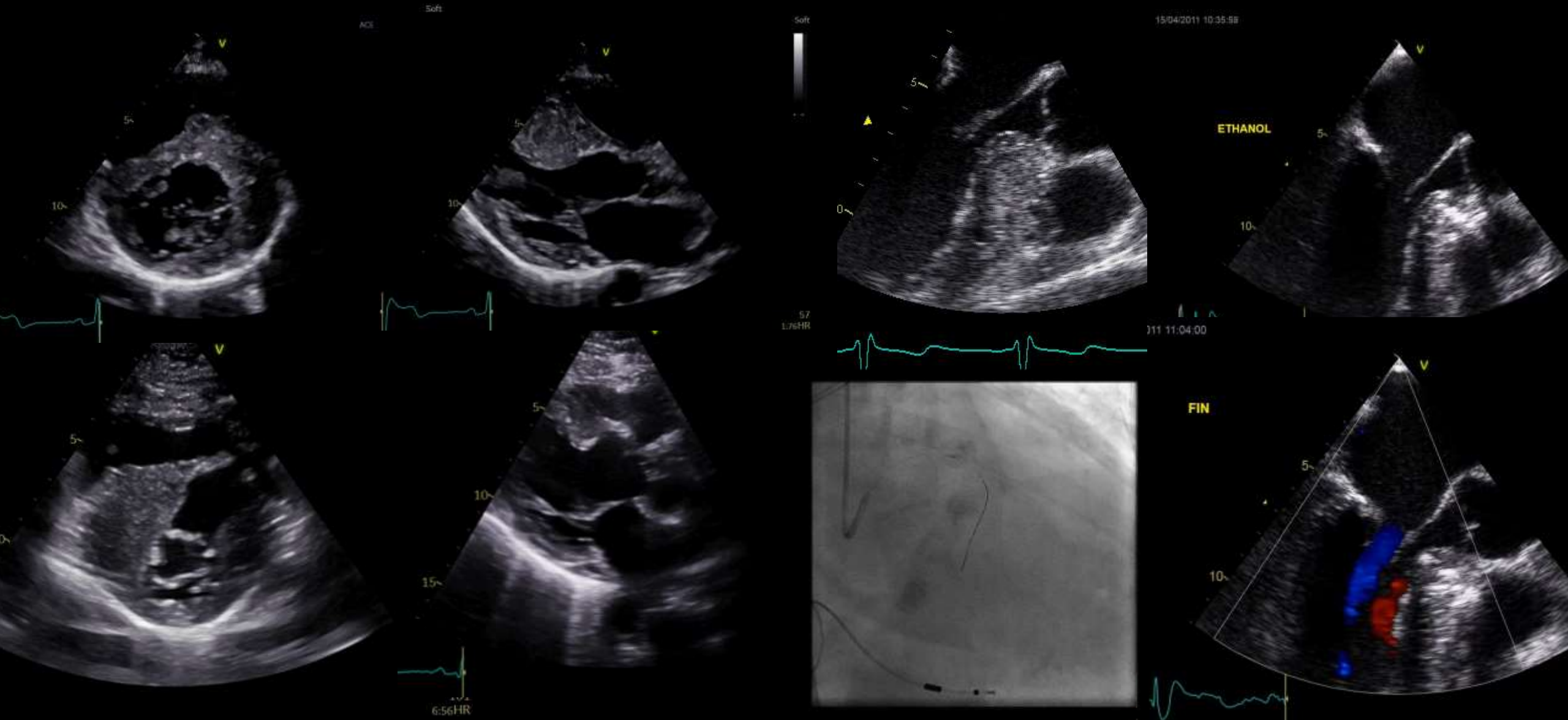
Mitral valve

Papillary Muscle

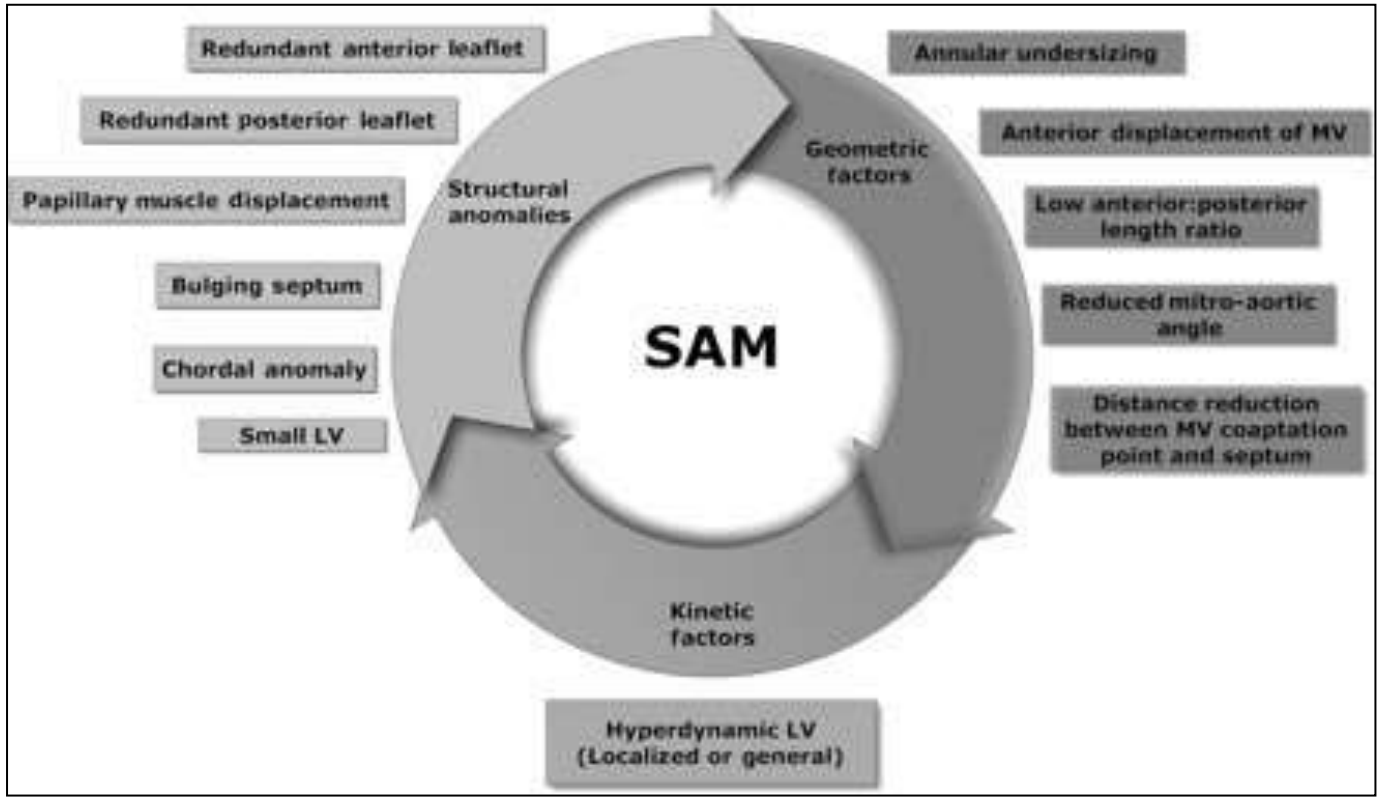
Crypte



Septum is no more a problem

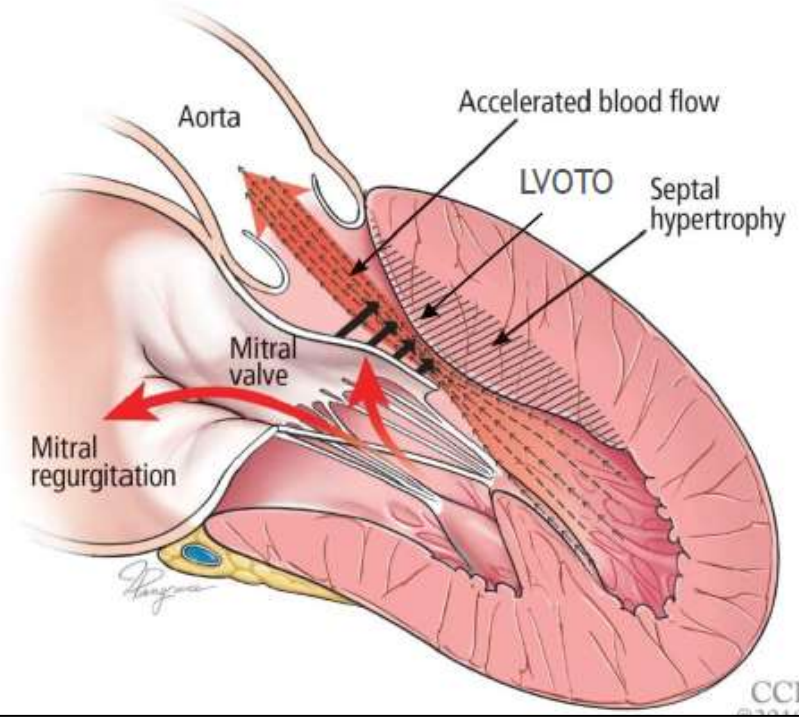


Pathophysiology of obstruction

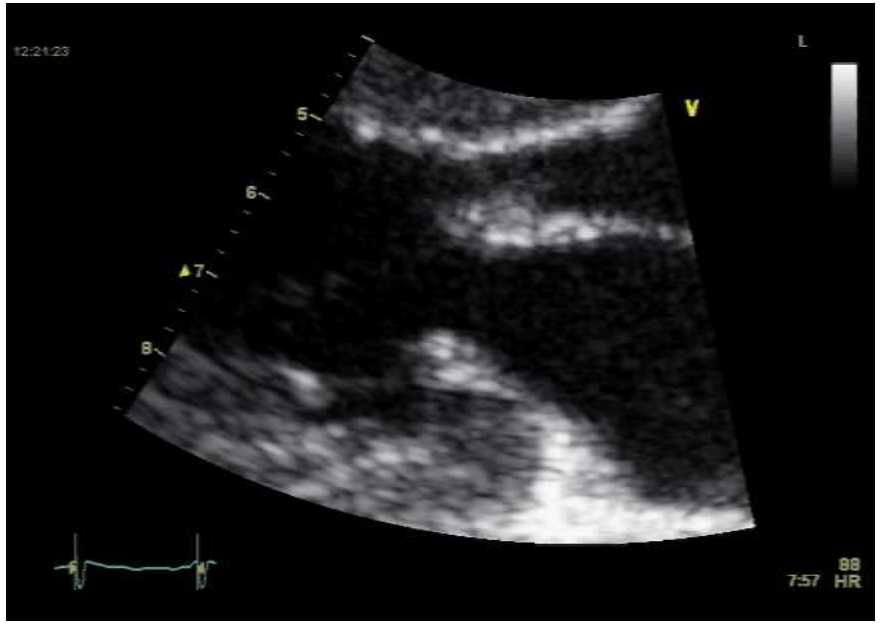


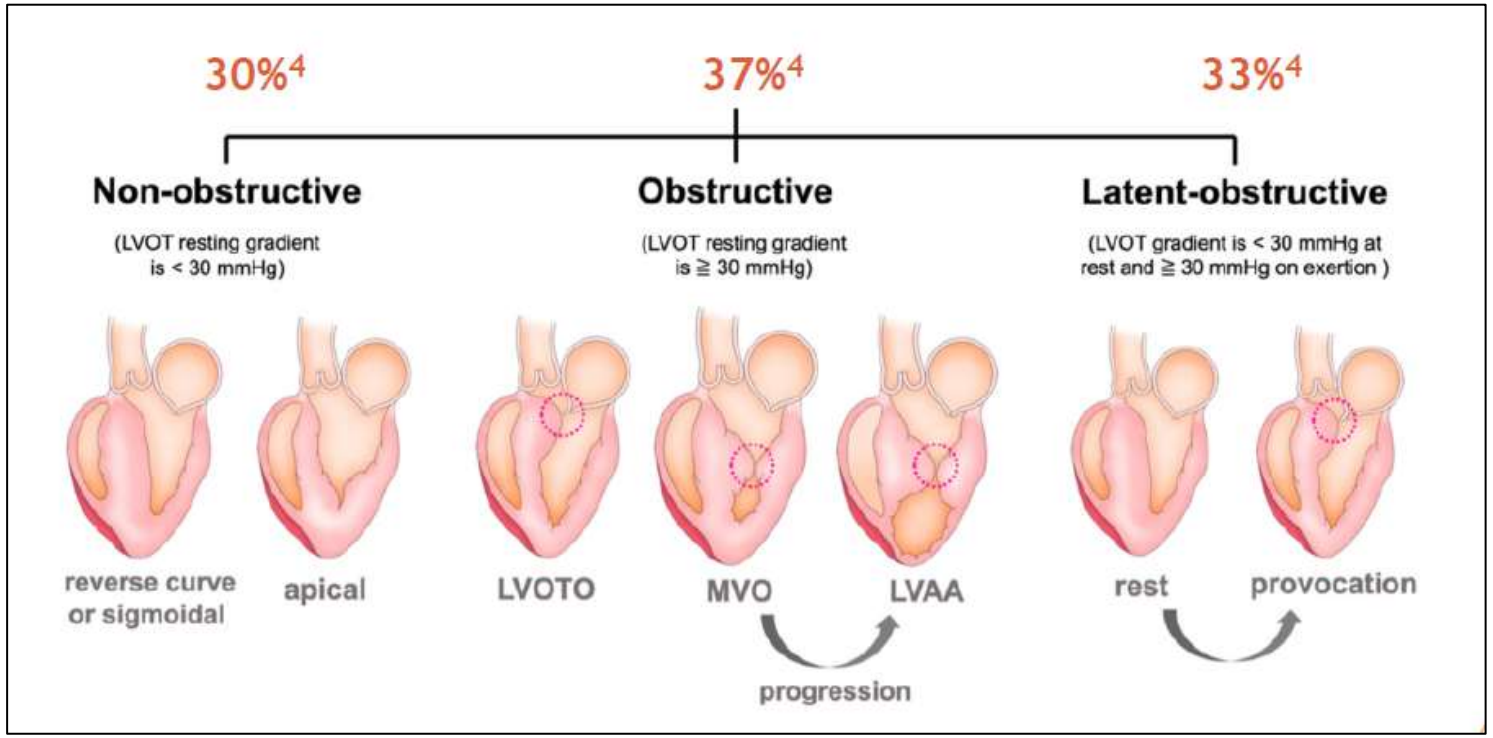
IVS hypertrophy ➔ narrowing of the LVOT

Mitral SAM toward the IVS (Venturi effect)



Obstruction





results from mitral-septal contact due to septal hypertrophy and/or SAM of the mitral valve

- SAM occurs when the mitral valve leaflets are pushed into the LVOT
  - During ventricular systole, flow against the abnormally positioned mitral valve causes drag on the mitral valve leaflets, pushing the leaflets into the outflow tract

SAM : Diagnosis

(A) 2D echo parasternal long-axis view with SAM of the anterior mitral valve leaflet

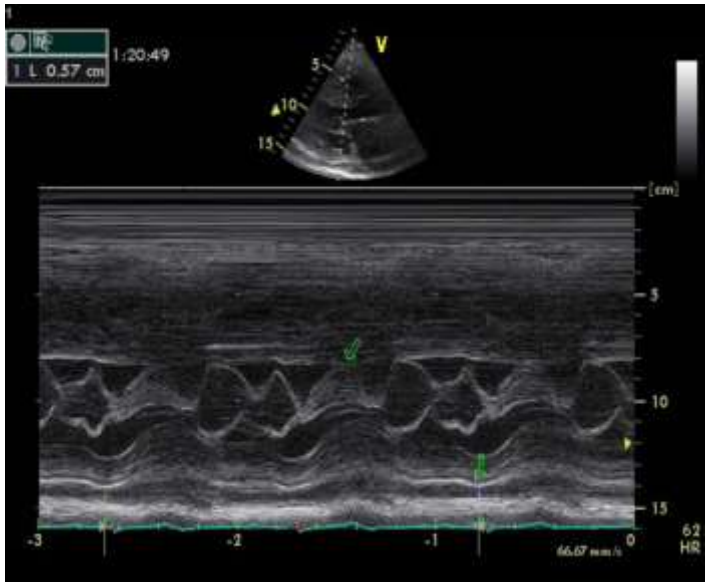
(B) SAM of the anterior mitral valve leaflet in M-mode

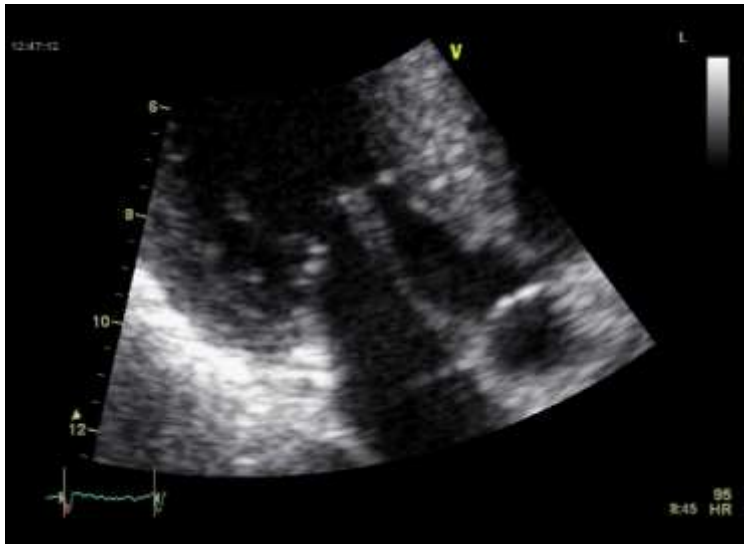
(C) M-mode colour Doppler flow imaging of the localisation of the obstruction

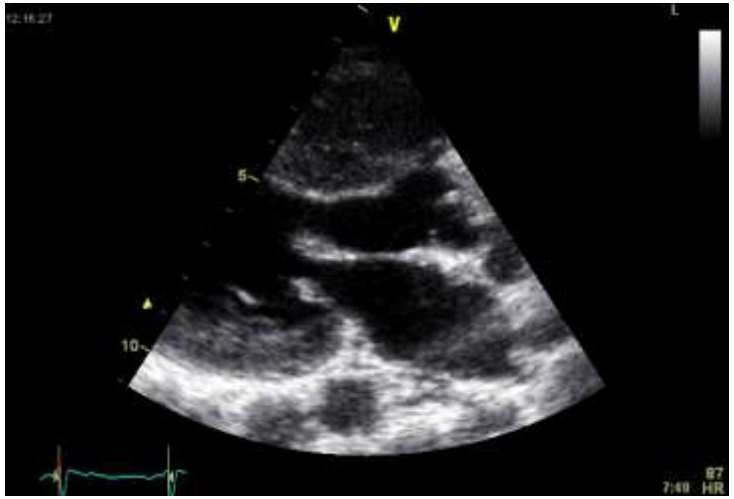
(D) 2D echo with colour Doppler flow imaging of the localisation of the obstruction in the LVOT

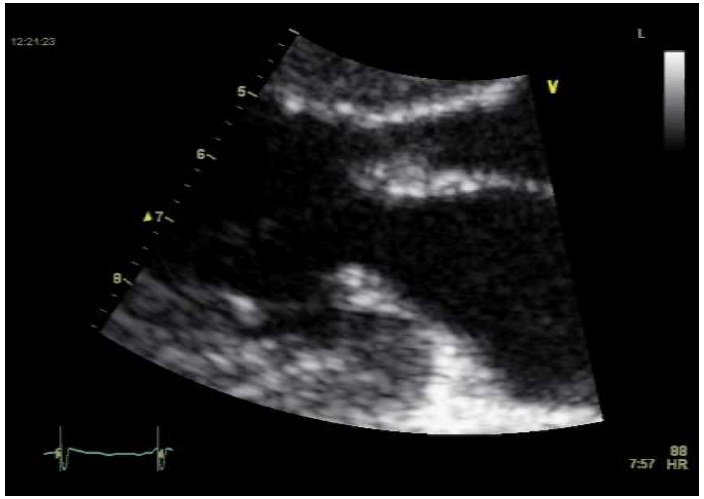
(E) M-mode partial mid-systolic closure of the aortic valve

(F) CW Doppler recording of the obstruction with a maximal peak gradient of 75 mmHg



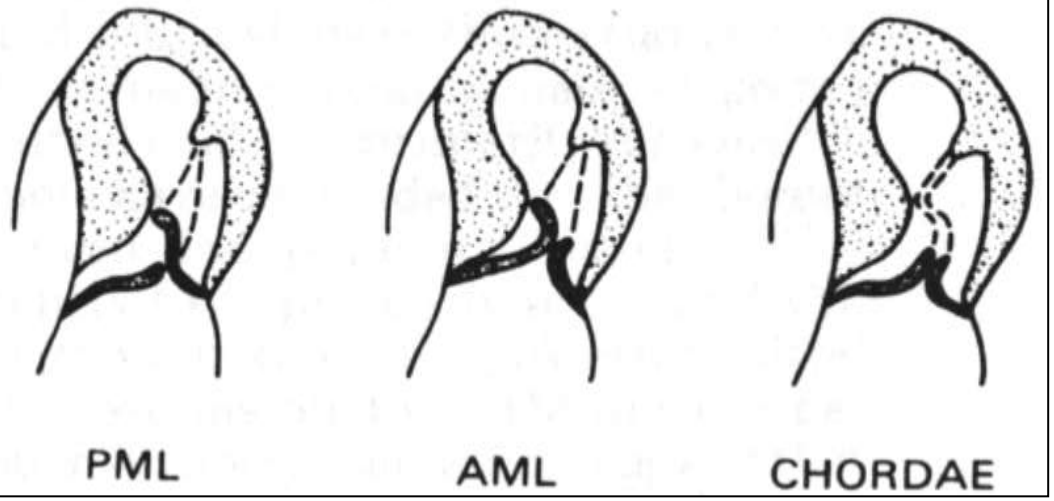




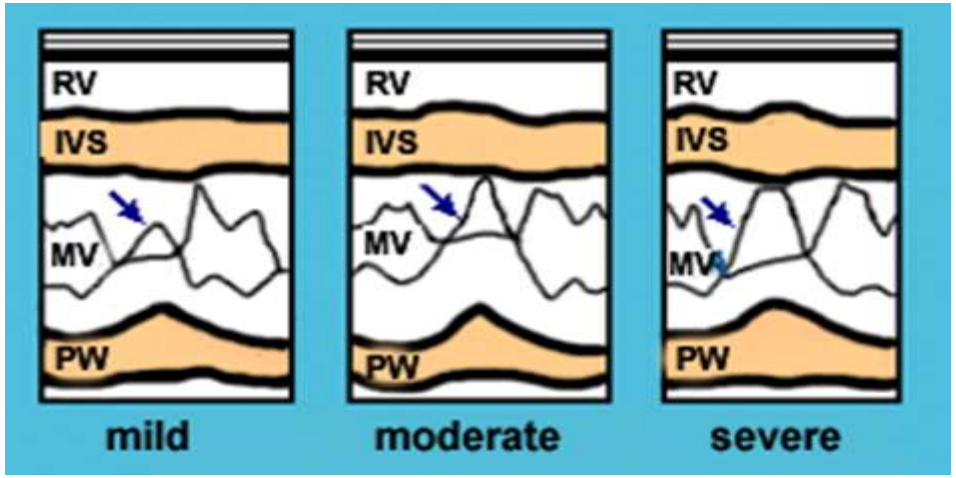


SAM and SAM

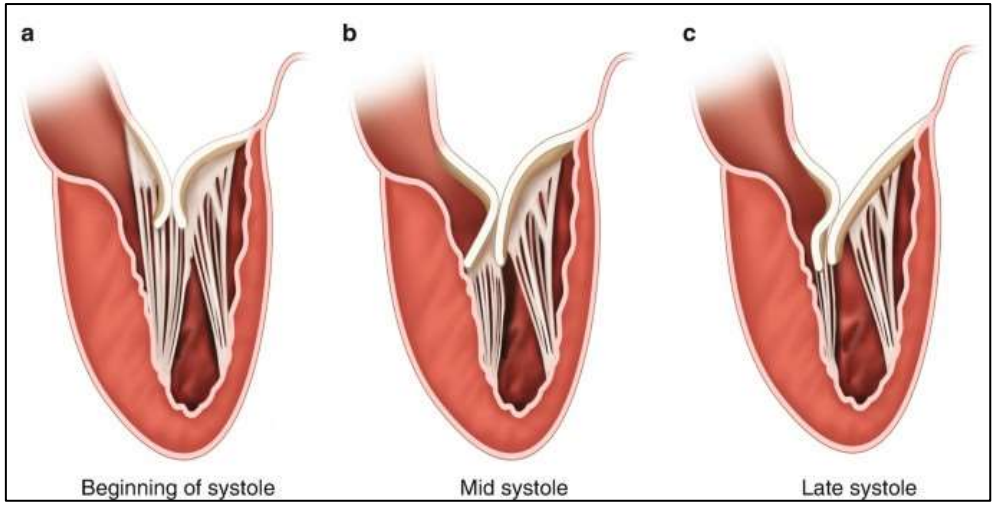
Chordal / valve / Both



Classification



complet / Brief / incomplet

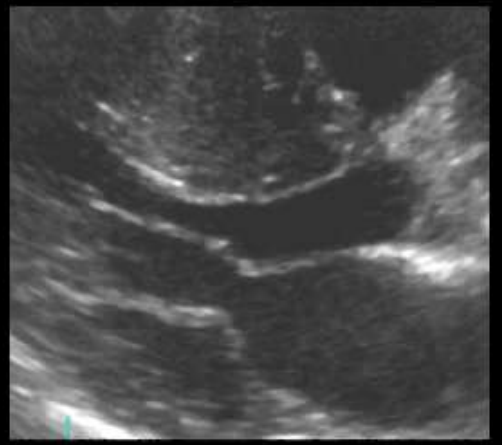



5
6
7
8
V

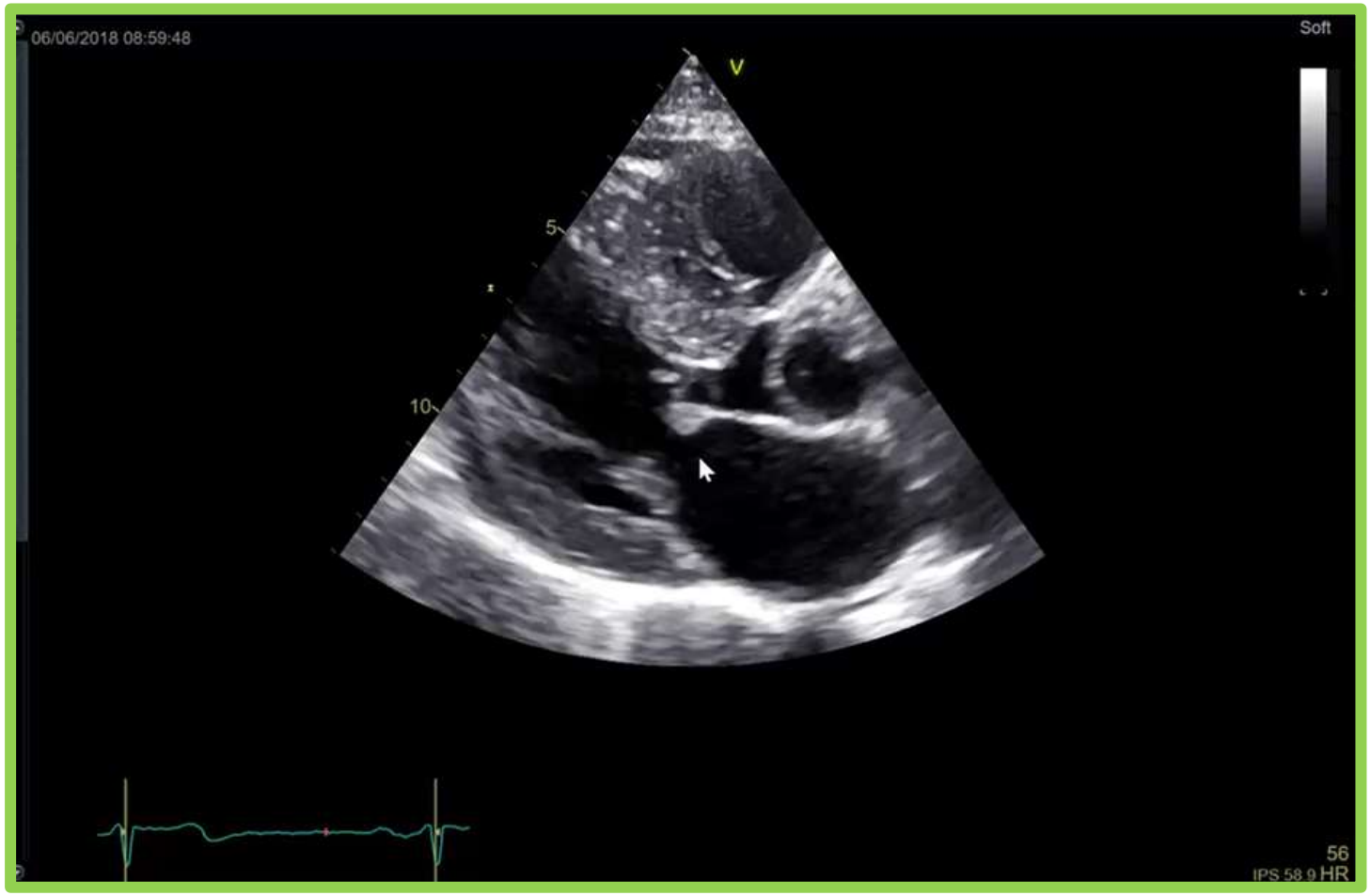

5
X
88
7:57 HR


5


5



Left ventricular obstruction : How to ?



Colour doppler

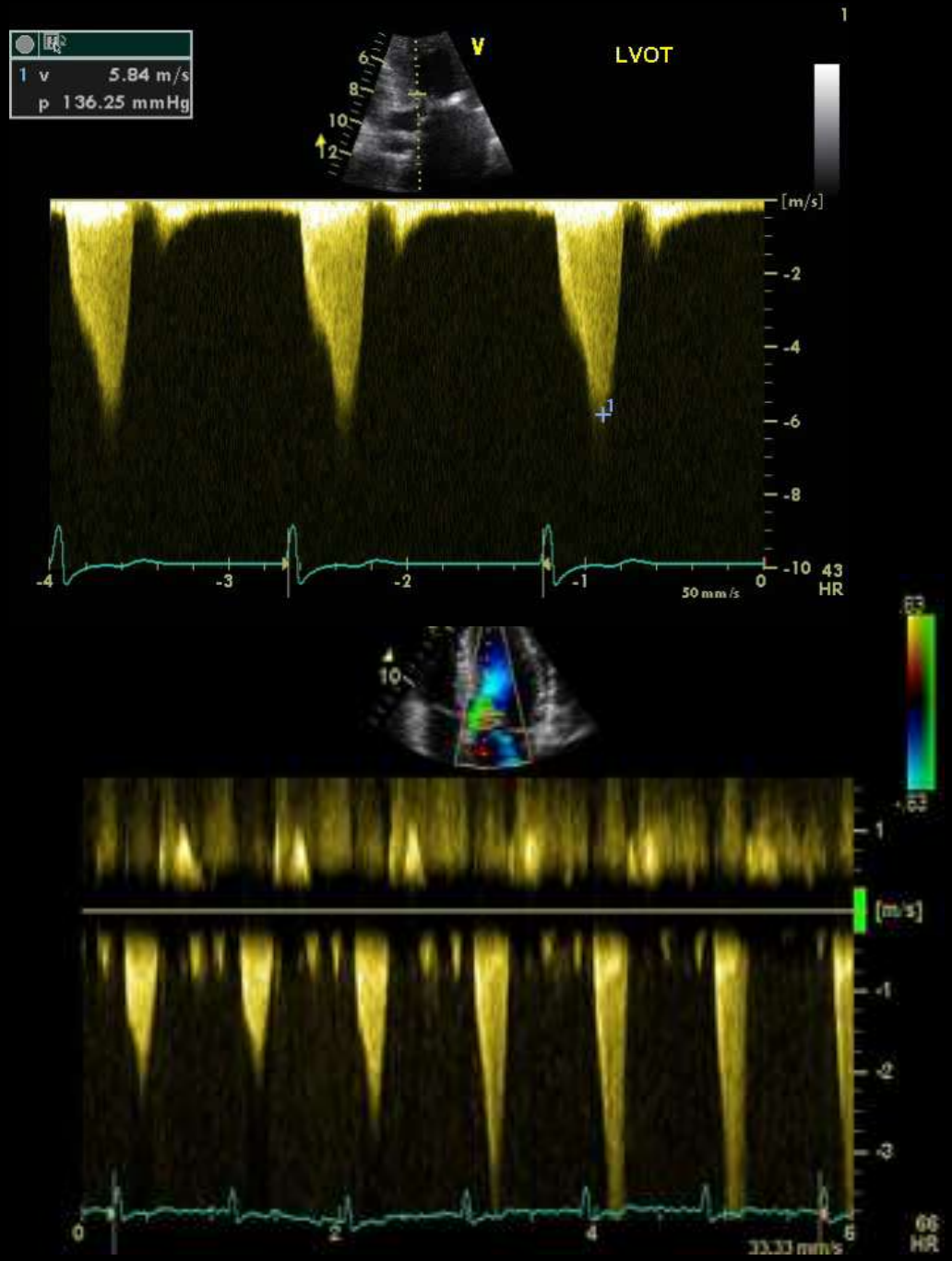
Level?

Medioventricular apical lvot

Pulsed doppler

Obstruction localizaition from base to apex

Continous doppler



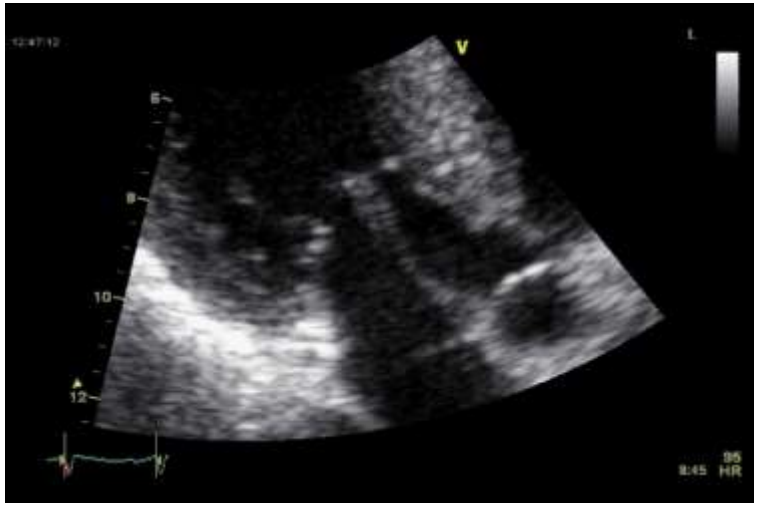
- Dynamique
- End systole
- May vary according to
  - Day time
  - Load condition
  - Medical treament

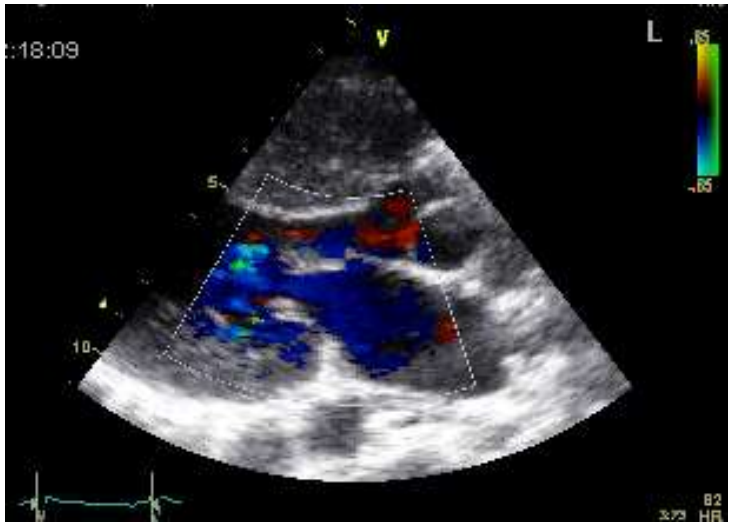
Mitral regurgitation

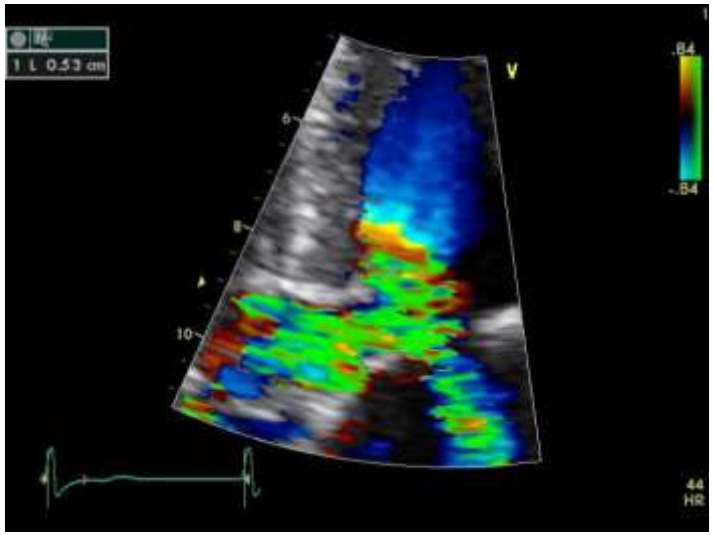
Secondary MR

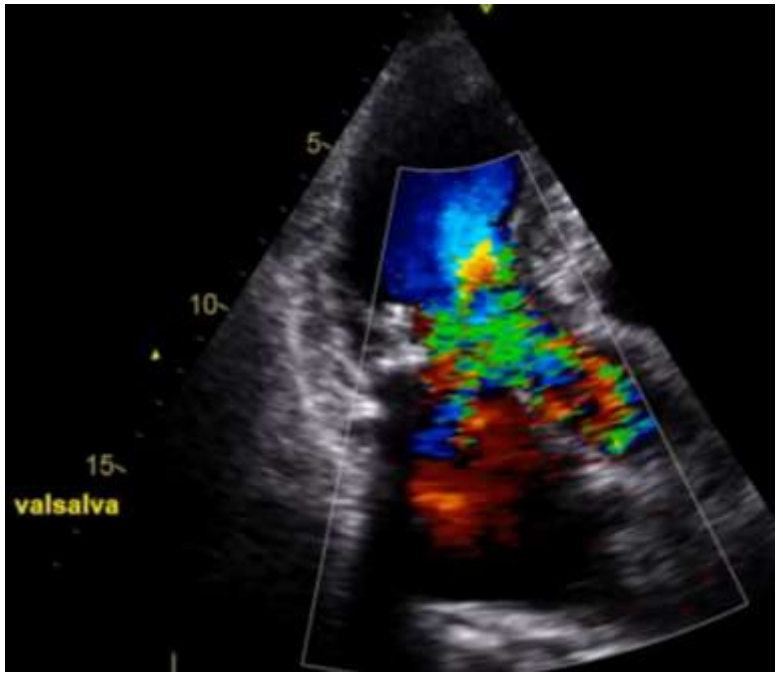
Related to SAM

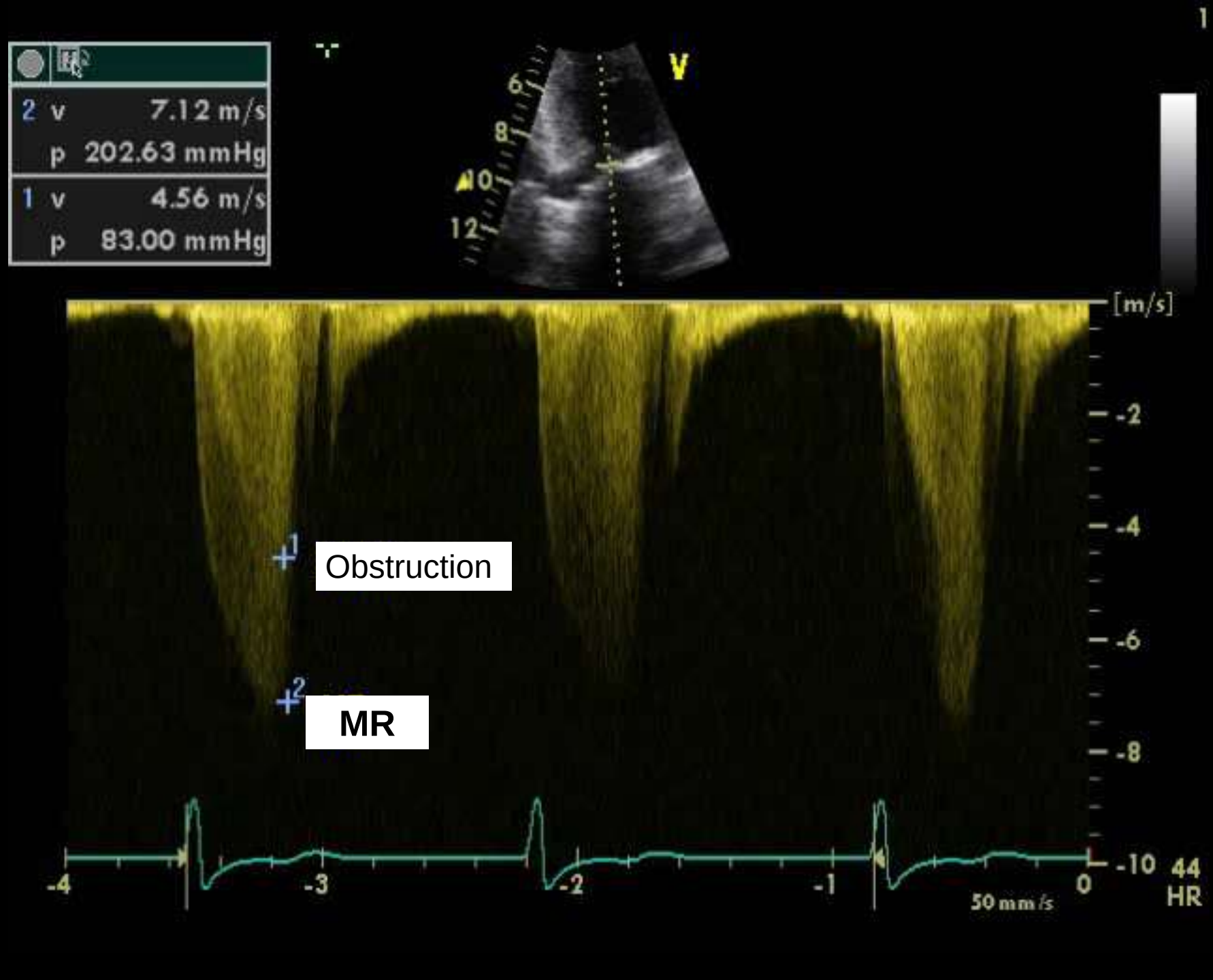
If central : discussion others causes





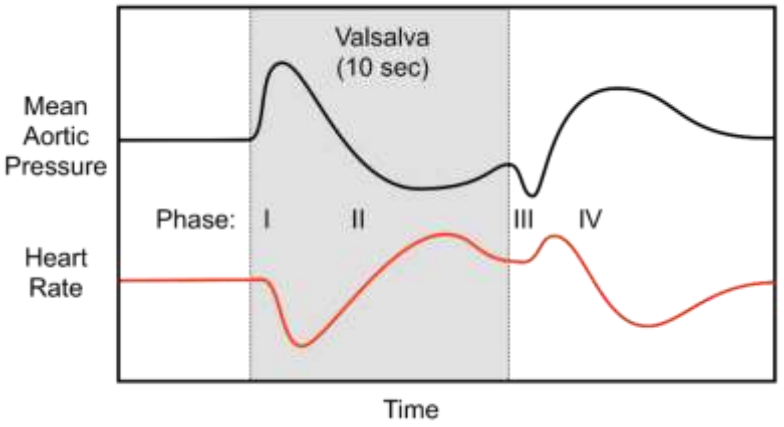





2 v 7.12 m/s
p 202.63 mmHg
1 v 4.56 m/s
p 83.00 mmHg
V
6
8
10
12
1
[m/s]
-2
-4
-6
-8
-10
Obstruction
MR
-4
-3
-2
-1
0
50 mm/s
44
HR

Provocative maneuver

valsalva , Standing and exercise echo (Gold Standard)



- Rest, Valsalva, upright
- EE using treadmill exercise is an important technique in the detection of inducible obstruction in HCM
- EE should be performed in symptomatic patients if bedside manoeuvres fail to induce LVOTO ≥ 50 mm Hg
- In asymptomatic patients, consider when the LVOT gradient is relevant to lifestyle advice and treatment decisions

EXERCISE DYNAMIC

SEMI RECUMBENT POSTURE

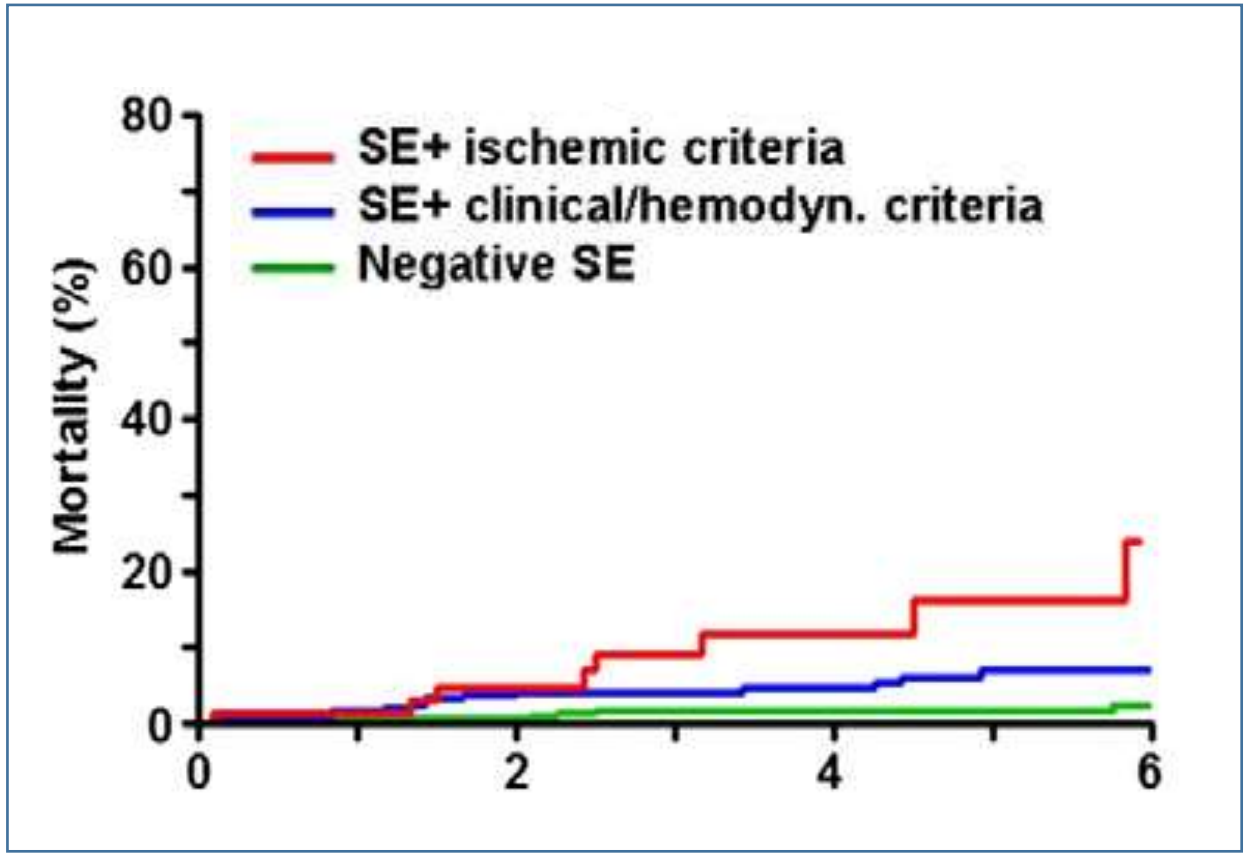
CONTINOUS + PEAK BUT NOT ONLY


Protocole écran: HCM EFFORT
A4C
A2C
PGAXE
PAXE-MP
SAM
GRADIENT
IM
REPOS
DEBUT EFF
DEBUT EFF
PIC
PIC
RECUP

Others causes of Dyspnea

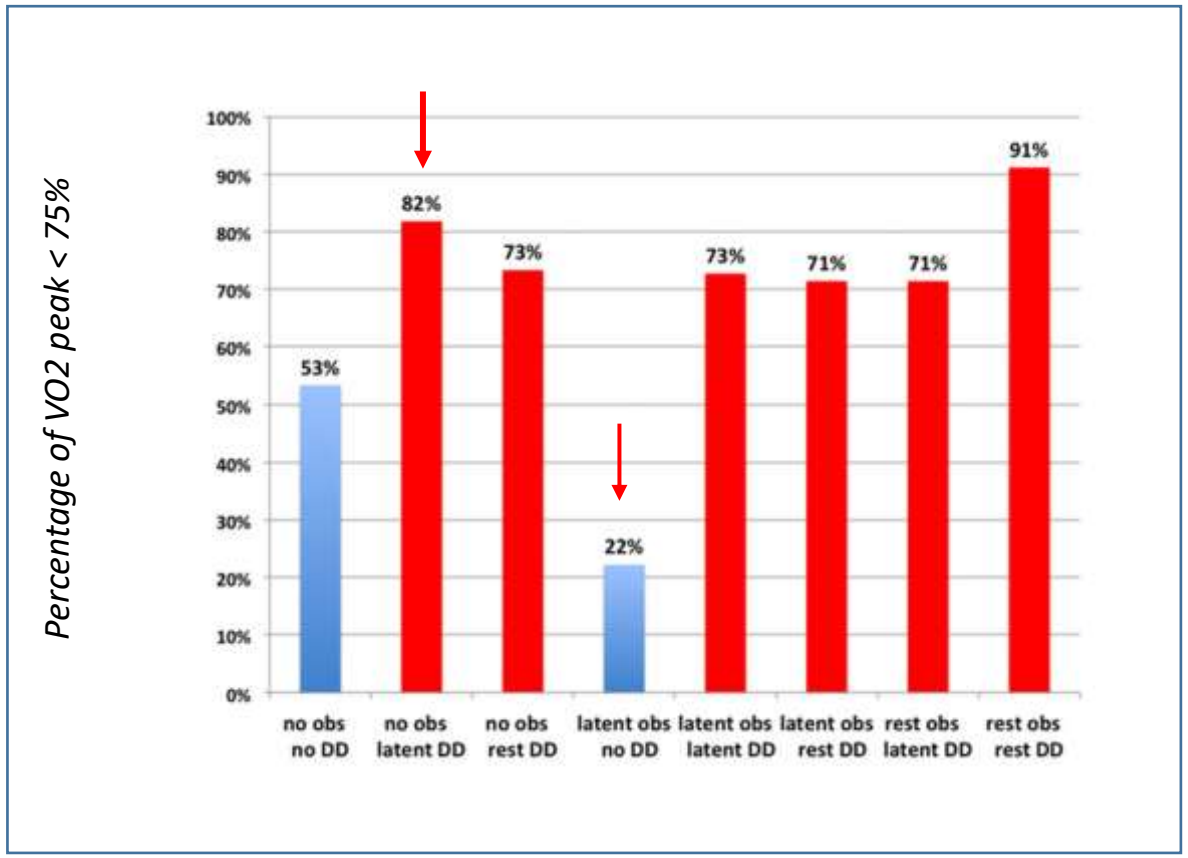
- *Not only hemodynamic endpoints and obstruction*

RULE OUT ISCHEMIA



Ciampi Q Int J Cardiol. 2016 Sep 15;219:331-8.

RULE OUT DIASTOLIC DYSFUNCTION



Re F Int J Cardiol. 2017 Jan 15;227:743-750

Nonobstructive	Rest	<30 mm Hg
	Physiologically provoked	<30 mm Hg

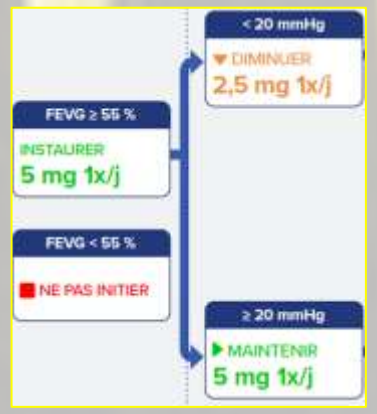
20mmHg

30mmHg

50mmHg

60mmHg

75mmHg



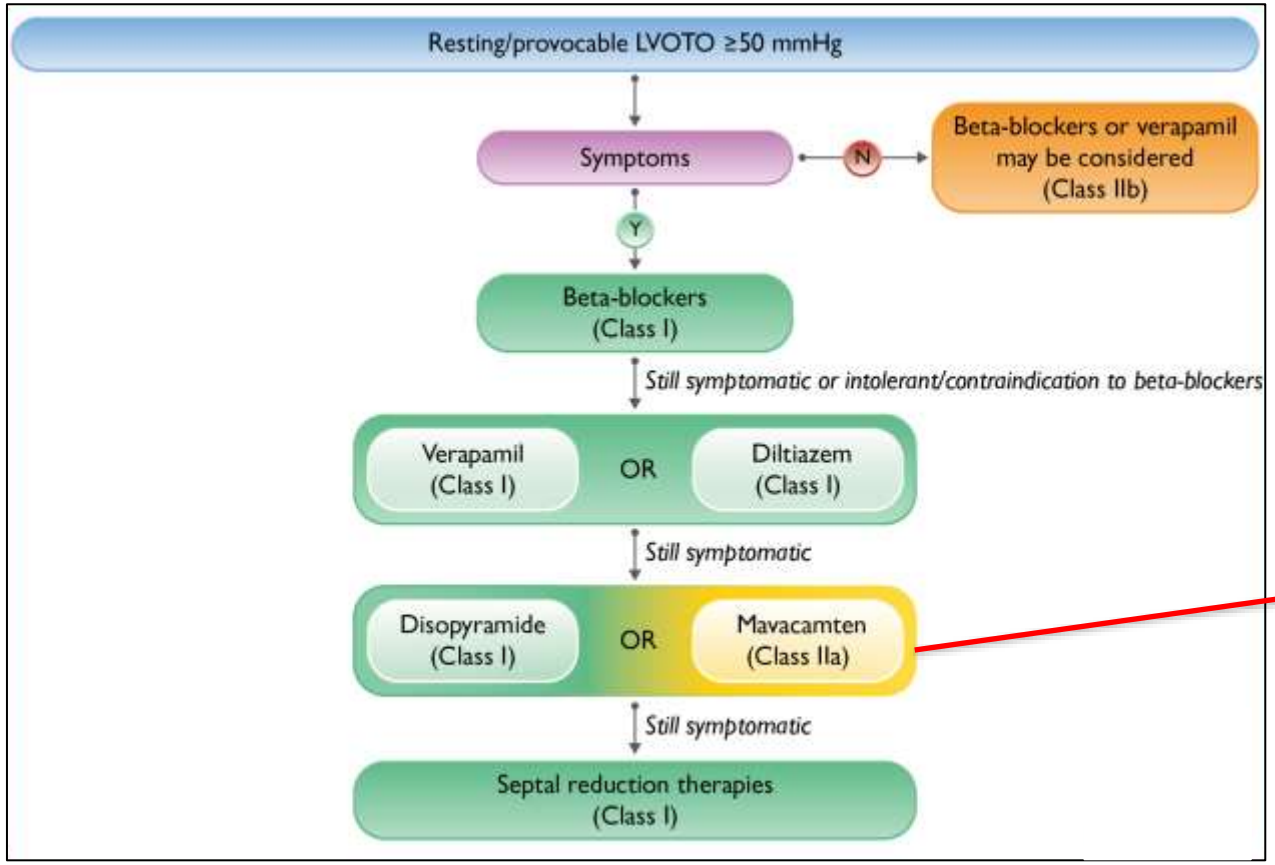
Labile obstruction	Rest	<30 mm Hg
	Physiologically provoked	≥30 mm Hg

Inclusion criteria explorer (i.e. gradient ≥30mmHg at rest or≥50mmHg after Valsalva manoeuvre or exercise)

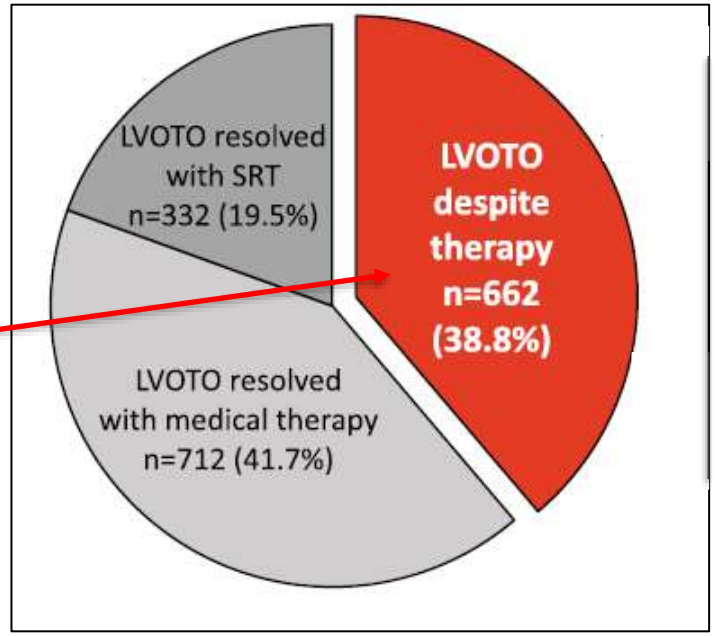
Recommendations	Class	Level
In all patients with HCM, at initial evaluation, transthoracic 2D and Doppler echocardiography are recommended, at rest and during Valsalva manoeuvre in the sitting and semi-supine positions – and then on standing if no gradient is provoked – to detect LVOTO.	I	B
In symptomatic patients with HCM and a resting or provoked peak instantaneous LV outflow tract gradient <50 mmHg, 2D and Doppler echocardiography during exercise in the standing, sitting (when possible), or semi-supine position are recommended to detect provocable LVOTO and exercise-induced mitral regurgitation.	I	B

Interpretation of LVOT

1. Obstruction at rest
2. No obstruction at rest but labile obstruction with bedside manoeuvres (standing, Valsalva)
3. No obstruction at rest nor with bedside manoeuvres but labile obstruction with exercise echo (performed because of high clinical suspicion)

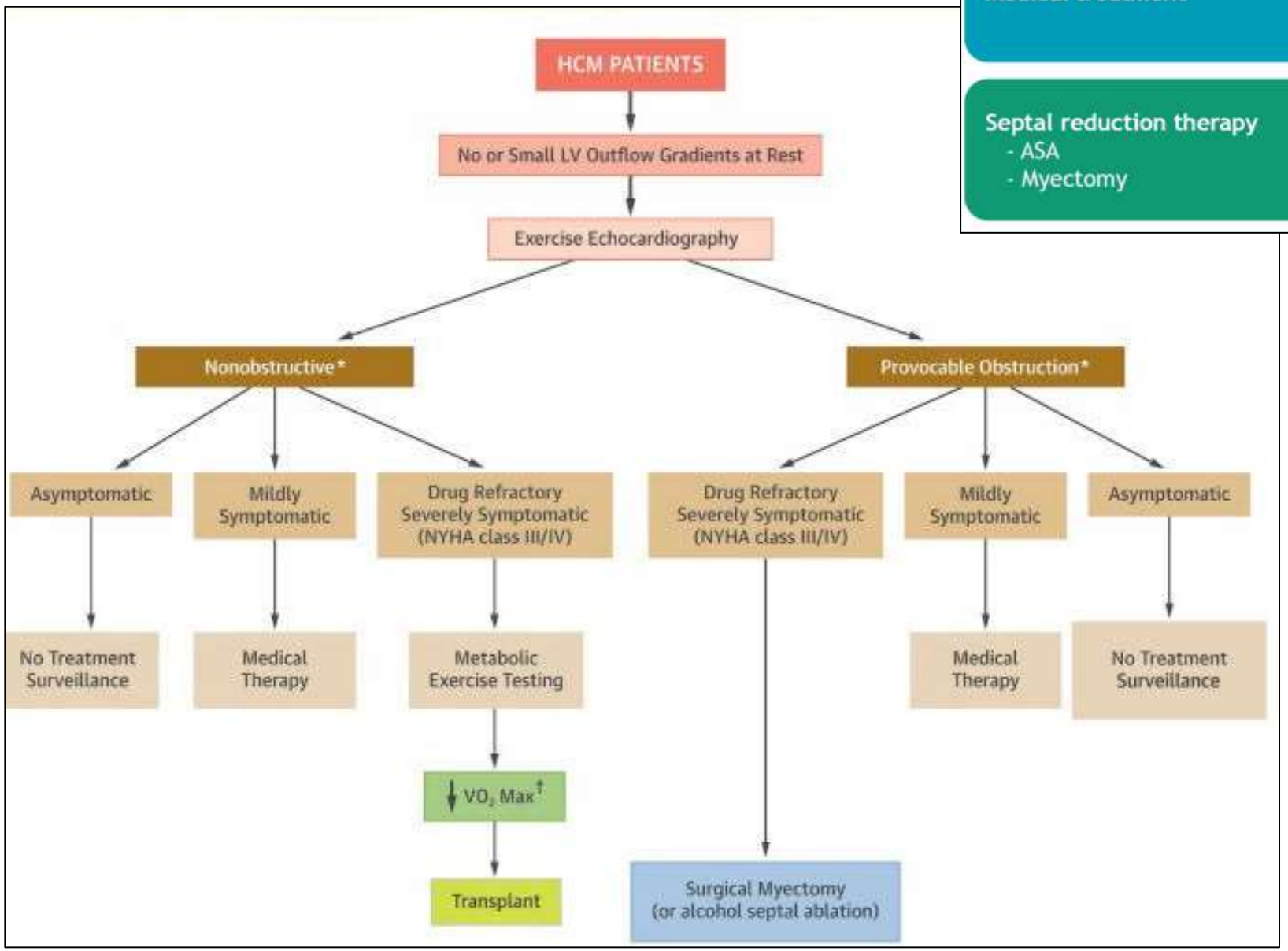


	Gradient au repos	Gradient à l'effort	Groupes
ETT de repos	<30mmhg	?	*Patient non obstructif au repos*
	>30mmhg	?	*Patient obstructif au repos*
	>60mmhg	non	*Patient très obstructif*
ETT de repos + effort	<30mmhg	<30mmhg	*Patient non obstructif*
	<30mmhg	>30mmhg	*Patient avec obstruction latente*



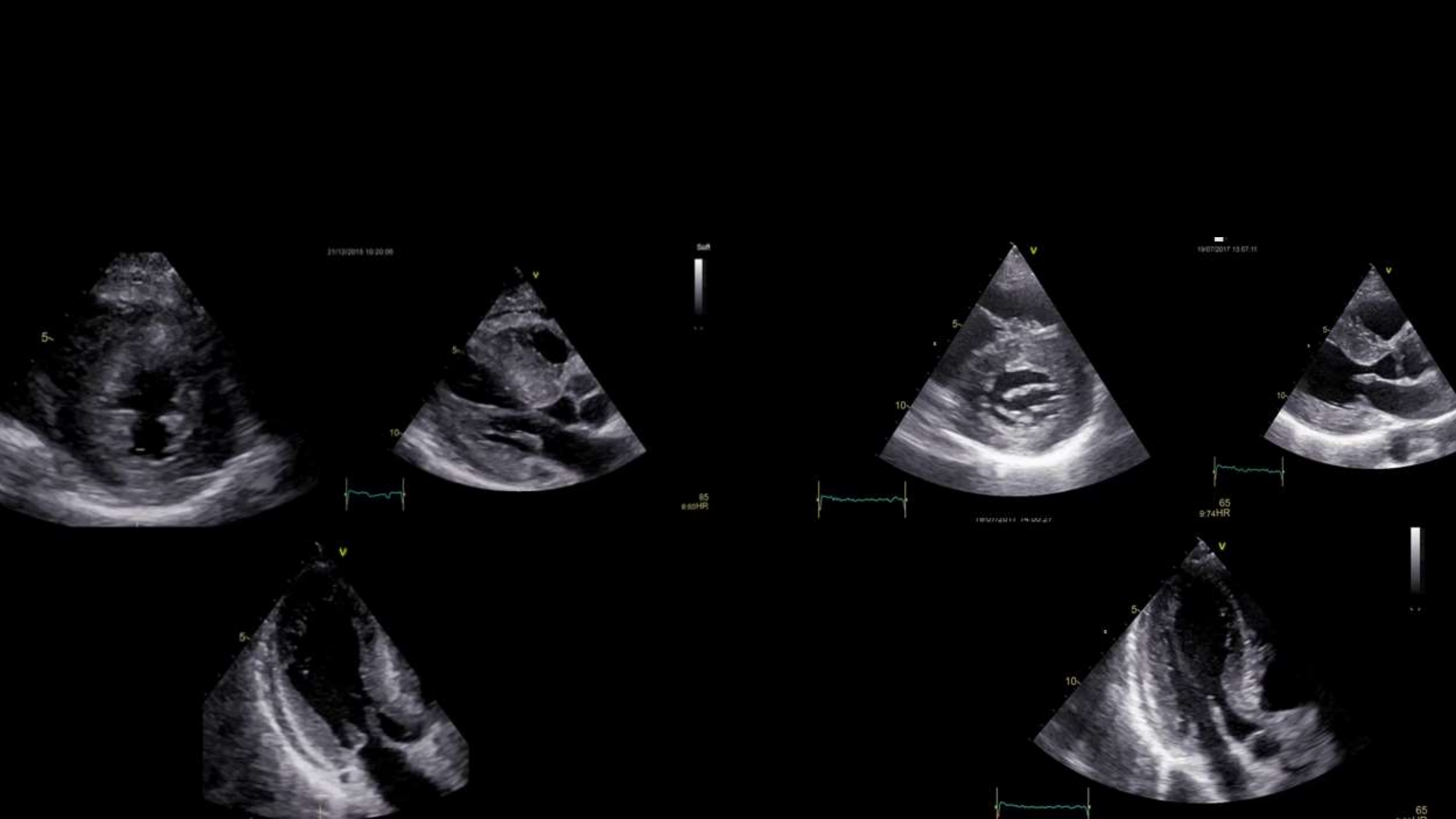
Edoardo Bertero European Journal of Heart Failure 2024

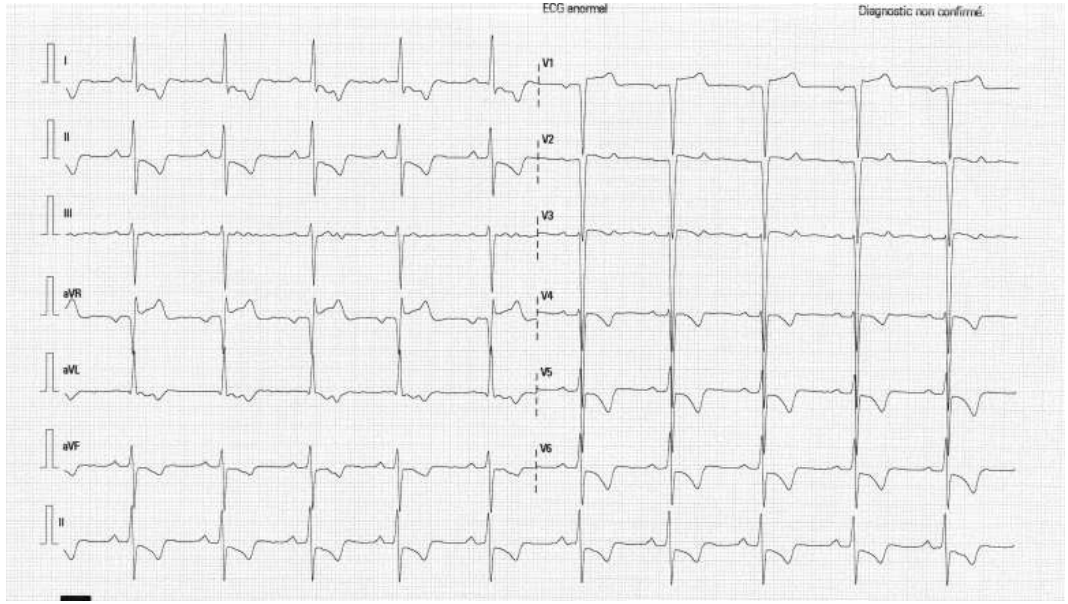
Management of LVOTO

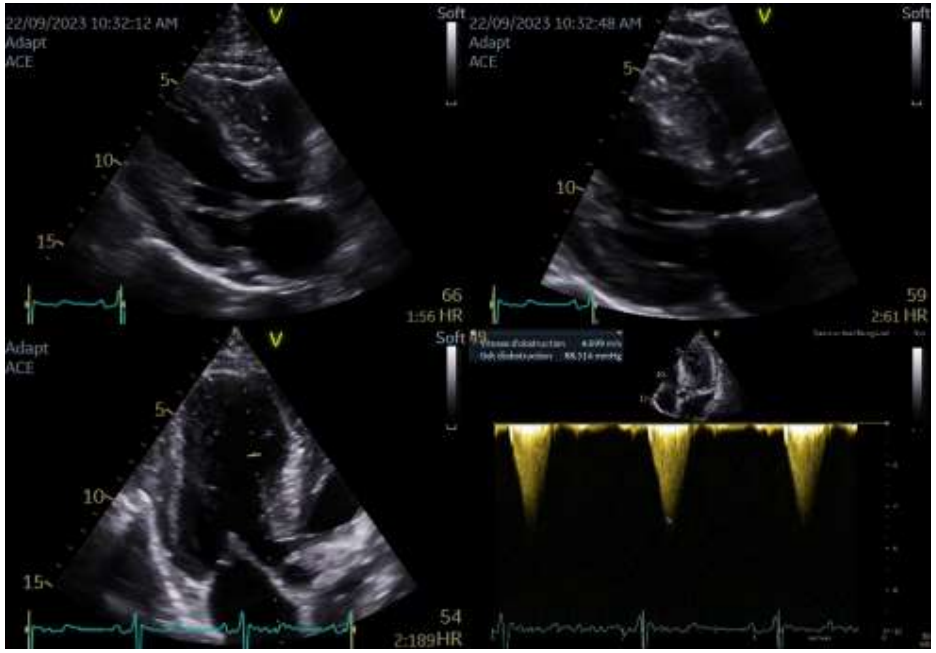


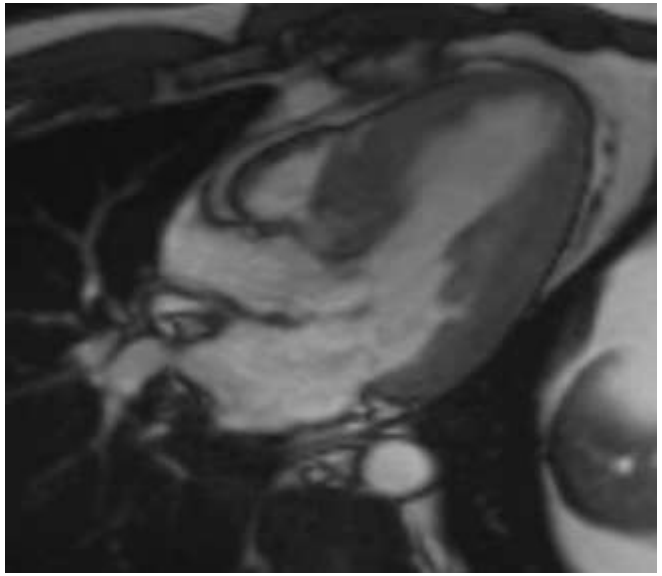
Medical treatment

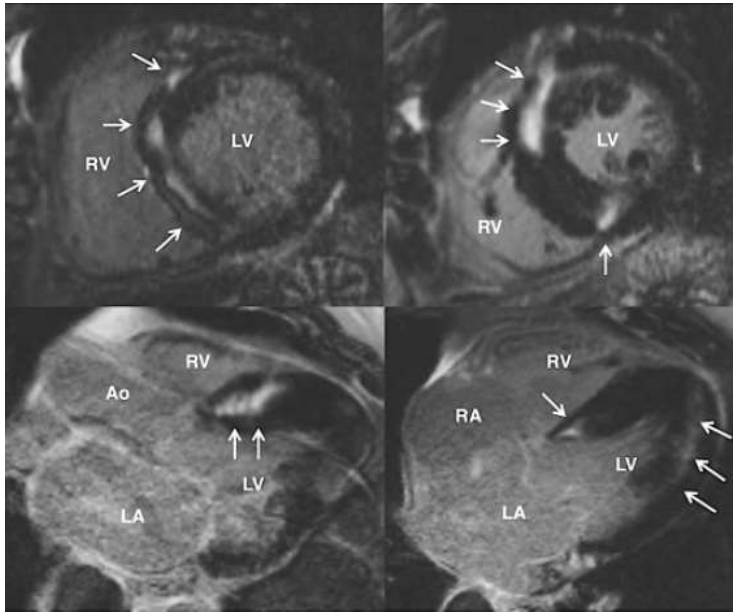
Septal reduction therapy
- ASA
- Myectomy

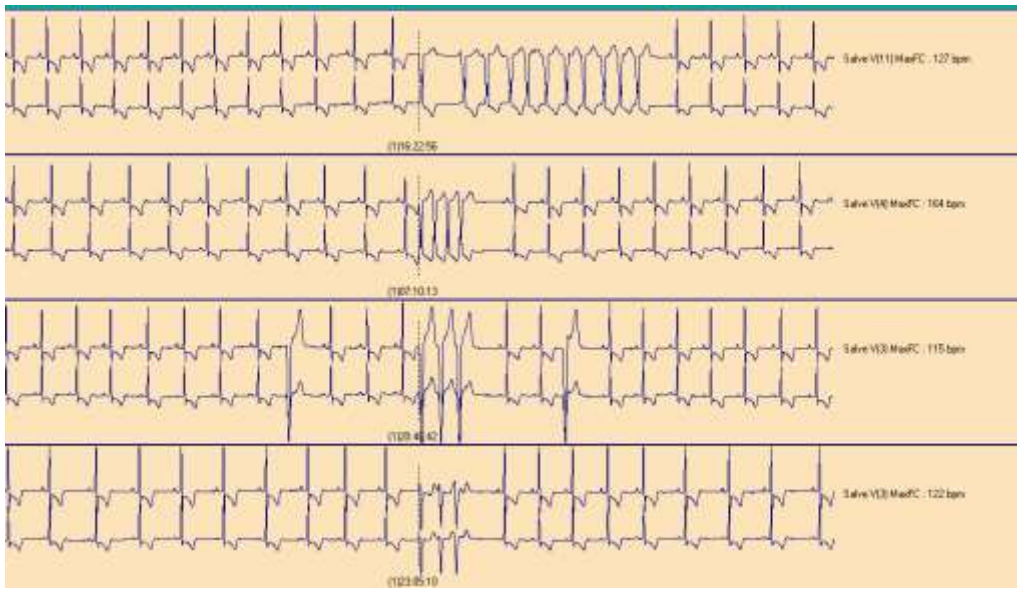

5
10
65
9.74HR

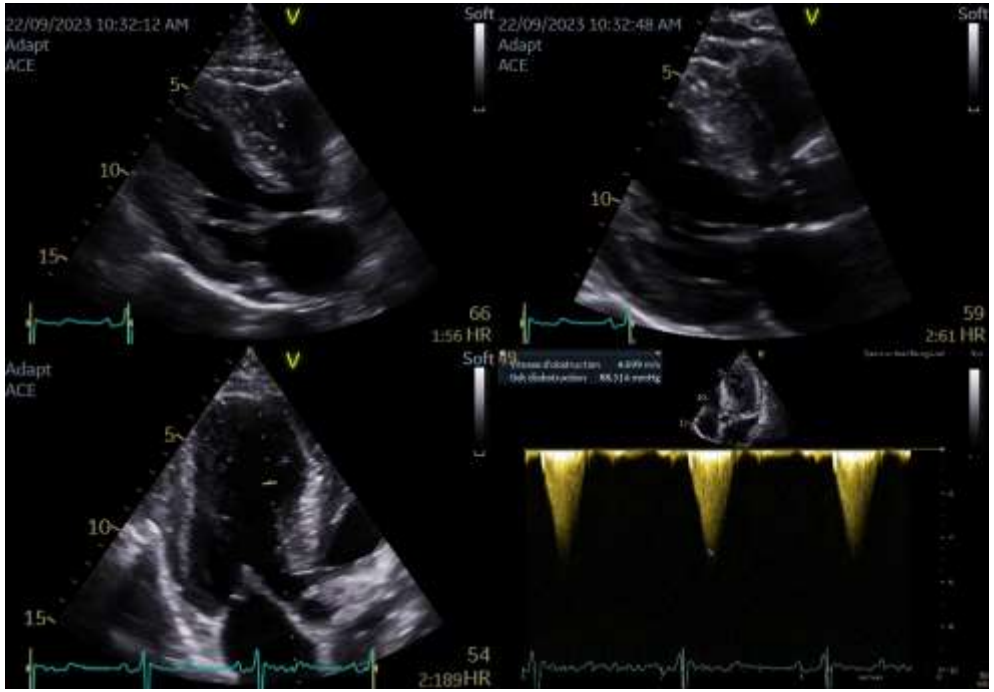

ECG anormal
Diagnostic non confirmé.
I
II
III
aVR
aVL
aVF
II
V1
V2
V3
V4
V5
V6


22/09/2023 10:32:12 AM
Adapt
ACE
22/09/2023 10:32:48 AM
Adapt
ACE
Soft
66
1:56 HR
59
2:61 HR
54
2:189HR




RV
LV
RV
LV
RV
Ao
LV
LA
RV
RA
LV
LA





CHRU NANCY

Parcours de soins Mavacamten ®

Screening

Echocardiographie de repos
+ valsalva +/- effort
-ECG - VO2 - TM6
Evaluation qualité de vie*
Prélèvement génétique
-Bilan biologique

Eligibilité
FEVG > 55%
gradient > 30mmHg

Résultats génétique

Validation de l'indication à l'accès précoce

Programmation suite parcours

IDE de coordination
Contact patient

HDJ introduction

Echocardiographie d'effort - ECG
IPA: TM6 / information patient
Béta HCG chez les femmes
Consultation Pharmacien

HDJ 4e semaine

ETT - ECG
IPA: Evaluation qualité de vie* / TM6
Prélèvement sanguin
Retrait traitement pharmacie

HDJ 8e semaine

ETT - ECG
IPA : Evaluation qualité de vie*/ TM6
Prélèvement sanguin
Retrait traitement pharmacie

HDJ 12e semaine

ETT - ECG - V02
IPA : Evaluation qualité de vie*/ TM6
Prélèvement sanguin
Retrait traitement pharmacie

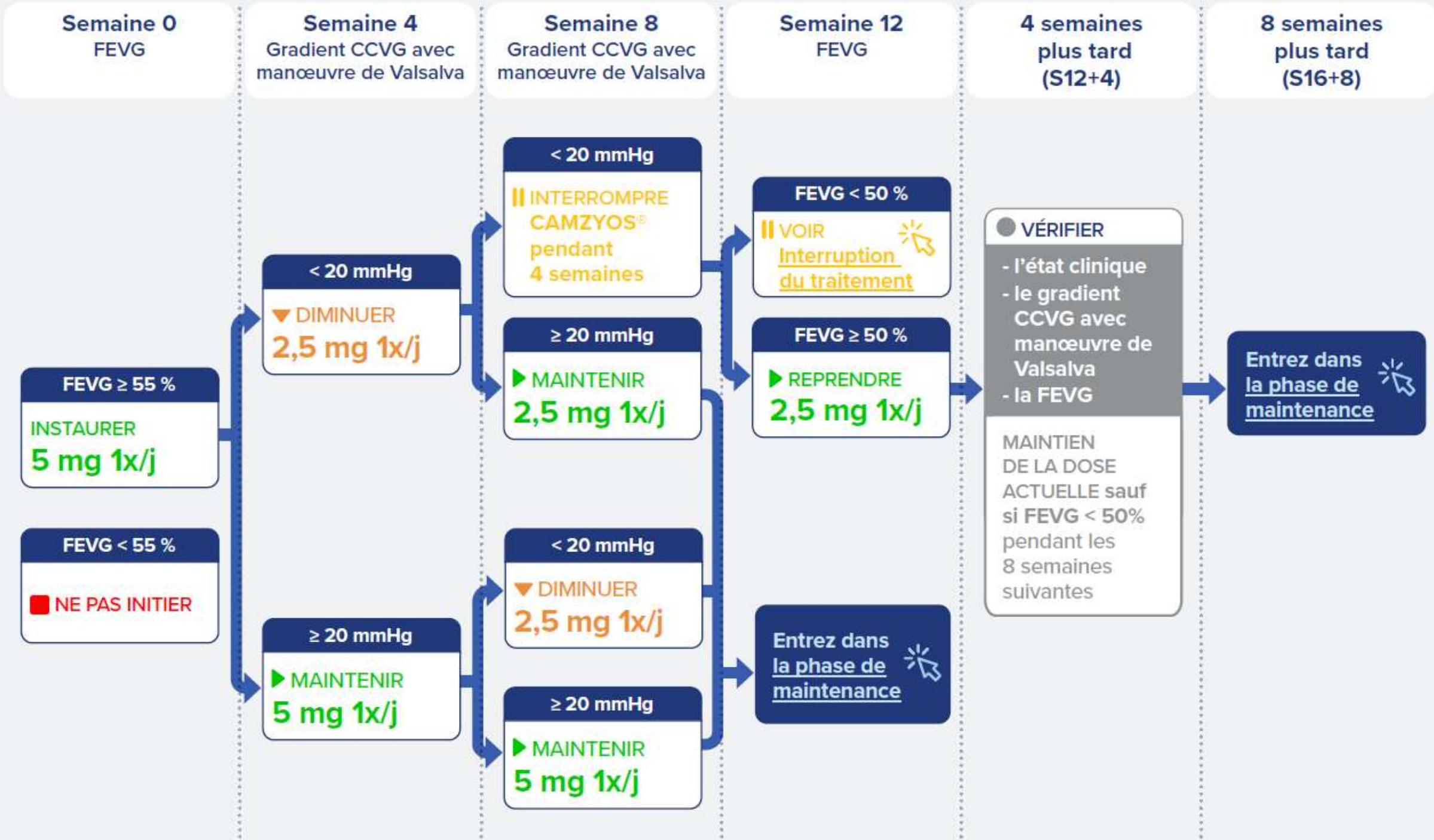
Changement posologie semaine 12

HDJ 16e semaine

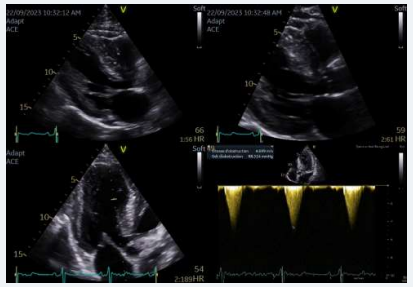
HDJ à 6 mois de suivi

ECG
Echocardiographie d'effort
IPA : Evaluation qualité de vie*/ TM6
Prélèvement sanguin
Retrait traitement pharmacie

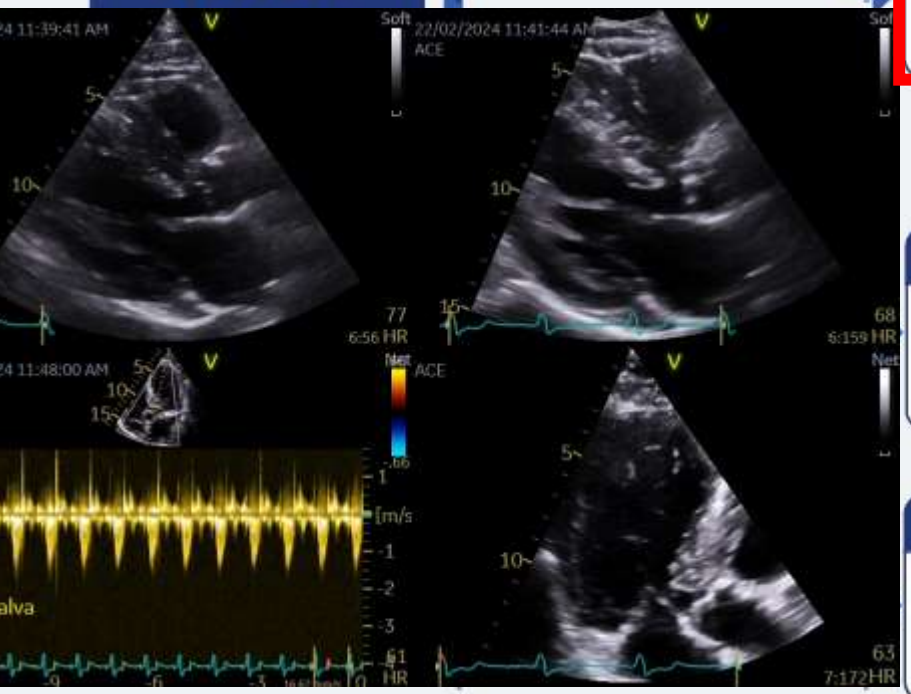
*Kansas city


Semaine 0
FEVG
Semaine 4
Gradient CCVG avec manœuvre de Valsalva
Semaine 8
Gradient CCVG avec manœuvre de Valsalva
Semaine 12
FEVG
4 semaines plus tard (S12+4)
8 semaines plus tard (S16+8)
FEVG ≥ 55 %
INSTAURER
5 mg 1x/j
FEVG < 55 %
NE PAS INITIER
< 20 mmHg
DIMINUER
2,5 mg 1x/j
≥ 20 mmHg
MAINTENIR
5 mg 1x/j
< 20 mmHg
INTERROMPRE CAMZYOS® pendant 4 semaines
≥ 20 mmHg
MAINTENIR
2,5 mg 1x/j
< 20 mmHg
DIMINUER
2,5 mg 1x/j
≥ 20 mmHg
MAINTENIR
5 mg 1x/j
FEVG < 50 %
VOIR Interruption du traitement
FEVG ≥ 50 %
REPRENDRE
2,5 mg 1x/j
Entrez dans la phase de maintenance
VÉRIFIER
- l'état clinique
- le gradient CCVG avec manœuvre de Valsalva
- la FEVG
MAINTIEN DE LA DOSE ACTUELLE sauf si FEVG < 50% pendant les 8 semaines suivantes
Entrez dans la phase de maintenance

Semaine 0	Semaine 4	Semaine 8	Semaine 12	4 semaines plus tard (S12+4)	8 semaines plus tard (S16+8)
FEVG	Gradient CCVG avec manœuvre de Valsalva	Gradient CCVG avec manœuvre de Valsalva	FEVG		



FEVG ≥ 55 %



< 20 mmHg

▼ DIMINUER
2,5 mg 1x/j

< 20 mmHg

‖ INTERROMPRE
CAMZYOS® pendant
4 semaines

≥ 20 mmHg

▶ MAINTENIR
2,5 mg 1x/j

< 20 mmHg

▼ DIMINUER
2,5 mg 1x/j

≥ 20 mmHg

▶ MAINTENIR
5 mg 1x/j

FEVG < 50 %

‖ VOIR
Interruption du traitement

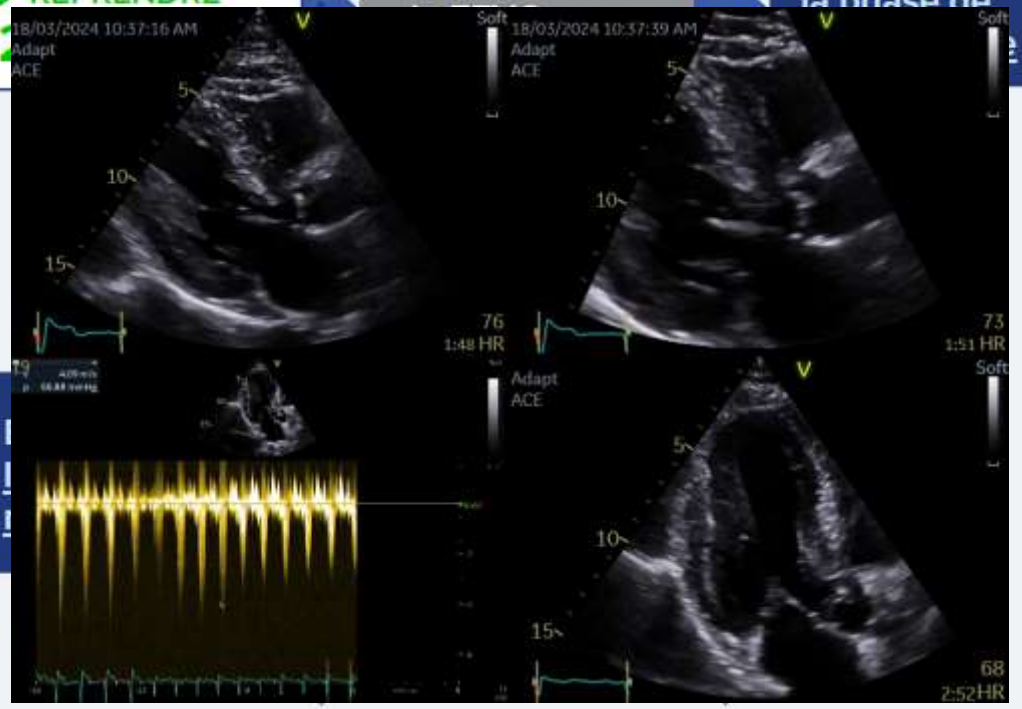
FEVG ≥ 50 %

▶ REPRENDRE

● VÉRIFIER

- l'état clinique
- le gradient CCVG avec manœuvre de Valsalva

Entrez dans la phase de



Open Bar sur le septum

- Septum
  - Focus on obstruction
  - But also increase Fibrosis > Diastolic HF and SCD
- Hypetrophic septum but not only
  - Location , severity
  - Impact on medical traitement
- Related LV obstruction
  - Accurate measurement
  - At rest but also after provocation
  - Different Cut Off 20 – 30 – 60 mmhg