

Amicale des **C**ardiologues de la **C**ôte d'**A**zur



Comment rechercher l'obstruction dans la cardiomyopathie hypertrophique?

F Levy

6 janvier 2026

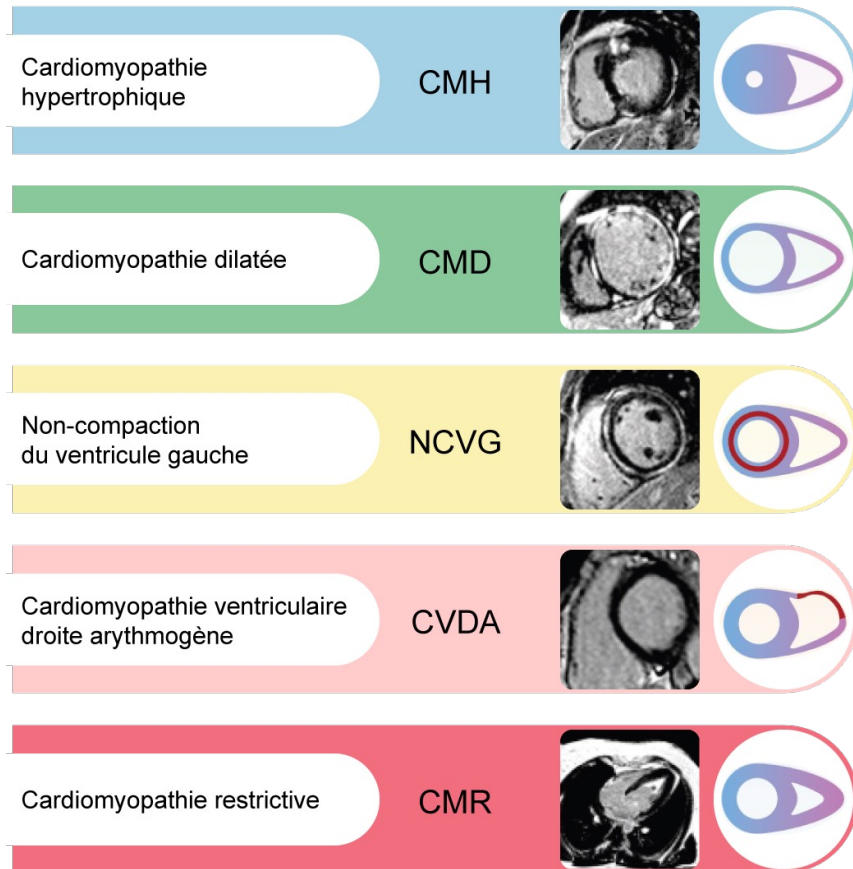
Nice

Sommaire

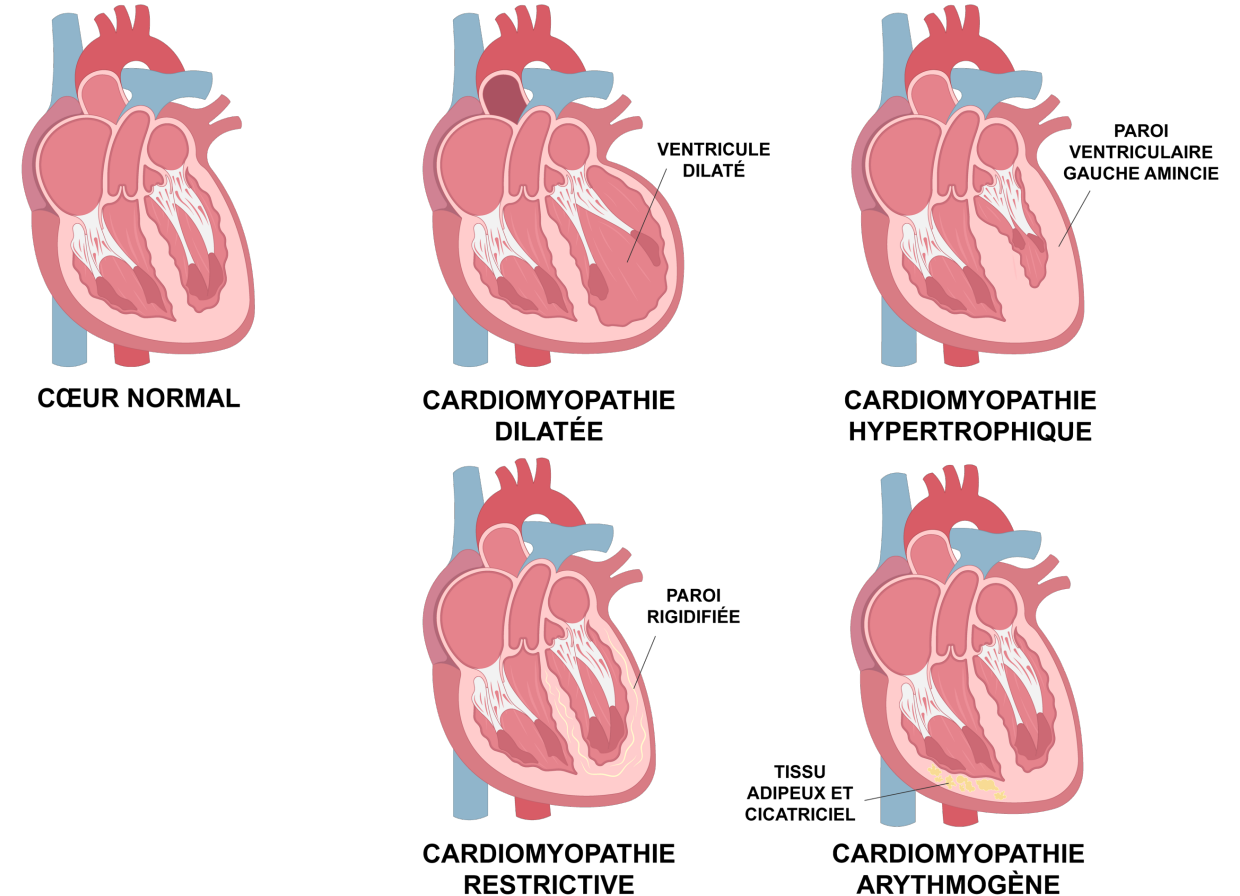
- Classification des cardiomyopathies
- Définition de l'obstruction intra-Ventriculaire Gauche (intra-VG)
- Méthodes de recherche de l'obstruction, de provocation
- Conclusion



Classification des cardiomyopathies



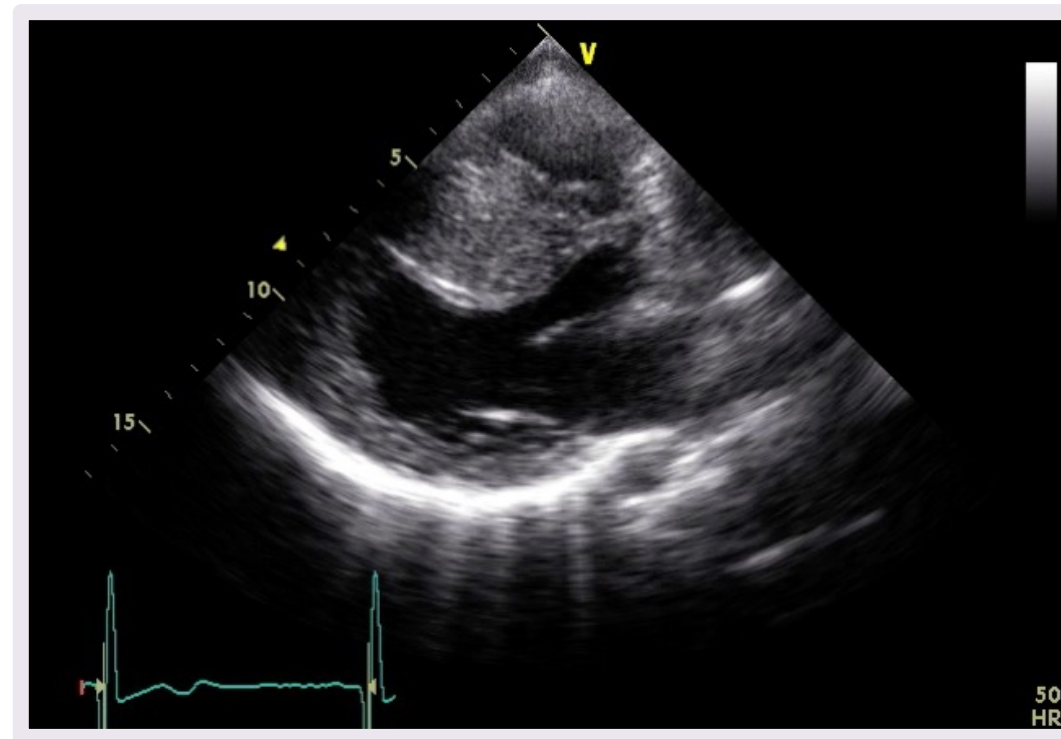
Classification par phénotype



Classification par sous-types morphologiques de cardiomyopathies héréditaires

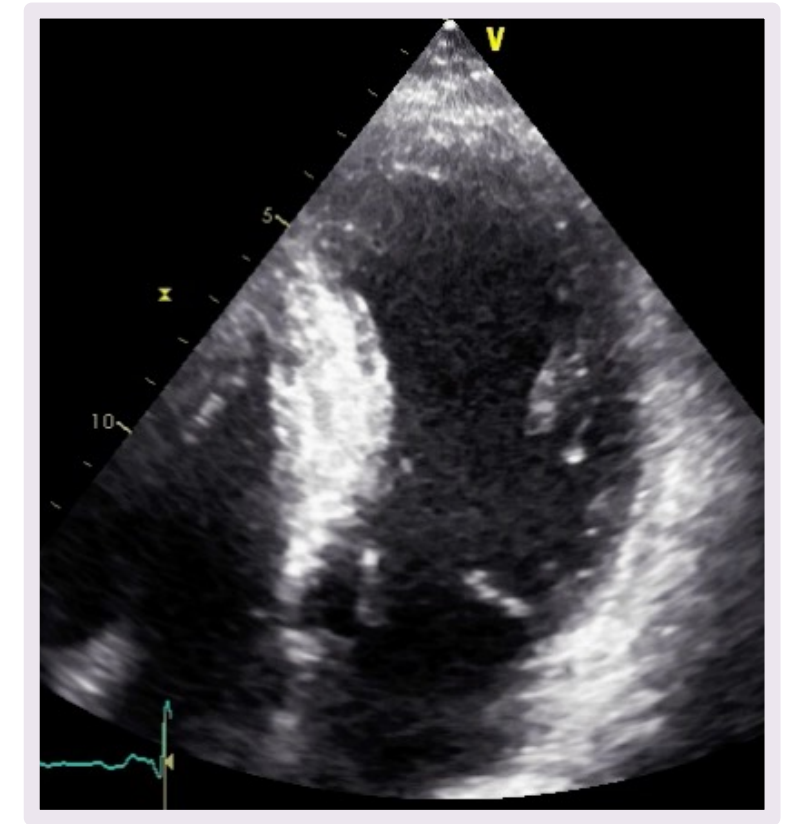
Définition de la CMH

La CardioMyopathie Hypertrophique (CMH) est caractérisée par un épaissement anormal des parois du cœur d'origine primitive sans cause clinique décelable (HTA sévère, de sténose aortique serrée, de maladie de surcharge...) capable de produire l'hypertrophie



Épidémiologie de la CMH

- **Prévalence estimée** : 1/200 à 1/500
- **Cause principale de mort subite** chez les jeunes notamment chez le sportif de moins de 35 ans
- **Mutation génétique** retrouvée dans 40 à 60% des cas
 - 42 % sur le gène MYBPC3
 - 40 % sur MYH7
 - Forme familiale **Autosomique Dominante (AD)** dans la majorité des cas
 - **Pénétrance incomplète liée à l'âge** :
 - 55 % avant 30 ans
 - 75 % entre 30 et 50 ans
 - 95 % après 50 ans



Définition de l'Hypertrophie Ventriculaire Gauche (HVG)

Cardiomyopathie hypertrophique (CMH) sarcomérique

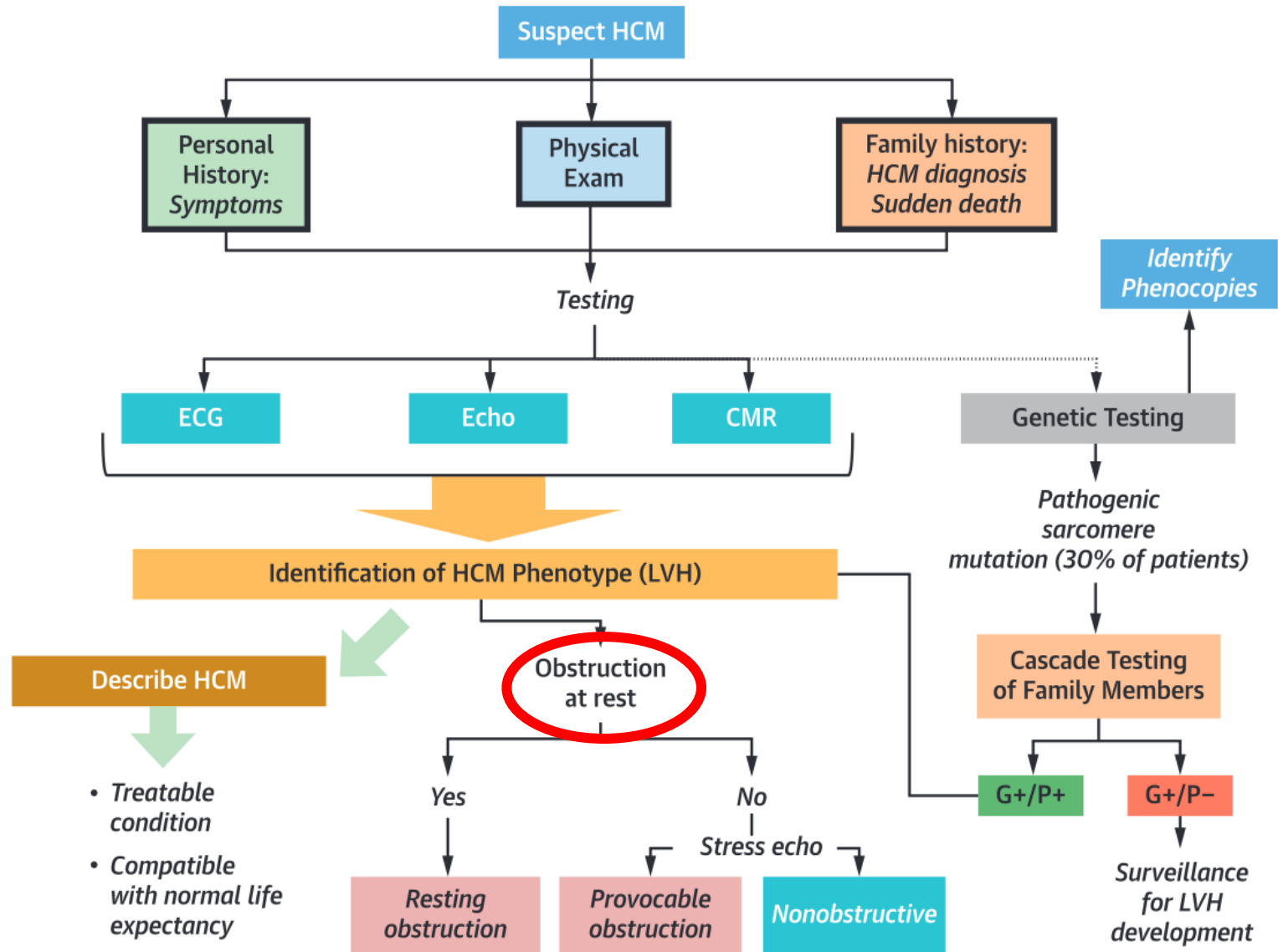
Cut-off de l'HVG

- Dans les formes sporadiques : ≥ 15 mm
- Dans les formes familiales : ≥ 13 mm

Amylose et Fabry

Cut-off de l'HVG

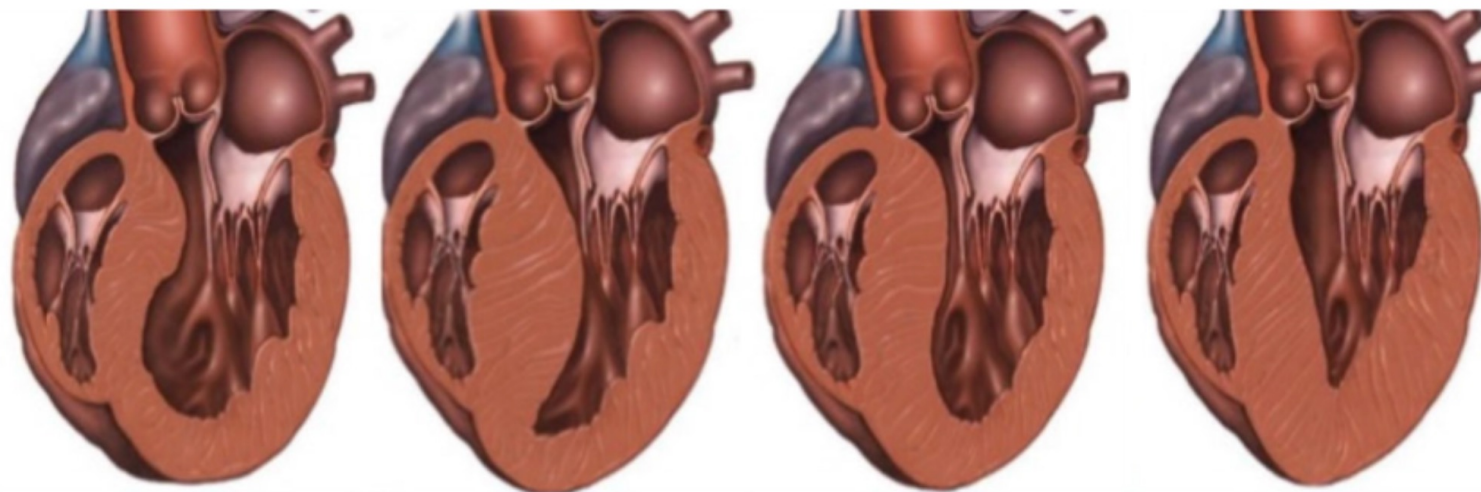
- 12 mm chez l'homme
- 11 mm chez la femme



Maron, B.J. et al. J Am Coll Cardiol. 2022;79(4):372-389.

Cardiomyopathy phenotype	
HCM	Sarcomeric
	Anderson–Fabry
	Danon
	TTR amyloidosis
	RASopathy
	Friedreich ataxia
	Mitochondrial
	Mitochondrial DNA
	Nuclear DNA

Cardiomyopathy phenotype	
HCM	Sarcomeric
	Anderson–Fabry
	Danon
	TTR amyloidosis
	RASopathy
	Friedreich ataxia
	Mitochondrial
	Mitochondrial DNA
DCM	Nuclear DNA
	<i>LMNA</i>
	<i>RBM20</i>
	Sarcomeric
	Dystrophin
	Emerin
	Barth syndrome
	Mitochondrial
	Mitochondrial DNA
NDLVC	Nuclear DNA
	<i>LMNA</i>
	<i>DES</i>
	<i>FLNC</i>
	<i>PLN</i>
	<i>TMEM43</i>
ARVC	<i>RBM20</i>
	<i>PLN</i>
	Desmosomal
	<i>TMEM43</i>
RCM	Sarcomeric
	<i>DES</i>
	<i>FLNC</i>
	<i>BAG3</i>
	RASopathy

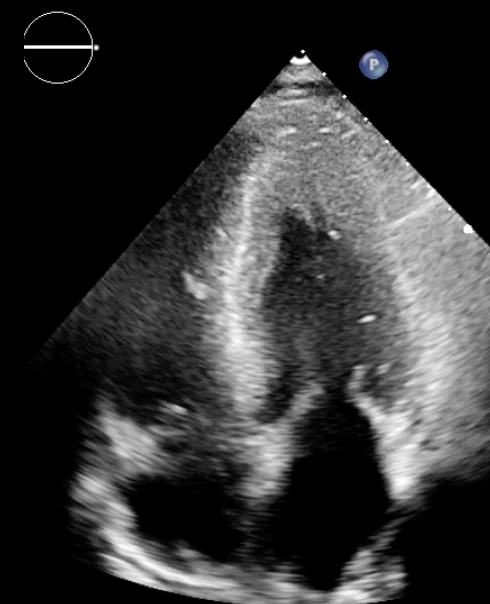
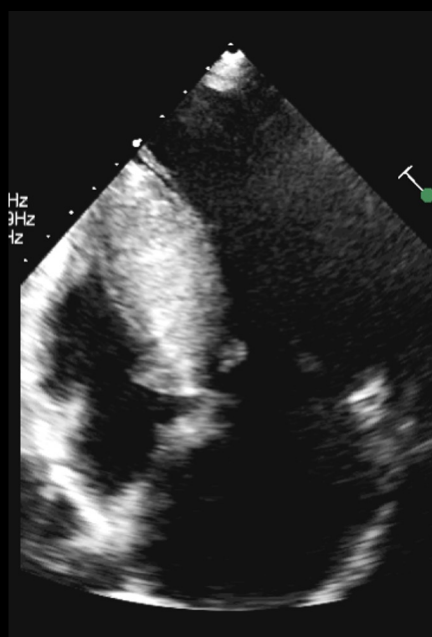


Sigmoid

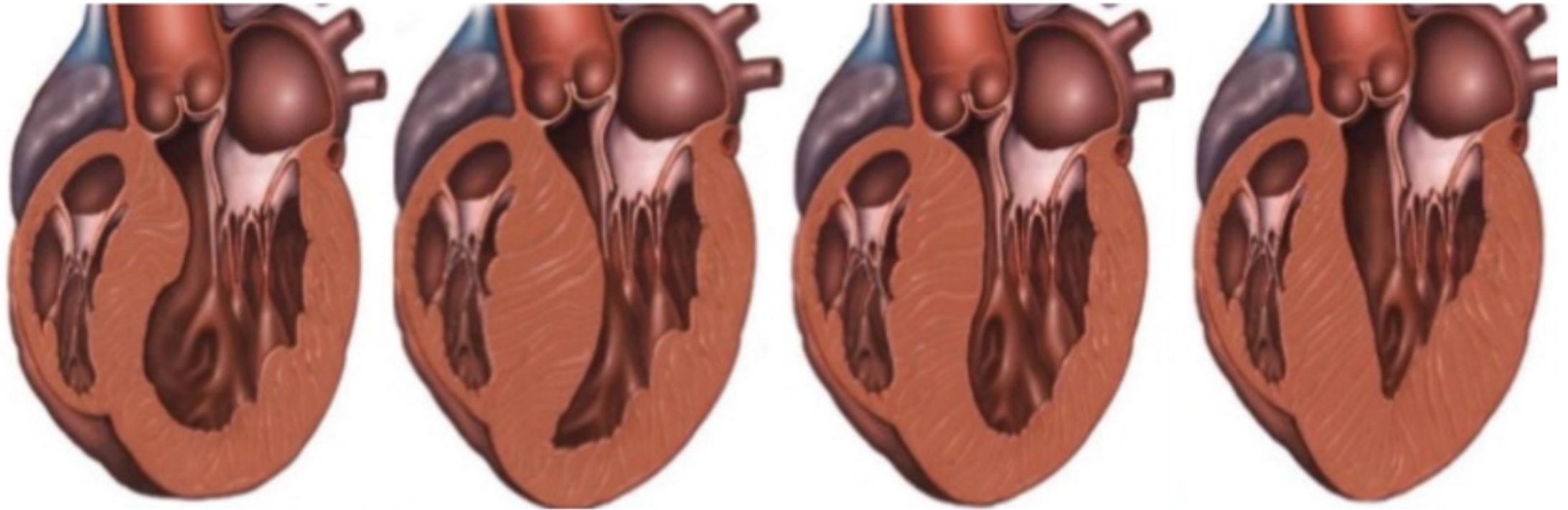
Reverse curve

Neutral

Apical



Variants/septal morphology



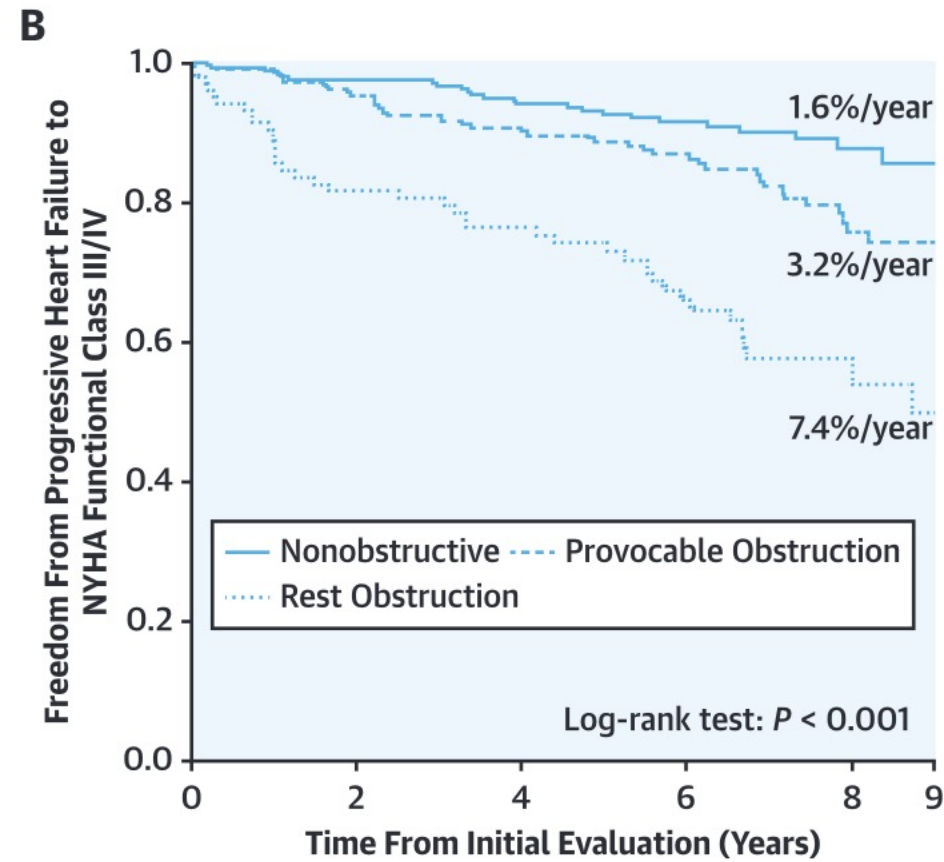
Sigmoid

Reverse curve

Neutral

Apical

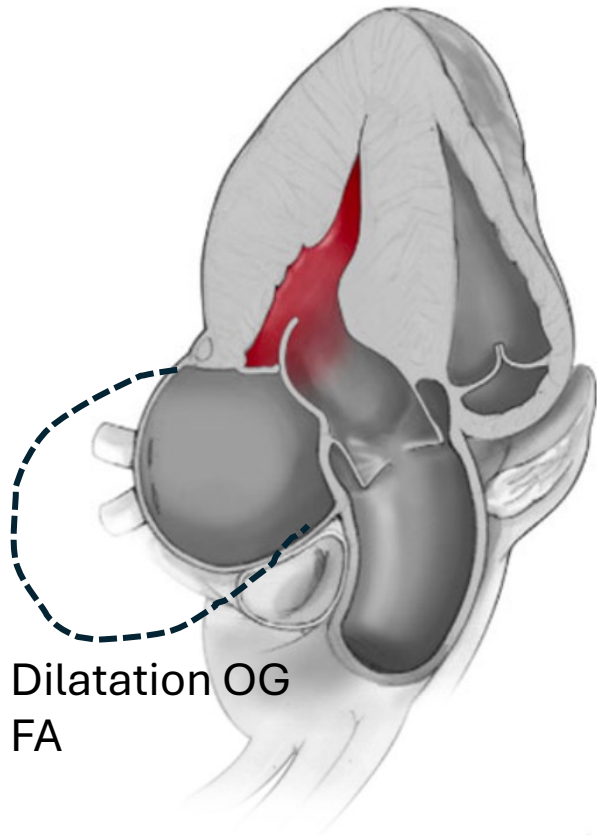
Rôle pronostic de l'obstruction



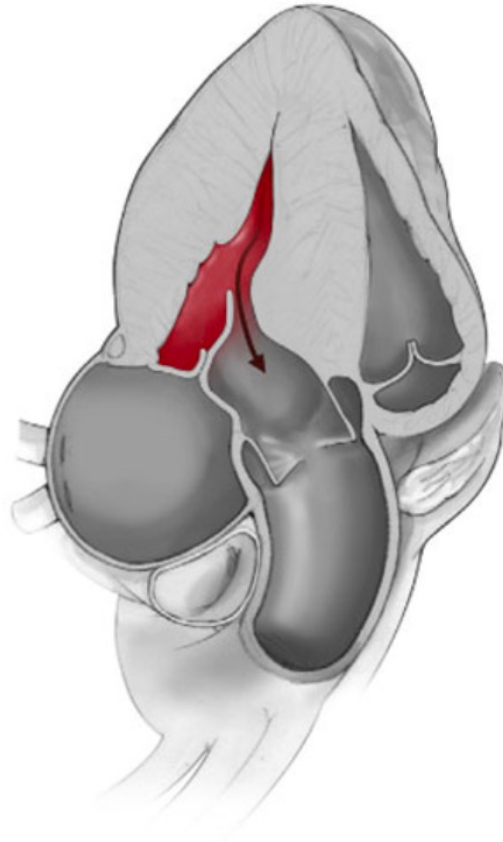
Conséquences de l'HVG

- 4 problèmes :

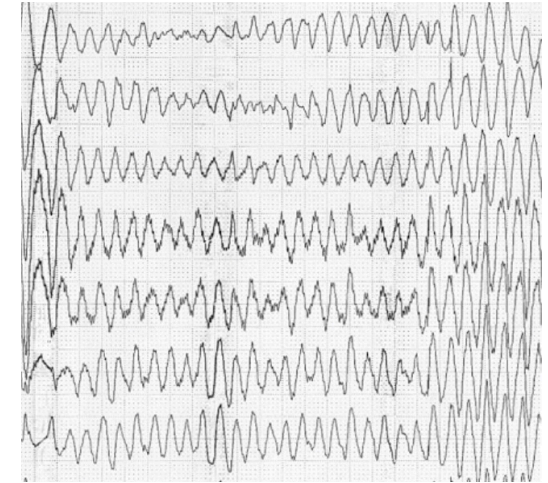
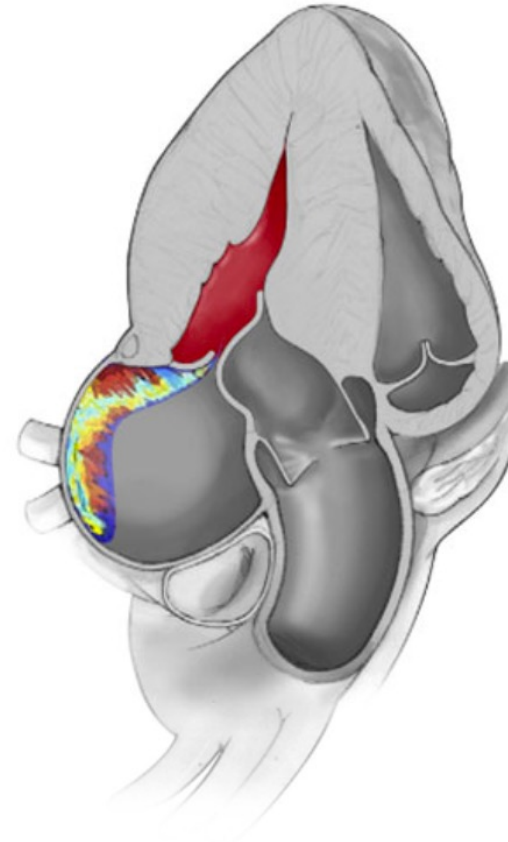
Remplissage VG

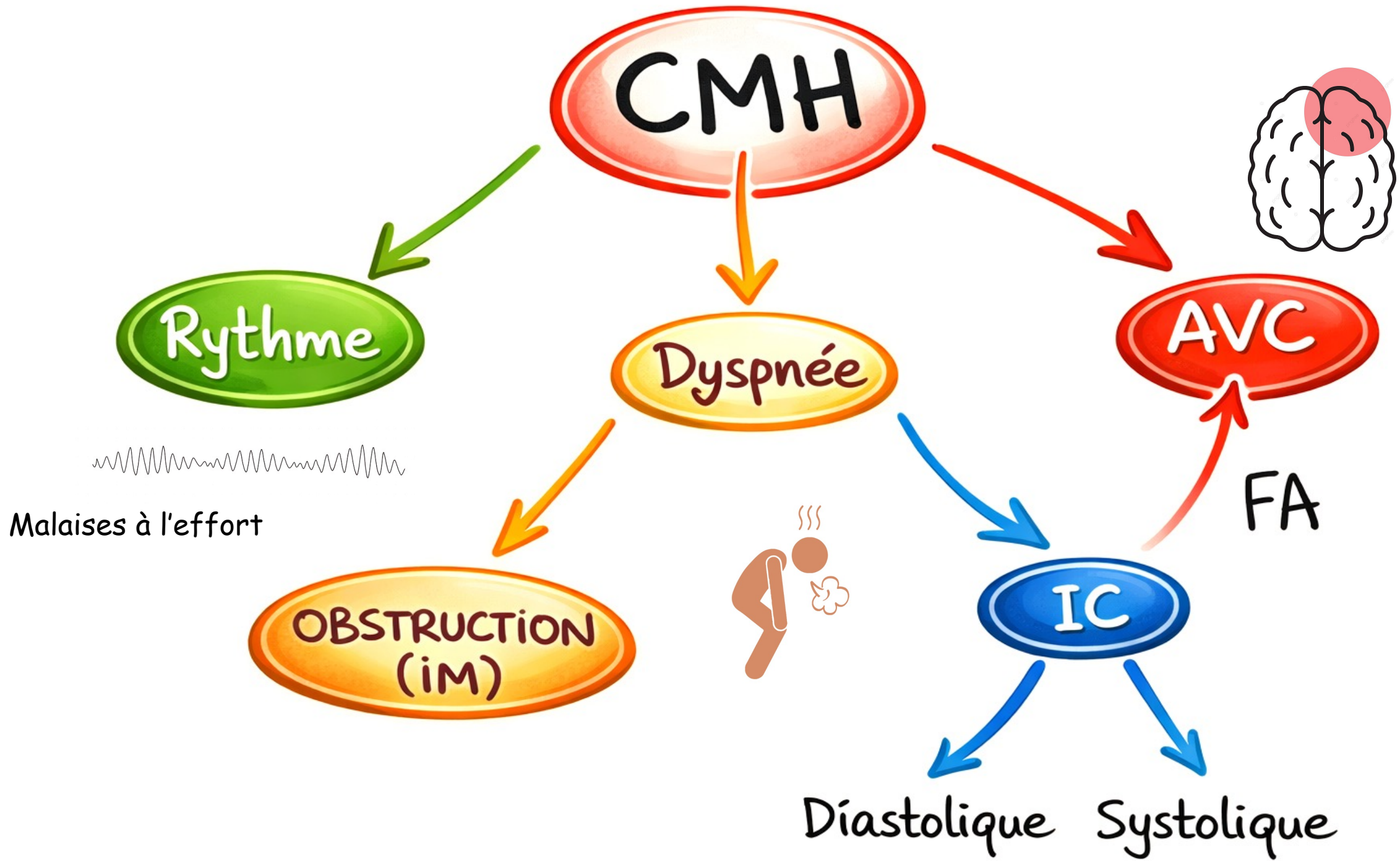


Obstruction



Fuite mitrale





Attention aux formes évoluées (end stage)

End-Stage HCM: LVEF <50%

ASE2023

- End stage HCM defined as EF < 50%.
 - ~3-5% of HCM population.
 - Younger at dx, family hx, MYH7
 - ~15-20% on long term f/up (SHaRe Registry)



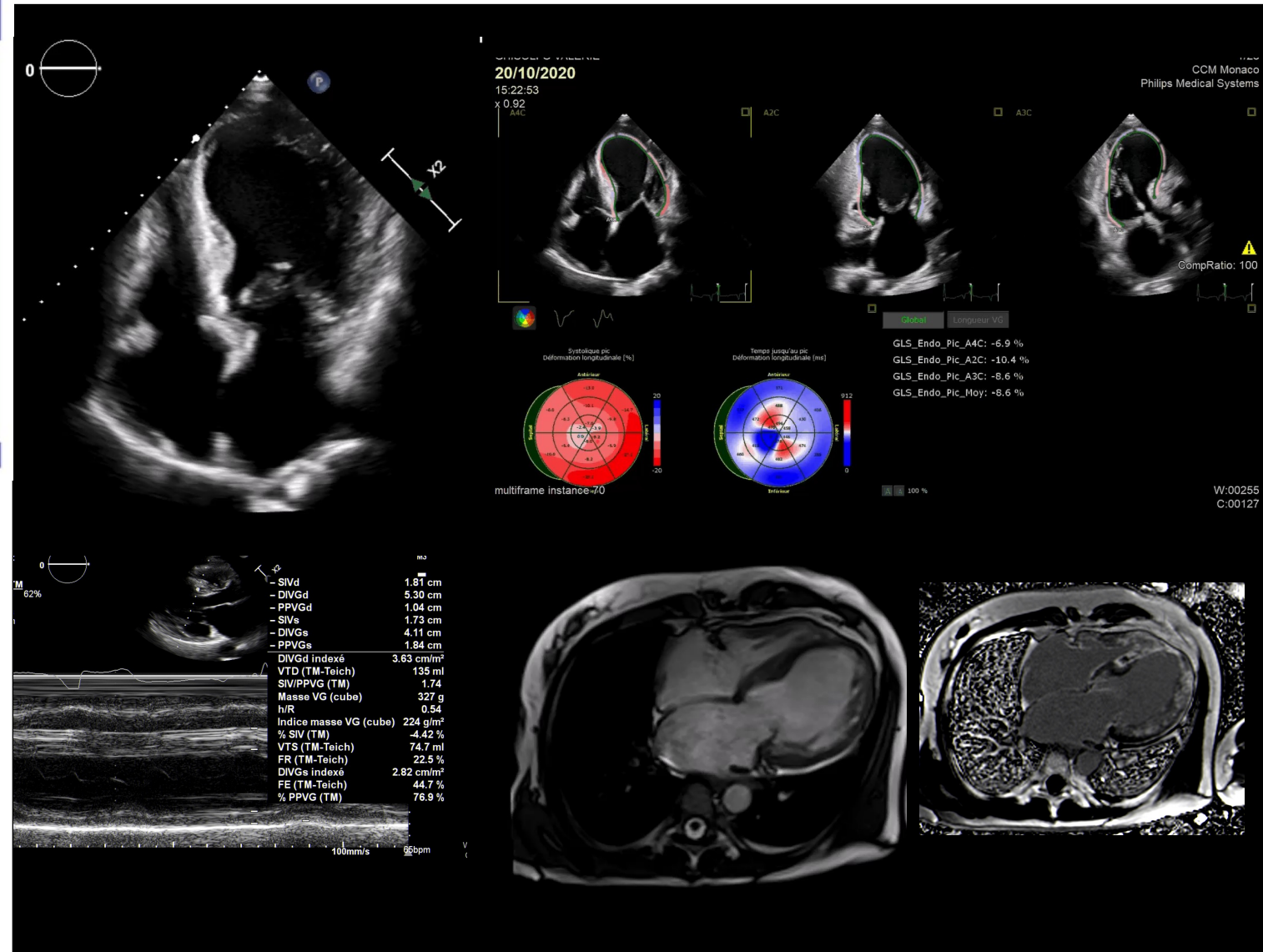
Harris KM, et al. Circulation 2006; Manstrand P et al. Circulation 2020; 141:1371-1383.
Ho CY et al. Circulation 2018

#ASE2023 ASEScientificSessions.org

Extensive LGE

- HCM a fibrotic disease:
 - Interstitial fibrosis in hypertrophied segments
- LGE present in >50% of HCM patients.
 - Progressive: avg \uparrow 0.5% / year
- Quantifiable: validated using 5-6 SD or "full width half-maximum" methods.

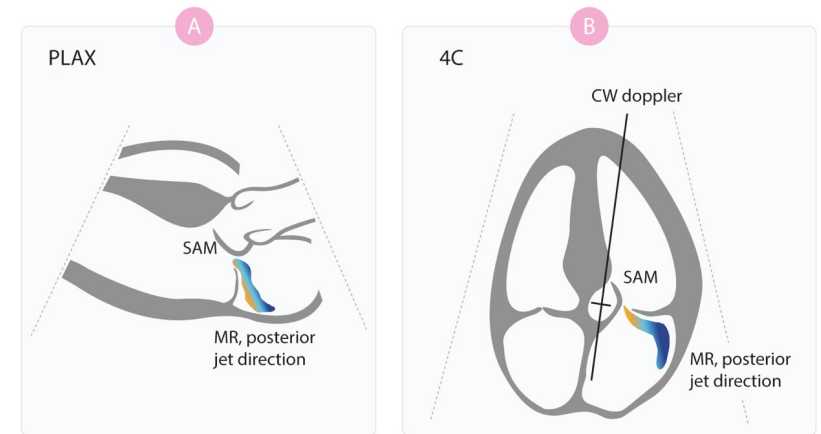
Habib M et al, JACC Cardiovasc Imaging 2021



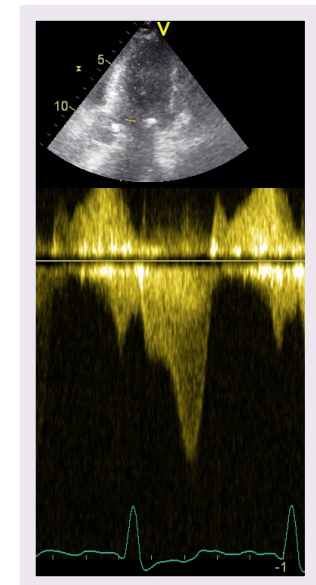
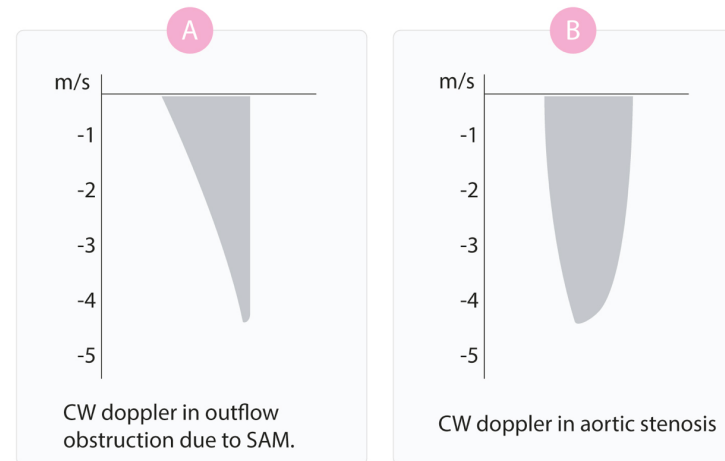
Définition de l'obstruction intra-VG

- **Échocardiographie de repos** : Doppler pulsé, couleur et continu ++ (cc VG++, apex, chambre de chasse VD), SAM ? En TM
- **Gradient max > 30 mmHg** (au repos) en Doppler continu
- Mais... phénomène labile, dynamique

Septal hypertrophy with SAM and mitral regurgitation (MR)



CW doppler in outflow obstruction vs. aortic stenosis



Qu'est-ce qu'une obstruction significative?

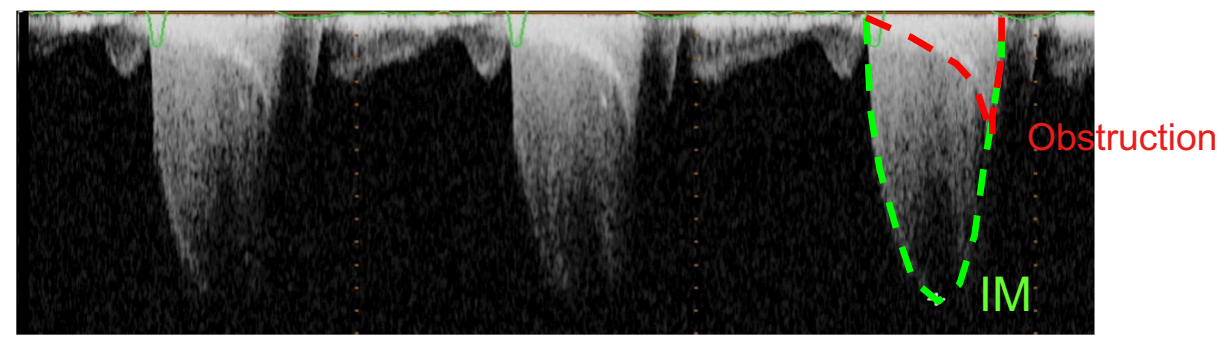
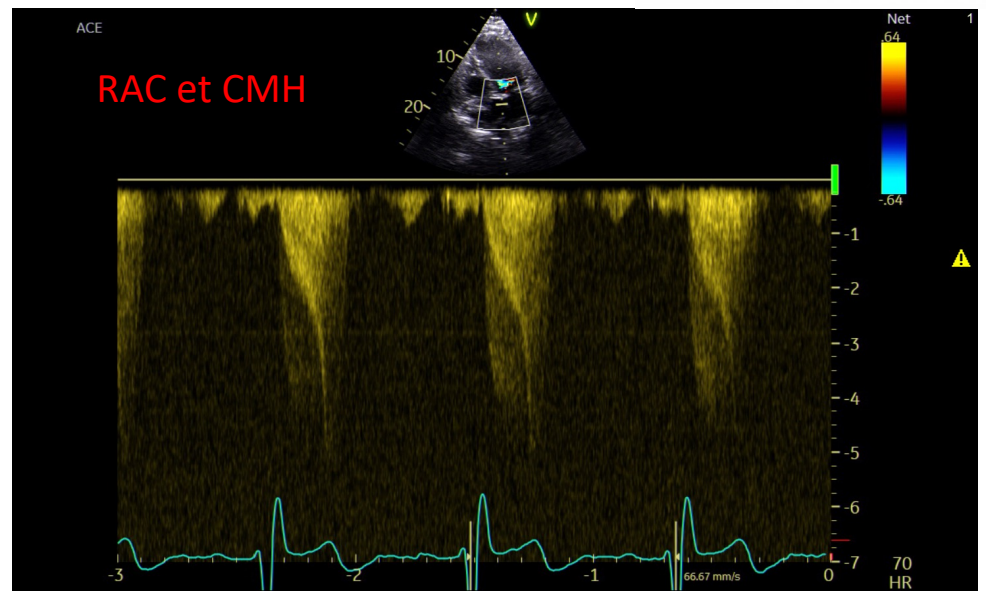
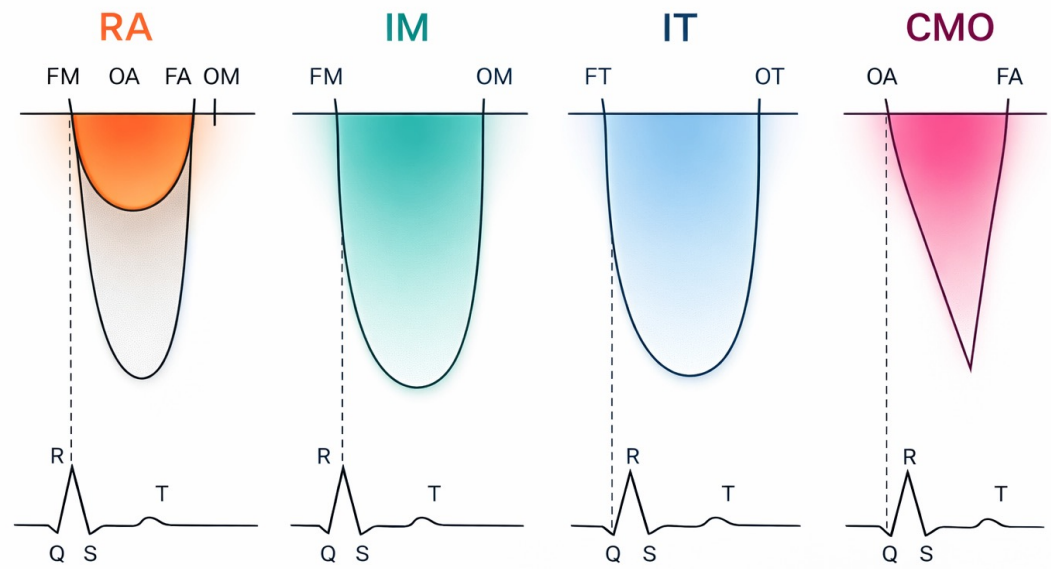
What is significant obstruction?

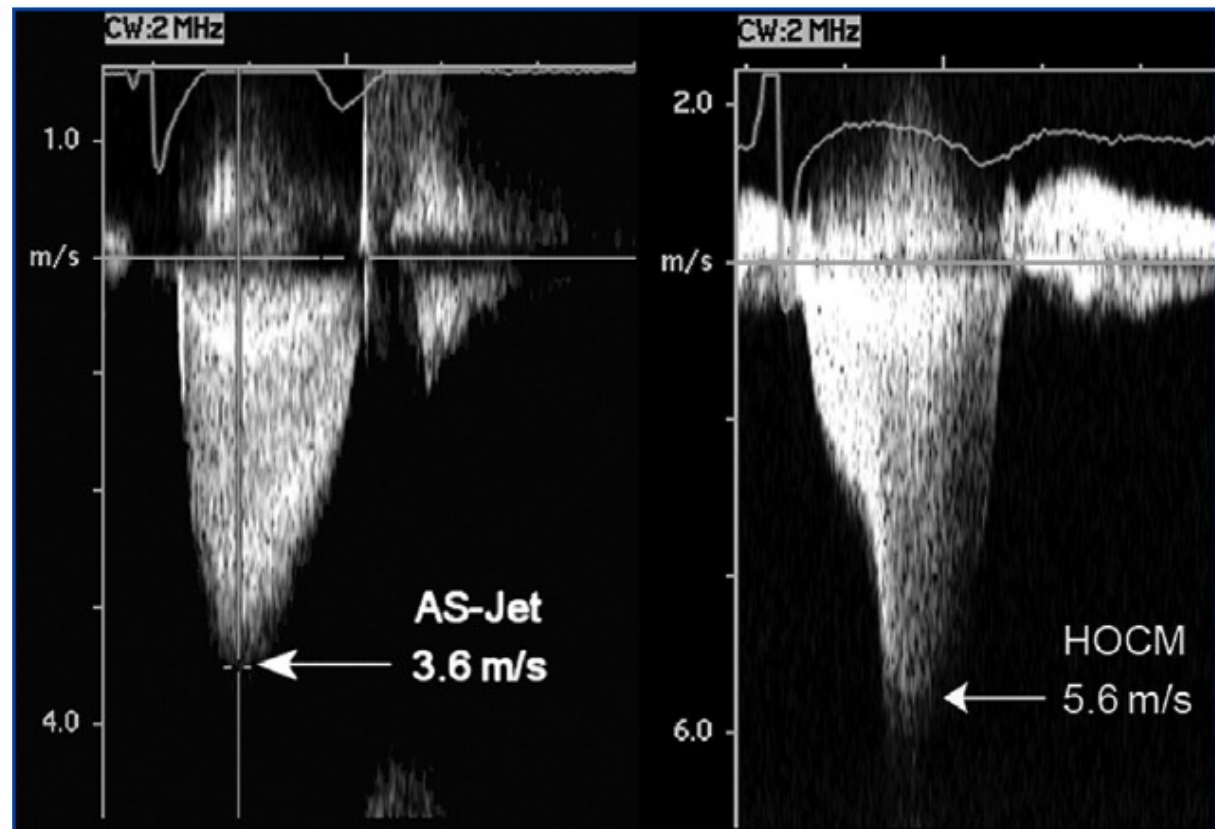
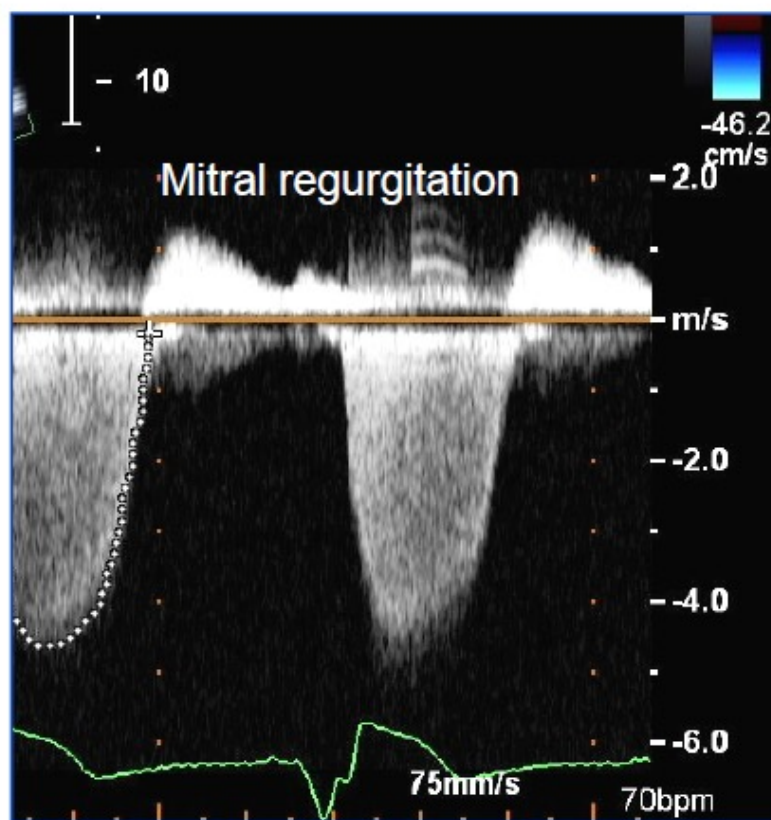
Obstructive HCM

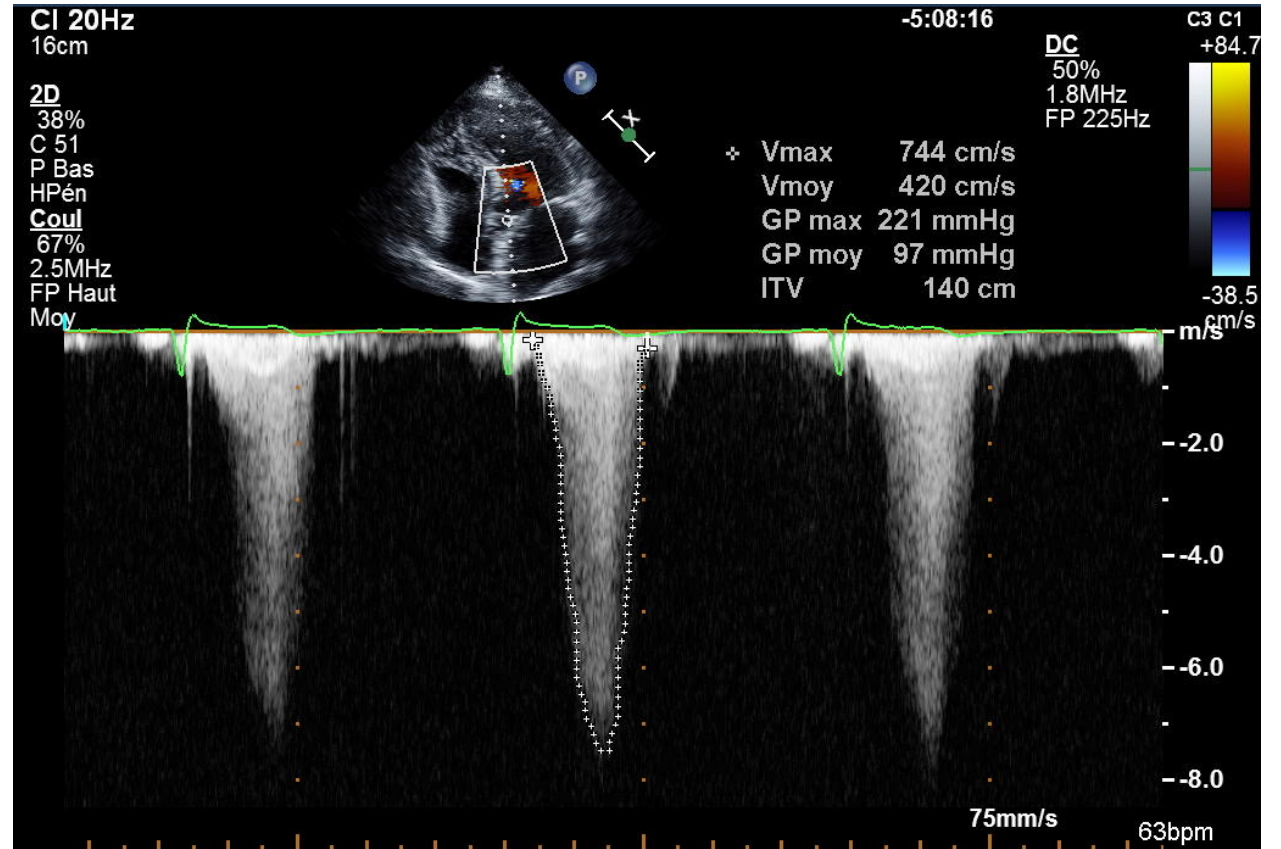
Peak Gradient > 30 at rest

Sufficient to result in symptoms

Peak Gradient >40-50

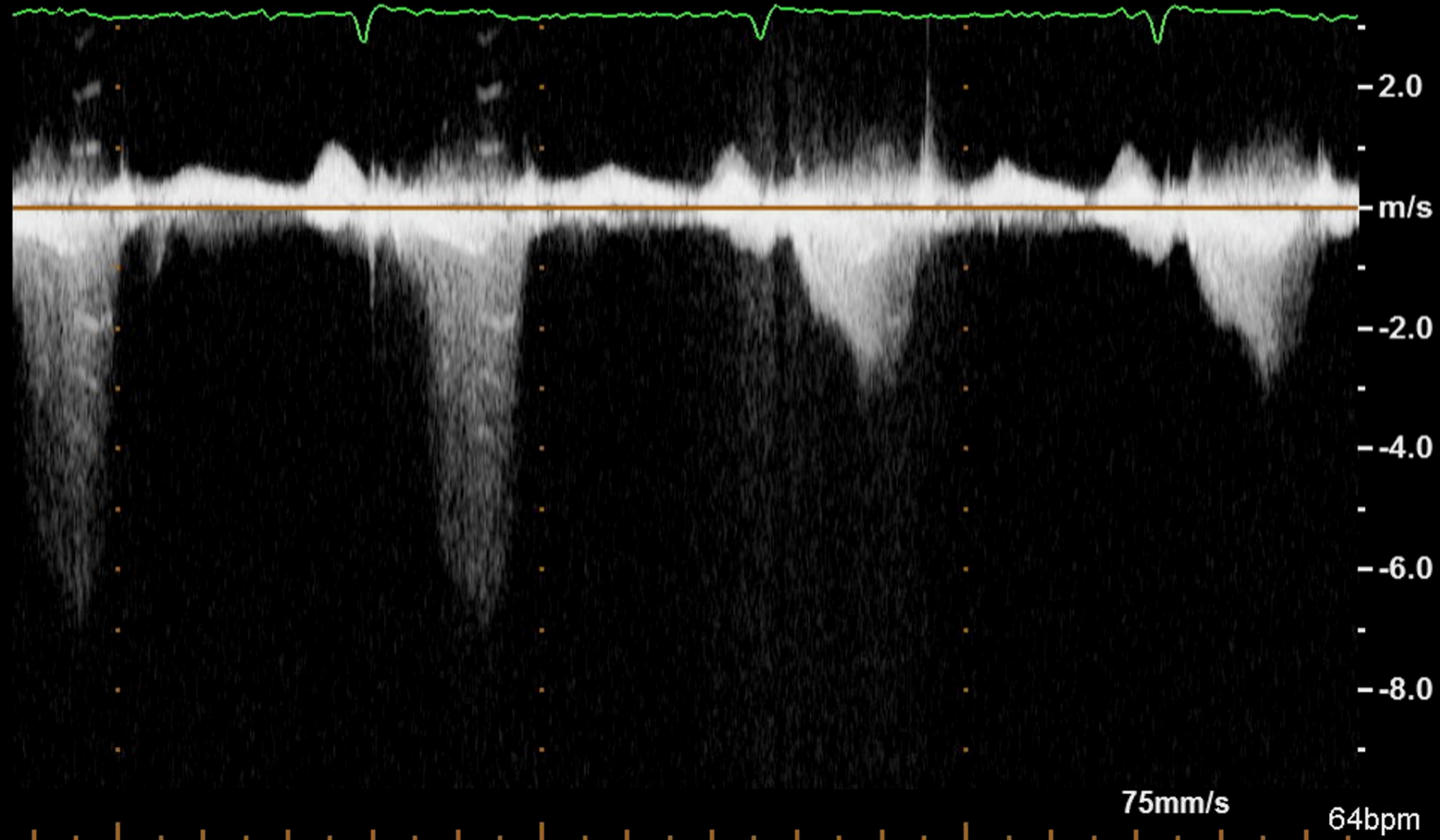






-5:08:16

DC
70%
2.0MHz
FP 225Hz



CI 24Hz
15cm

2D

32%

C 51

P Bas

HPén

Coul

67%

2.5MHz

FP Haut

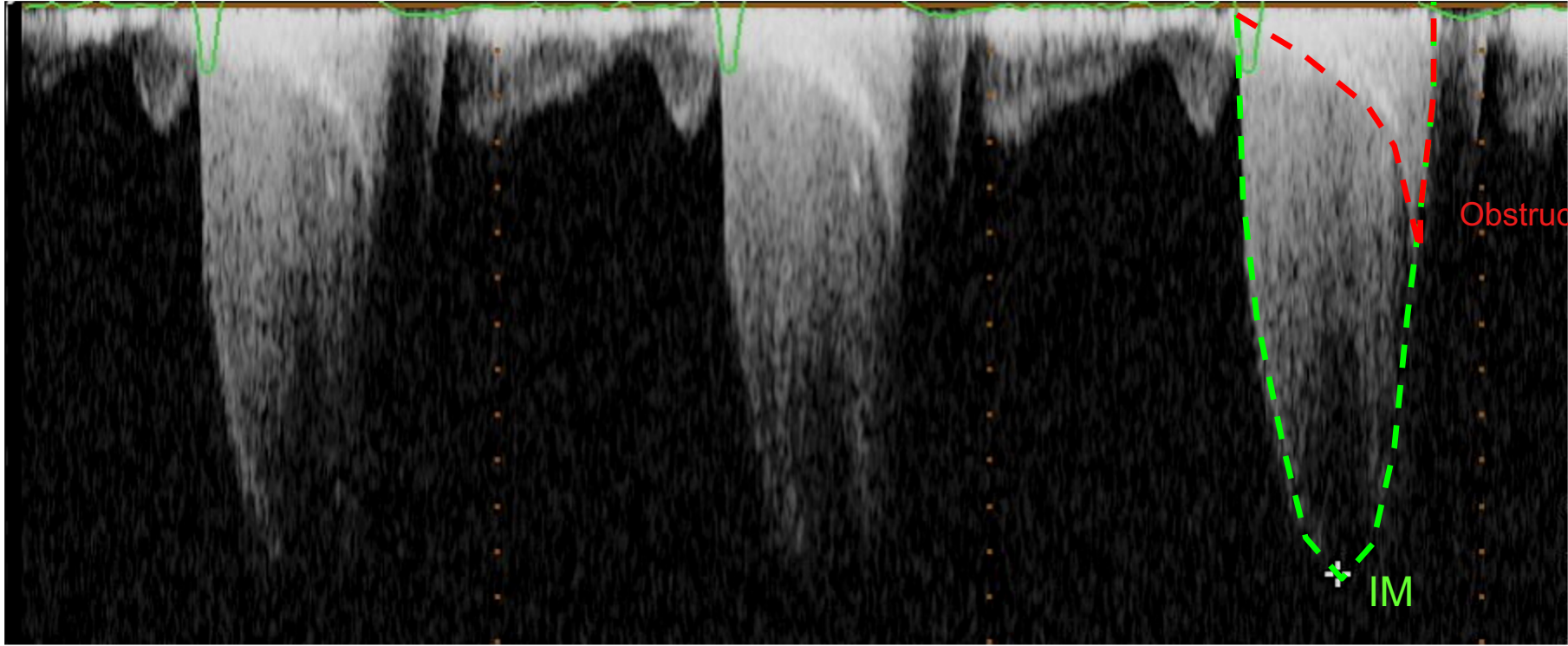
Moy

C3 C1
+61.6

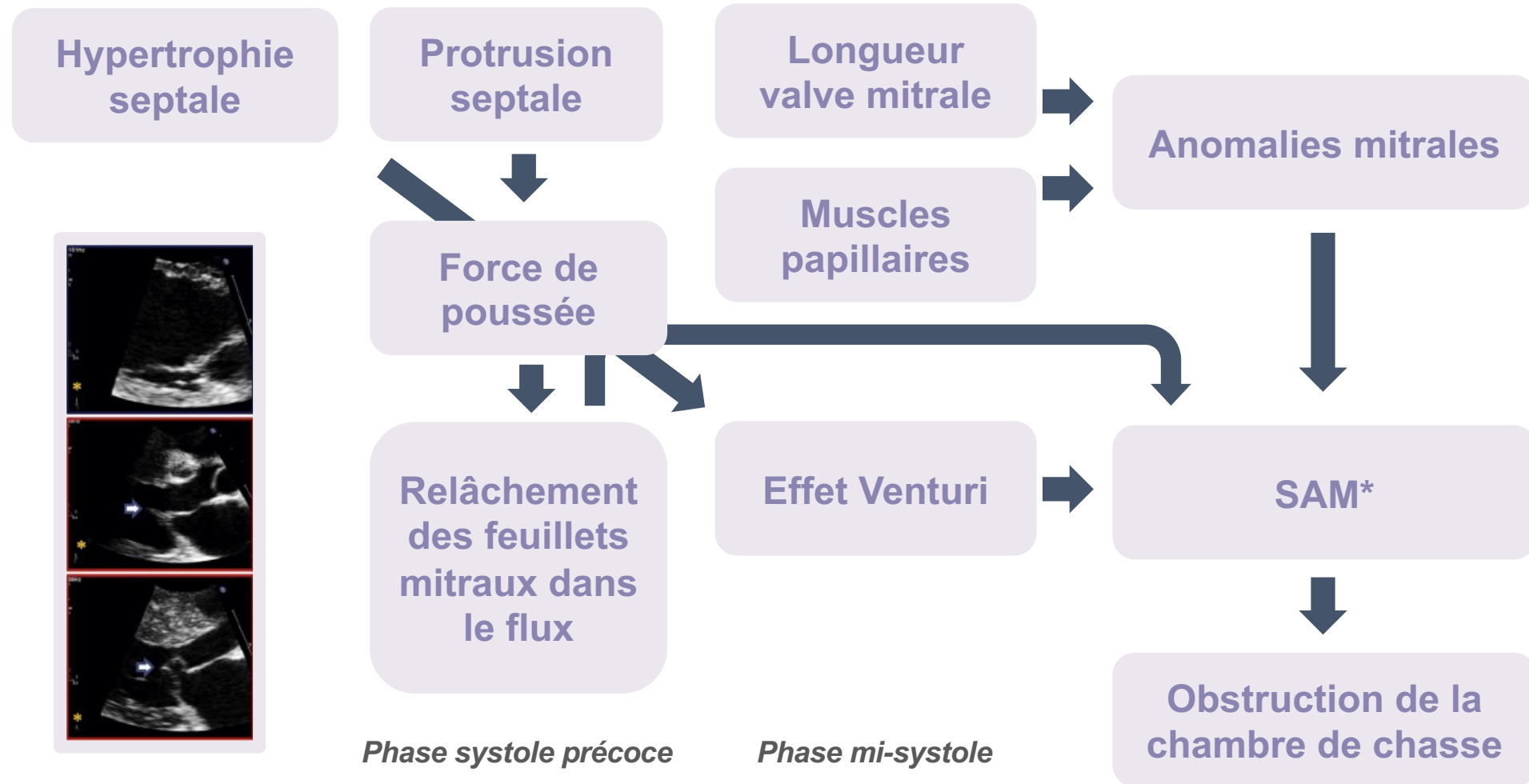


JPEG

65 bpm

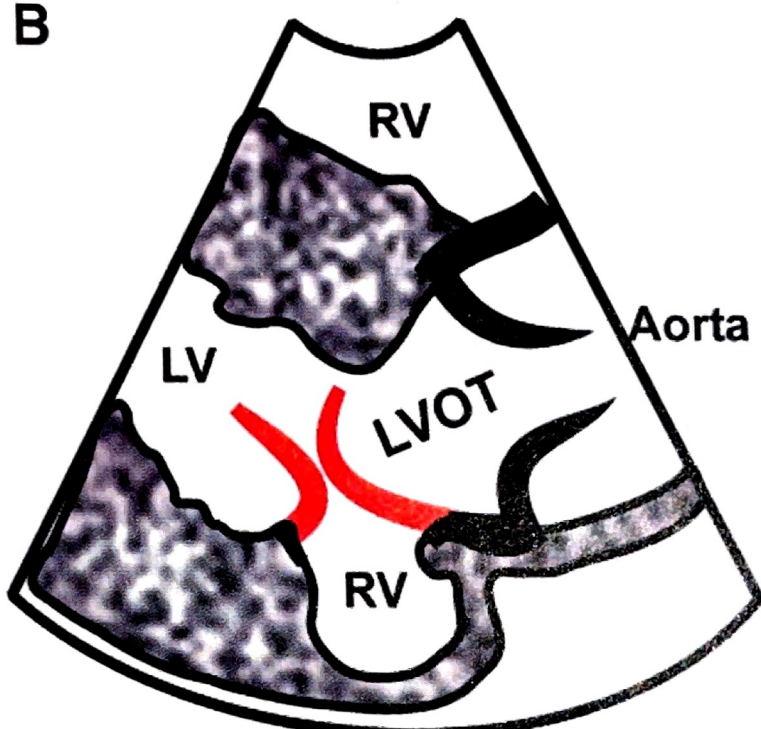
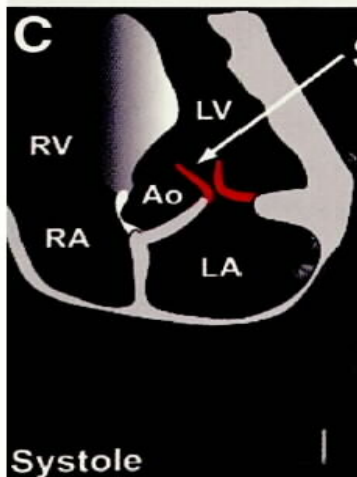
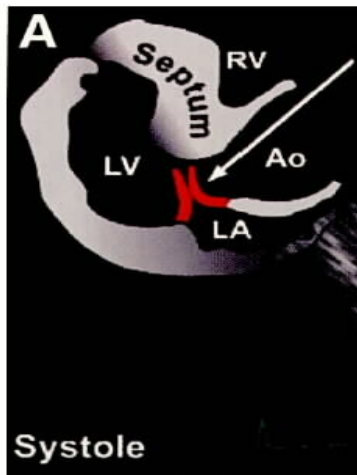


Physiopathologie : un mécanisme complexe

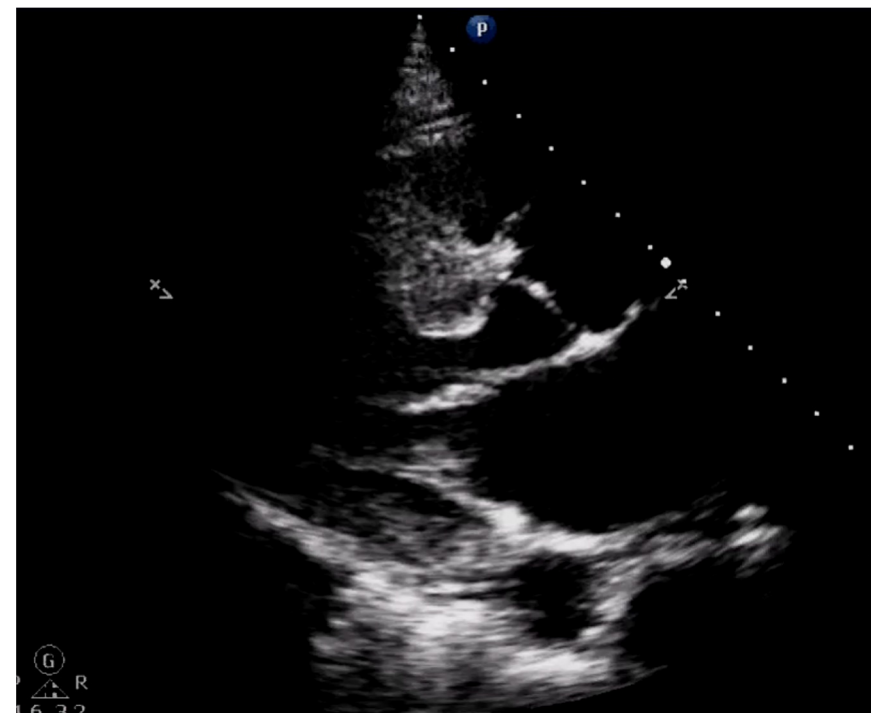
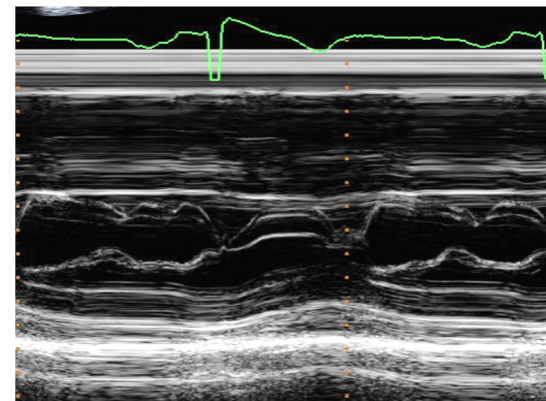
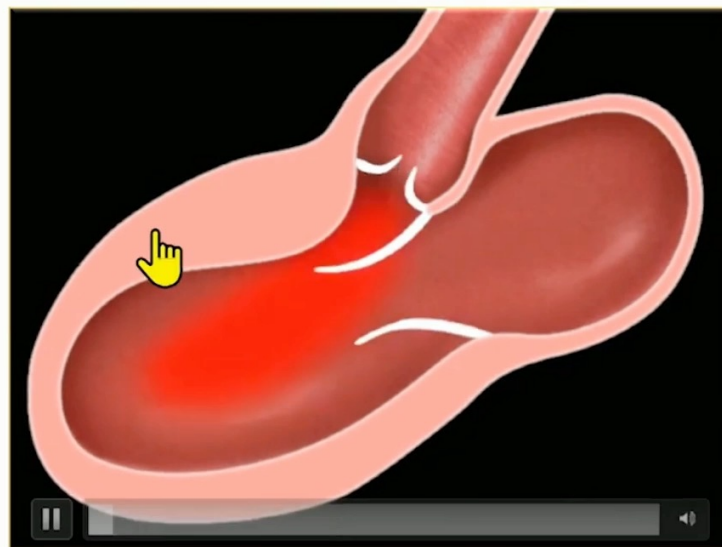


* SAM : Systolic Anterior Motion = Mouvement Systolique Antérieur.

11. Abbasi M *et al.* J Am Soc Echocardiogr 2024;37:613-25.



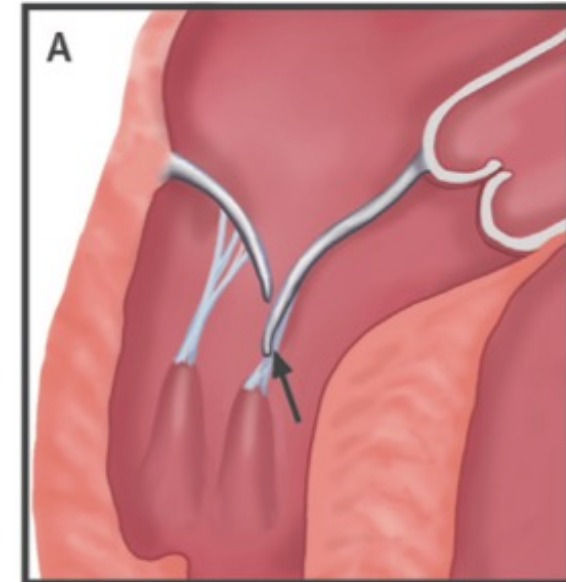
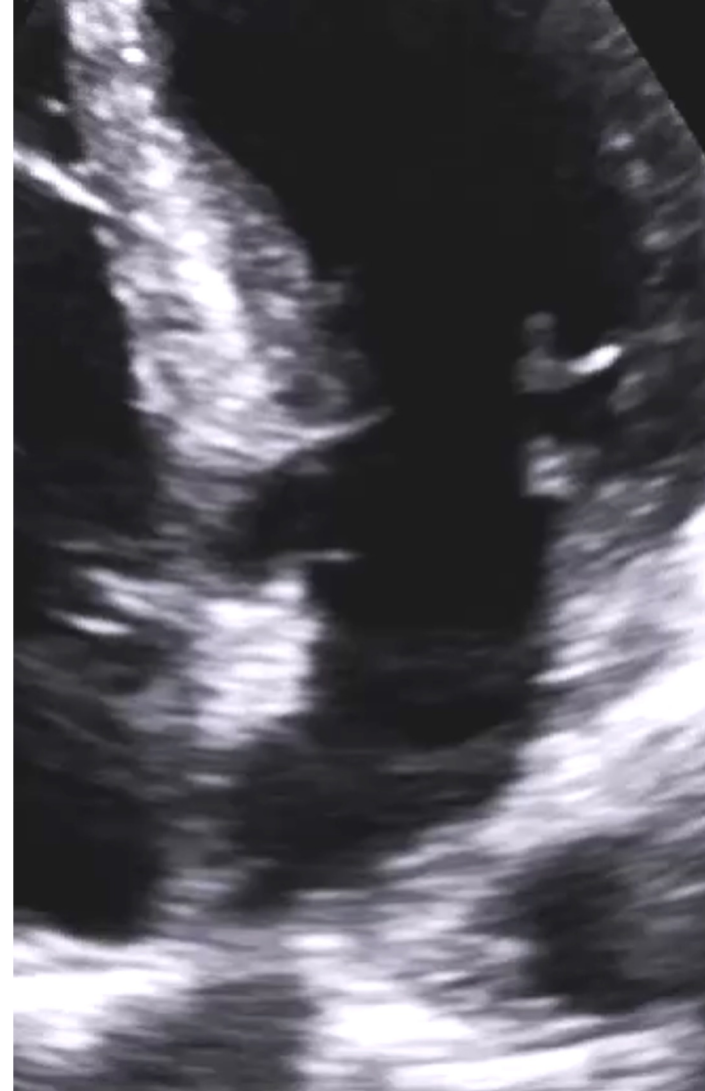
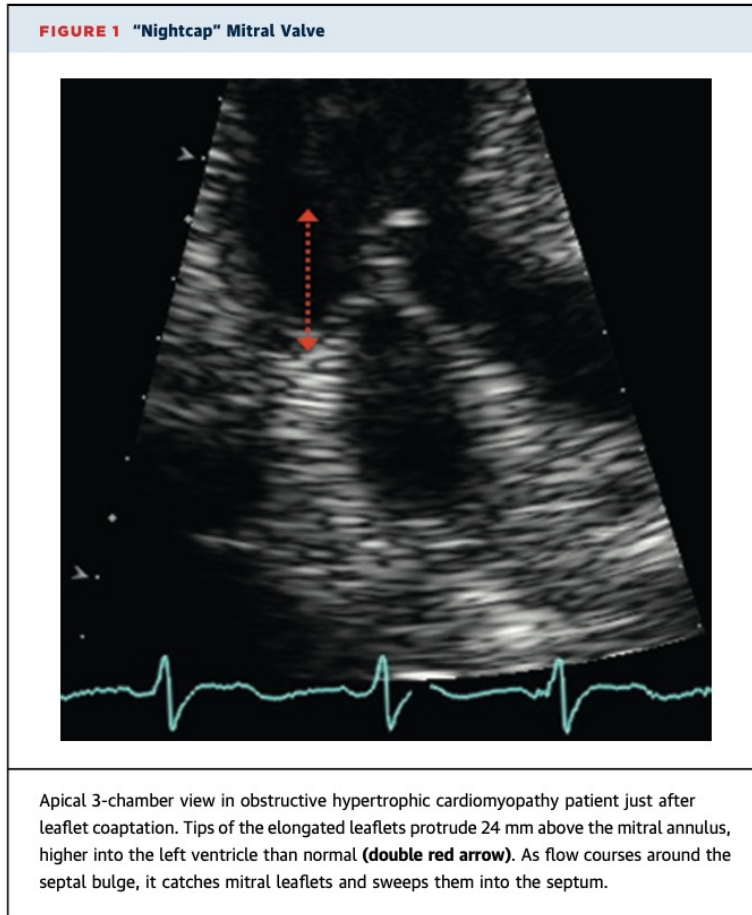
Obstruction



Le SAM un excès de longueur des valves ?

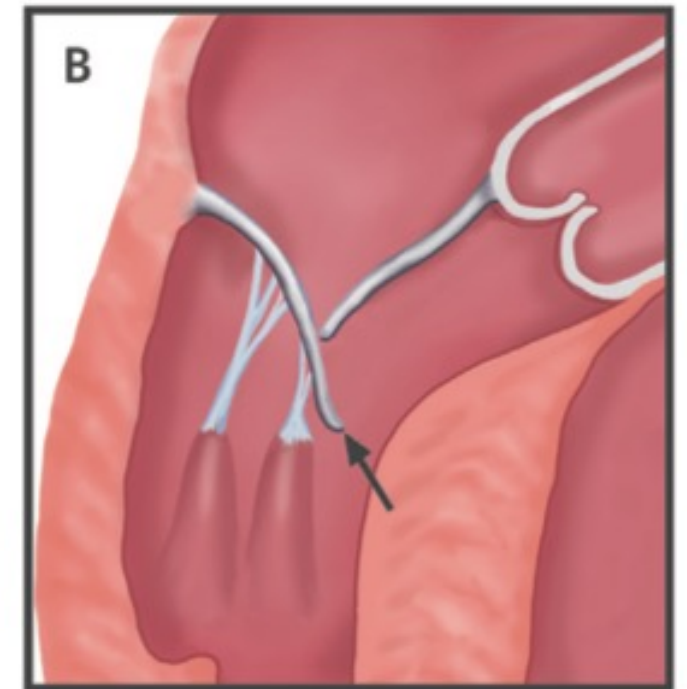
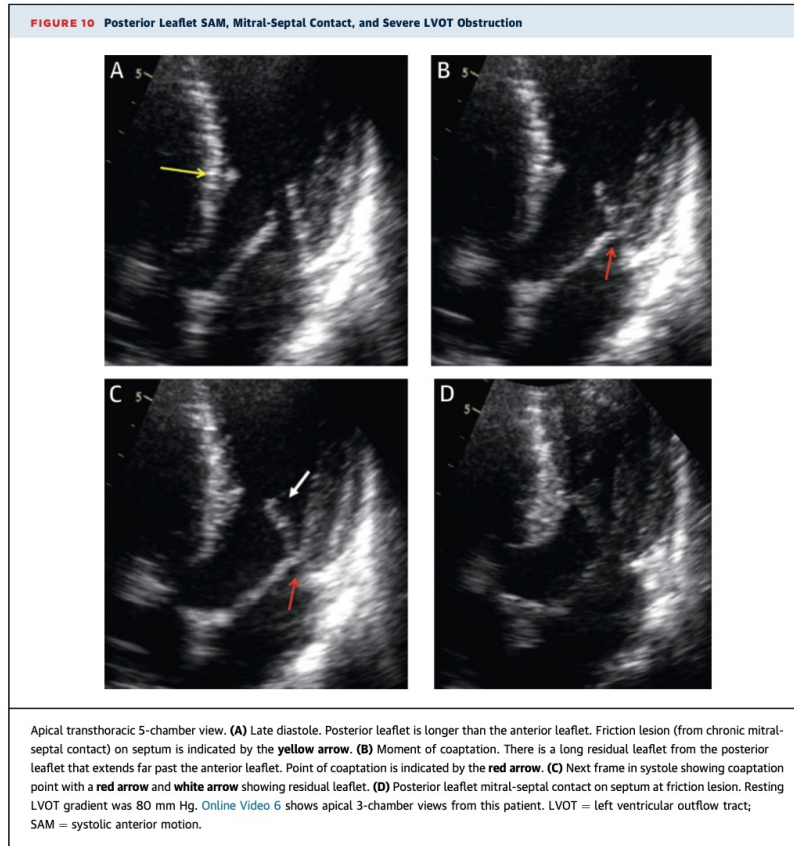
La majorité des patients avec CMHO ont des feuillets ant et post mitraux **allongés** :

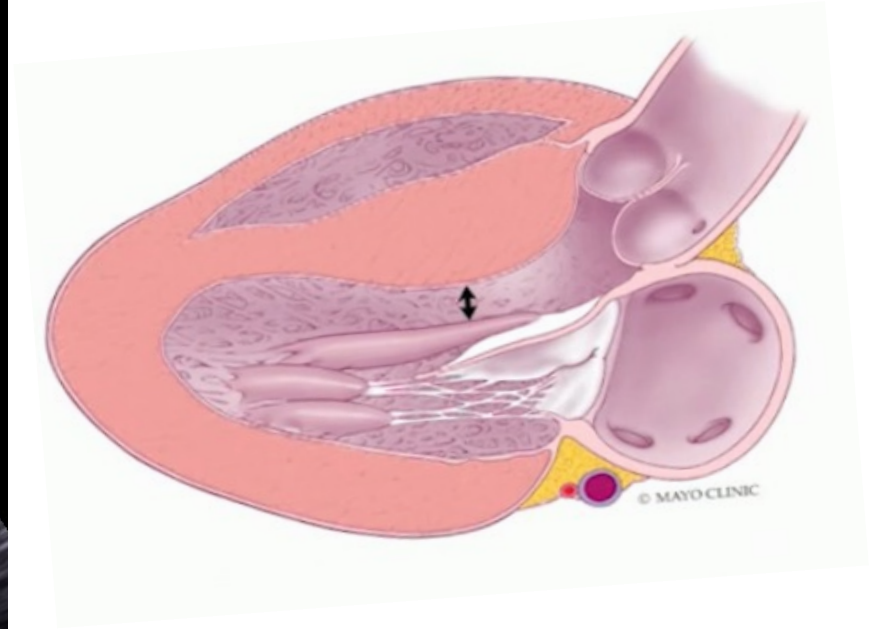
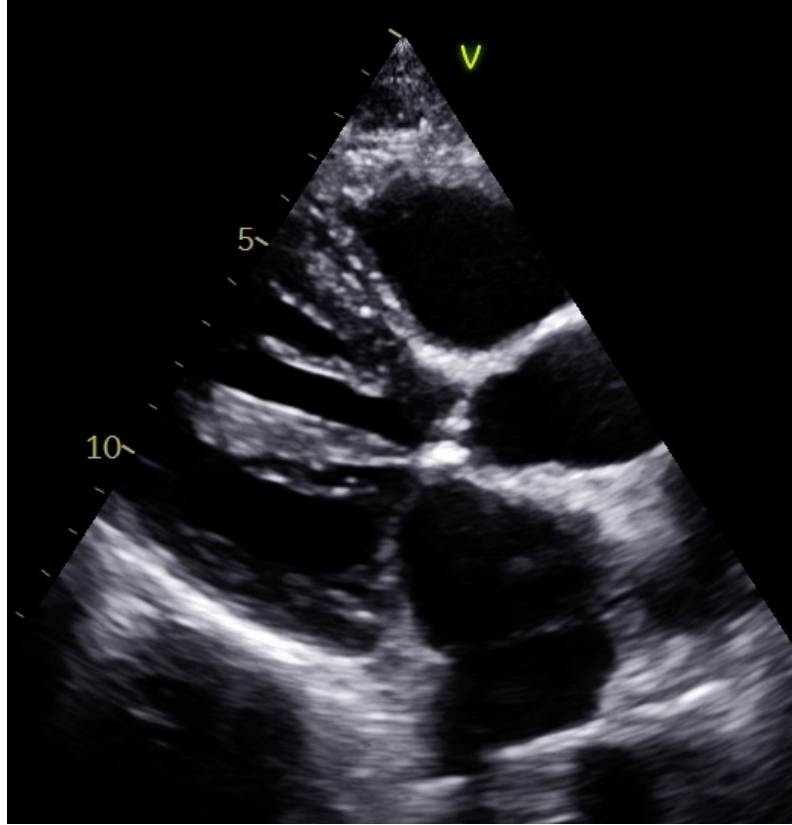
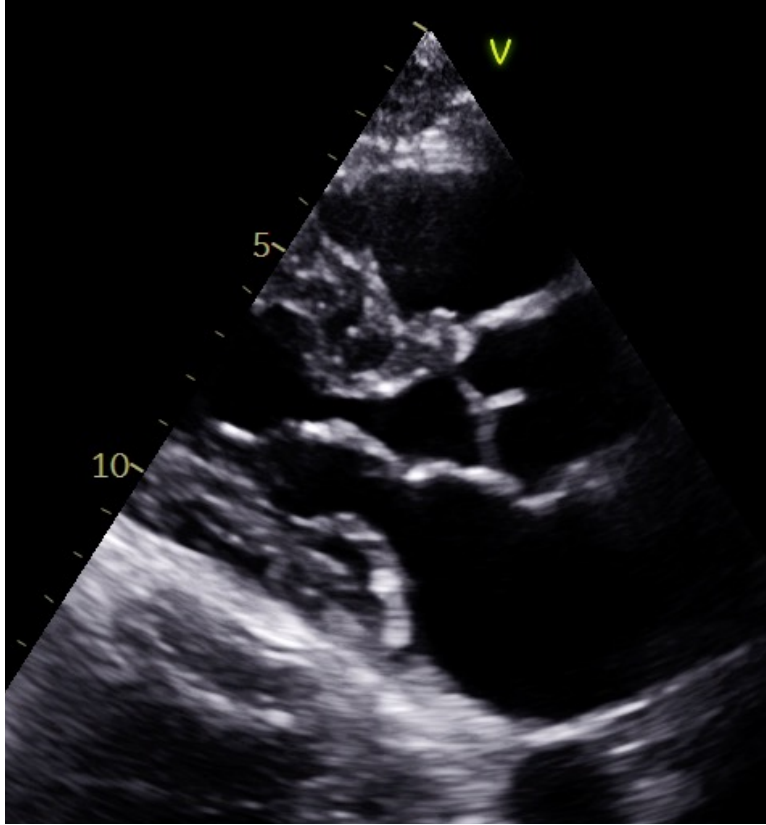
Feuillet ant **34mm vs 24mm** chez les patients normaux

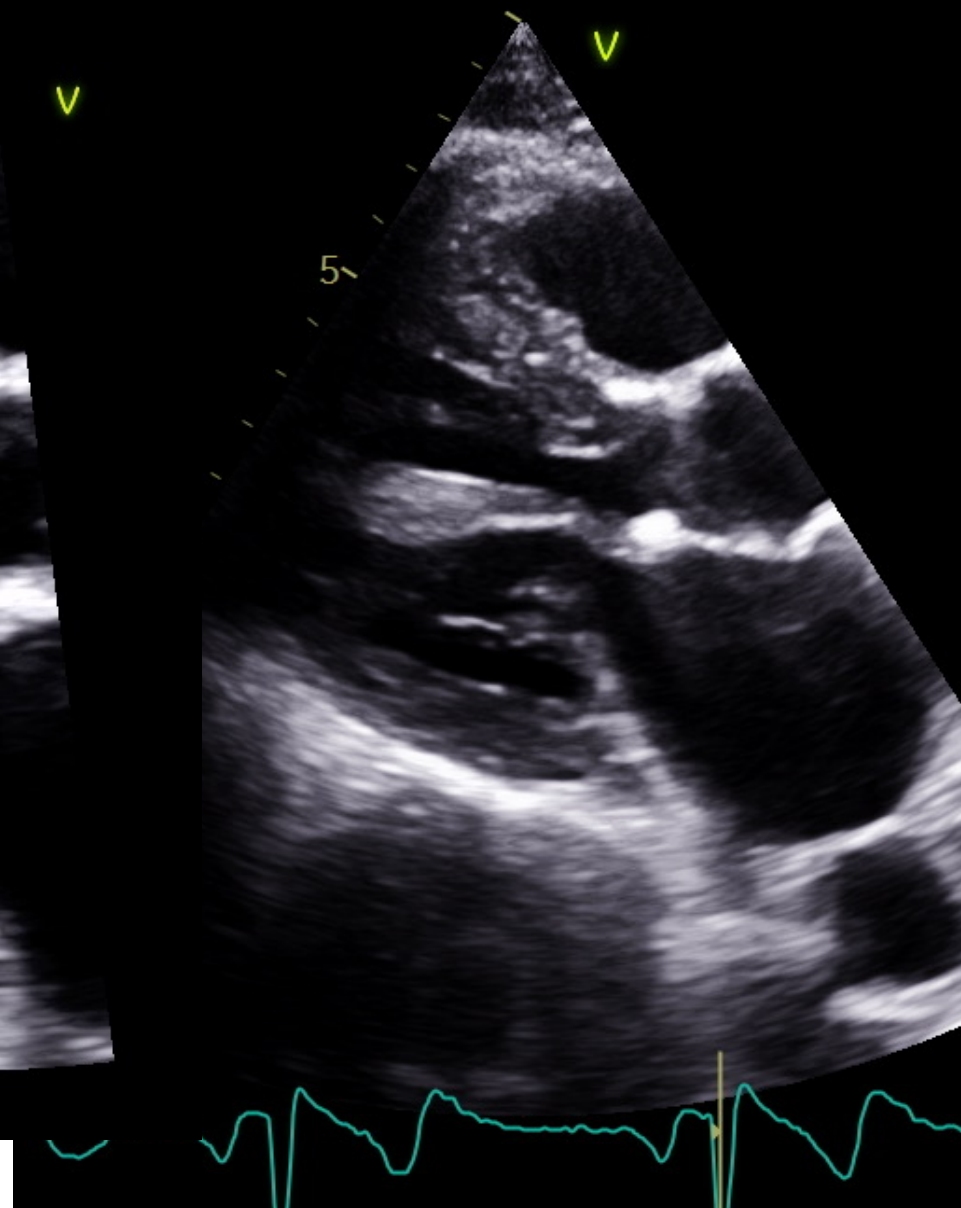
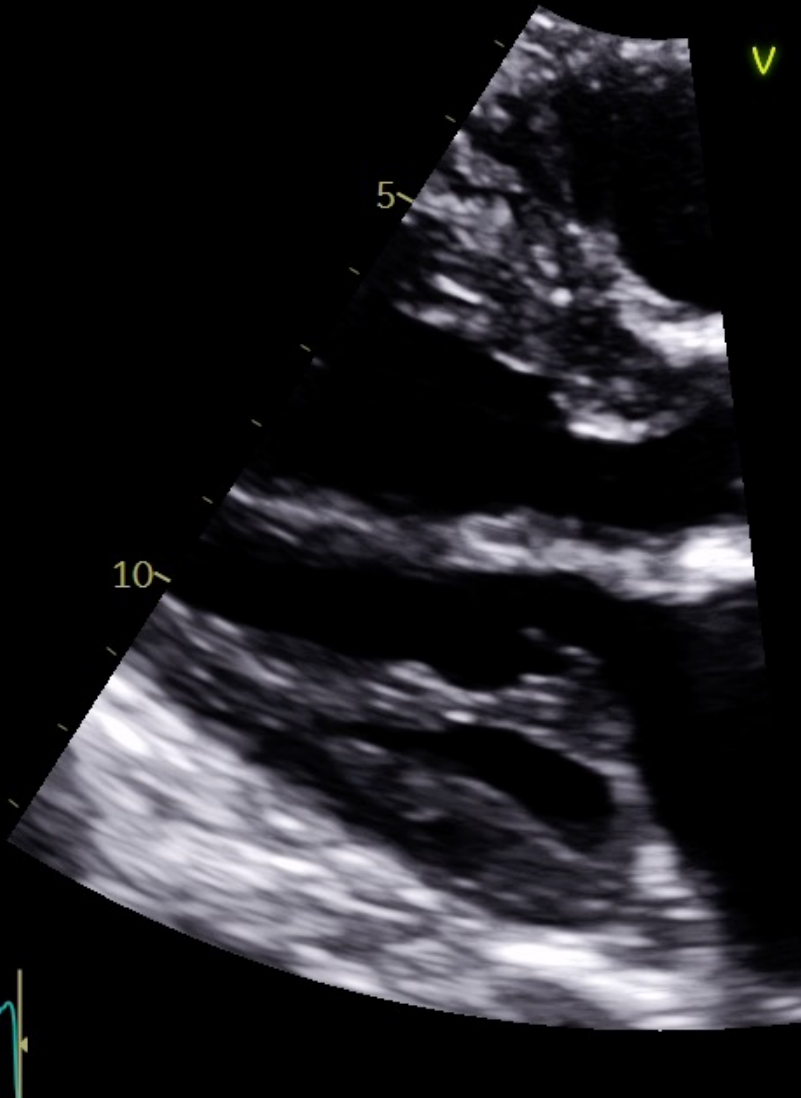
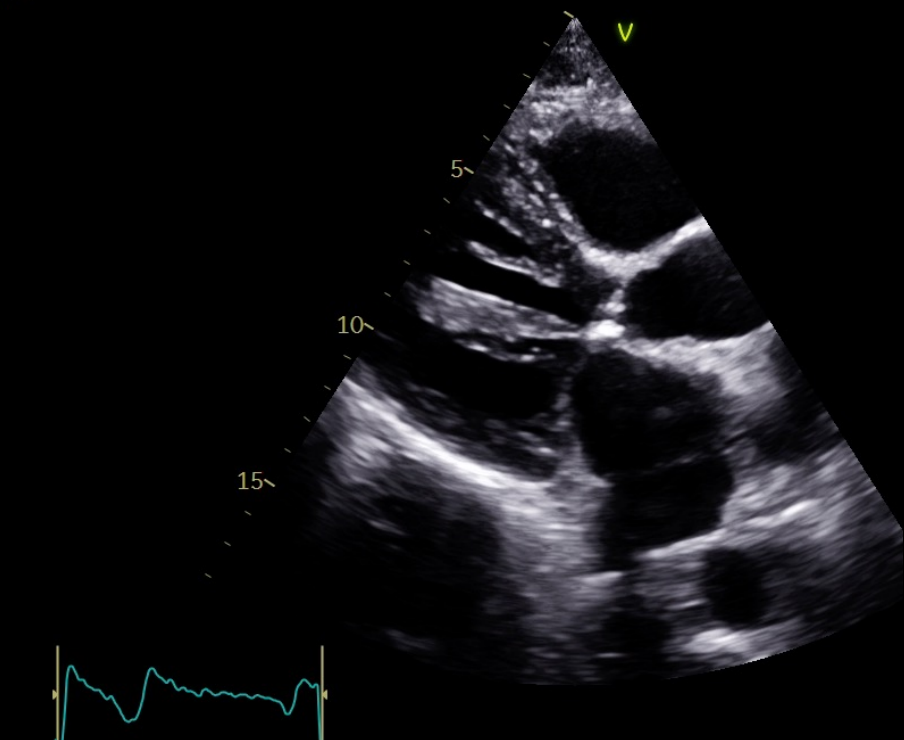


Signe du
bonnet de nuit

La posterieure aussi peut être très longue



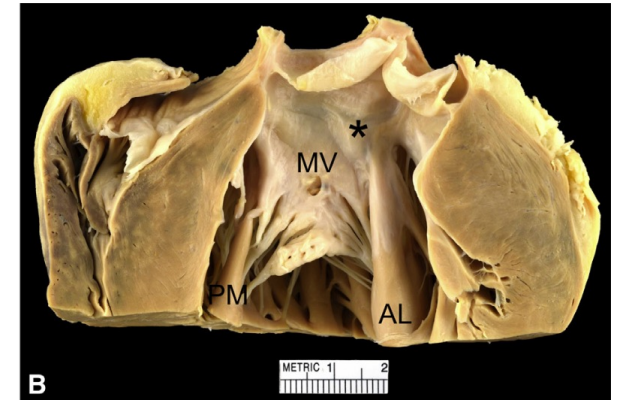




Anomalies d'insertion du pilier sur la valve mitrale

Anomalous papillary muscles—Implications in the surgical treatment of hypertrophic obstructive cardiomyopathy

Juliano Lentz Carvalho, MD,^a Hartzell V. Schaff, MD,^a Catherine S. Morris, MD,^b Rick A. Nishimura, MD,^c Steve R. Ommen, MD,^c Joseph J. Maleszewski, MD,^b and Joseph A. Dearani, MD^a



Type 1

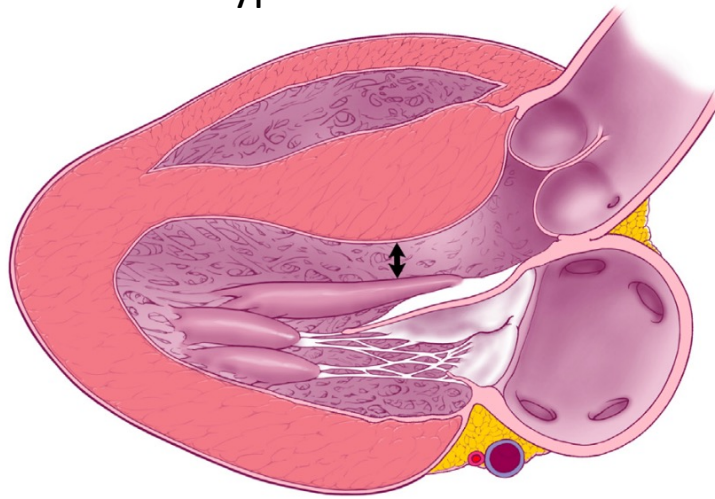


FIGURE 1. Long-axis view of a type I anomalous PM. The *arrow* indicates the site of LVOT obstruction between the hypertrophied interventricular septum and the anomalous PM.

Type 2

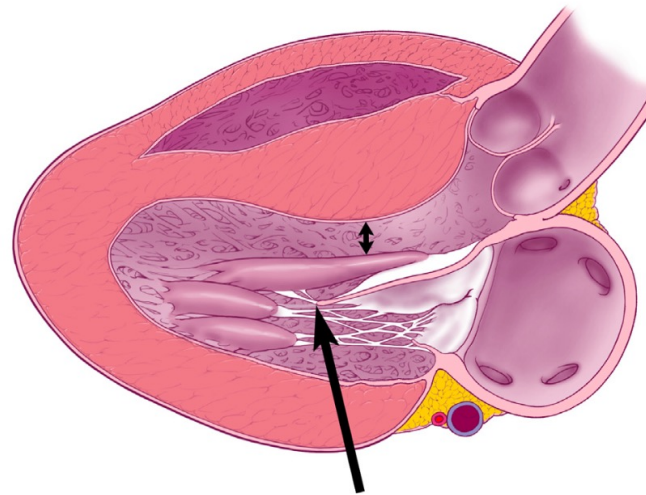


FIGURE 2. Long-axis view of a type II anomalous PM. The *short arrow* indicates the site of LVOT obstruction between the hypertrophied interventricular septum and the anomalous PM. In contrast to the type I anomalous PM, this morphologic subtype has chordal attachments to the free edge of the anterior MV leaflet (*long arrow*), which lend support to the valve.

Type 3

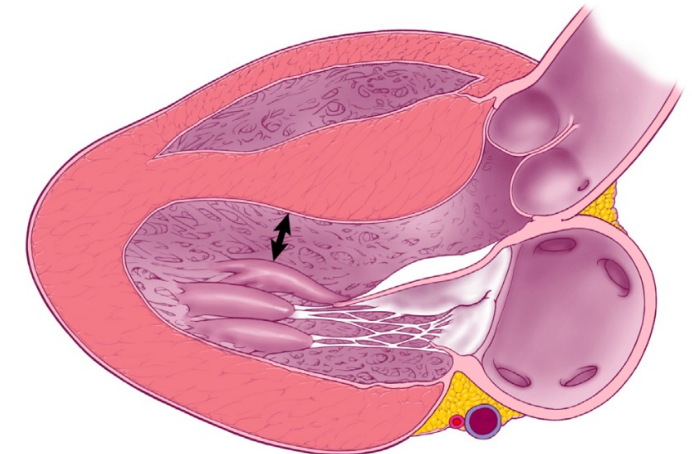
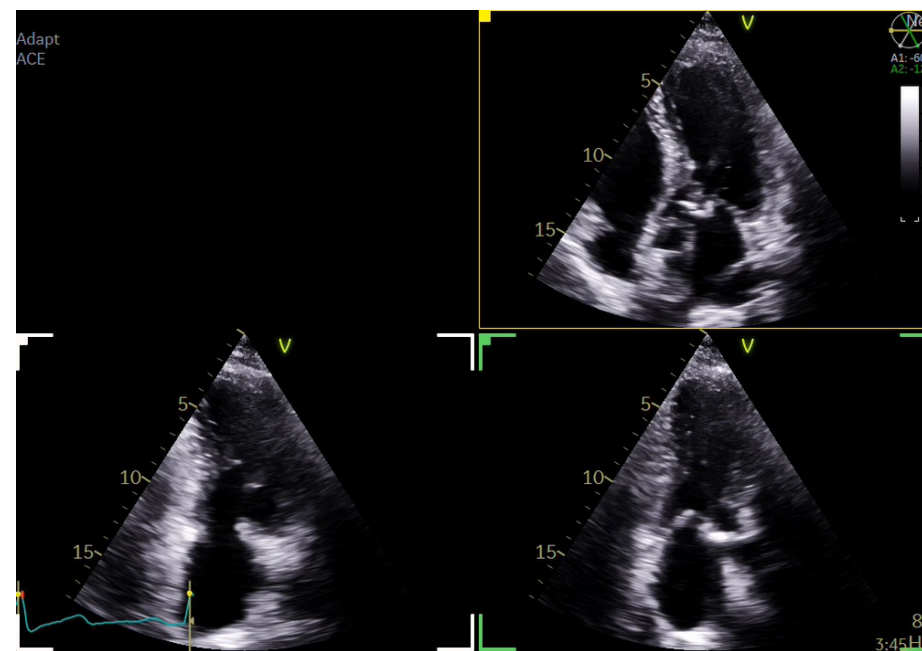
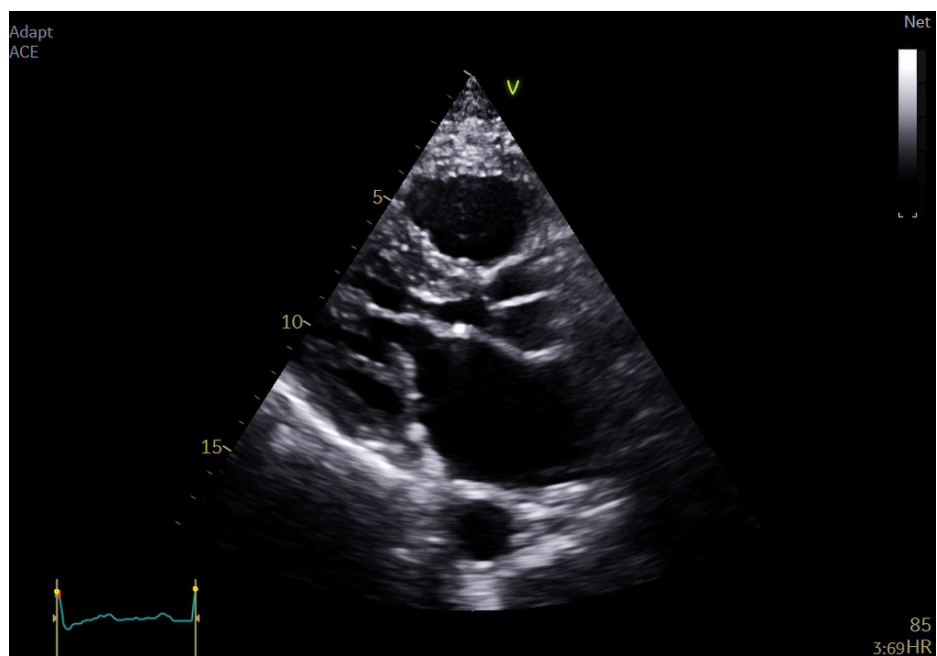
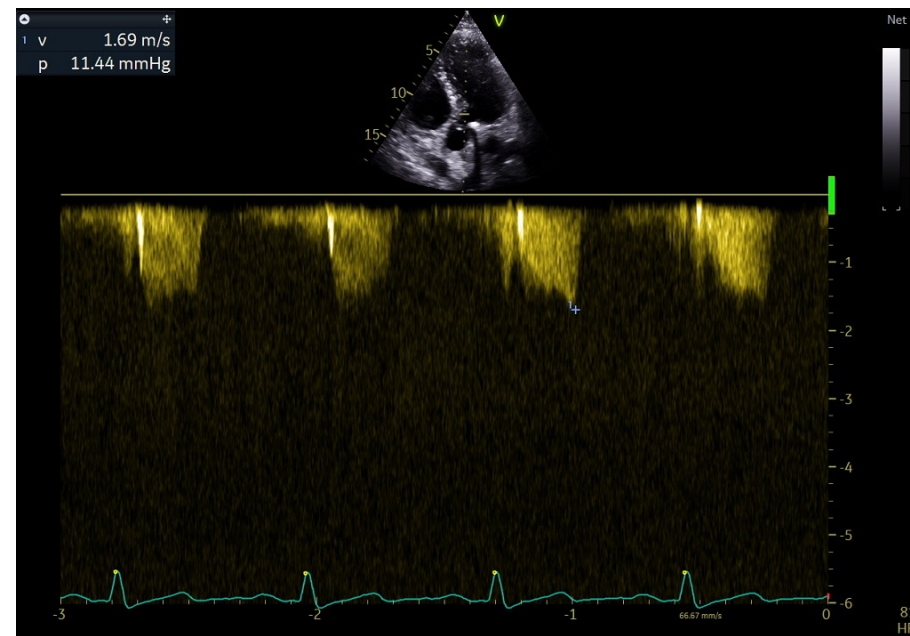
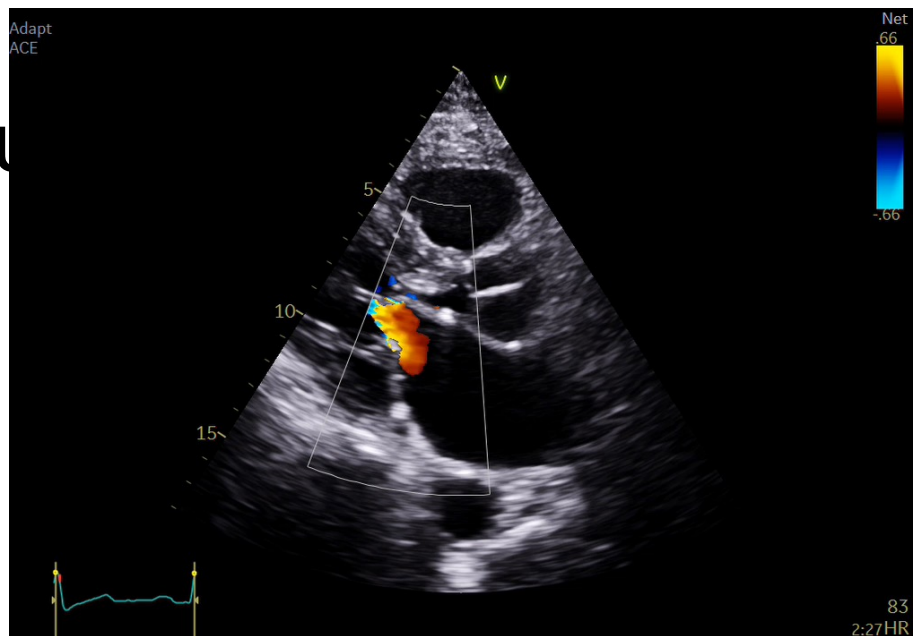
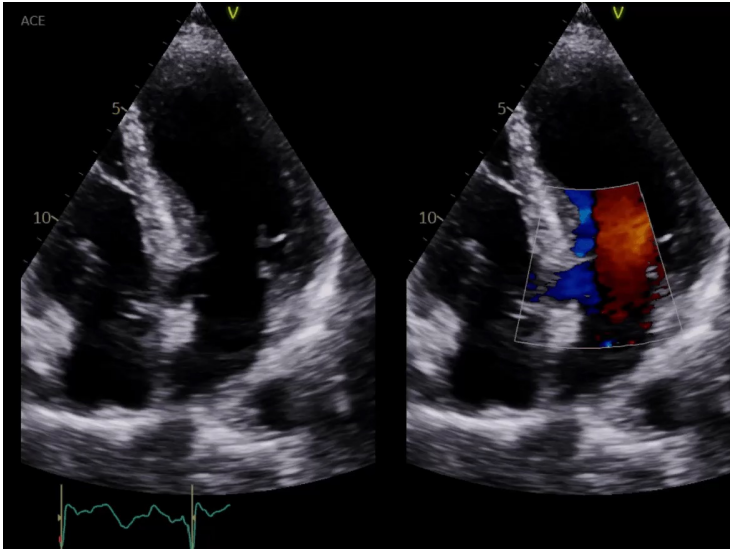


FIGURE 3. Long-axis view of a type III anomalous PM. This variant does not cause subaortic obstruction but may contribute to midventricular obstruction at the level indicated by the *arrow*.

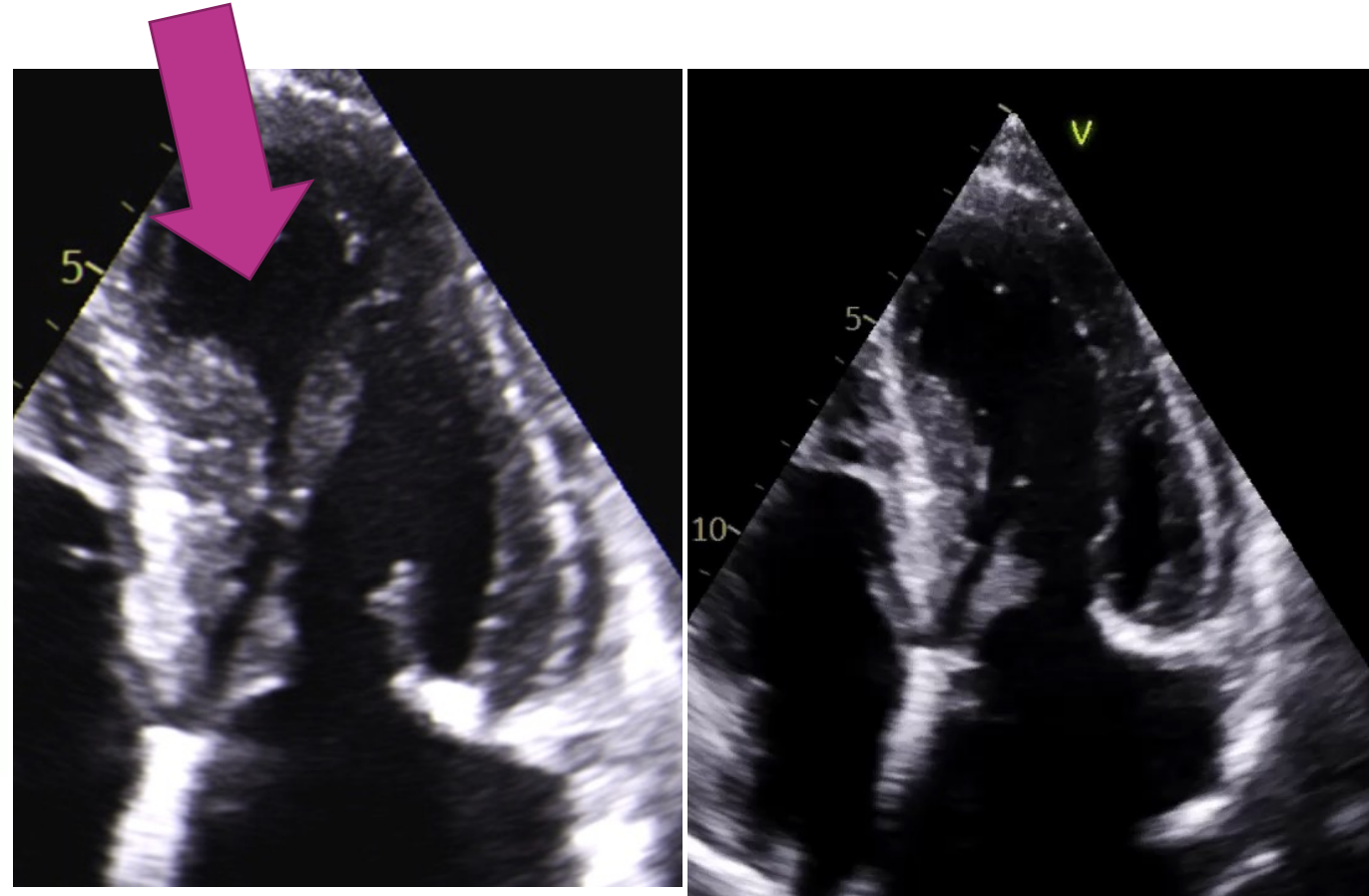
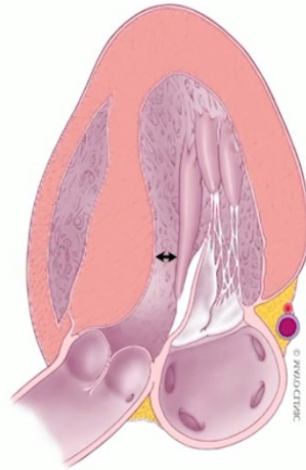
Su



Role des piliers dans l'obstruction

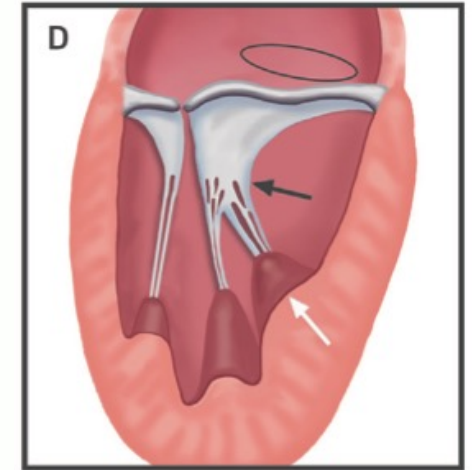
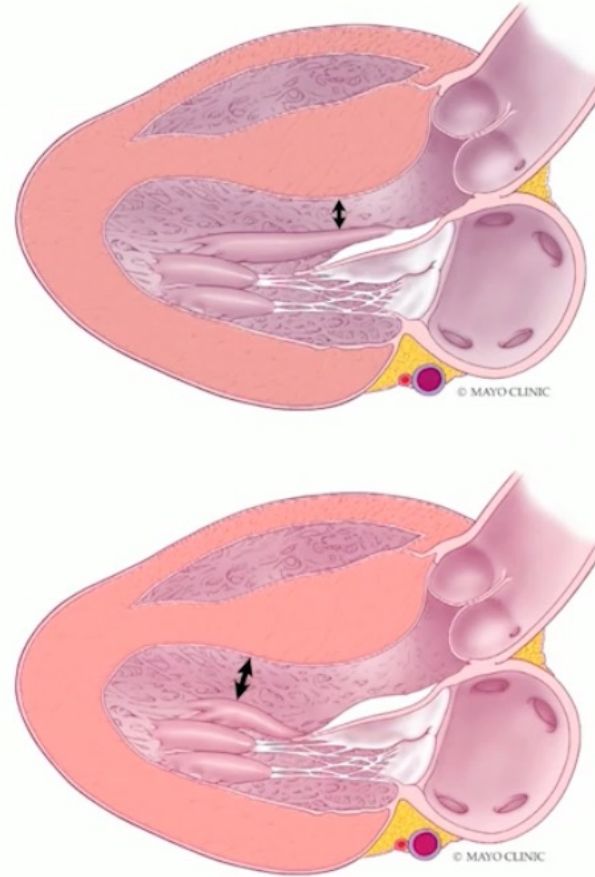


Pas de pilier



Mitral Valve Abnormalities in HCM

- Anterior/apical displacement of papillary muscle
- Hypertrophy of papillary muscles
- False chordae, muscle bundles
- Abnormal insertion of papillary muscle
- Elongation of leaflets and chordae



Insertion ant du pilier
qui est bifide

Est-ce fréquent ?



- Abnormal PM insertion : 4 à 13% des CMHO

*On ne trouve que ce que l'on cherche
On ne cherche que ce que l'on connaît*

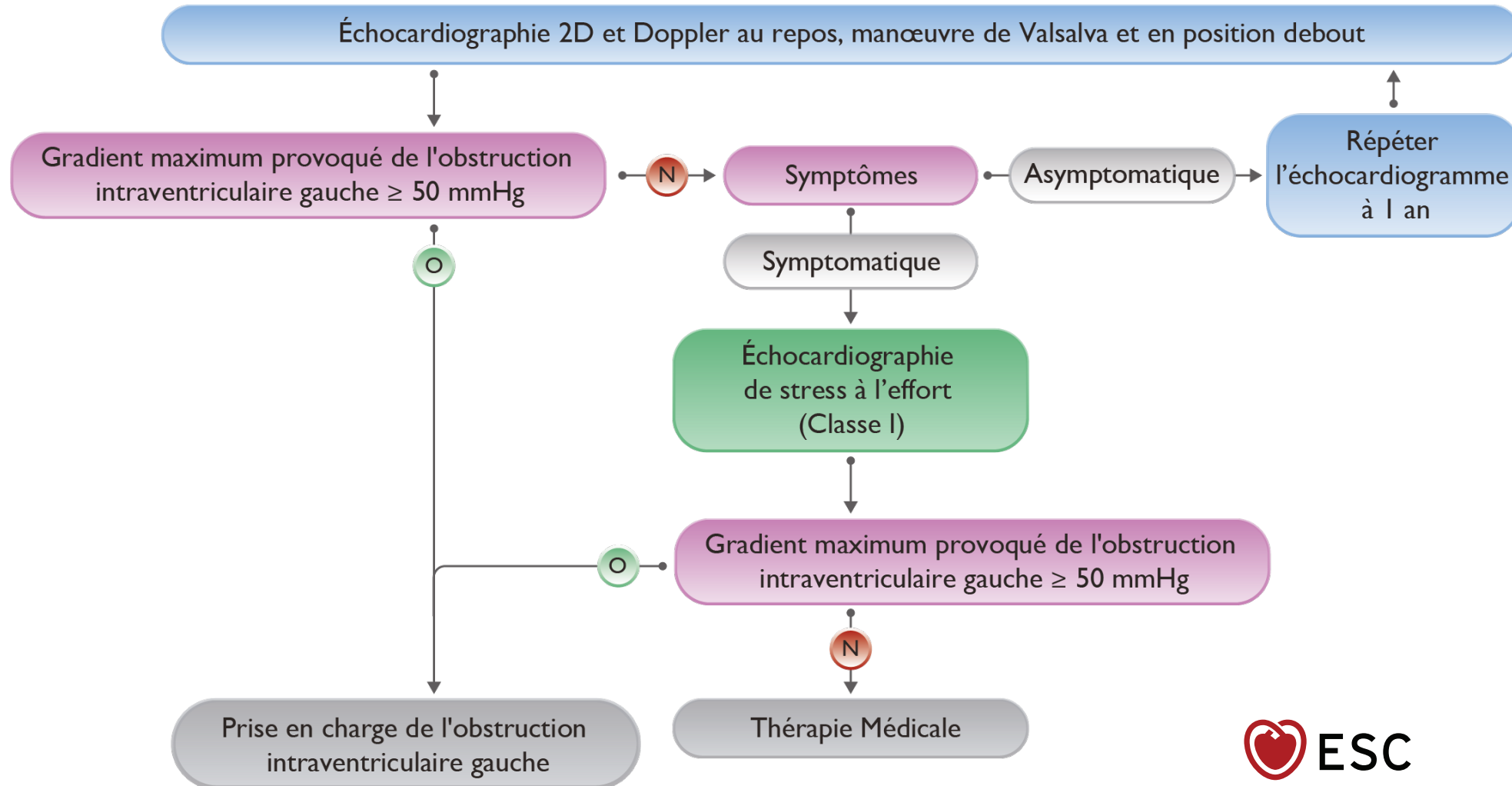
Anomalous papillary muscles—Implications in the surgical treatment of hypertrophic obstructive cardiomyopathy



Juliano Lentz Carvalho, MD,^a Hartzell V. Schaff, MD,^a Catherine S. Morris, MD,^b Rick A. Nishimura, MD,^c Steve R. Ommen, MD,^c Joseph J. Maleszewski, MD,^b and Joseph A. Dearani, MD^a

The anomalous papillary muscle was detected on preoperative transthoracic echocardiography in **11%** of patients and by intraoperative transesophageal echocardiography in **27.4%** of patients.

Quand rechercher l'obstruction intra-VG ?

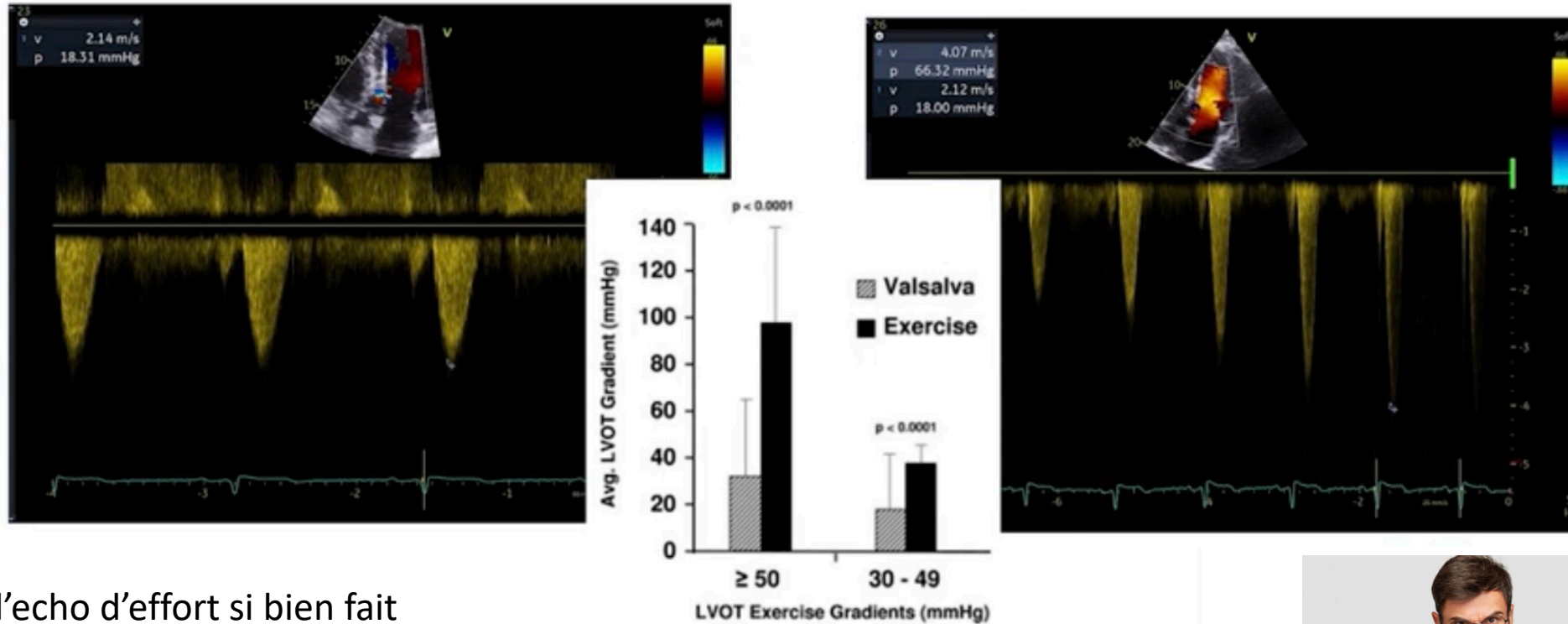


Méthodes de provocation

- Échocardiographie de **repos + Valsalva** ± lors de la mise en **orthostatisme**
 - ➡ **Obstruction significative si gradient max (DC) > 50 mmHg**
- **Valsalva** : à faire mais insuffisant si patient symptomatique et négatif
- Selon Maron, **sensibilité de 40 %** pour prédire l'obstruction à l'effort
- Réalisation en **post-prandial ++++**

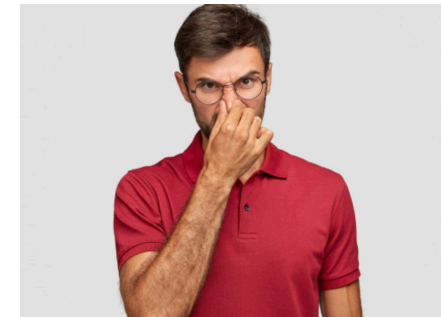
Manoeuvre de Valsalva

Détection du gradient par la manœuvre de Valsalva



Proche de l'écho d'effort si bien fait

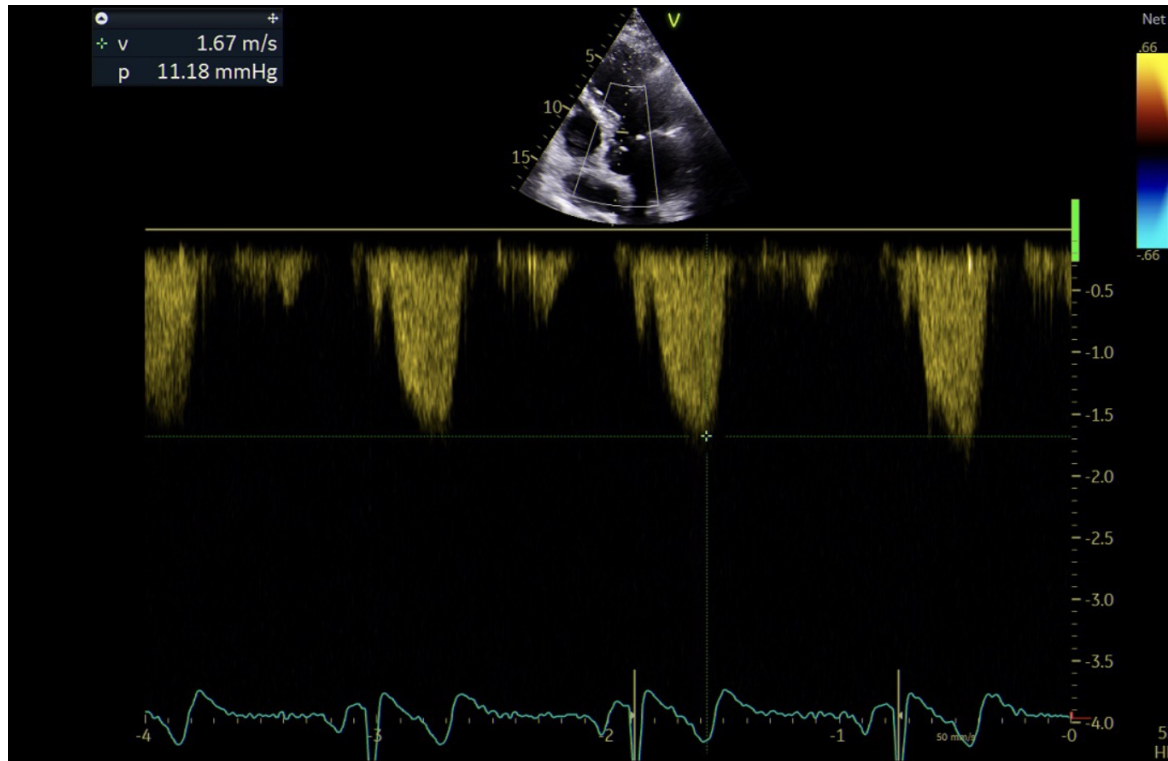
Attention :
Le Valsalva nécessite expertise et précision



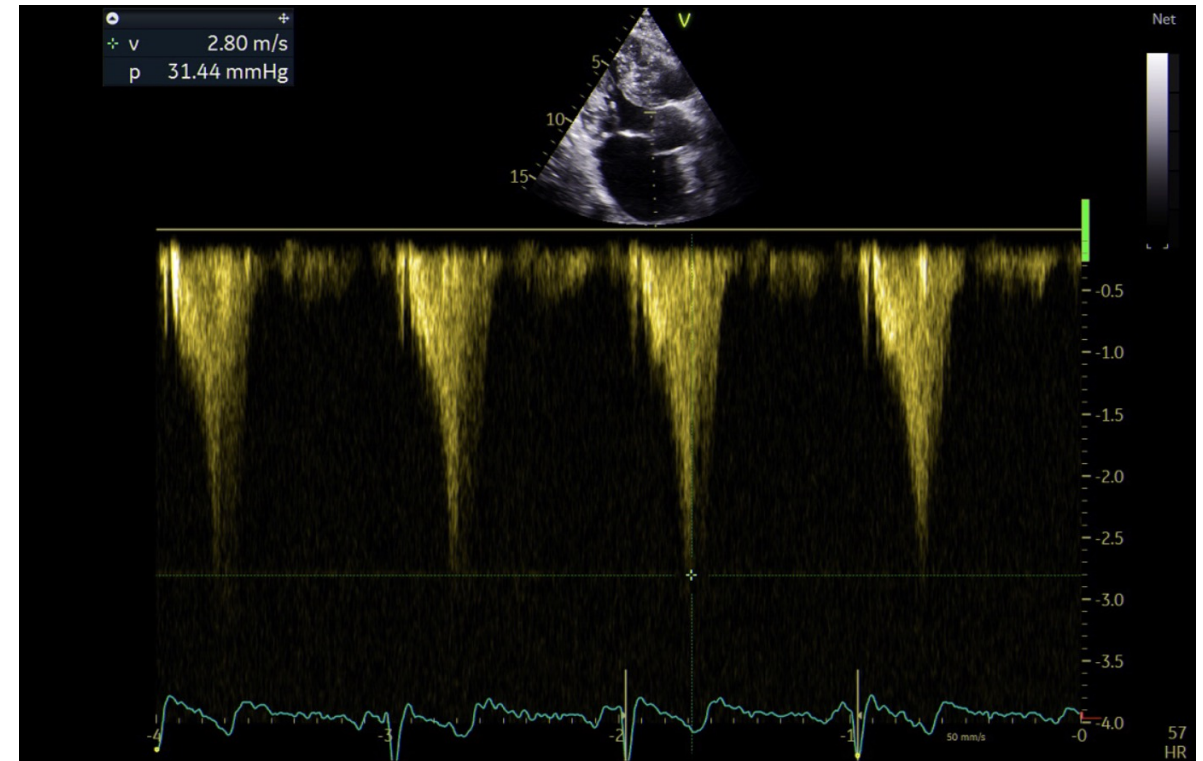
Ne pas trop gonfler les poumons

Manoeuvre de valsalva

Respiration libre



Manoeuvre de Valsalva



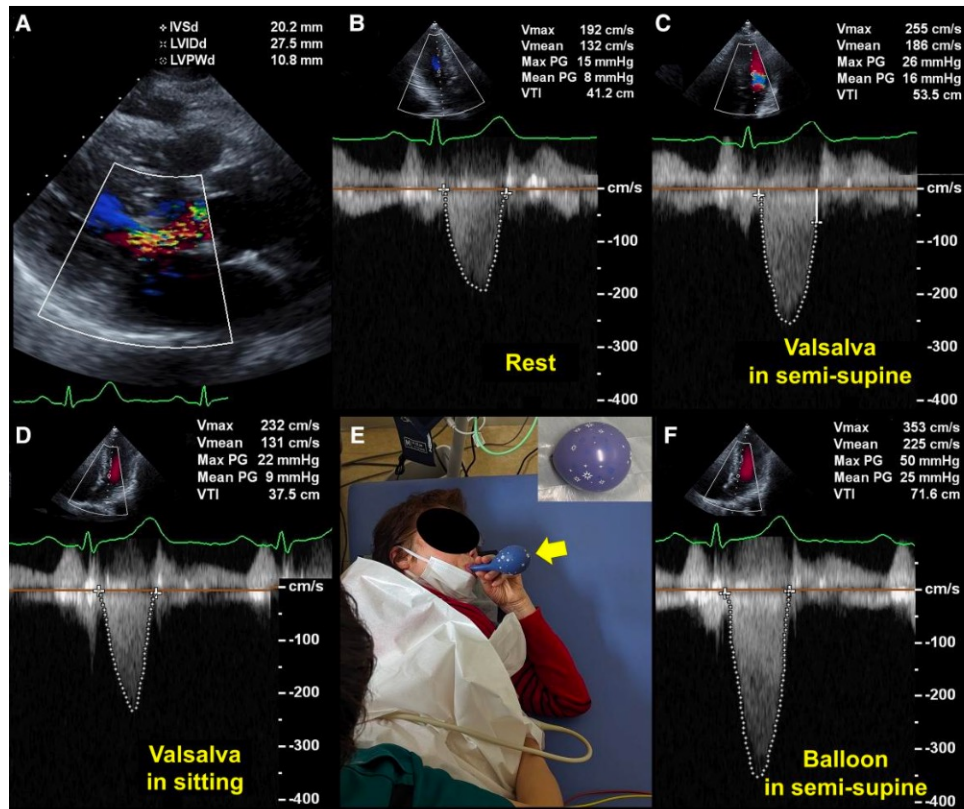


Left ventricular outflow tract obstruction induced by party balloon inflation manoeuvre in transthoracic echocardiography

Kento Kito, Akihisa Kataoka *, Taiga Katayama, Yusuke Watanabe, and Ken Kozuma

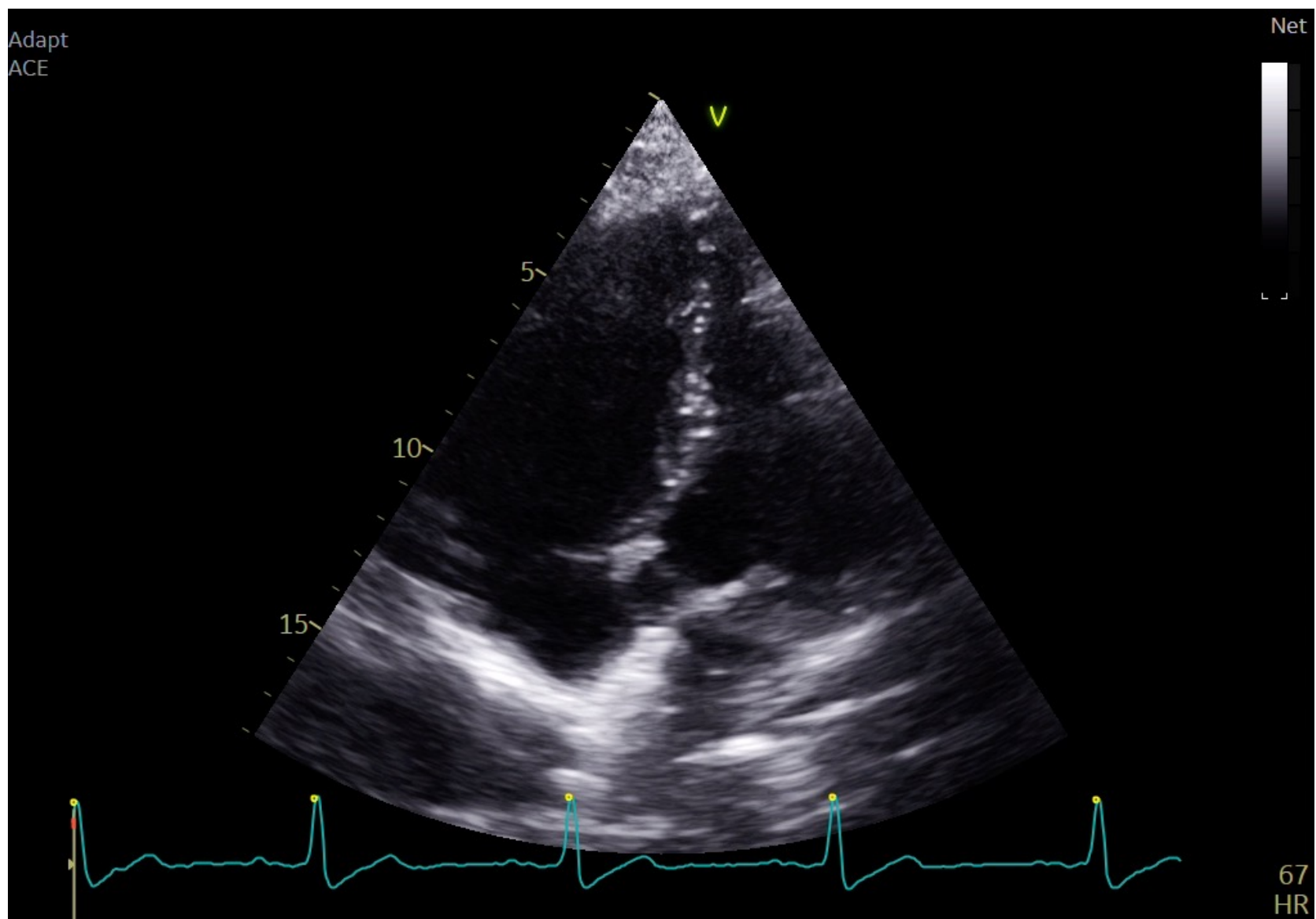
Division of Cardiology, Department of Internal Medicine, Teikyo University, 2-11-1 Kaga Itabashi-Ku, Tokyo 173-8606, Japan

Received 8 February 2023; first decision 16 February 2023; accepted 31 March 2023; online publish-ahead-of-print 3 April 2023



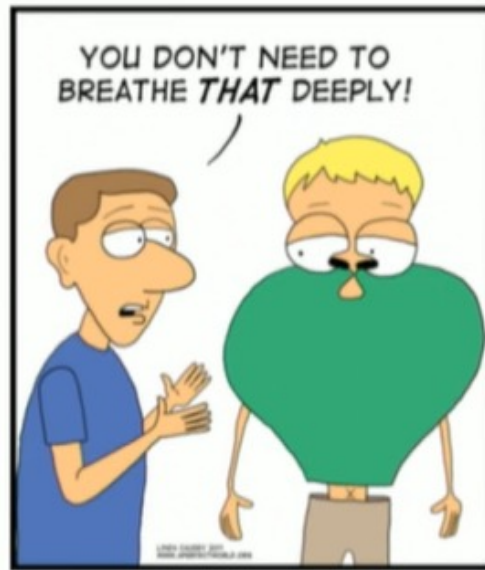
Balloon inflation provided 20–25 mmHg positive end-expiratory pressure to the patient's airway, increasing intrathoracic pressure and decreasing venous return, leading to narrowed left ventricular cavity and exacerbation of SAM, which finally increased the LVOT pressure gradient.

Compared to conventional loading methods, this novel manoeuvre is easier to understand and objectively assesses the performance;



Instructed Valsalva

- Unable to measure efficacy
- Variability in description
- Image degradation



Source: Linda Casau 2011

Goal Directed Valsalva

CARDIAC HEMODYNAMIC ASSESSMENTS

Standardized Goal-Directed Valsalva Maneuver for Assessment of Inducible Left Ventricular Outflow Tract Obstruction in Hypertrophic Cardiomyopathy

Suven Kumar, MD, Grace Van Ness, MPH, Aron Bender, MD, Mrinal Yadava, MD, Jessica Minnier, PhD,
Sriram Ravi, MD, Lidija McGrath, MD, Howard K. Song, MD, PhD, and Stephen B. Heitner, MD,
Portland, Oregon



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Methods of Provocation

Routine clinical practice at our HCM center includes the use of the Valsalva maneuver and postprandial exercise for provocation of LVOTO. The GDV was additionally performed as a part of the study.

Self-Directed Valsalva Maneuver. While lying in the left lateral decubitus position for echocardiography, patients were instructed to “bear down” or “strain as if attempting to move your bowels.” Patients were encouraged to hold the strain phase of the maneuver for as long as they could tolerate.

Goal-Directed Valsalva Maneuver. To measure intraoral pressure, an aneroid manometer was connected to a disposable 10-mL syringe (plunger removed) with rubber tubing (Figure 1). While lying in the left lateral decubitus position for echocardiography, patients were instructed by the sonographer to blow into the syringe and maintain a pressure of >40 mm Hg for >10 sec.

Exercise. Postprandial HSE was performed with an upright treadmill exercise per standard Bruce protocol. Subjects taking cardiac medications (β -blockers, calcium channel blockers, and/or disopyramide) were instructed to continue their medications as prescribed and asked to eat a hearty meal 2 hours before the test. Echocardiographic and Doppler data on LVOTO were acquired at rest and immediately following peak exercise.

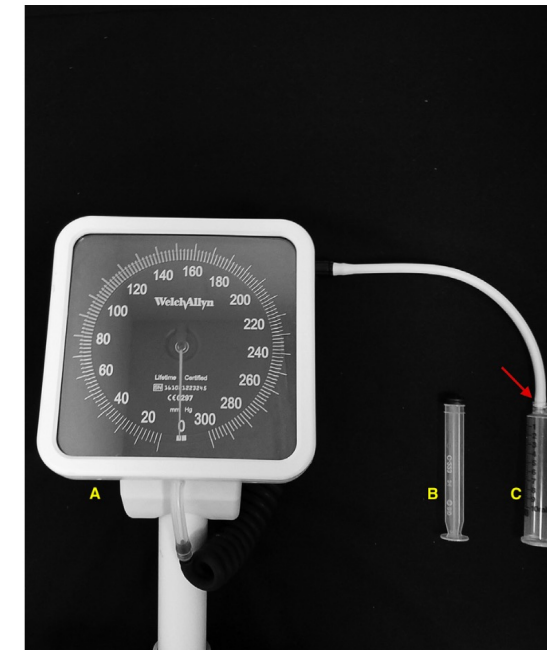


Figure 1 Apparatus to conduct the GDV. Aneroid manometer (A) connected to a 10-mL syringe (C) through the tubing (arrow) after removing the plunger (B).

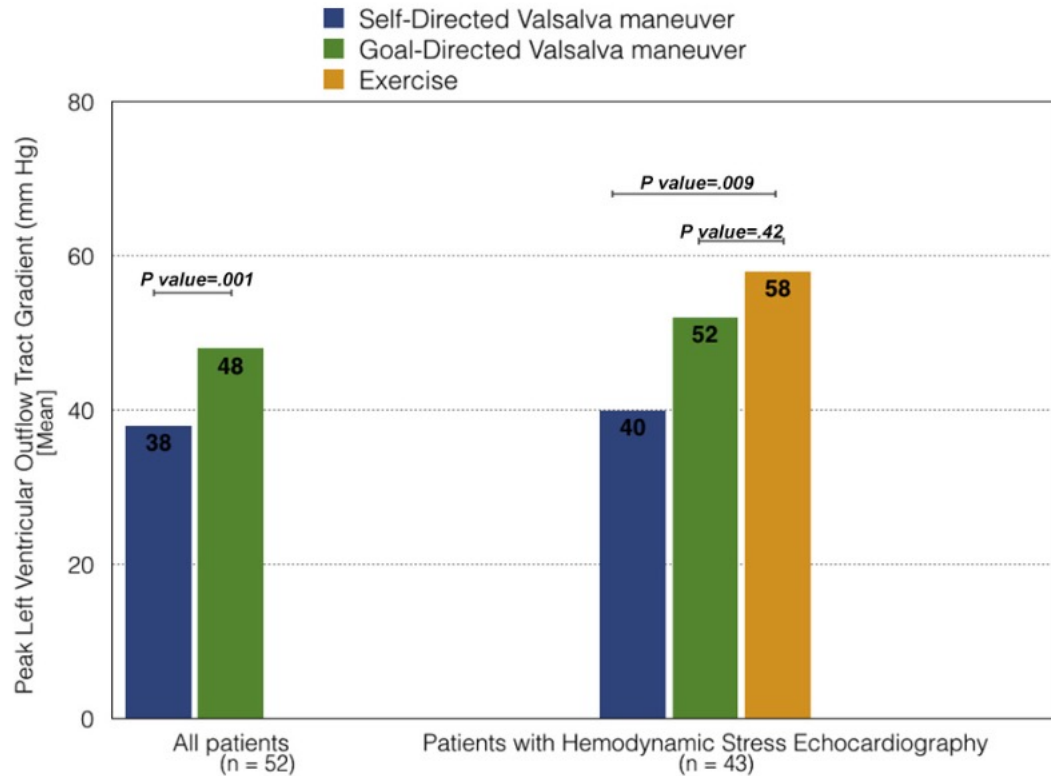
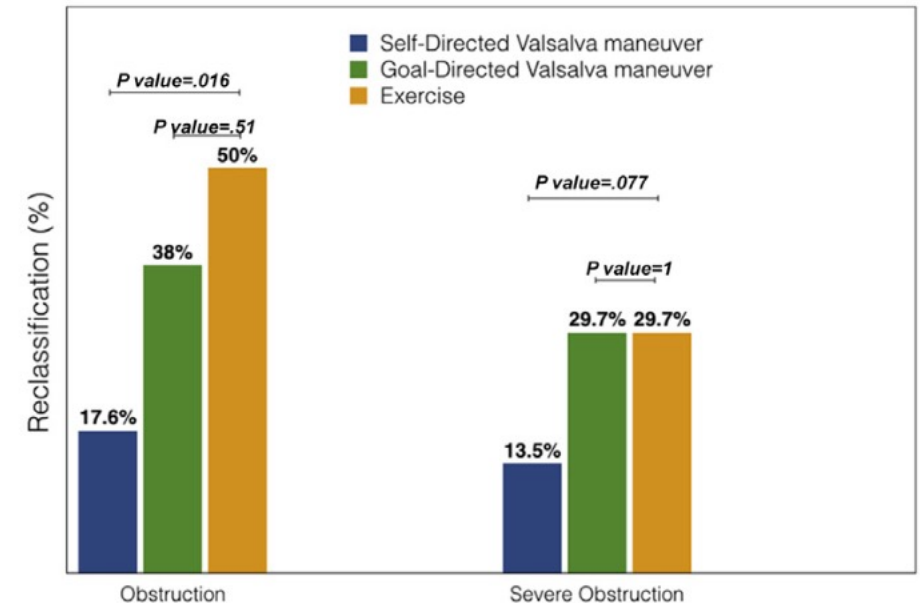


Figure 2 Comparison of pLVOTG with SDV, GDV, and exercise. GDV had significantly higher pLVOTG compared with SDV ($P = .001$) but was similar to exercise ($P = .042$). Similarly, exercise had higher pLVOTG compared with SDV ($P = .009$).

HIGHLIGHTS

- The Valsalva maneuver is widely used to provoke outflow tract gradient in obstructive HCM.
- It is performed subjectively, introducing wide variability and inconsistency.
- A goal-directed method using intraoral pressure >40 mm Hg for 10 sec can standardize this maneuver.
- GDV is more effective in provoking outflow tract gradient than SDV.
- GDV can significantly alter patient management by reclassifying disease severity.

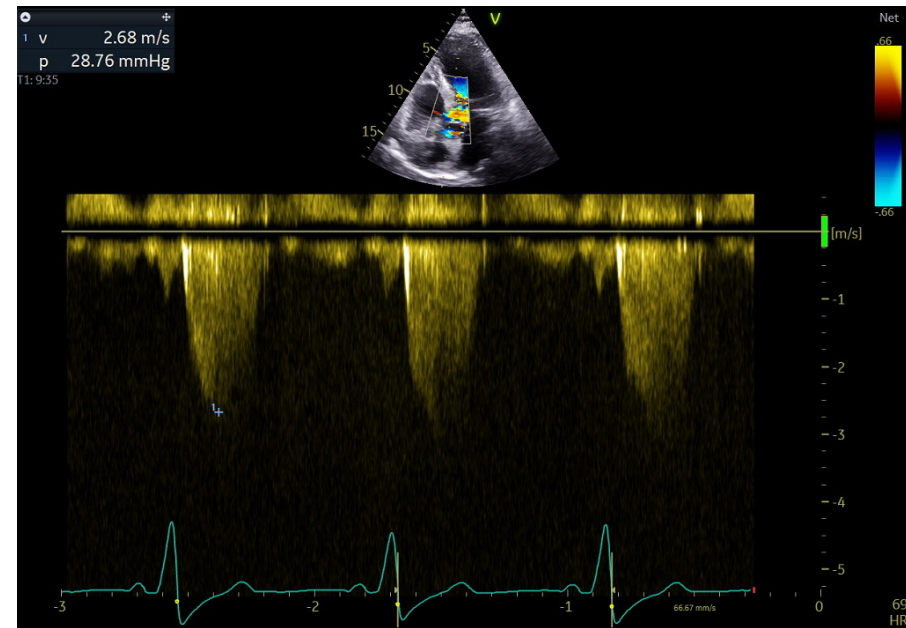
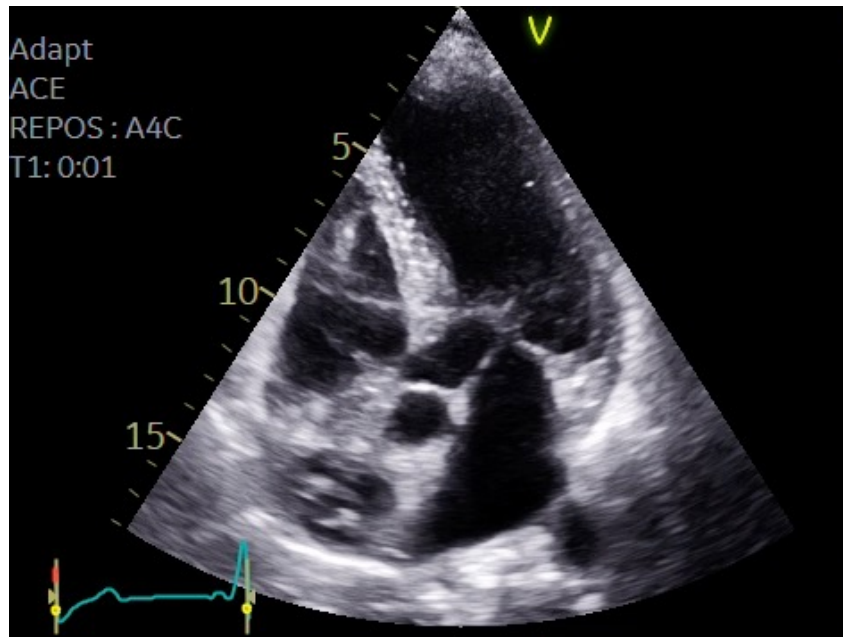
A

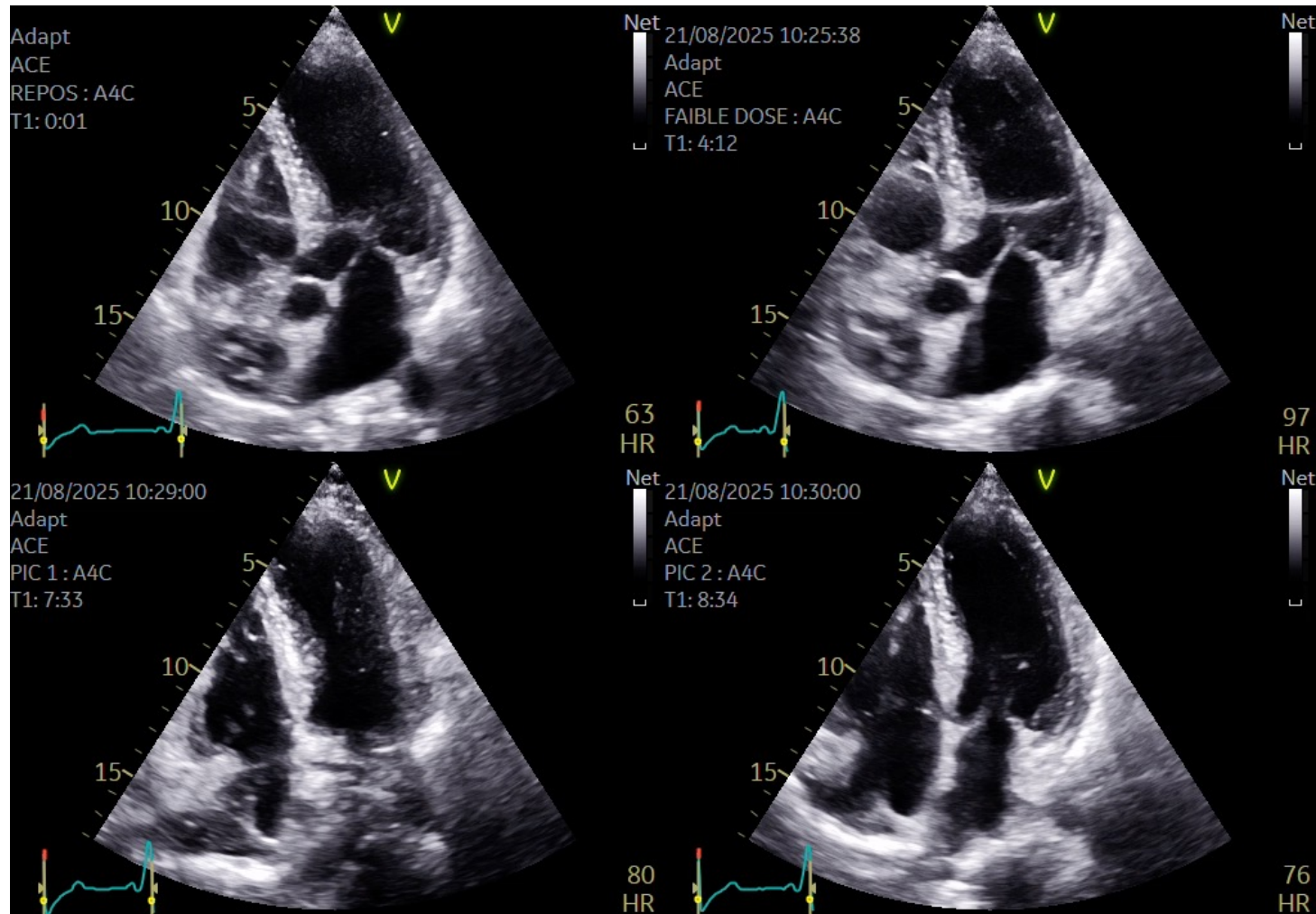


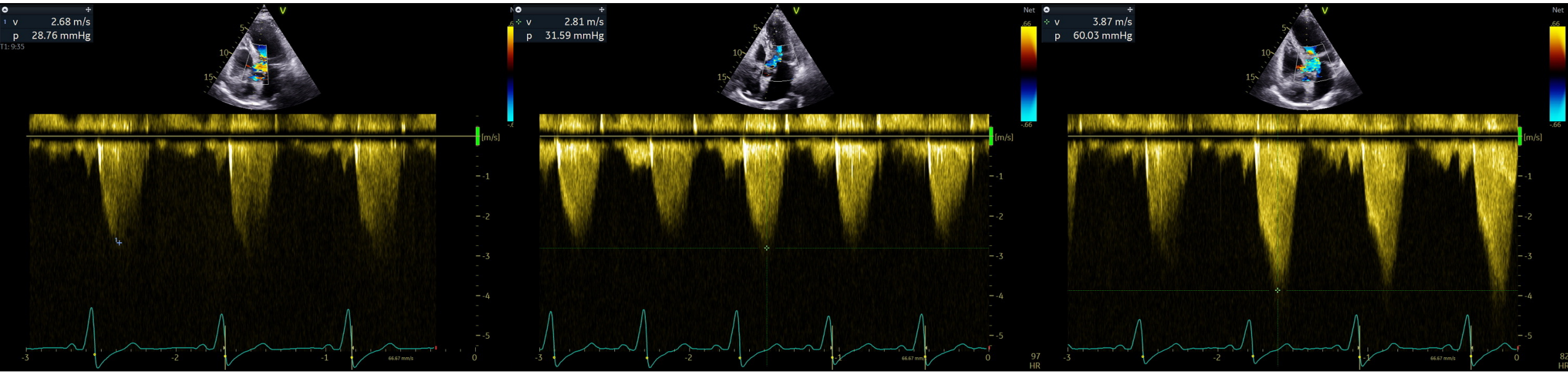
Cas de Mr X, 50 ans

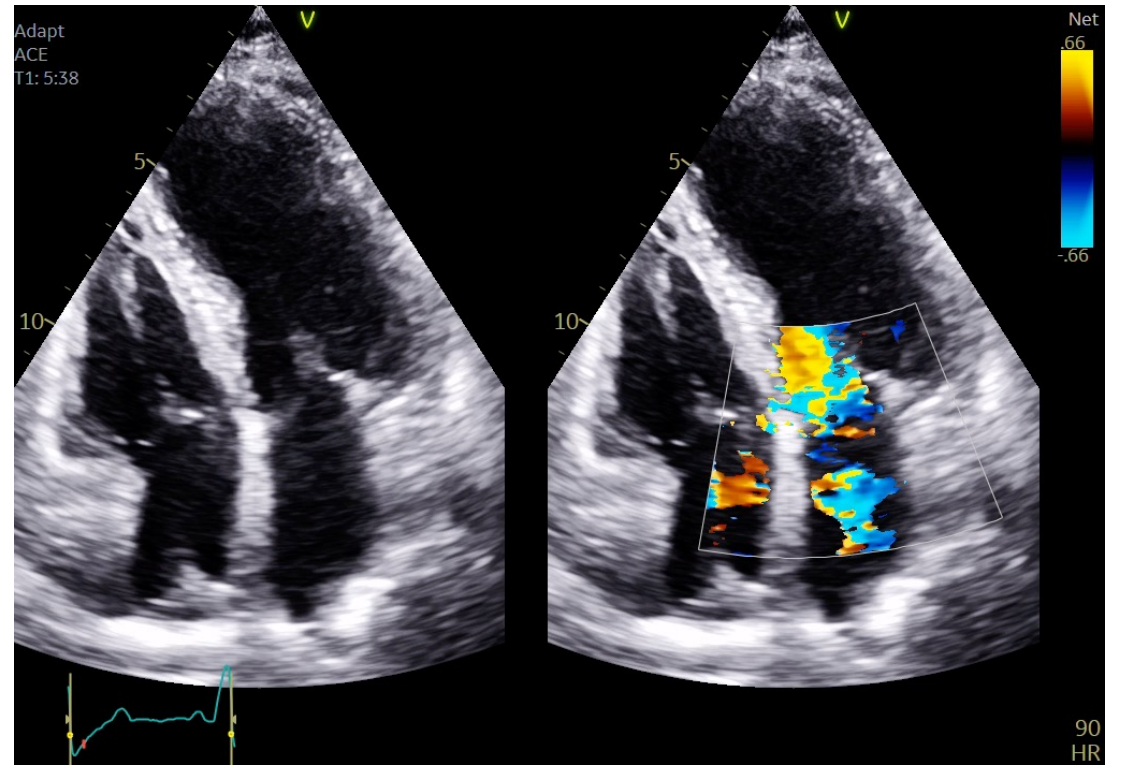
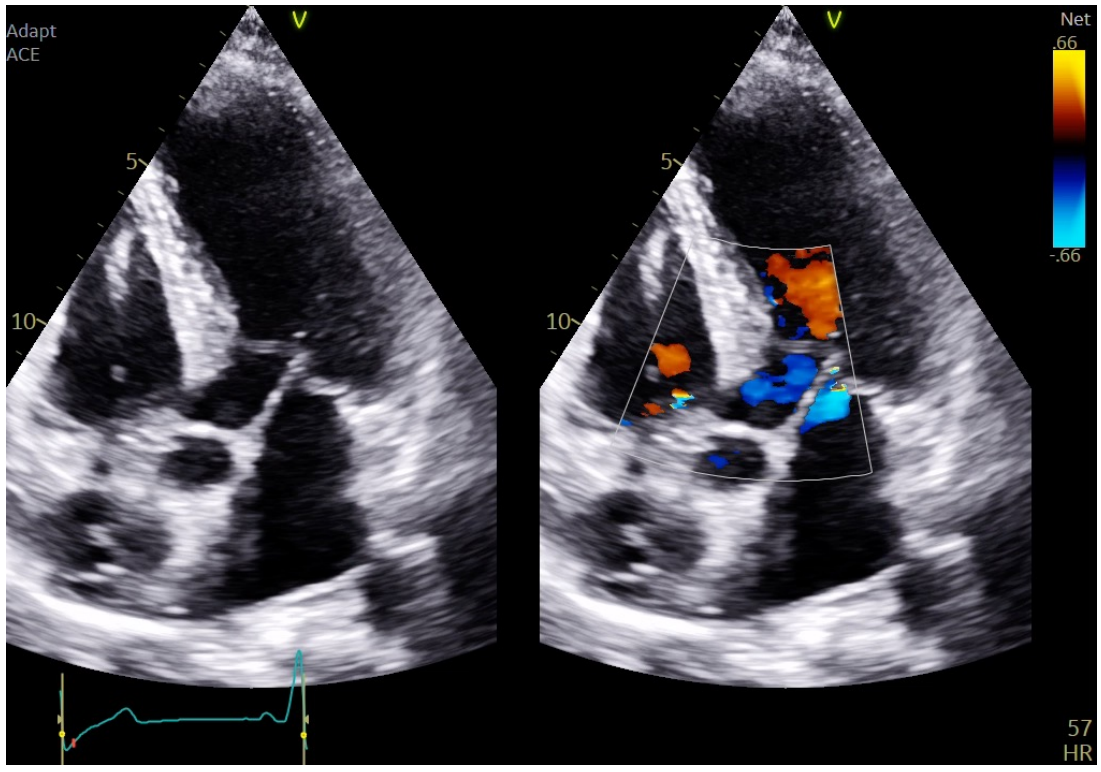
- Dyspnée d'effort (NYHA II)
- CMH asymétrique découverte récemment











Provocation par l'effort physique



- A l'effort : **échocardiographie d'effort**

- Per-effort ++++ et post-effort

- **Vélo assist+++**, le plus vertical possible

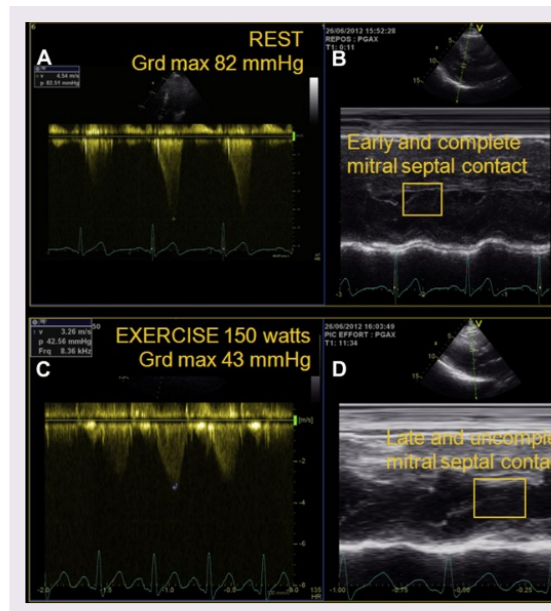
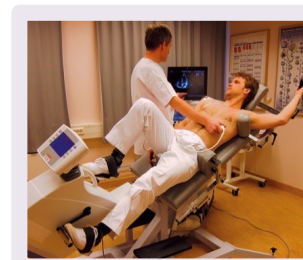
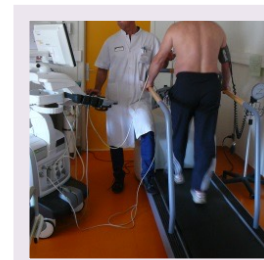
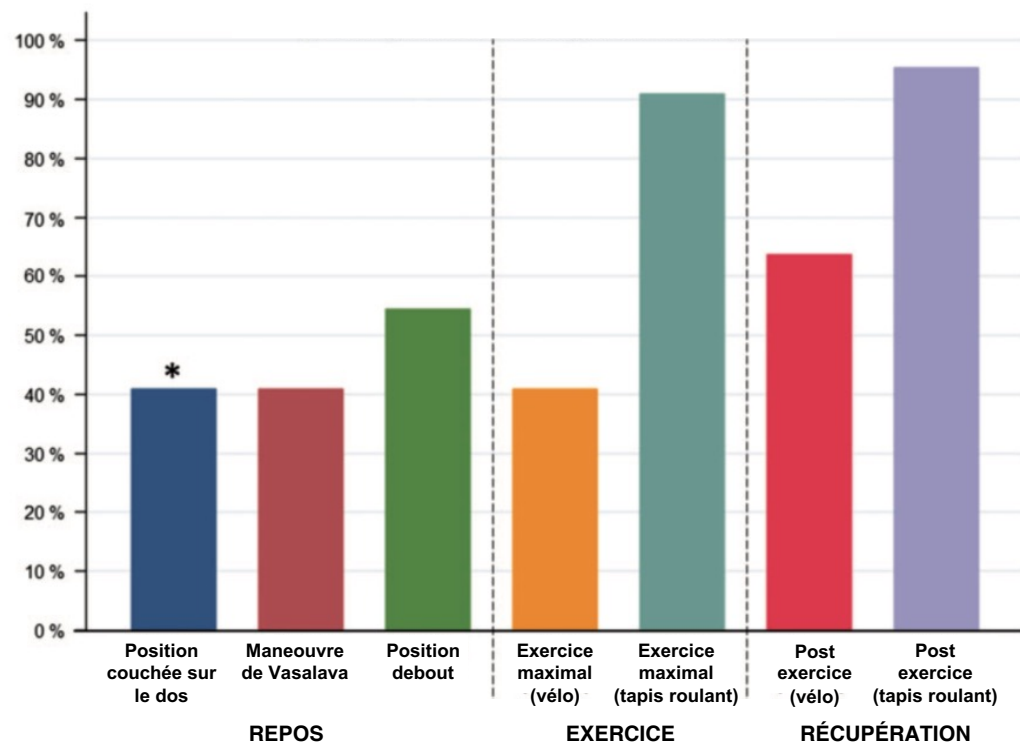
- Impact du retour veineux à prendre en compte



Impact de la position sur la provocation de l'obstruction

Étude pilote prospective 22 patients CMH NYHA II

Pourcentage de patients CMH
avec gradient max sous-aortique ≥ 50 mmHg



Upright treadmill vs. semi-supine bicycle exercise echocardiography to provoke obstruction in symptomatic hypertrophic cardiomyopathy: a pilot study

Patricia Reant^{1,2,3,4*}, Maxence Dufour^{1,2}, Jerome Peyrou^{1,2}, Amelie Reynaud^{1,2,3}, Caroline Rooryck^{1,2}, Marina Dijos^{1,2,3}, Cecile Vincent^{1,2}, Claire Cornolle^{1,2,3}, Raymond Roudaut^{1,2,3}, and Stephane Lafitte^{1,2,3}

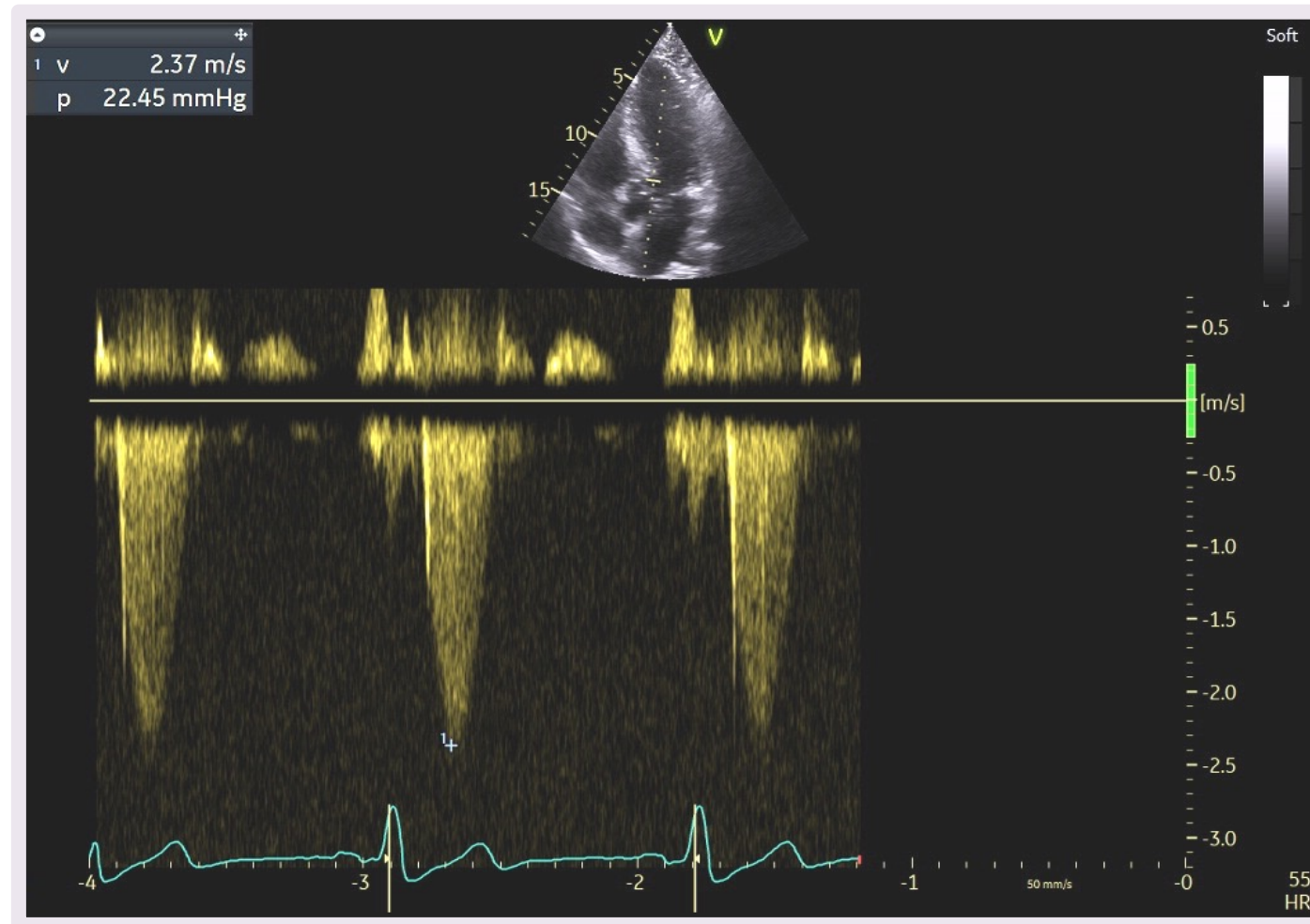
Paradoxical Response to Exercise in Asymptomatic Hypertrophic Cardiomyopathy

A New Description of Outflow Tract Obstruction Dynamics

Stéphane Lafitte, MD, PhD,* Patricia Reant, MD, PhD,* Cecile Touche, MD,* Xavier Pillois, PhD,* Marina Dijos, MD,* Florence Arsac, MD,* Jerome Peyrou, MD,* Michel Montaudon, MD, PhD,* Philippe Ritter, MD,* Raymond Roudaut, MD,* Anthony DeMaria, MD, PhD†

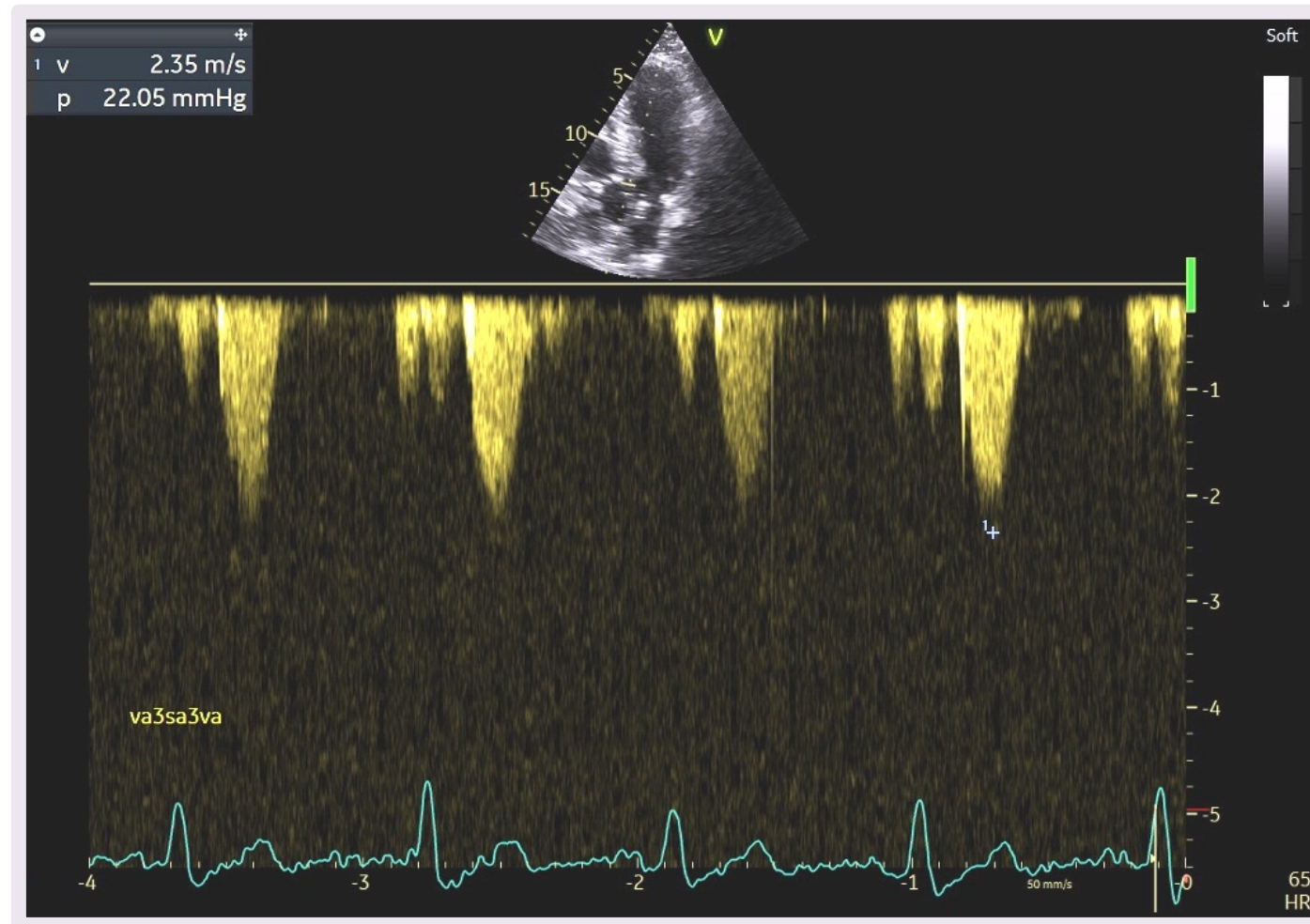
Bordeaux, France; and San Diego, California

Recherche de l'obstruction



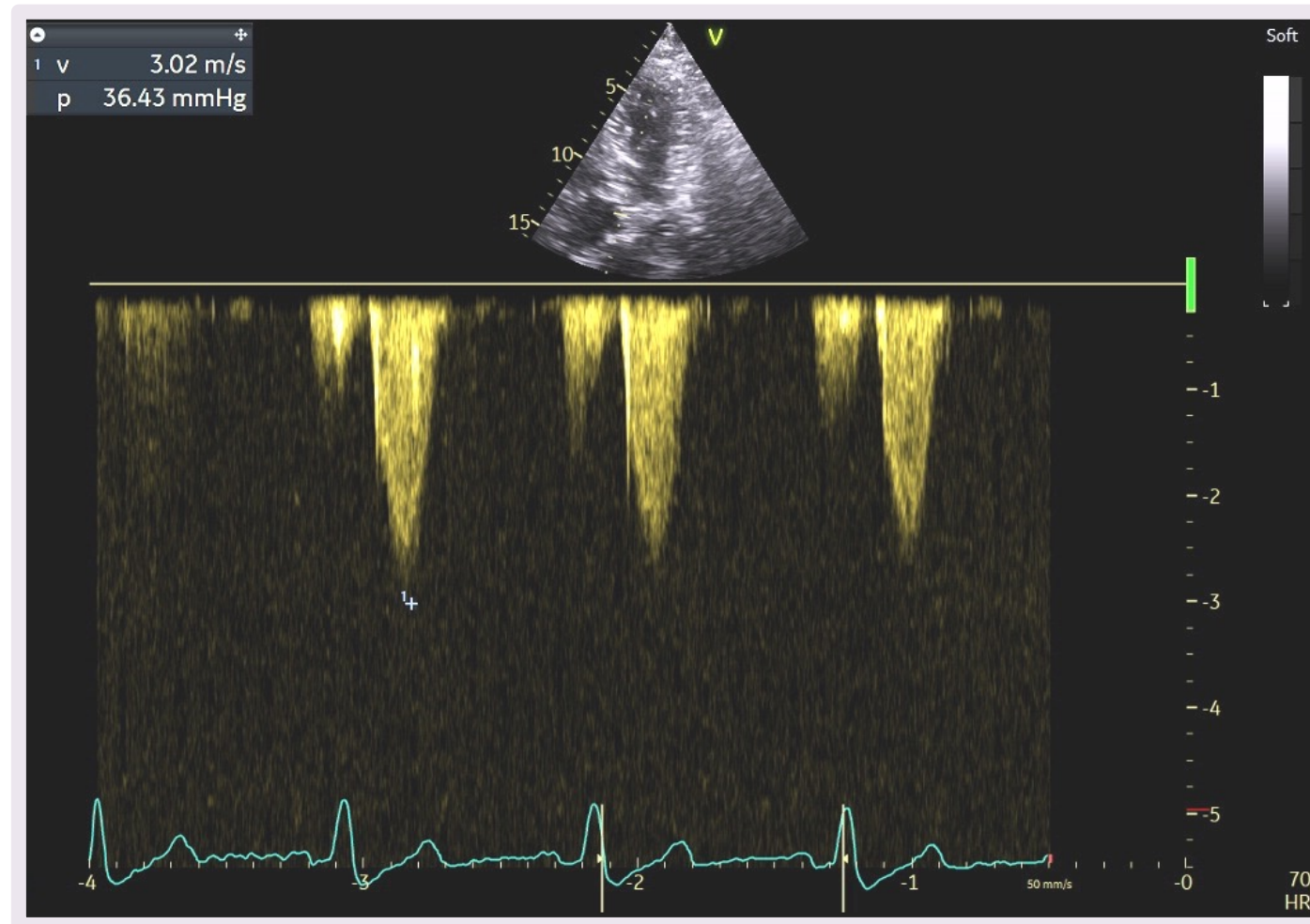
Repos couché

Recherche de l'obstruction



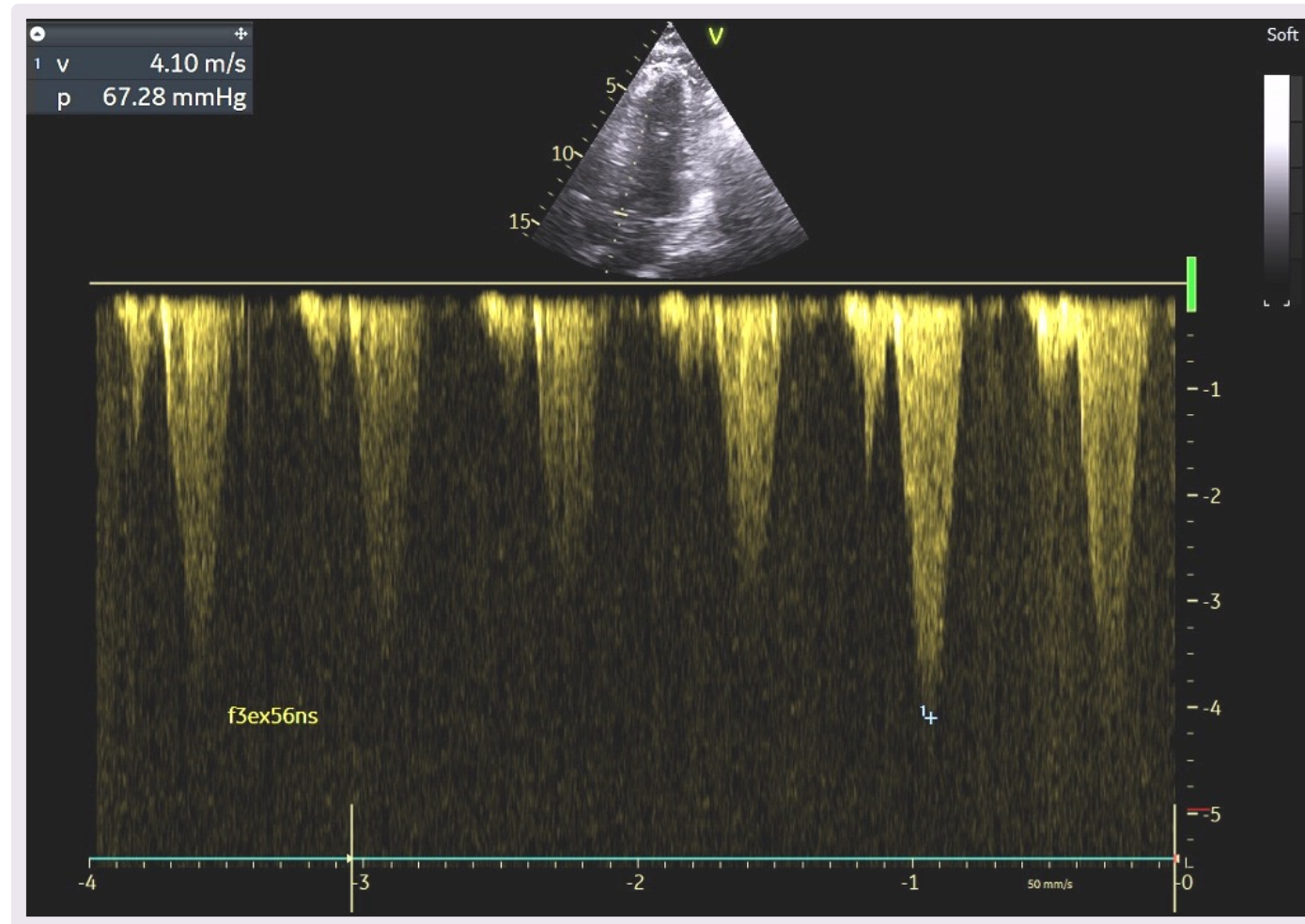
Valsalva

Recherche de l'obstruction



Après mise à l'orthostatisme

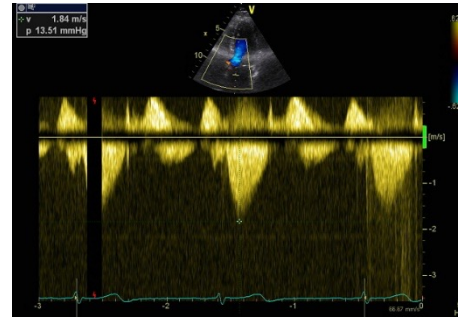
Recherche de l'obstruction



Après
10 flexions

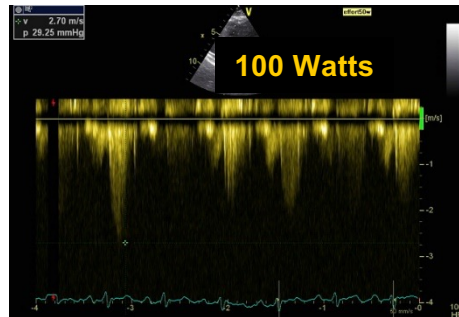
Exemple d'effet de la position

Évaluation **au repos**, position couchée



Gradient max sous-aortique = 13 mmHg

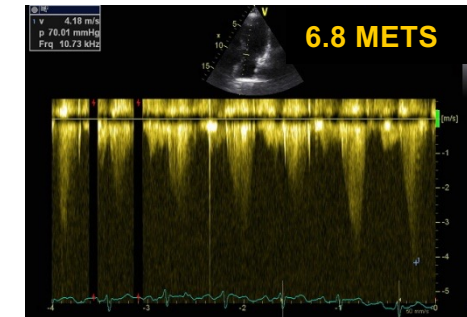
Effort sur bicyclette semi-inclinée



Gradient max sous-aortique = 29 mmHg

Stop pour
dyspnée

Effort sur tapis Protocole Bruce modifié



Gradient max sous-aortique = 70 mmHg

OBSTRUCTION LATENTE

Manœuvres
hémodynamiques

VALSALVA



SQUAT



CHANGEMENT
DE POSITION

ECHO D'EFFORT

VELO



TAPIS



Manœuvres hémodynamiques



VALSALVA

+

SQUAT



Repetitive Squat-to-Stand Provocation of Dynamic Left Ventricular Outflow Tract Obstruction in Hypertrophic Cardiomyopathy



Journal of the American Society of Echocardiography
Volume 35 Number 3

Brief Research Communications 325

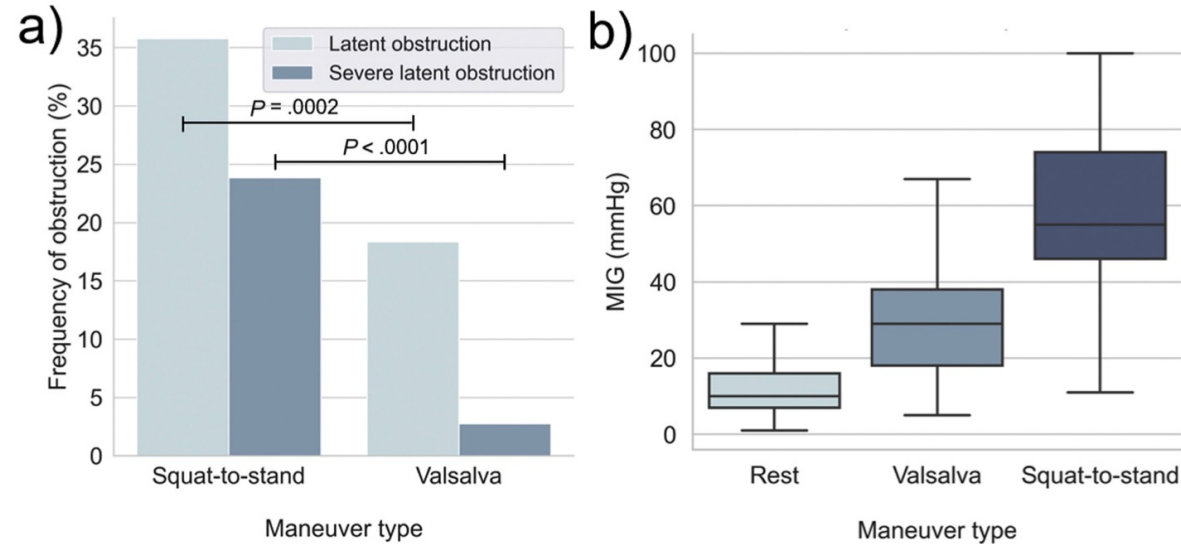
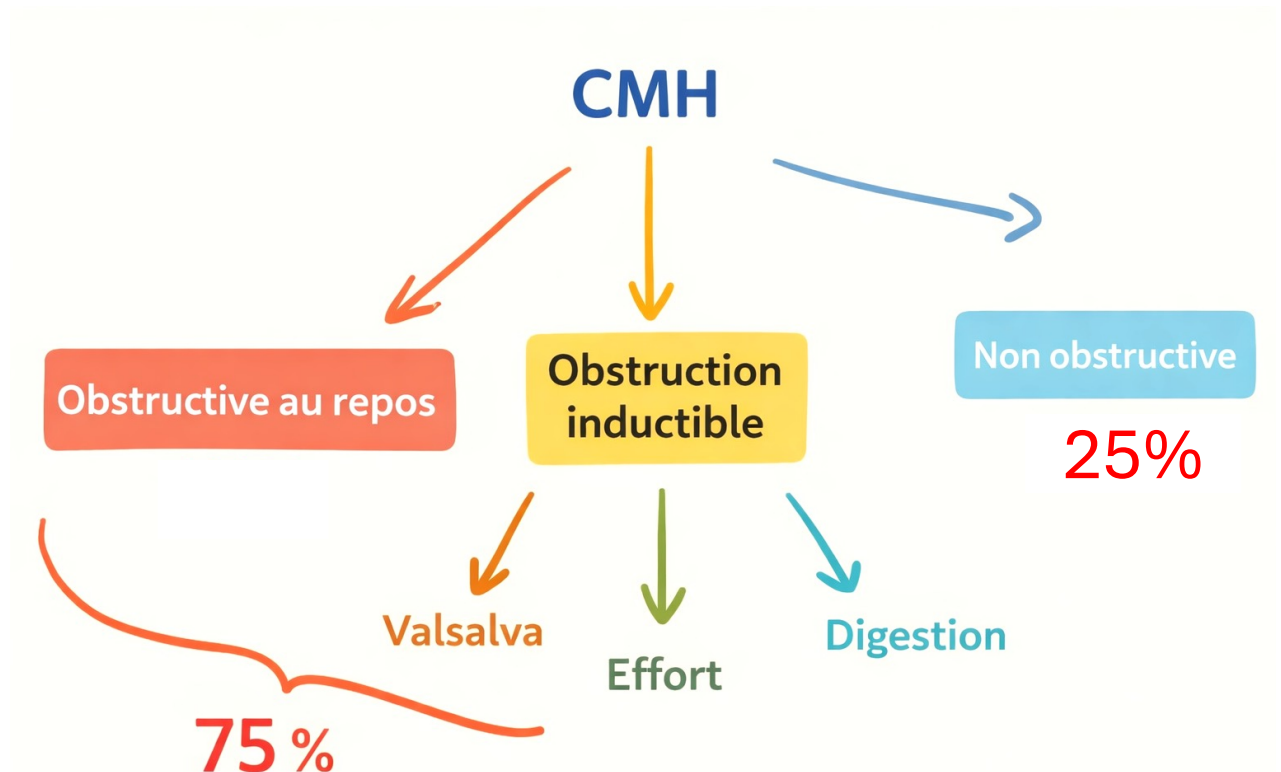


Figure 1 (A) Frequency of obstruction and severe obstruction provoked by each technique. The squat-to-stand maneuver elicited latent obstruction and severe latent obstruction in a higher proportion of patients (36.1% and 25.2%, respectively) than the Valsalva maneuver (18.3% and 2.8%, respectively). (B) Maximal instantaneous gradient (MIG) by provocation techniques. MIG (median and interquartile range) at rest, with Valsalva maneuver, and with squat-to-stand maneuver among patients with latent obstruction who performed both the Valsalva and squat-to-stand maneuvers ($n = 41$). The squat-to-stand maneuver elicited a higher median MIG (55 mm Hg) than the Valsalva maneuver (29 mm Hg).

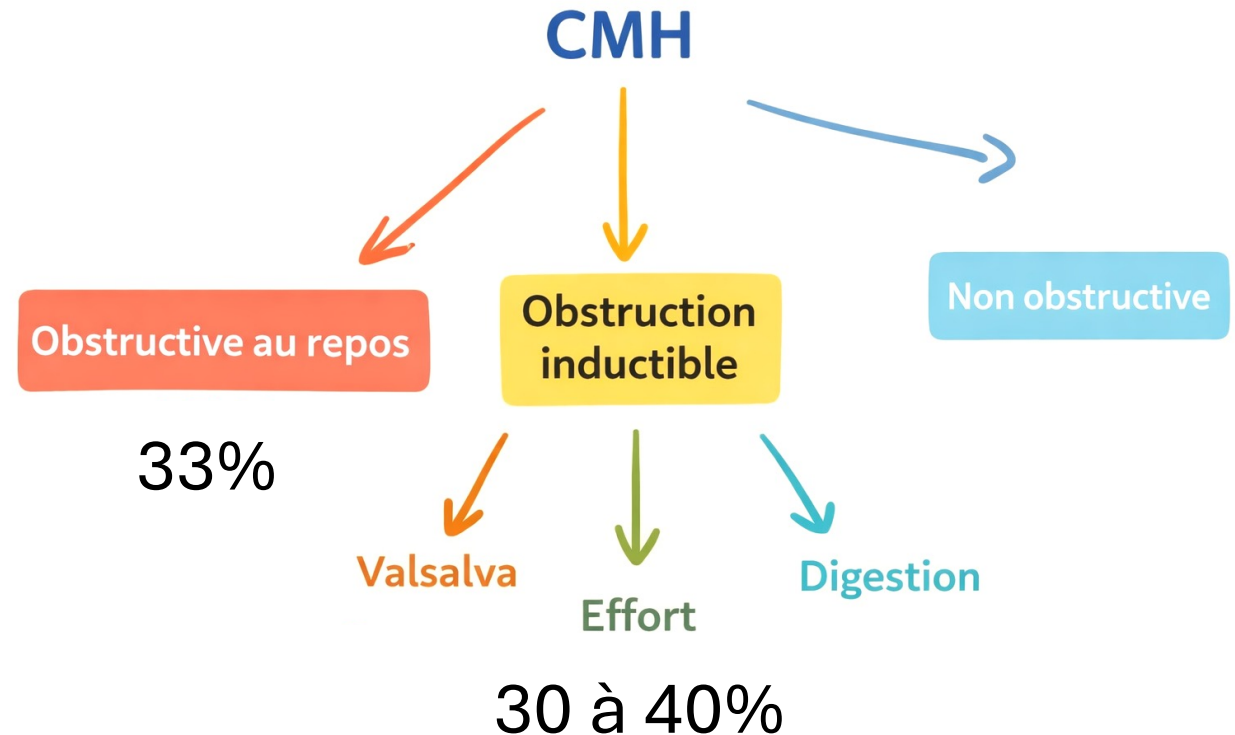
En synthèse:

- 75% (3/4) des cmh sont obstructives spontanément ou après provocation
- 33% sont obstructives spontanément au repos (1/3)
- Mais labile +++



En synthèse:

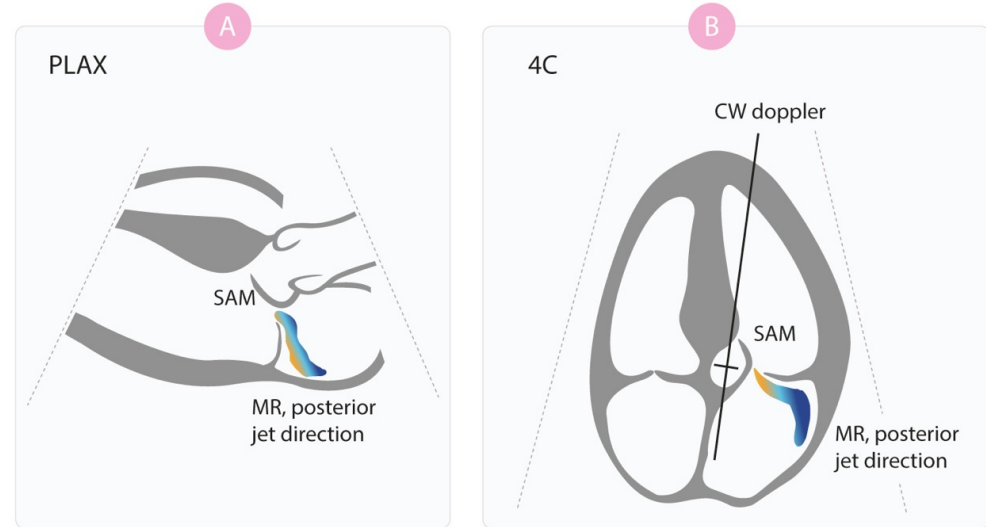
- 75% (3/4) des cmh sont obstructives spontanément ou après provocation
- 33% sont obstructives spontanément au repos (1/3)
- Mais labile +++



phénotypes de CMH

- Obstruction sous aortique
- Médio ventriculaire
- Les deux

Septal hypertrophy with SAM and mitral regurgitation (MR)



Midventricular and apical hypertrophy

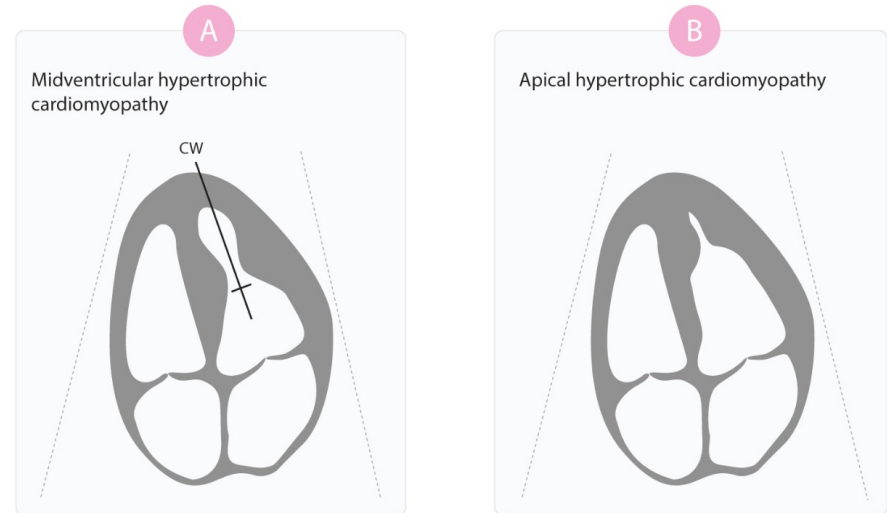
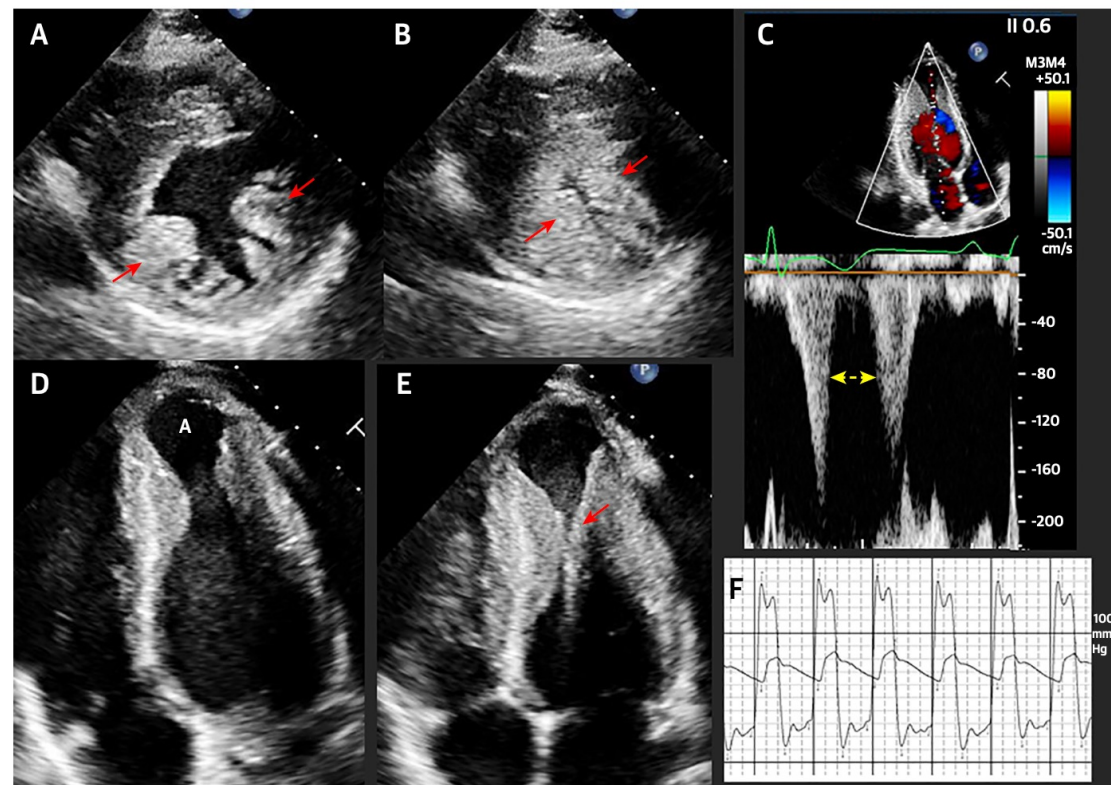
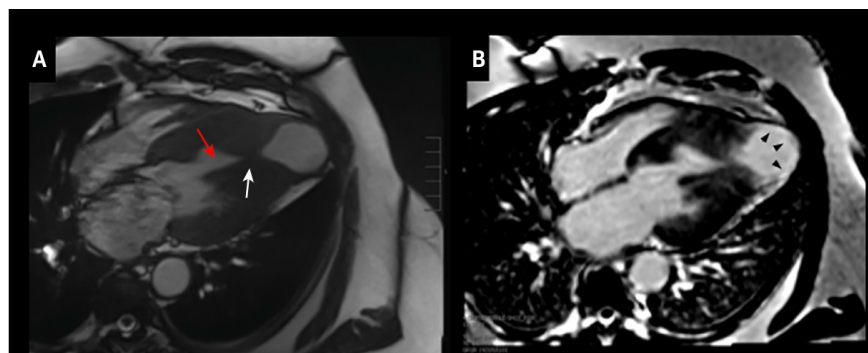


Figure 4. Cardiomyopathie hypertrophique apicale et médio-ventriculaire.

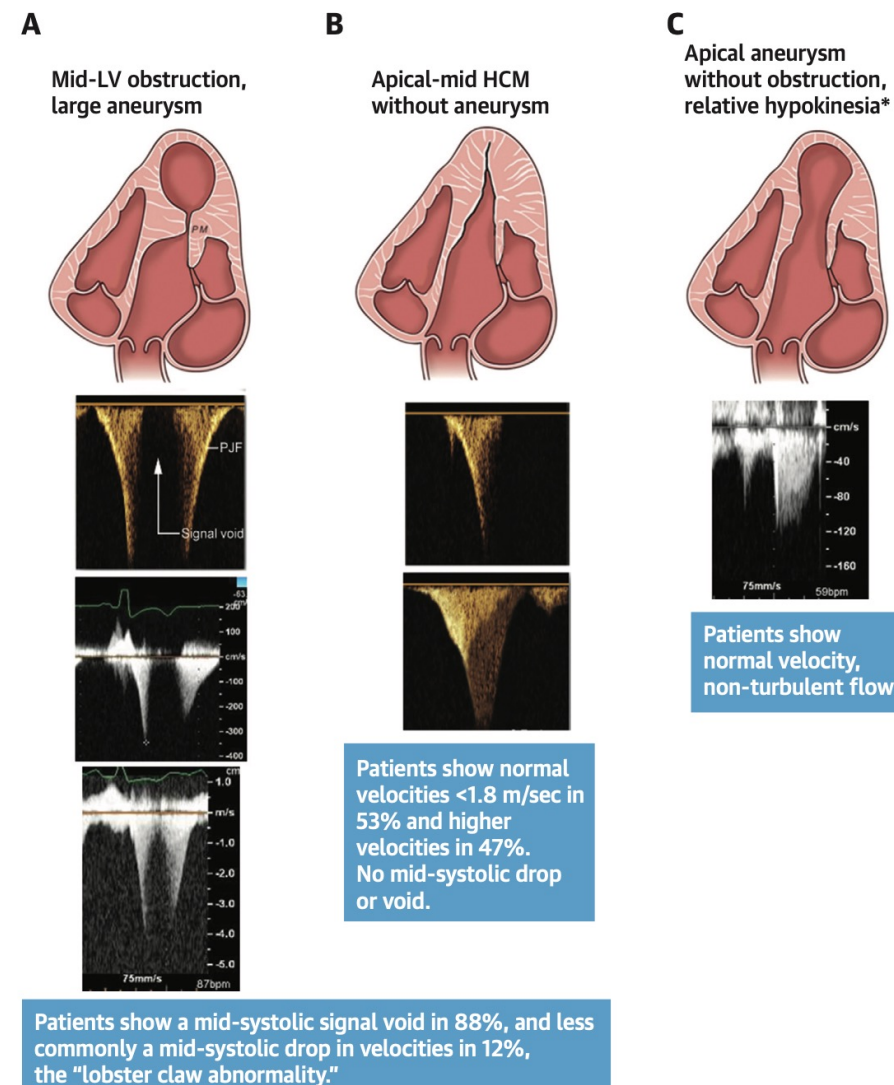
FIGURE 1 Mid-LV Obstruction and Large Aneurysm



Short-axis views in (A) diastole and (B) systole and 4-chamber views in (D) diastole and (E) systole show complete systolic emptying of the mid-left ventricle at the papillary muscle (PM) level. The PMs are hypertrophied and contribute to complete emptying (red arrows). (C) Pulsed wave Doppler at the level of obstruction shows a mid-systolic signal void caused by complete cessation of flow (double yellow arrow). (F) Presurgical catheterization shows a gradient of 65 mm Hg between the apical left ventricle and the ascending aorta despite consistently low velocities across the mid-LV. Doppler underestimates the prevalence and magnitude of gradients.^{10,11} A = aneurysm; LV = left ventricular.



CENTRAL ILLUSTRATION Comparison of 3 Apical-Mid Hypertrophic Cardiomyopathy Phenotypes in Systole



Sherrid MV, et al. J Am Coll Cardiol Img. 2023;16(5):591-605.

Le « PIZZA test »



Ne pas dépasser la dose prescrite...

Le « PIZZA test »

L alcool et le gras augmentent l'obstruction...

Hypovolémie relative...

Avez-vous des symptômes plus intenses en post prandial?

Hydratation?

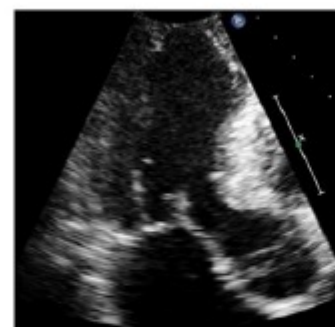
Rendez vous à 14h après un repas un peu lourd....



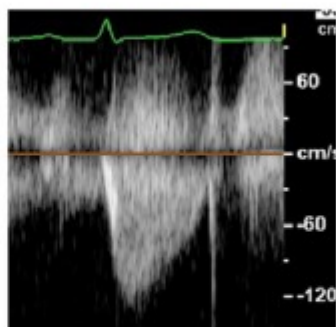
Le « pizza test »

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

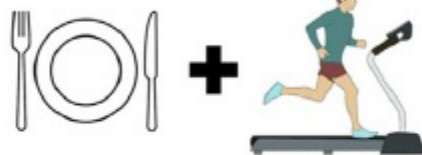
Daniele Massera, MD, MSc, Clarine Long, MD, Yuhe Xia, MS, Les James, MD, MPH, Elizabeth Adlstein, BA,
Isabel C. Alvarez, BS, MPH, Woon Y. Wu, FNP, Maria C. Reuter, AGACNP, Milla Arabadjian, PhD,
Eugene A. Grossi, MD, Muhamed Saric, MD, PhD, and Mark V. Sherid, MD, *New York and Mineola, New York*



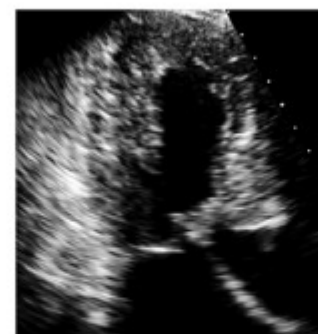
SAM without mitral-septal contact under fasting conditions



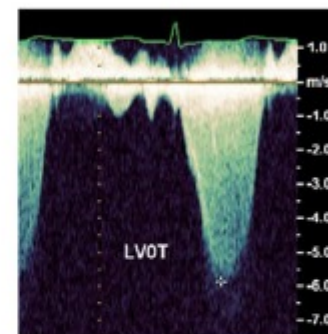
No dynamic LVOT gradient



Postprandial resting / stress echocardiogram



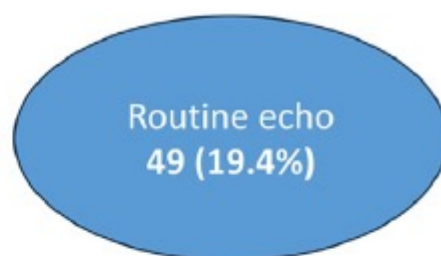
SAM with mitral-septal contact



LVOT gradient 138 mmHg



of patients referred for evaluation of HCM without LVOTO



of patients with LVOT gradient ≥ 50 mmHg on routine echo (rest or provocation)



of patients with LVOT gradient ≥ 50 mmHg only on postprandial echo (rest, provocation or exercise) or only with exercise



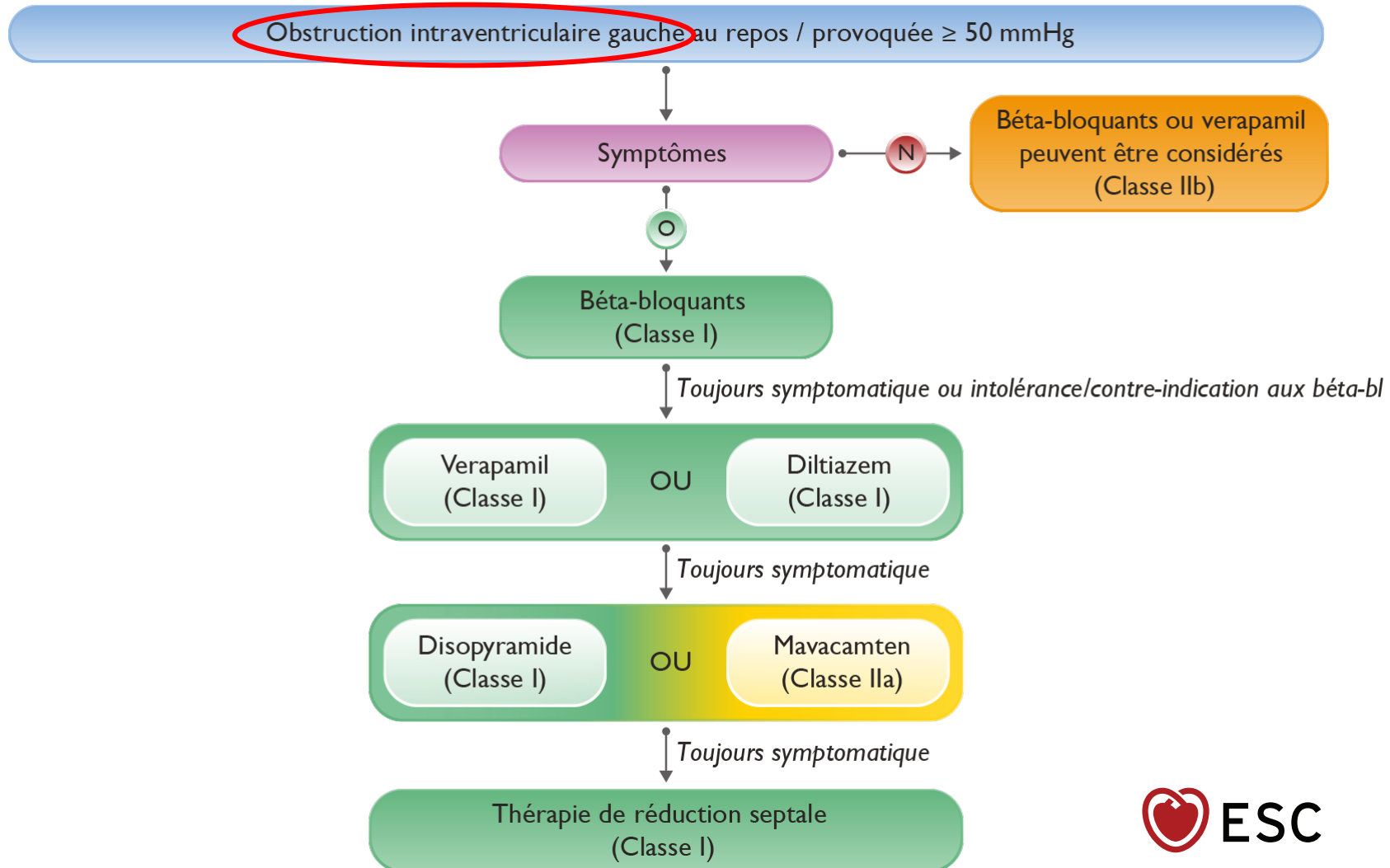
of patients treated with invasive or enhanced drug therapies and who only had postprandial LVOTO ≥ 50 mmHg

Central Illustration Postprandial resting and treadmill stress echocardiography unmasked LVOTO (gradient ≥ 50 mm Hg) in 35.7% of patients who did not have high gradients under routine conditions, with 15.1% achieving it only on postprandial stress echo. More than half of patients who were treated with invasive or enhanced drug therapies (surgical myectomy, alcohol septal ablation, disopyramide, and myosin inhibitors) only had postprandial LVOTO ≥ 50 mm Hg.

Quel traitement ?



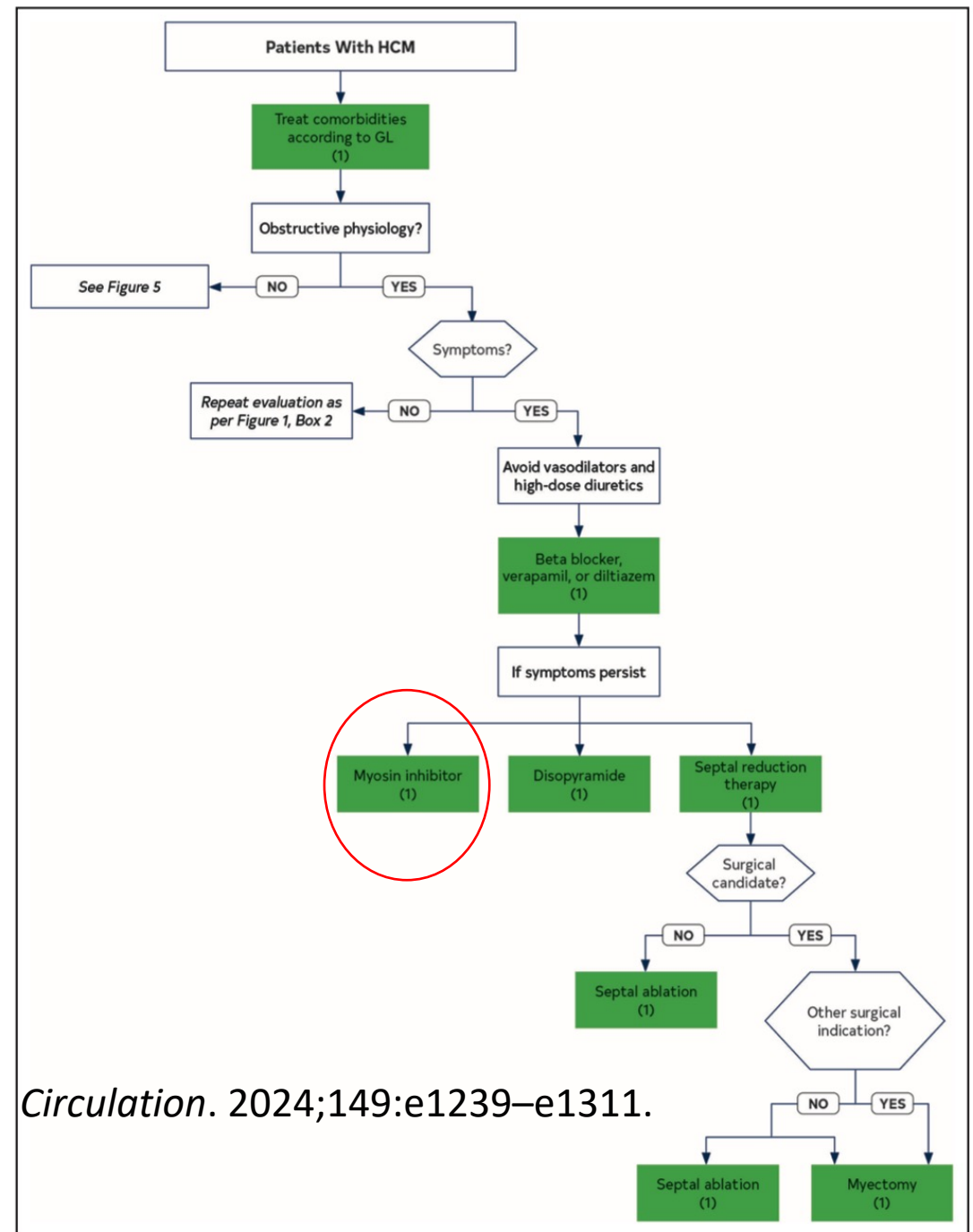
Algorithme thérapeutique CMHo recommandations 2023



Reco ACC/AHA juin 2024

CLINICAL PRACTICE GUIDELINES

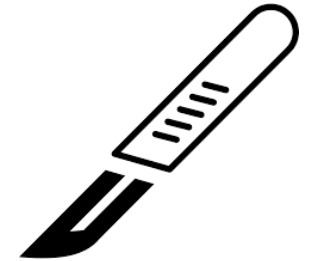
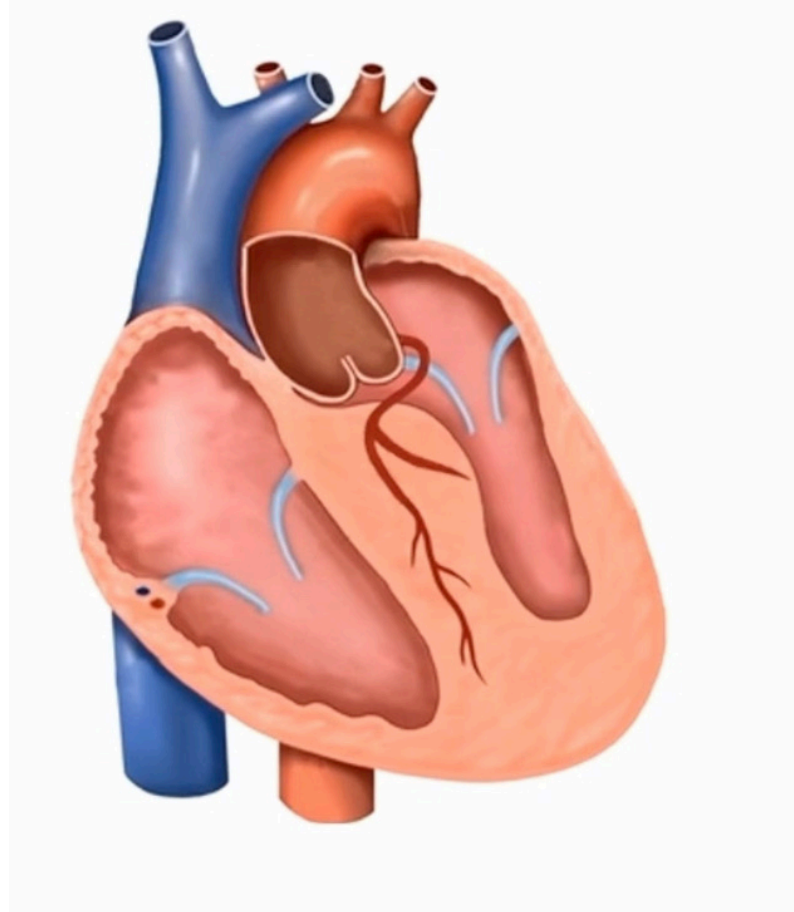
2024 AHA/ACC/AMSSM/HRS/PACES/SCMR
Guideline for the Management of Hypertrophic
Cardiomyopathy: A Report of the American Heart
Association/American College of Cardiology
Joint Committee on Clinical Practice Guidelines



Principes du traitement

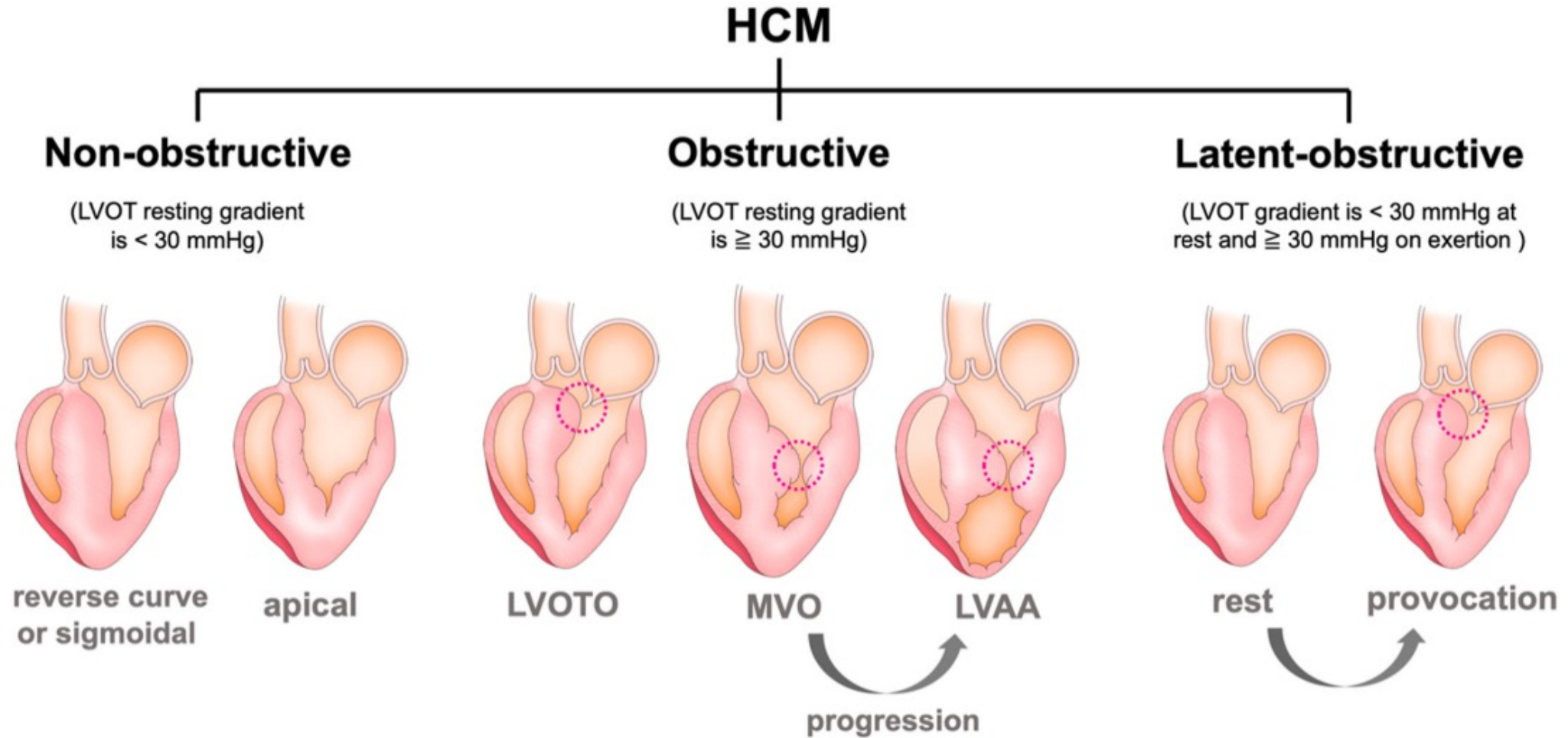


Diminuer la contractilité



Diminuer l'épaisseur septale

CMH et obstruction



Merci de votre attention

- Traitement de la CMH si patient symptomatique +++
- Évaluation complète du patient
- Recherche de l'obstruction à l'état de base
± manœuvres de provocation
(Valsalva, orthostatisme, effort)
- Indications/contre-indications à l'écho d'effort
- Tenir compte des autres facteurs pouvant
participer aux symptômes