GRCI 2017
France

6 AU 8 DÉCEMBRE 2017
Novotel Paris Tour Eiffel

Passion Communication Education

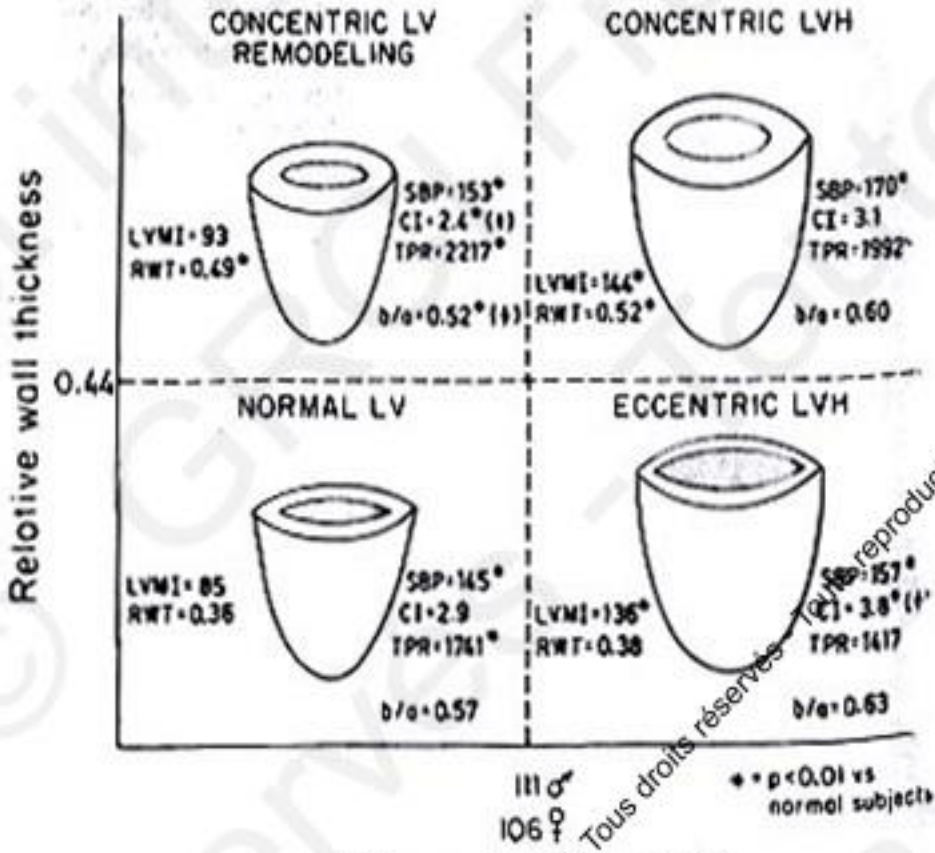
Echographie de la CMH

Dr Thierry LAPERCHE
Centre Cardiologique du Nord
Saint-Denis

- Pas de conflit d'intérêts

2017 © GRCI, Tous droits réservés - Toute reproduction même partielle est interdite.

Géométrie ventriculaire gauche chez l'hypertendu



Ganau A. JACC 1992;19:1550-8

Définition

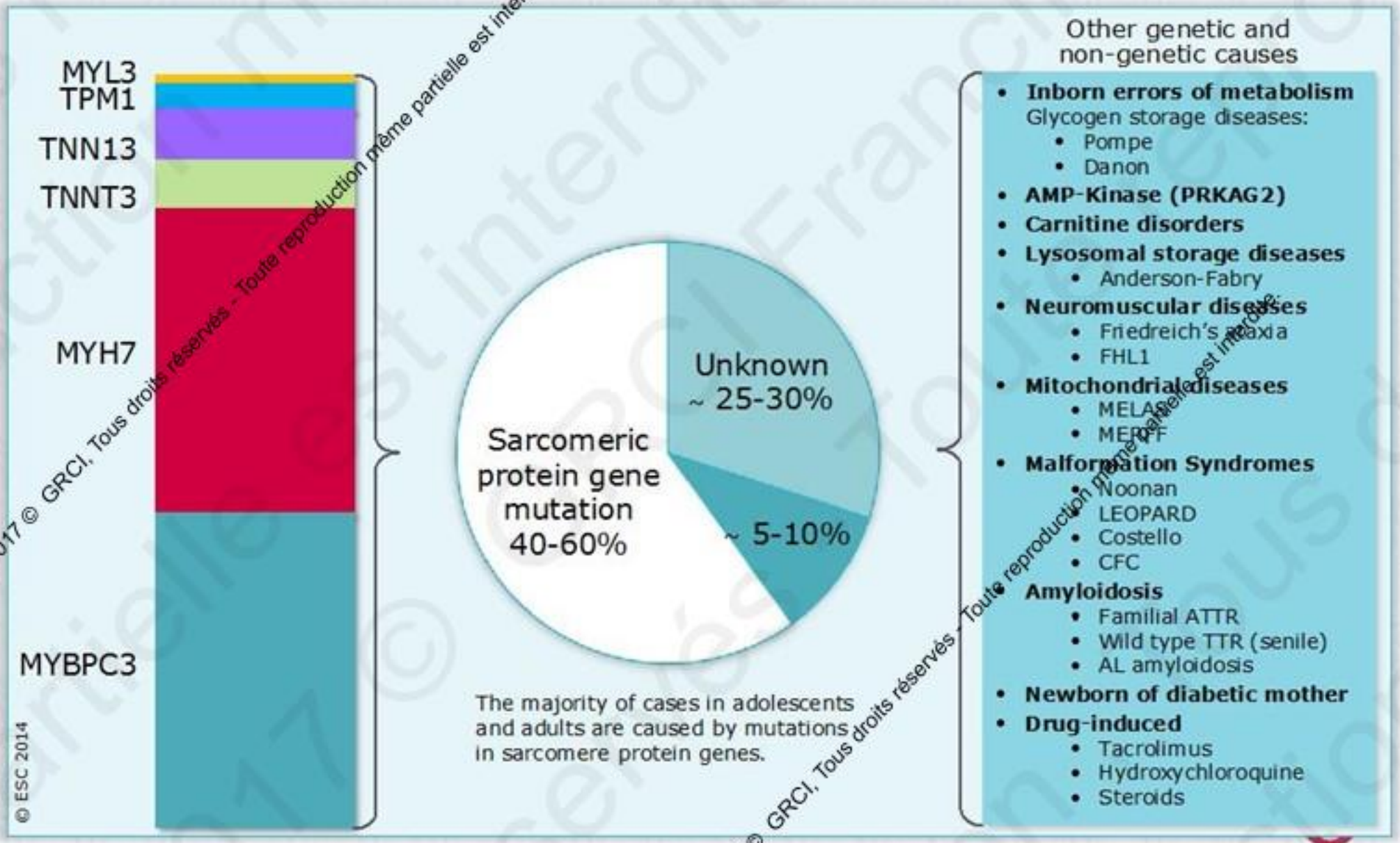
- Présence d'une épaisseur pariétale ventriculaire gauche augmentée (>15mm), pas uniquement expliquée par une augmentation des conditions de charge

- Prévalence : 0.02-0.23% de la population adulte

- A différencier de l'hypertrophie physiologique du sportif, des conséquences de l'HTA ou de certaines valvulopathies, de l'hypertrophie septale du sujet âgé.

ESC 2014

Diverse aetiology of hypertrophic cardiomyopathy



European Heart Journal (2014);35:2733–2779 - doi:10.1093/eurheartj/ehu284

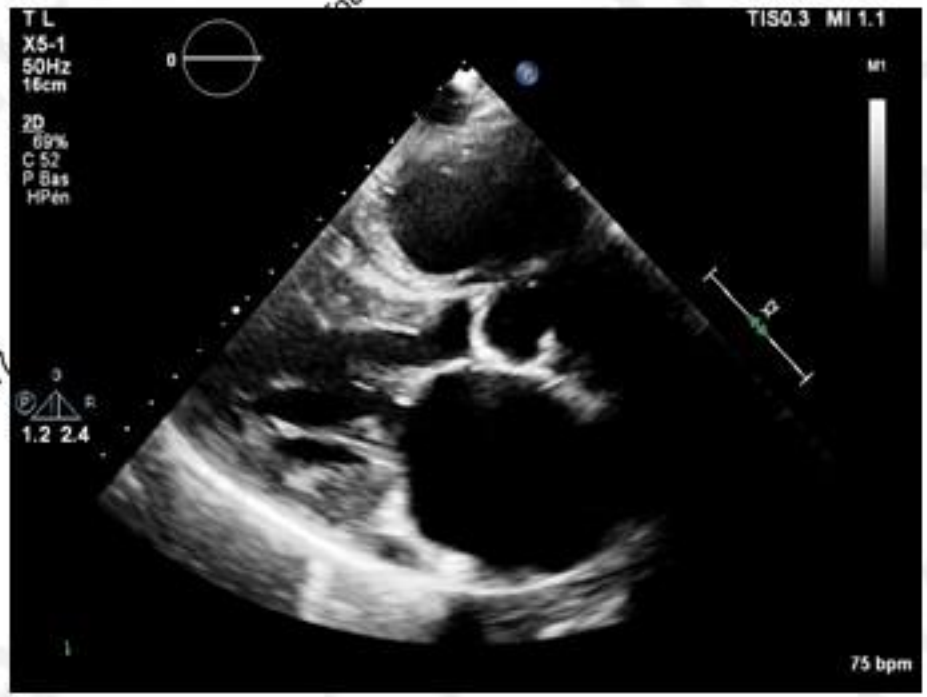
EUROPEAN SOCIETY OF CARDIOLOGY®

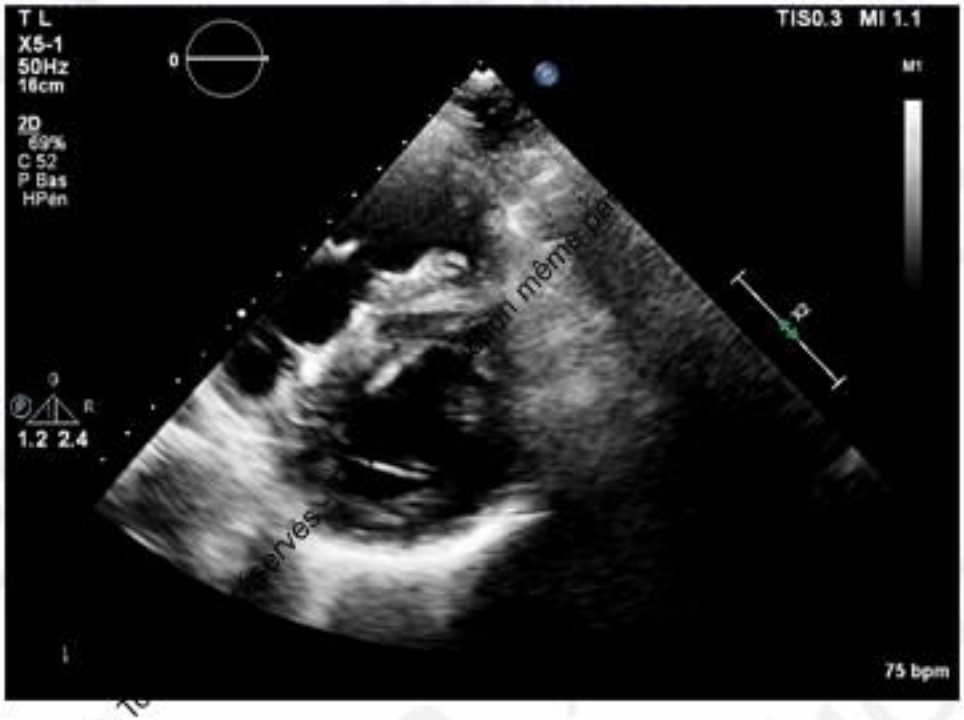
Echocardiographic features that suggest specific aetiologies

Finding	Specific diseases to be considered
Increased interatrial septum thickness	Amyloidosis
Increased AV valve thickness	Amyloidosis; Anderson-Fabry disease
Increased RV free wall thickness	Amyloidosis, myocarditis, Anderson-Fabry disease, Noonan syndrome and related disorders
Mild to moderate pericardial effusion.	Amyloidosis, myocarditis
Ground-glass appearance of ventricular myocardium on 2-D echocardiography	Amyloidosis
Concentric LVH	Glycogen storage disease, Anderson-Fabry disease, PRKAG2 mutations
Extreme concentric LVH (wall thickness ≥30 mm)	Danon disease, Pompe disease
Global LV hypokinesia (with or without LV dilatation)	Mitochondrial disease, TTR-related amyloidosis, PRKAG2 mutations, Danon disease, myocarditis, advanced sarcomeric HCM, Anderson-Fabry disease
Right ventricular outflow tract obstruction	Noonan syndrome and associated disorders

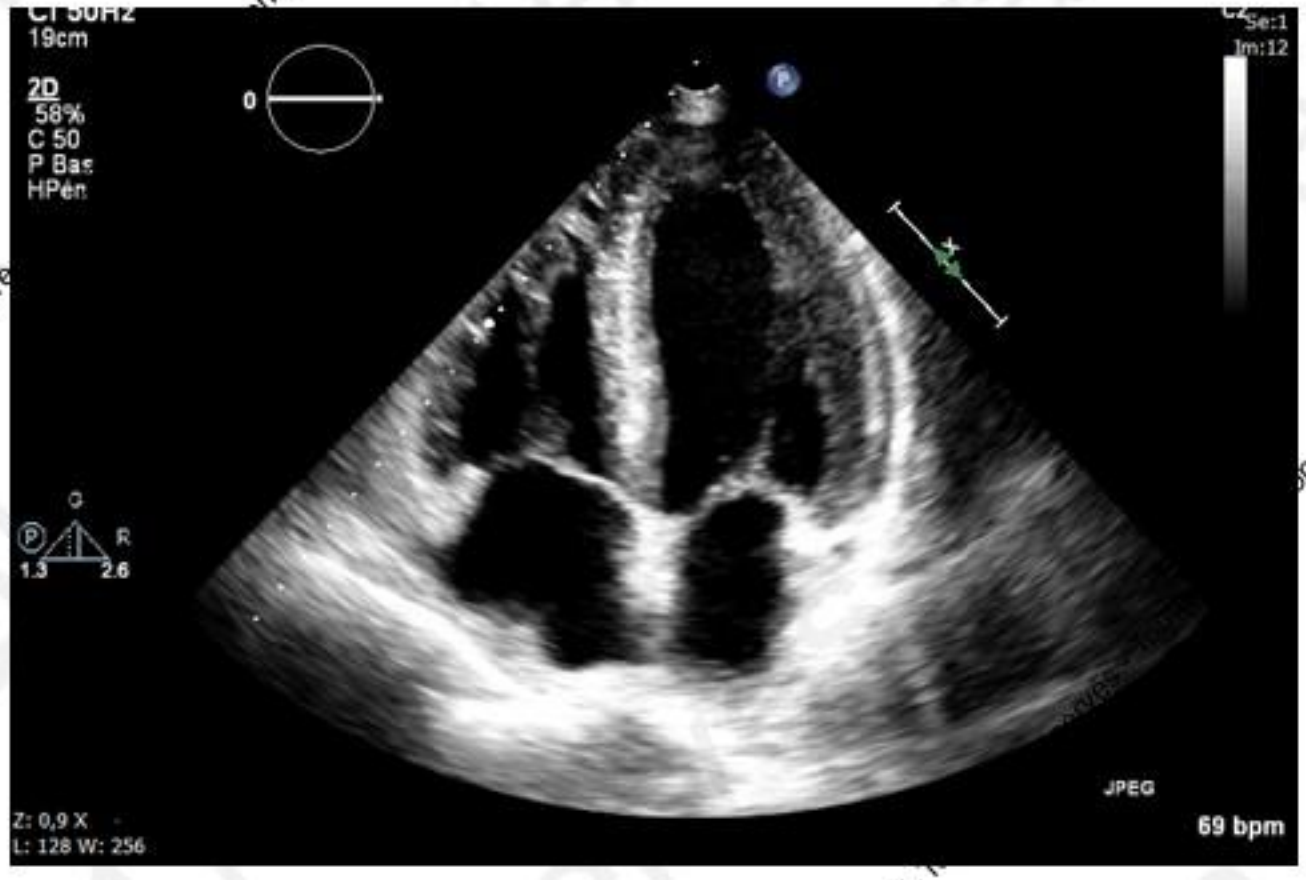
PRKAG2 = gamma-2 subunit of the adenosine monophosphate-activated protein kinase;

Pièges

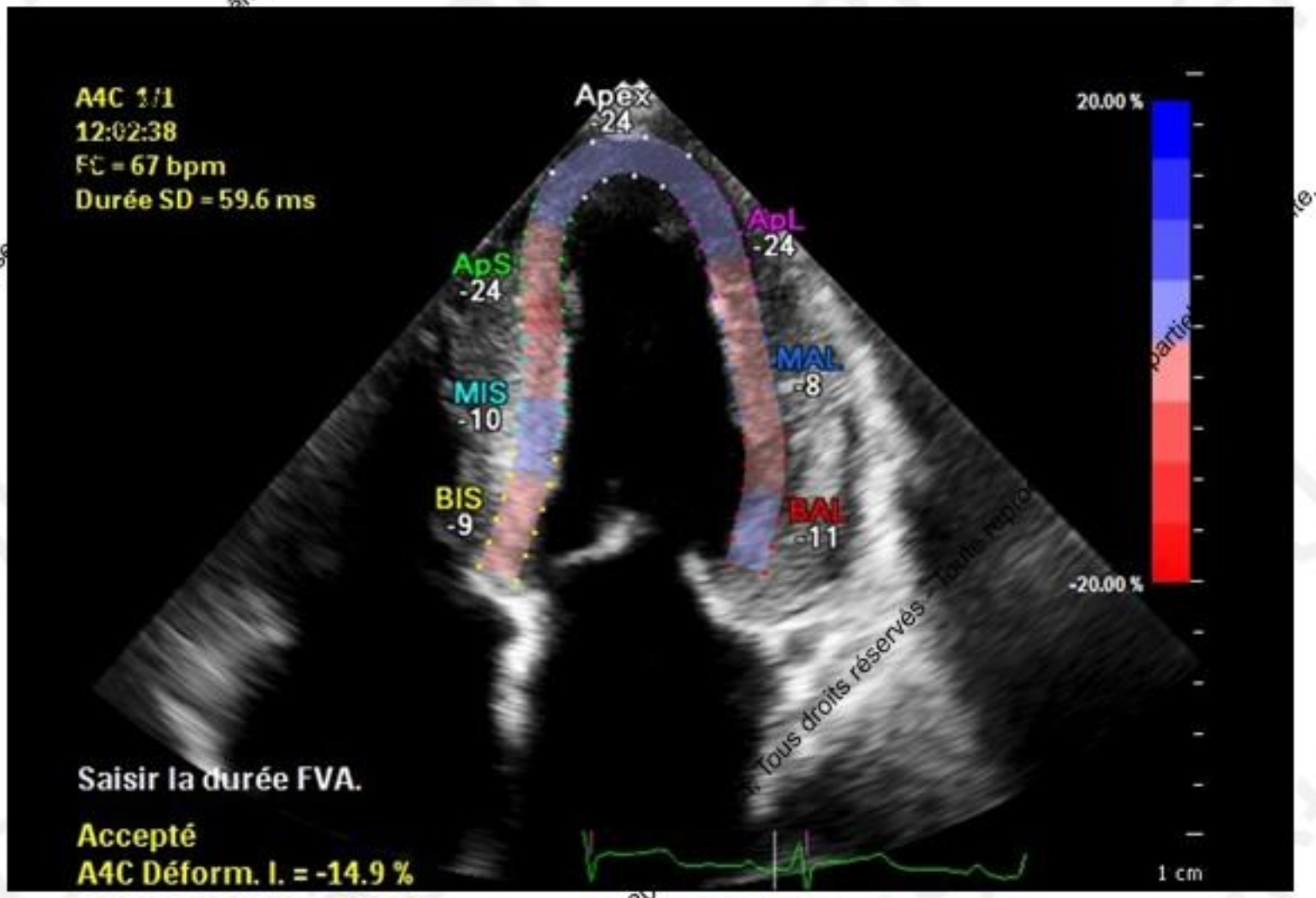




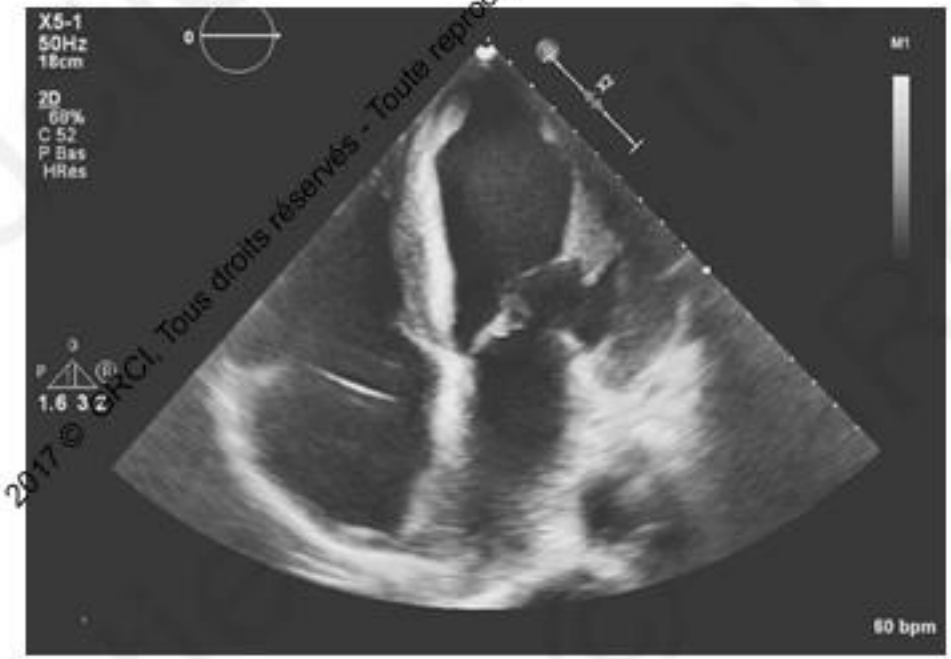
Amylose

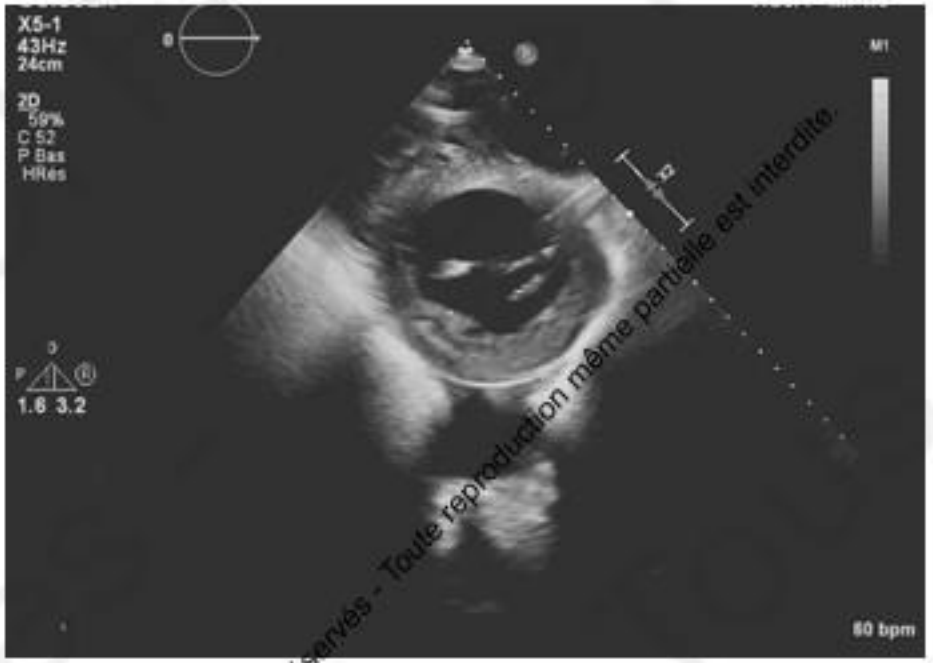


Amylose : préservation de la contractilité apicale.

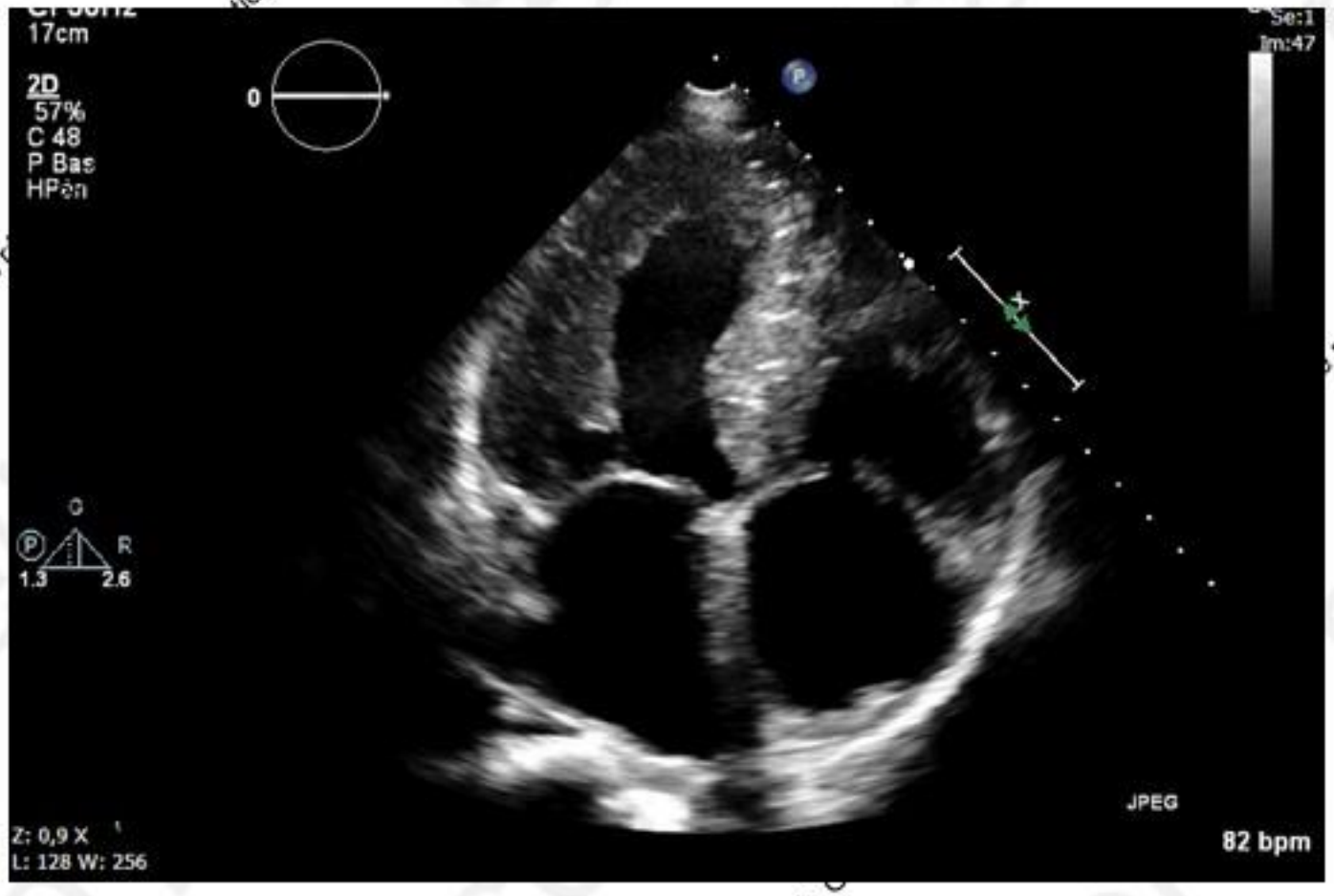


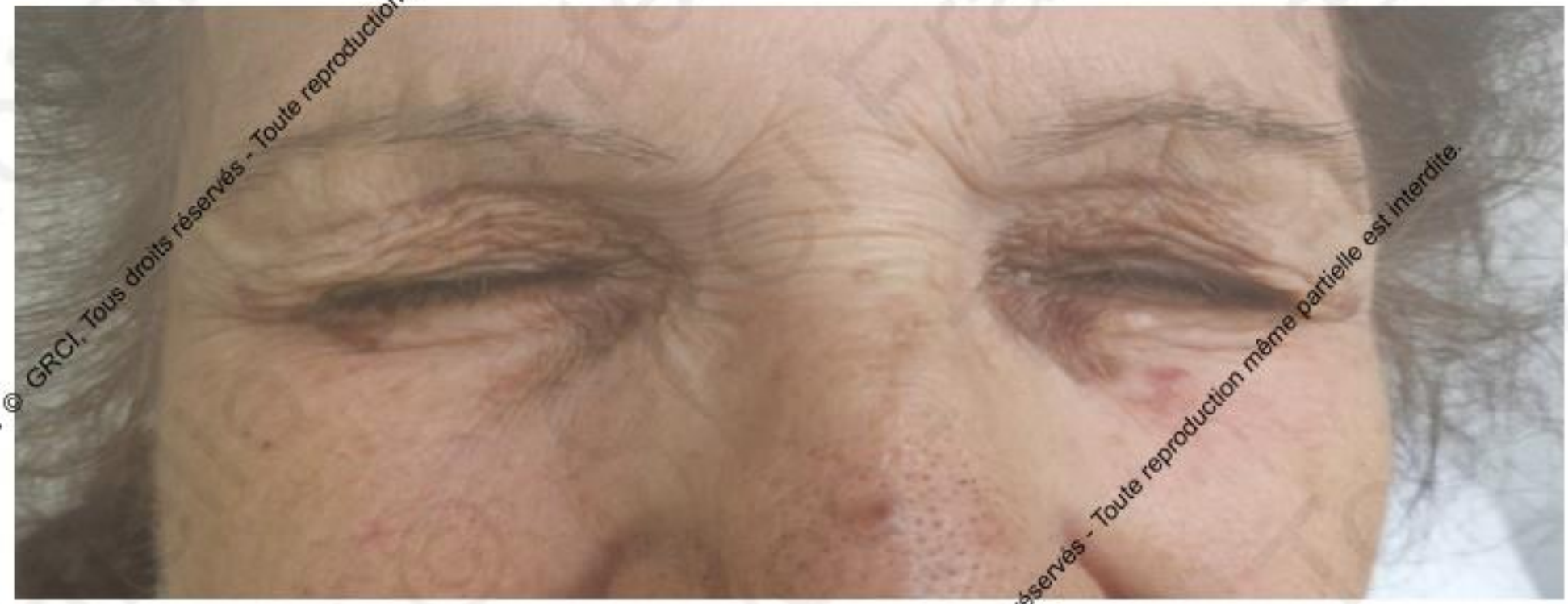
Amylose





Amylose cardiaque. Forme avancée





2017 © GRCI, Tous droits réservés - Toute reproduction même partielle est interdite.

Maladie de Fabry

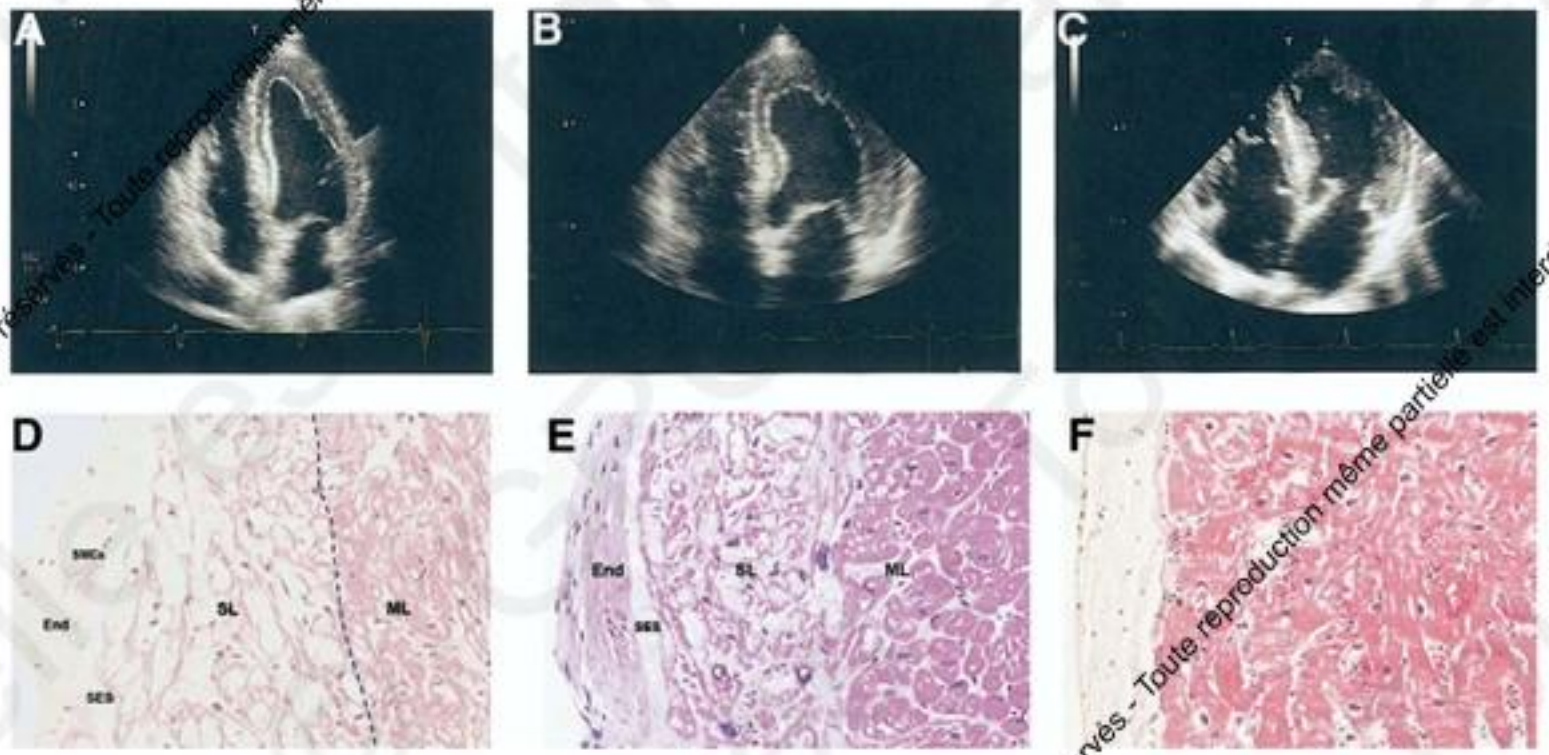
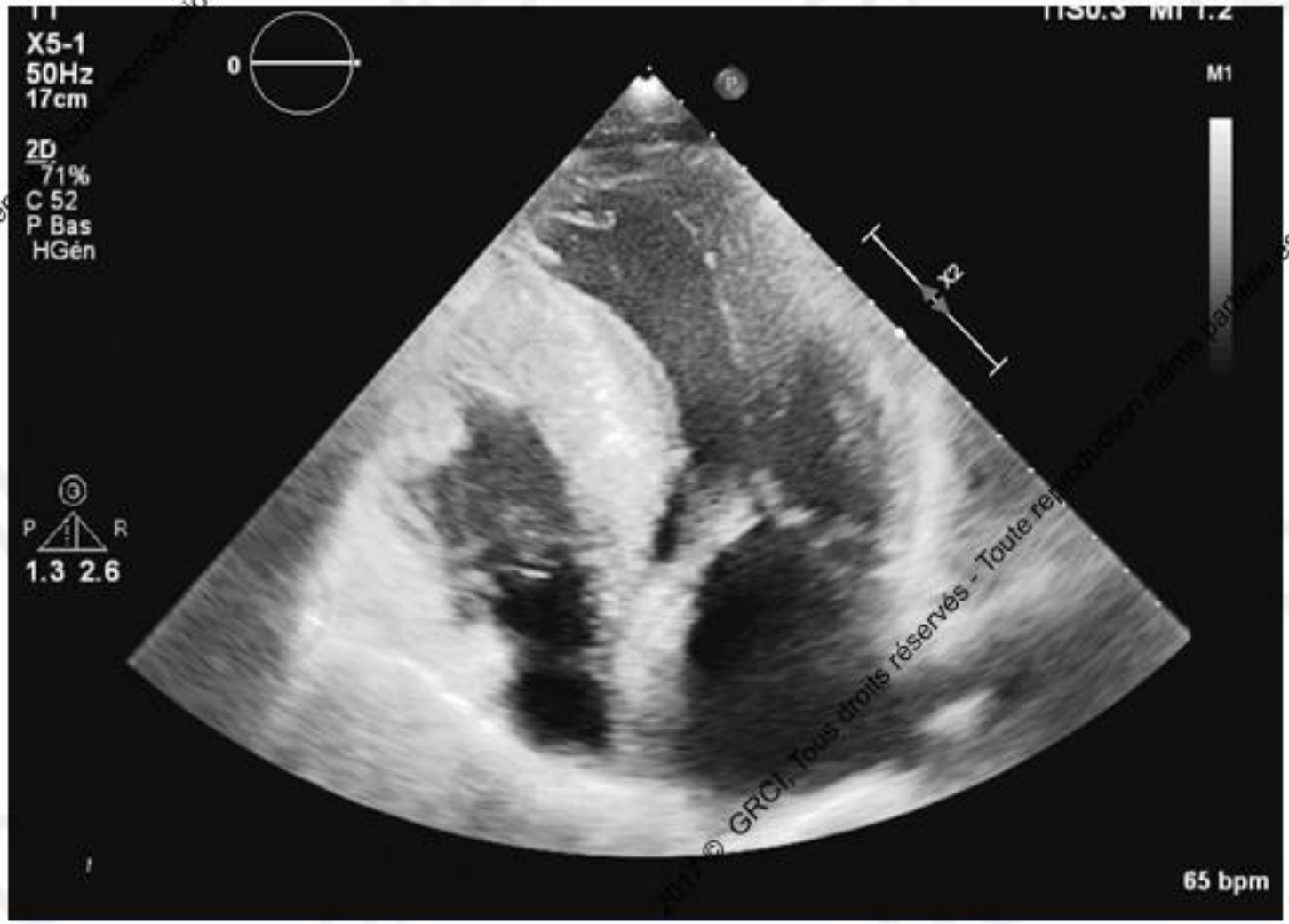


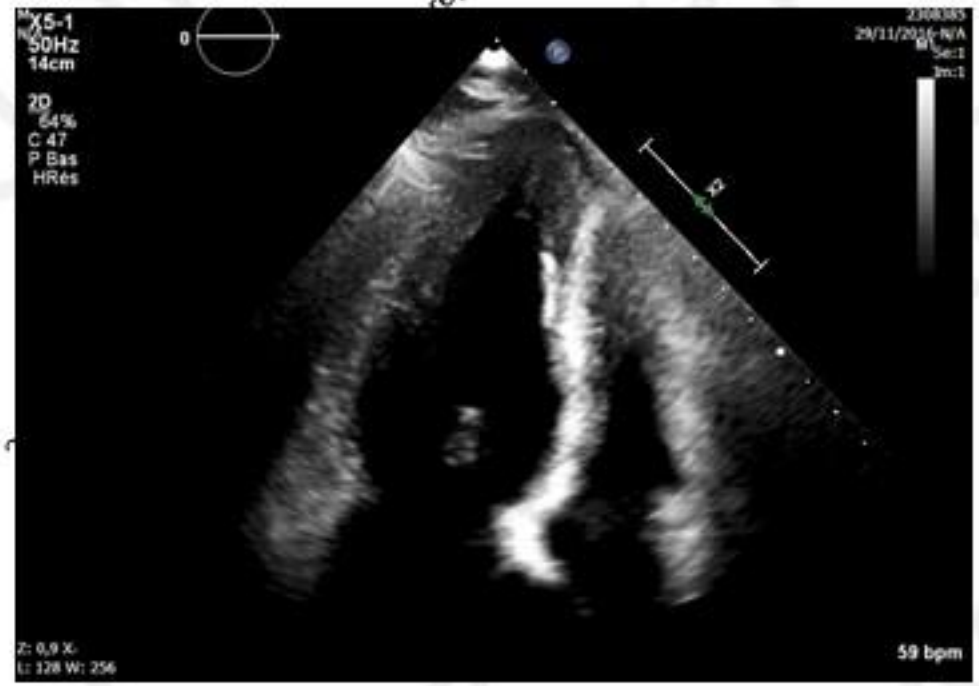
Figure 1. Two-dimensional echocardiography in four-chamber apical view and left ventricular endomyocardial biopsy from two patients (Patient #4 and Patient #18 of Table 2) with Fabry's disease cardiomyopathy (**A,D** and **B,E**, respectively) and a patient with hypertrophic cardiomyopathy **(C,F)**. Comparison of the three echocardiographic frames reveals the presence of a binary appearance of left ventricular endocardial border in the two Fabry patients **(A,B)**. This echocardiographic finding reflects the glycosphingolipids compartmentalization involving a thickened endocardium (End) with enlarged and engulfed smooth muscle cells (SMC), a subendocardial empty space (SES), and a prominent involvement of subendocardial myocardial layer (SL), while the middle layer (ML) appears partially spared **(D,E)**. The echocardiographic pattern is absent in hypertrophic cardiomyopathy **(C)**, despite a similar thickening of the endocardium **(F)**.

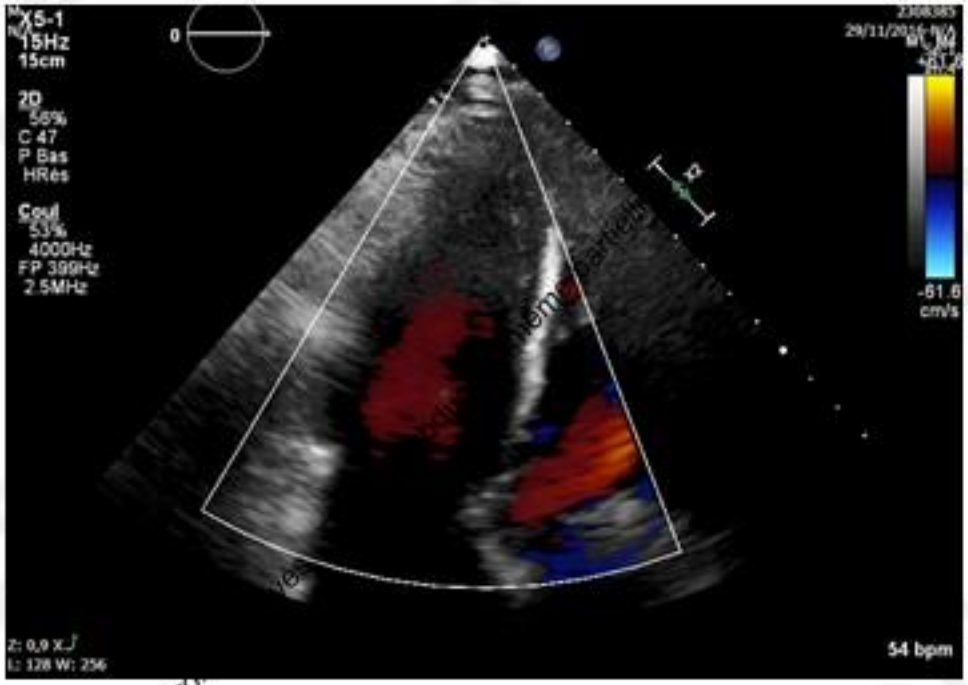
Pieroni M et al. J Am Coll Cardiol 2006;47:1663-71

CMH diffuse

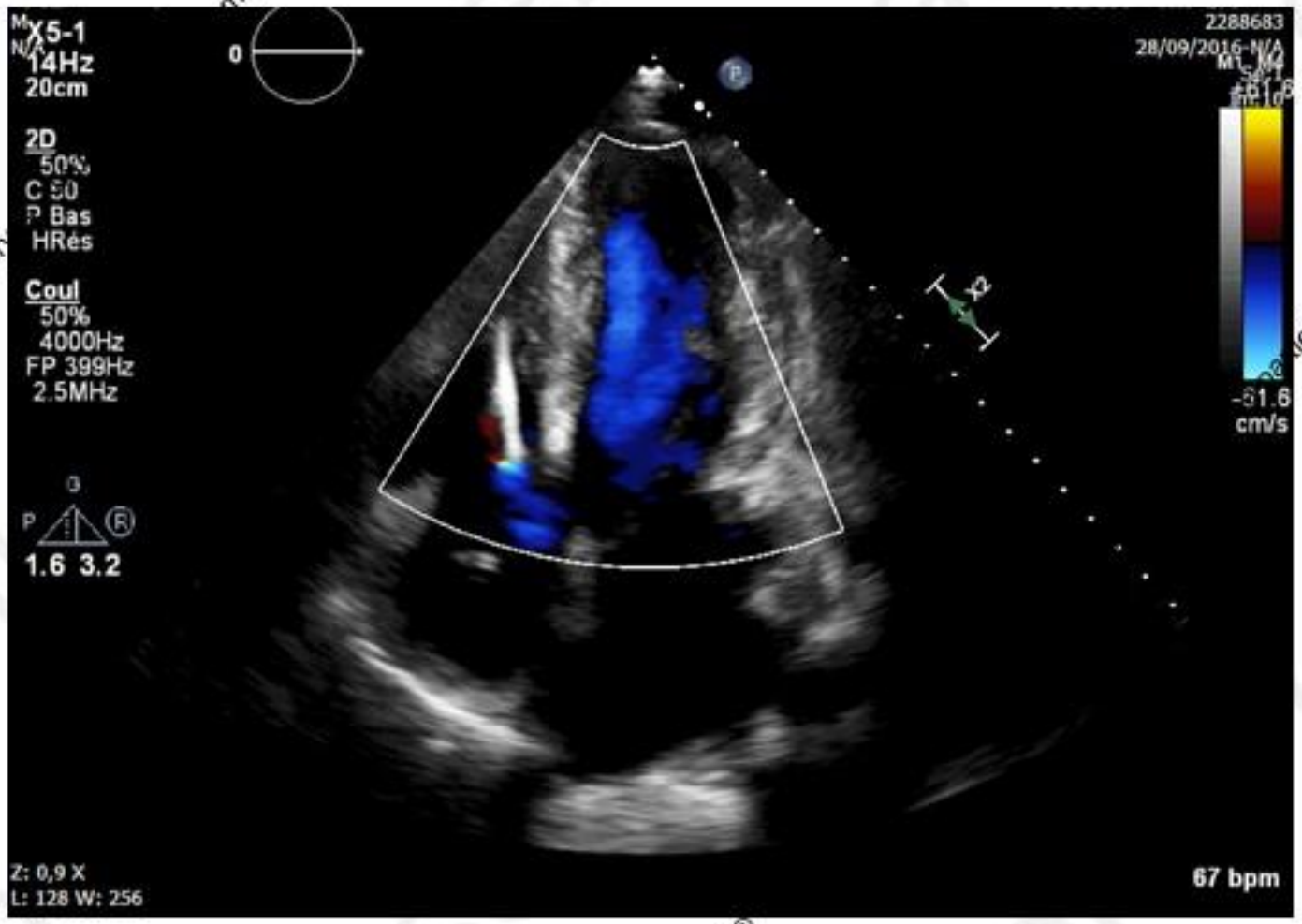


CMH apicale

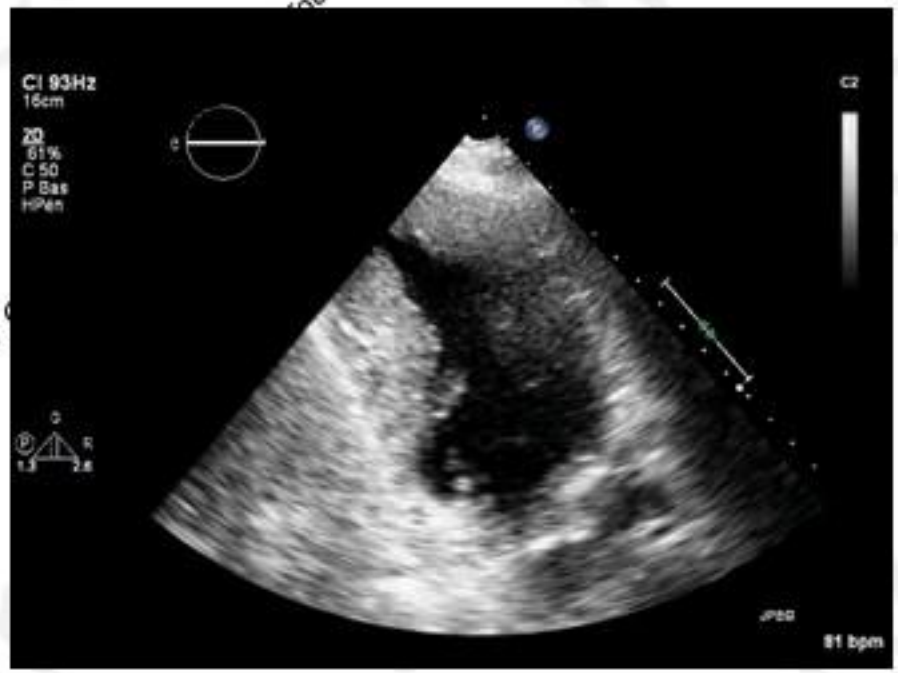


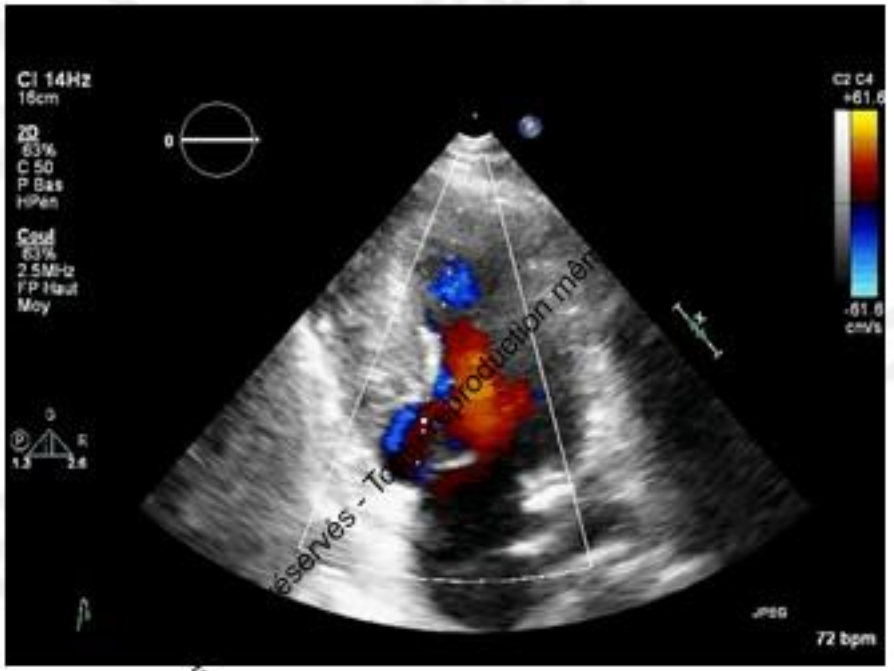


CMH médioventriculaire

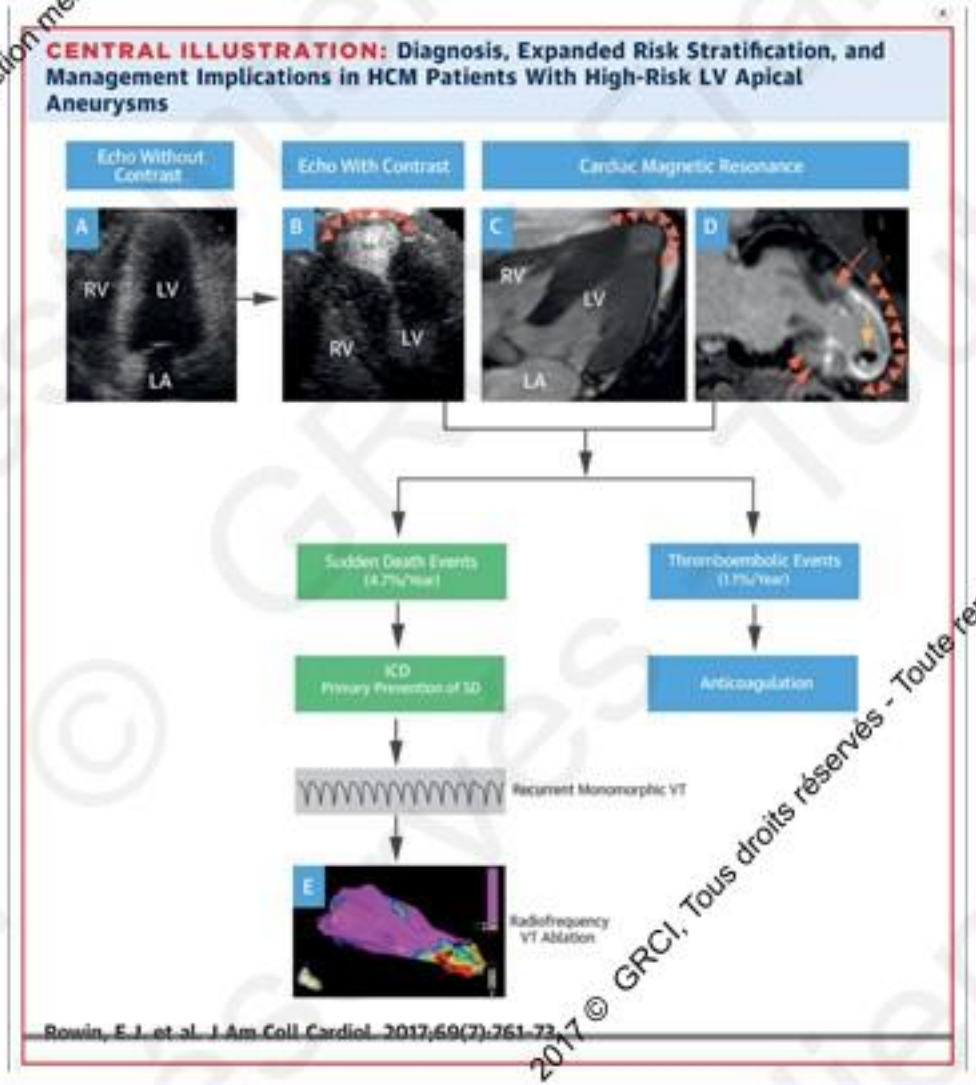


CMH. Obstacle médioventriculaire



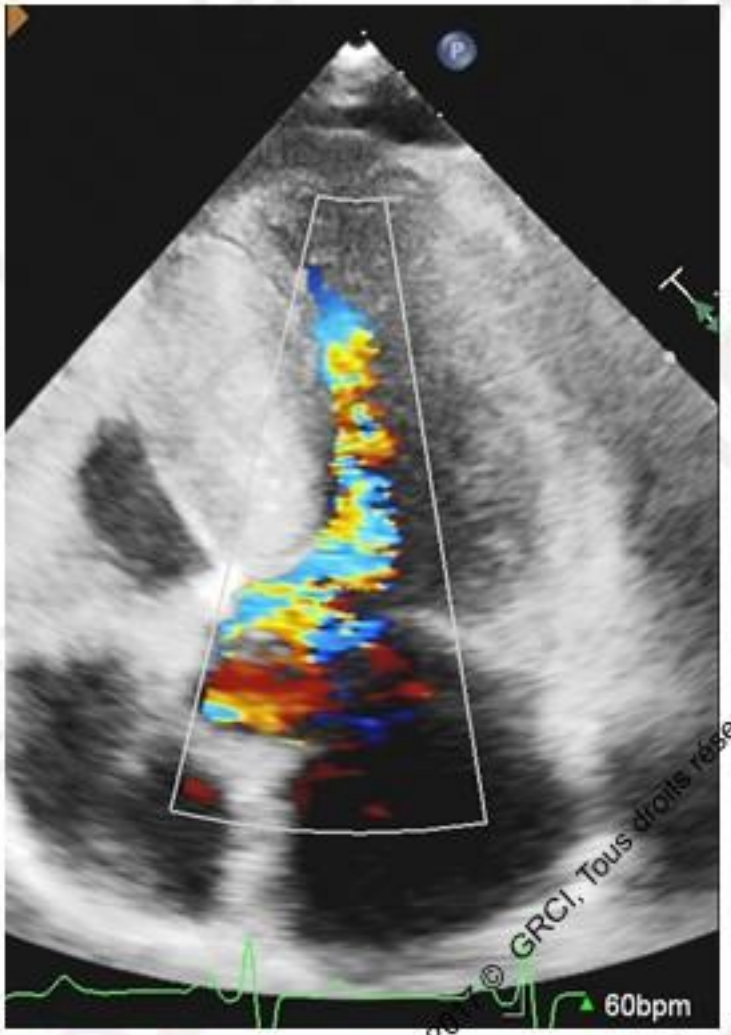


Formes avec anévrysme apical

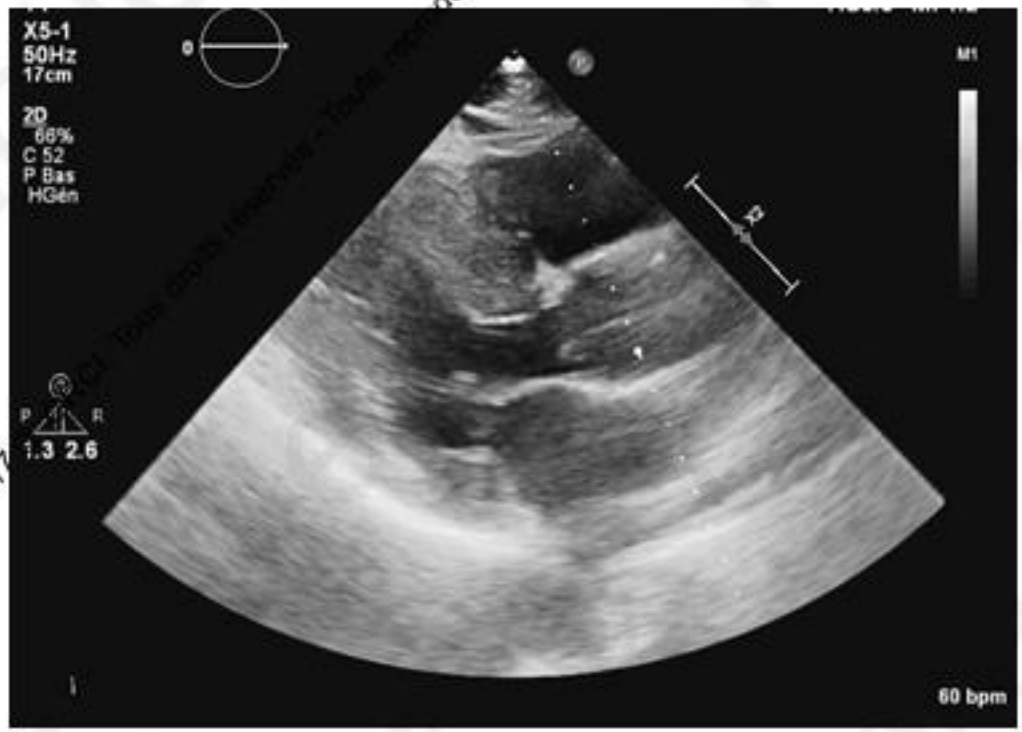


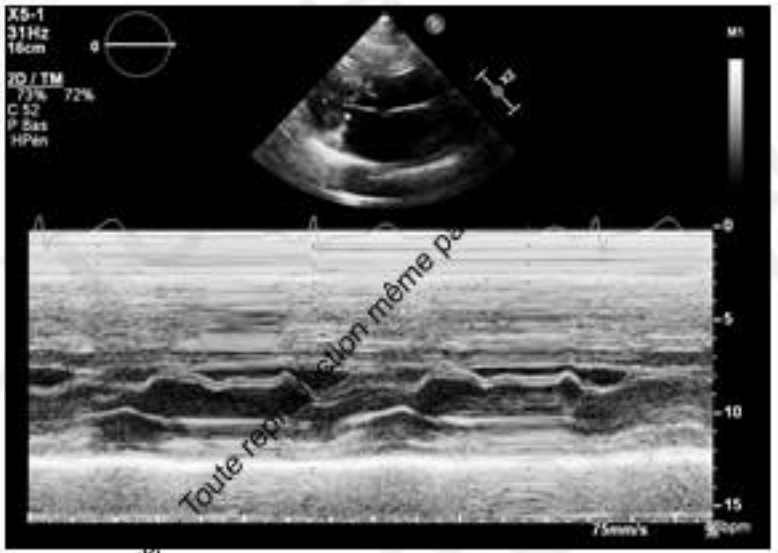
2017 © GRCI, Tous droits réservés - Toute reproduction même partielle est interdite.
2017 © GRCI, Tous droits réservés - Toute reproduction même partielle est interdite.

CMH. Obstacles étagés

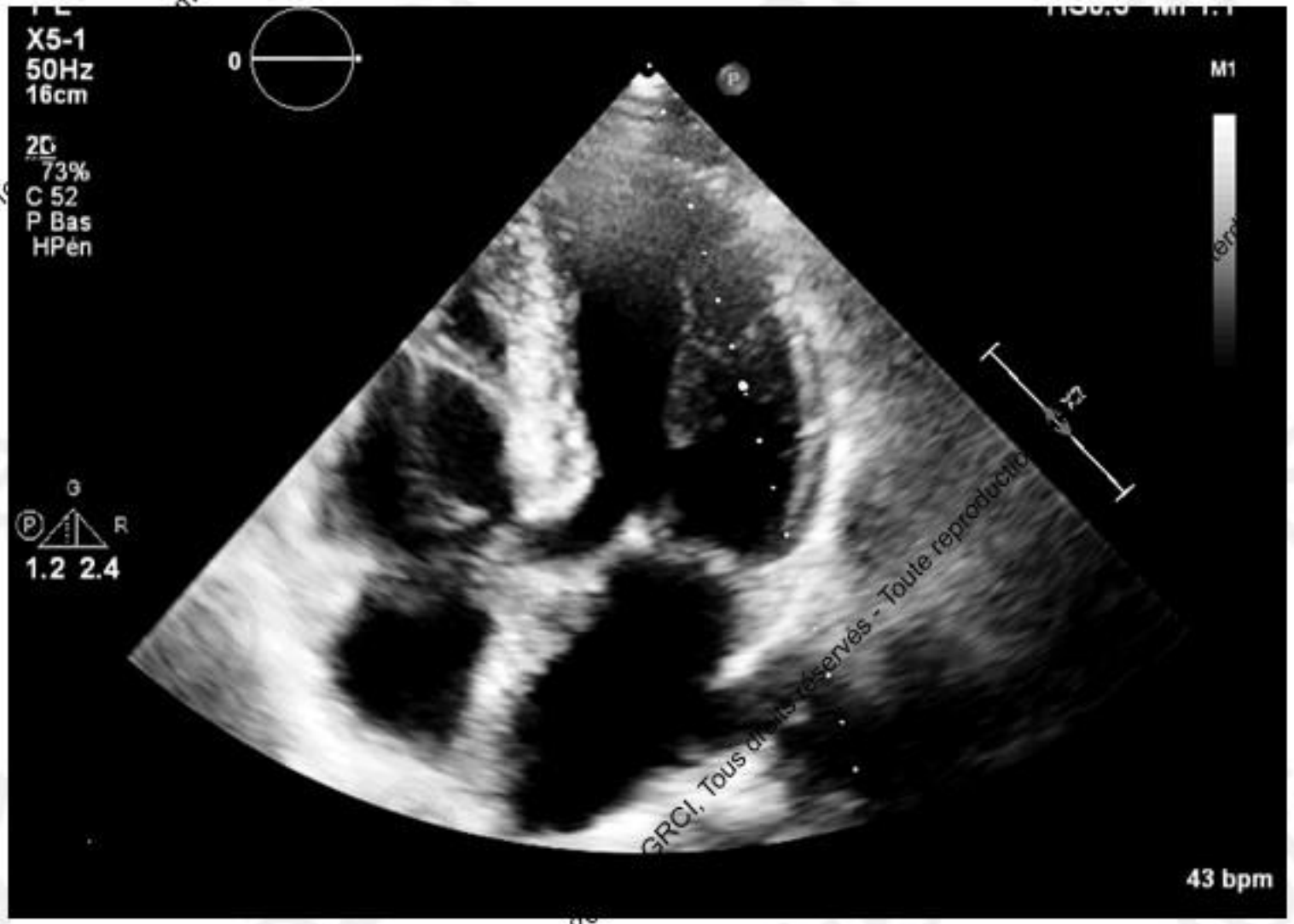


CMH. SAM

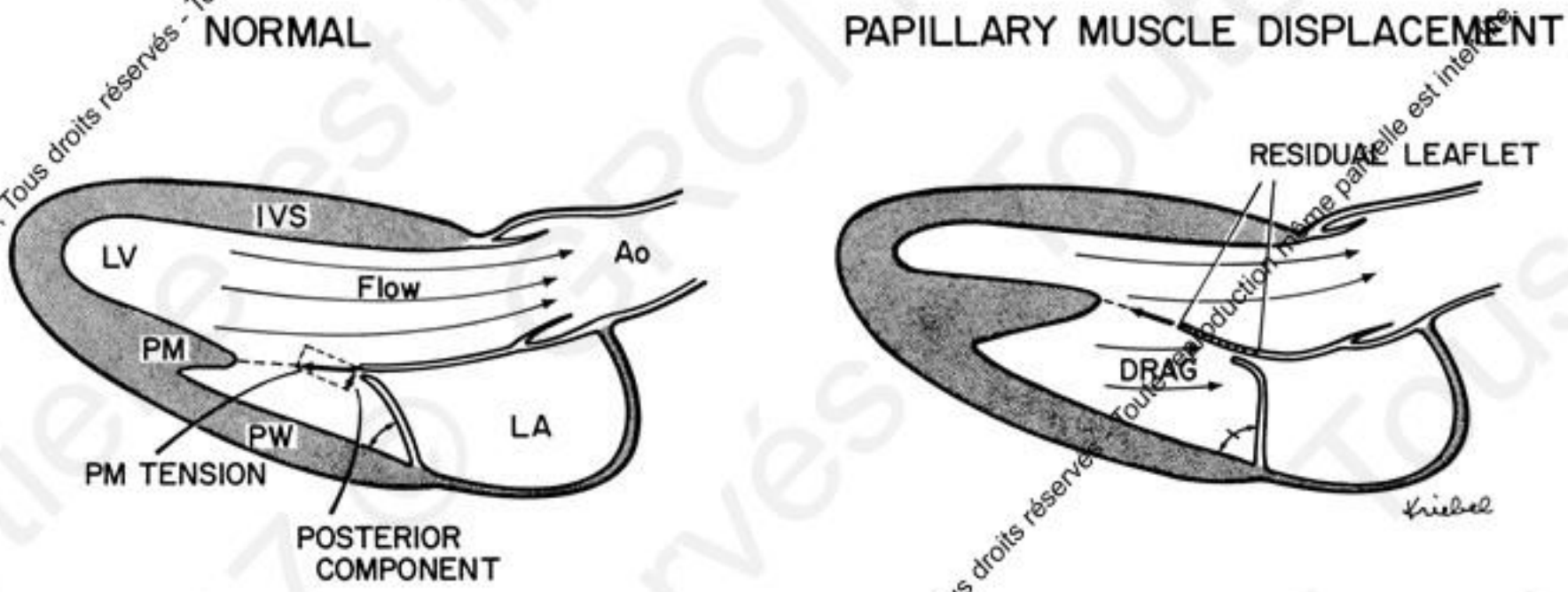




CMH. SAM

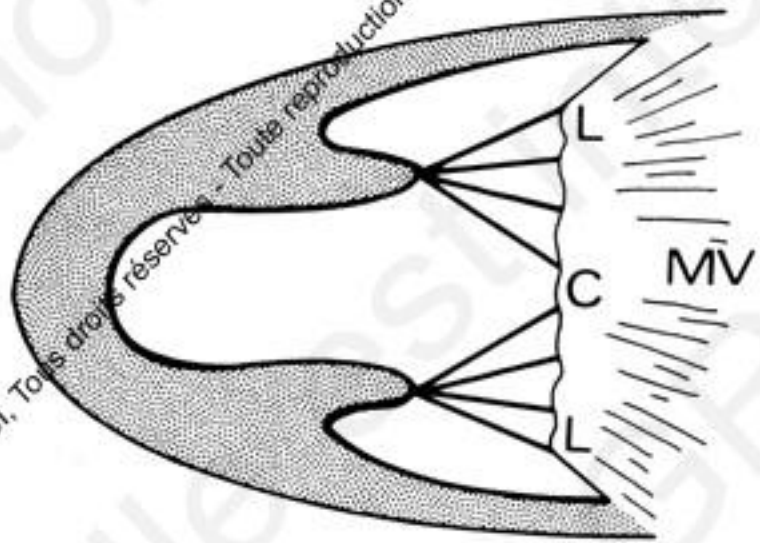


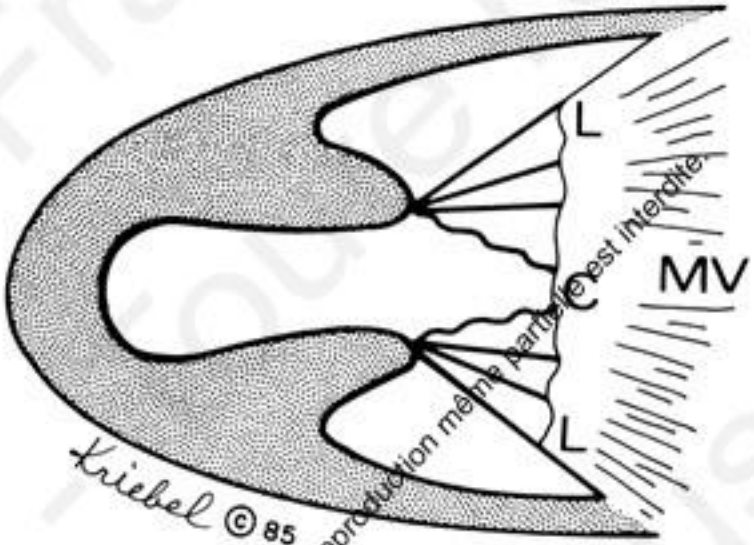
Drawings of possible mechanisms for systolic anterior motion with anterior displacement of the papillary muscles (PM): (1) the normal posterior component of PM tension is reduced by anterior displacement of the muscle tips; (2) interposing the leaflets into the streamlines of flow causes drag forces with an anterior component; and (3) pulling up the posterior leaflet so that it meets the anterior leaflet closer to its base creates a long, overlapping residual leaflet, as seen clinically.



Robert A. Levine et al. Circulation. 1995;91:1189-1195

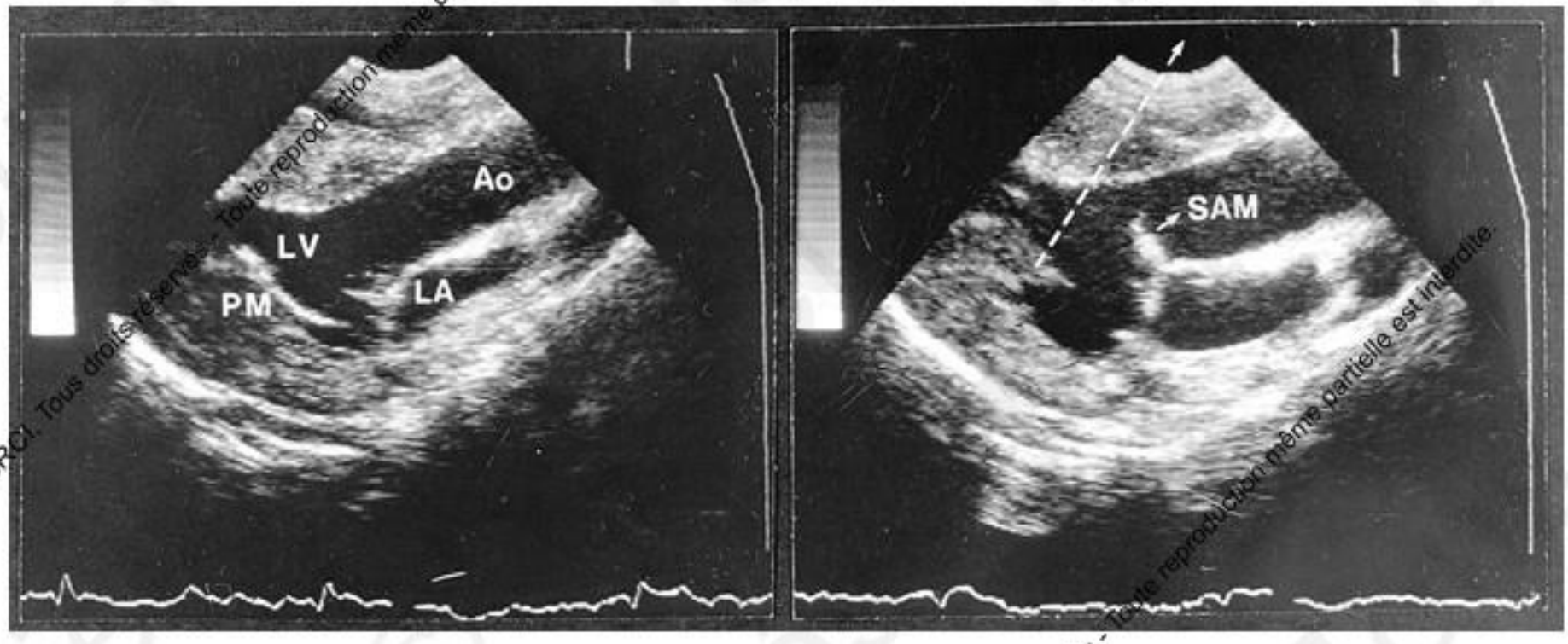
Diagrams of chordal geometry illustrating effects of papillary muscle malposition on distribution of tension to the mitral leaflets.





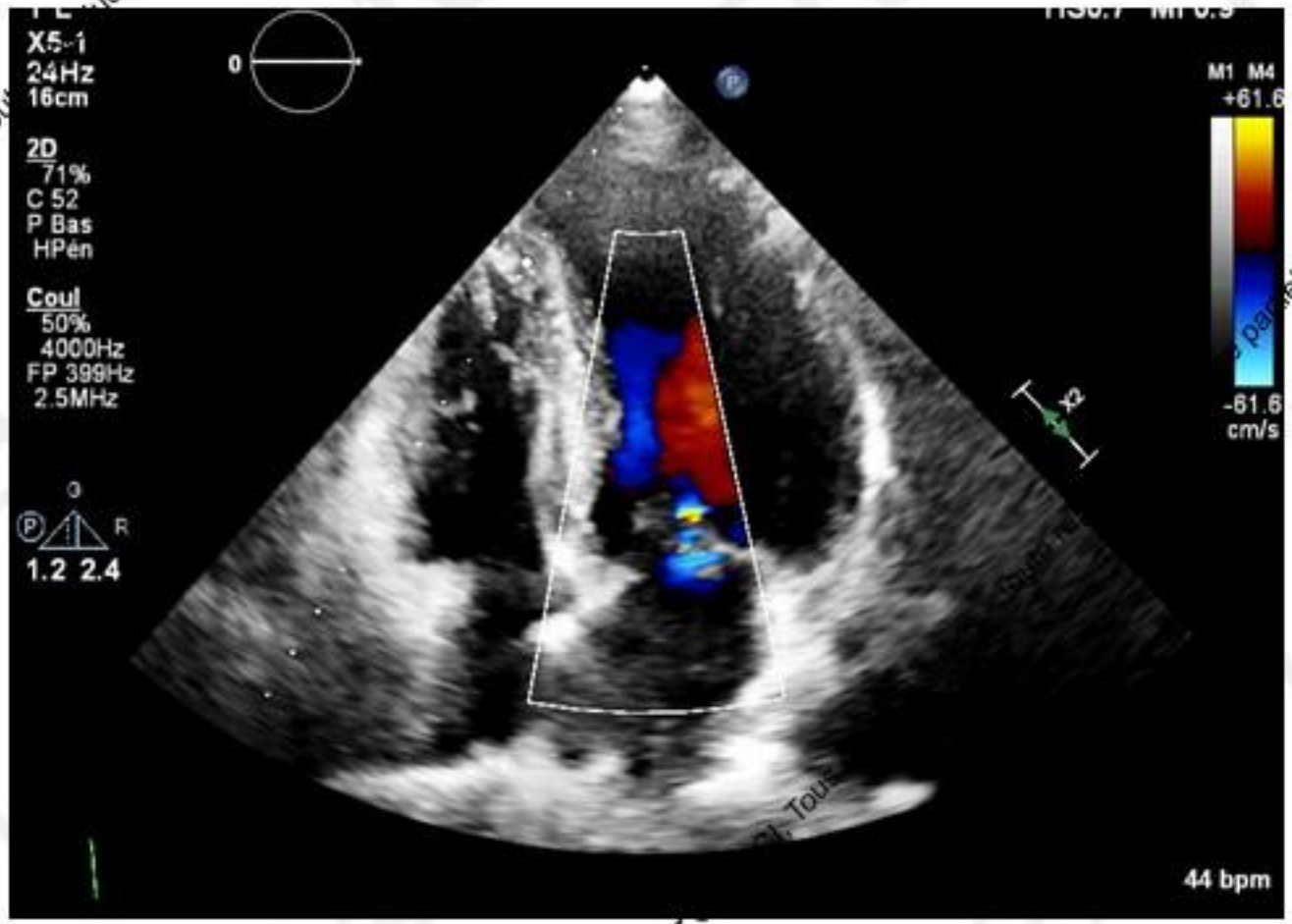
Robert A. Levine et al. Circulation. 1995;91:1189-1195

Echocardiographs of systolic anterior motion (SAM, right) created in vivo by anterior displacement of the papillary muscles (PM).

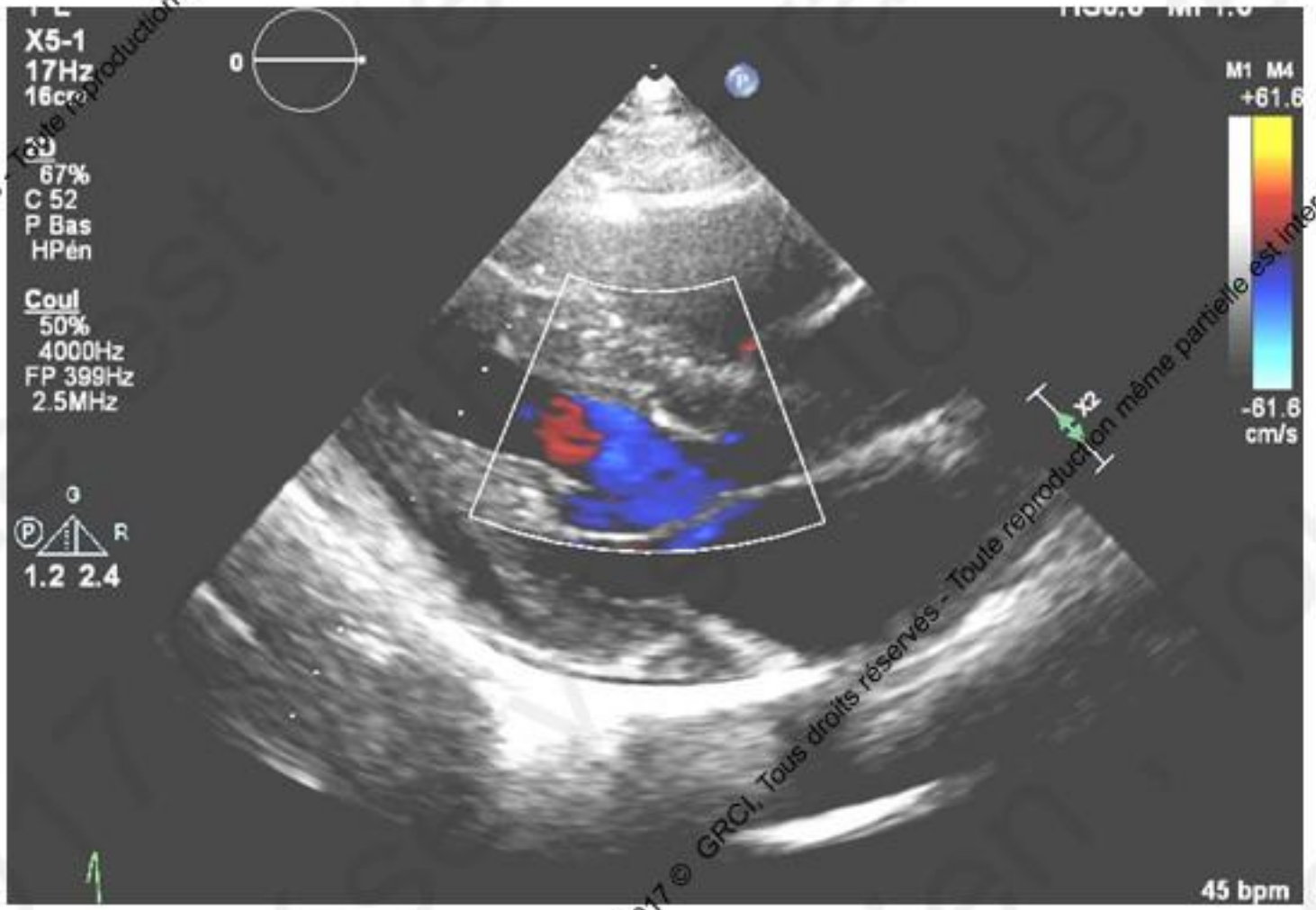


Robert A. Levine et al. Circulation. 1995;91:1189-1195

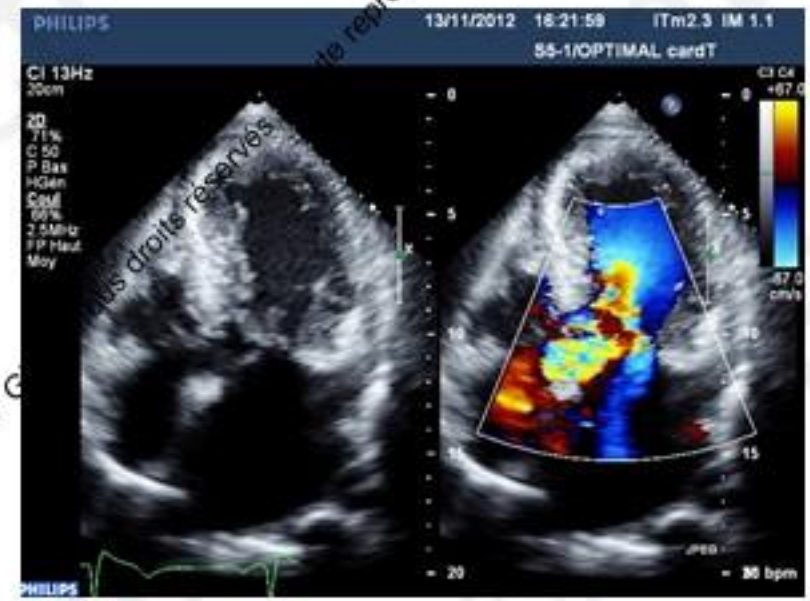
CMH. Obstruction sous-aortique et IM

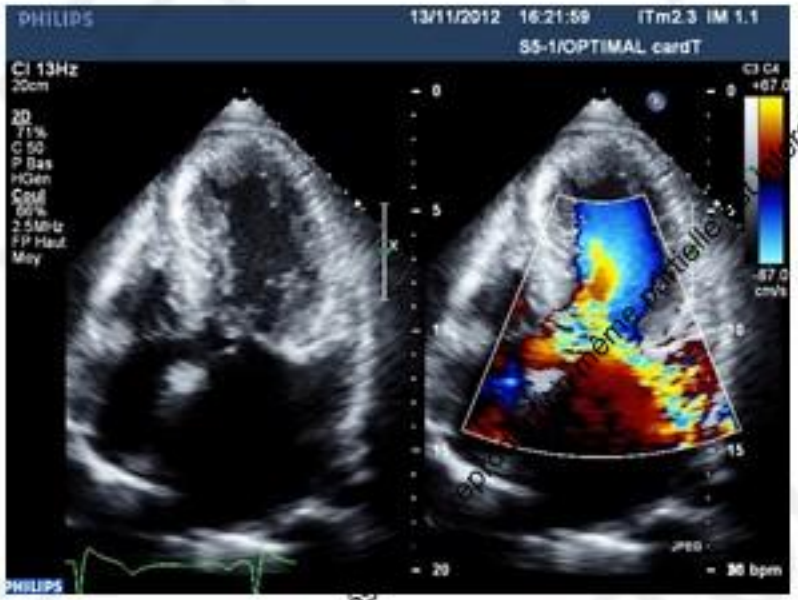


CMH: Obstruction sous-aortique et IM

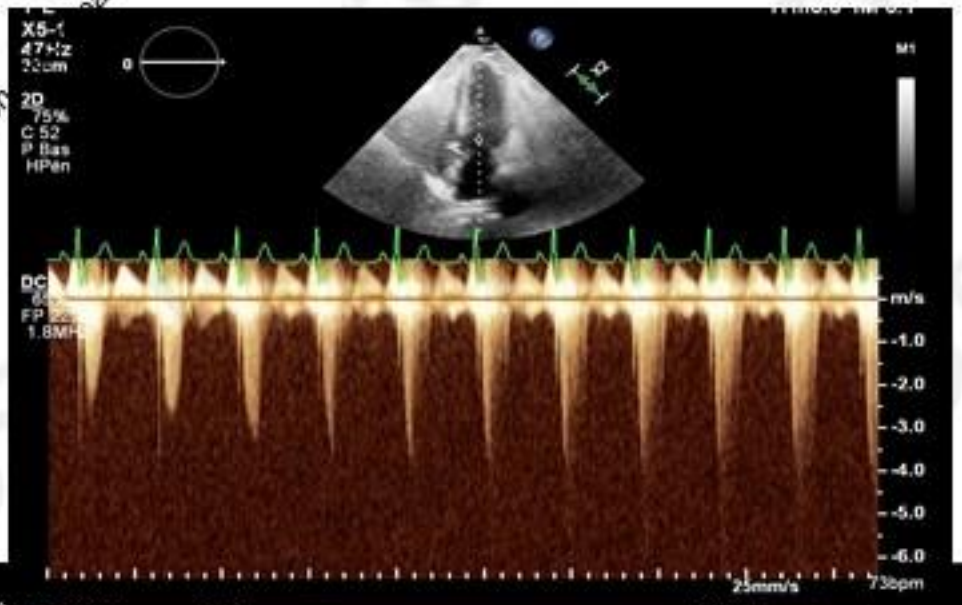


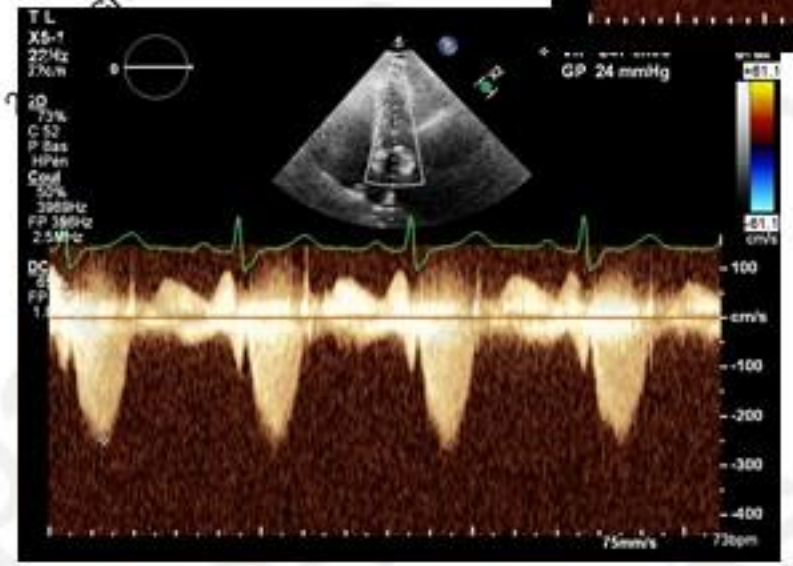
CMH. Obstruction sous-aortique et IM

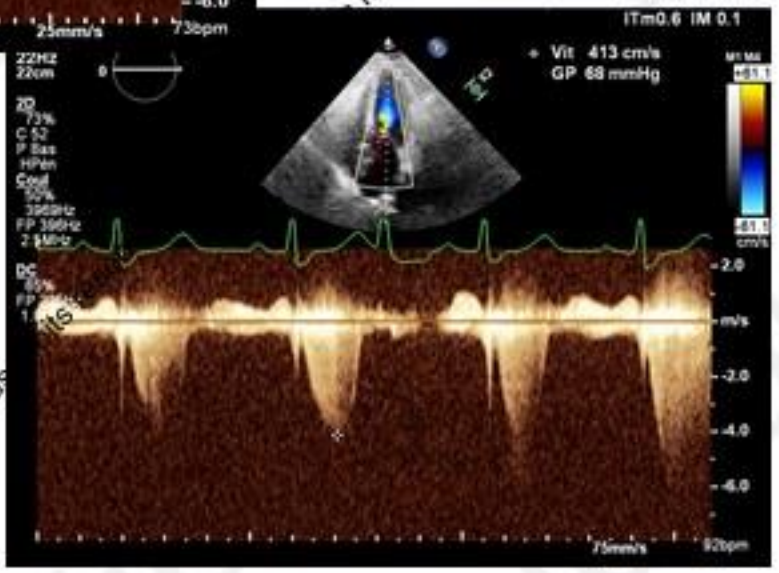




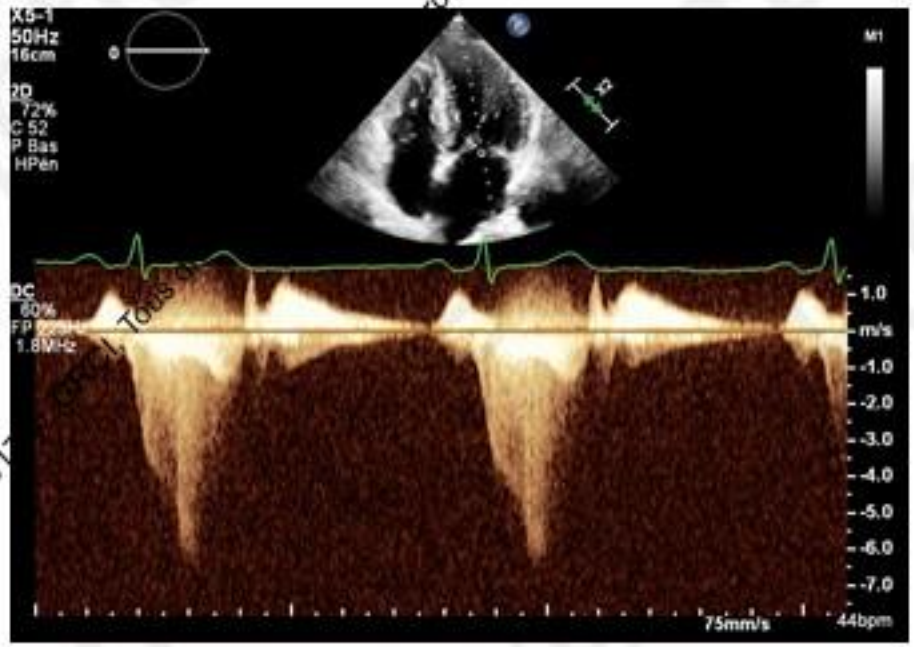
Manoeuvre de Valsalva sur CMH obstructive.

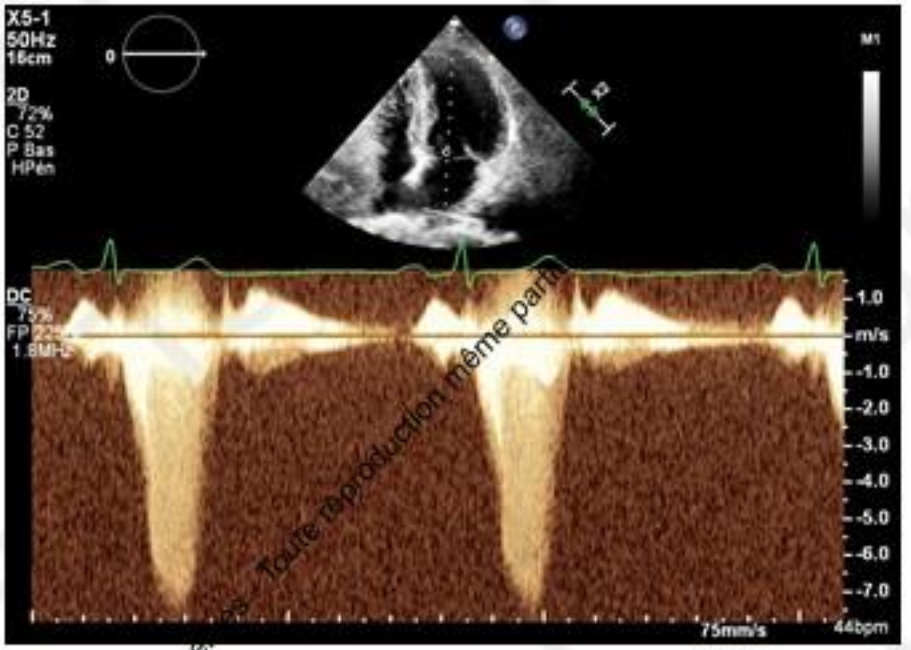




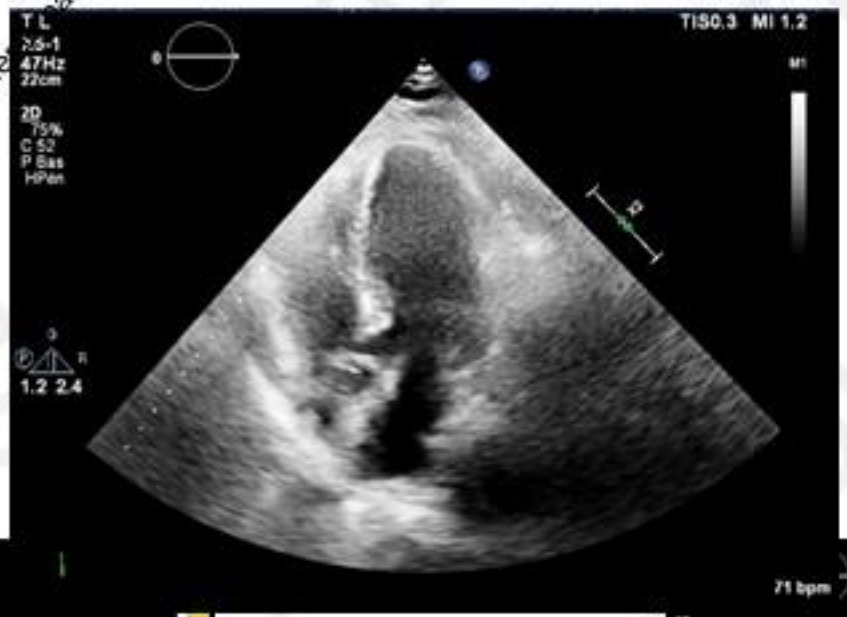


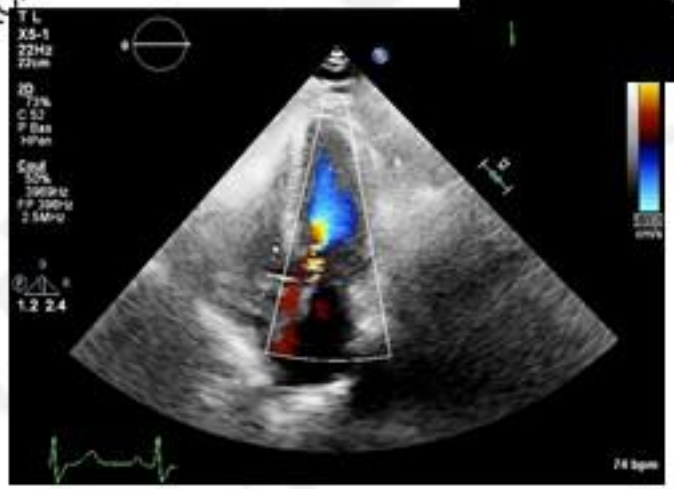
Manoeuvre de Valsalva sur CMH obstructive.

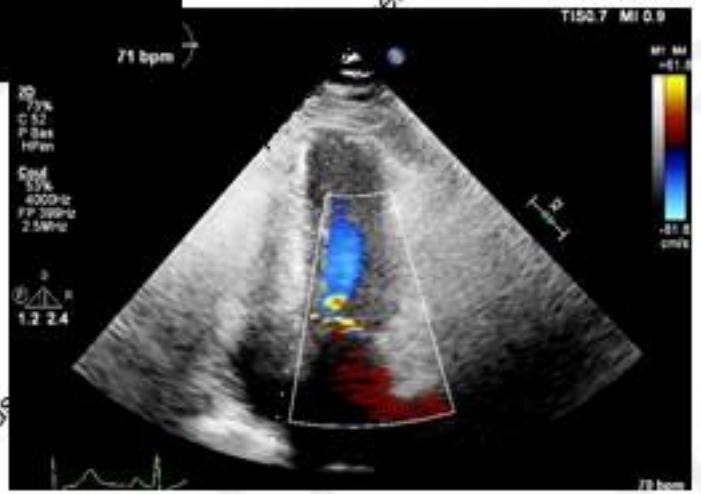




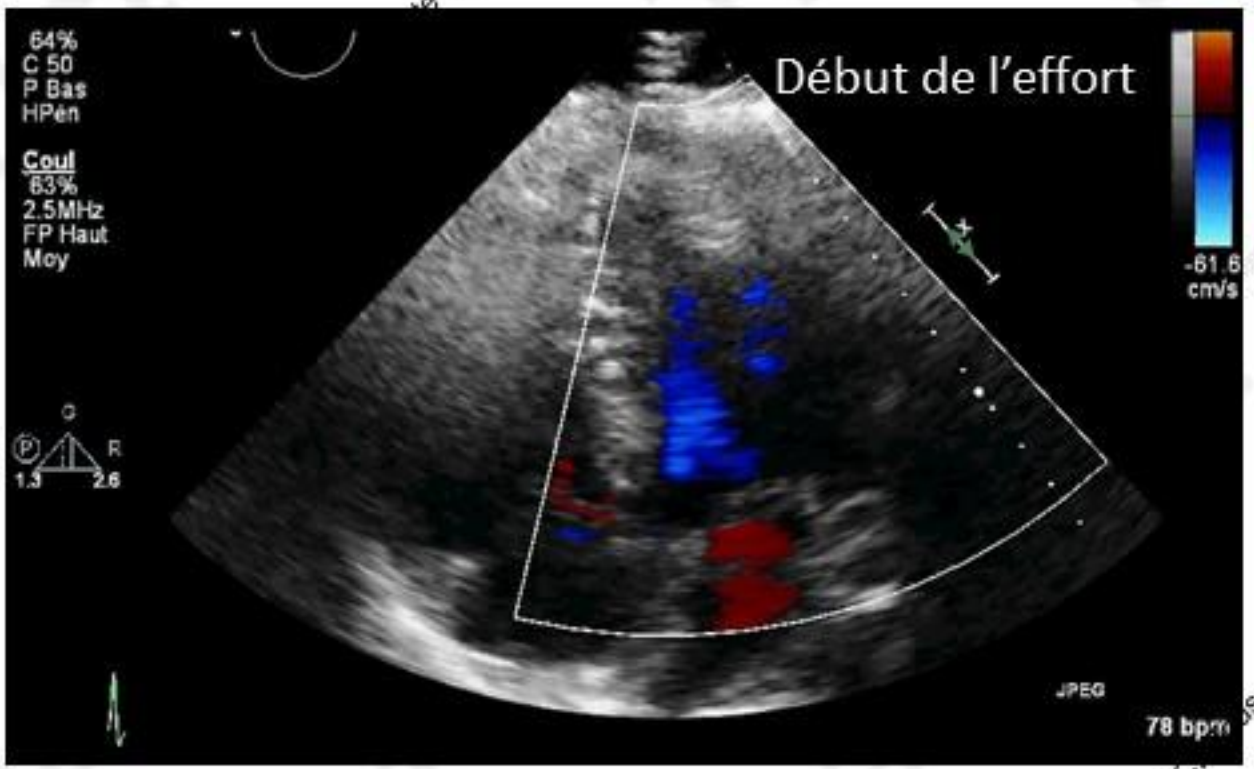
Manoeuvre de Valsalva sur CMH obstructive.

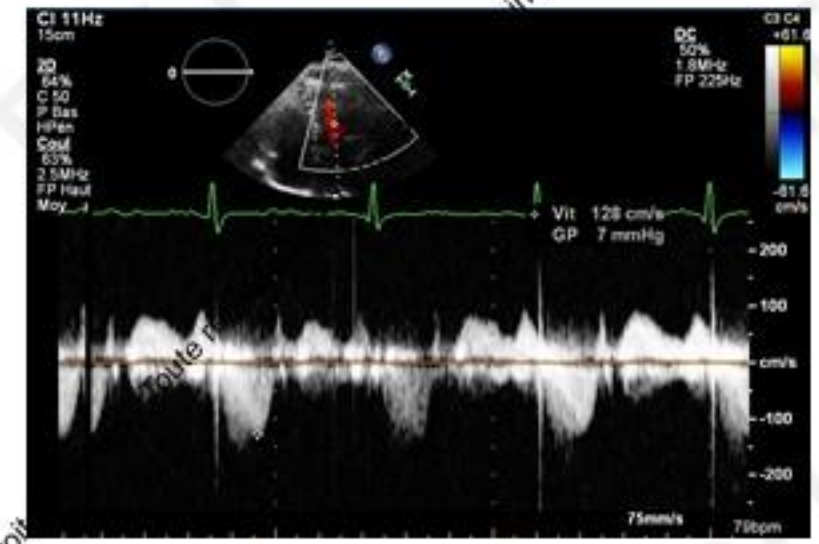




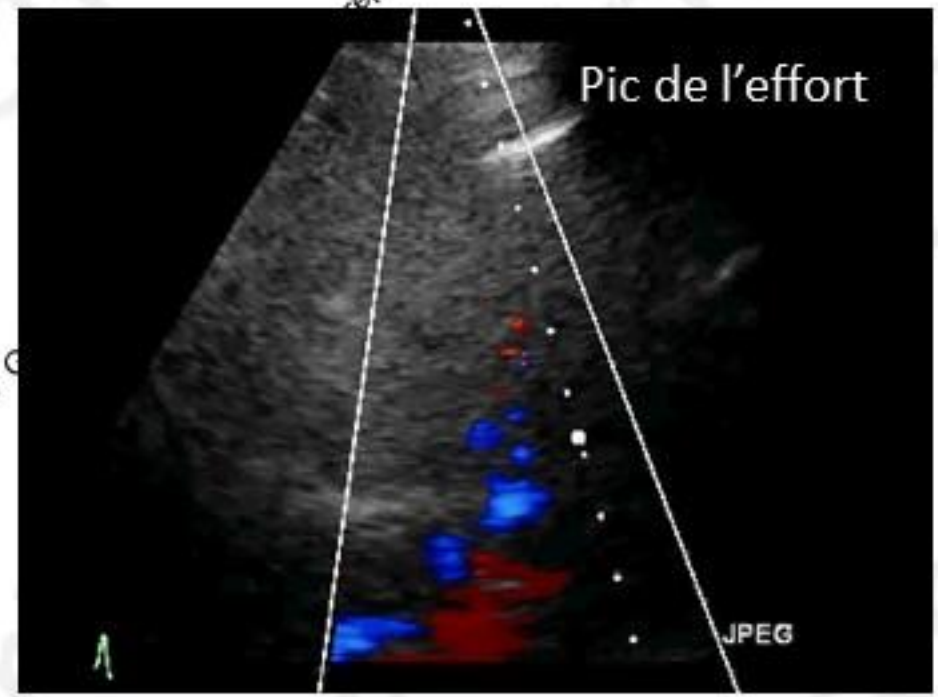


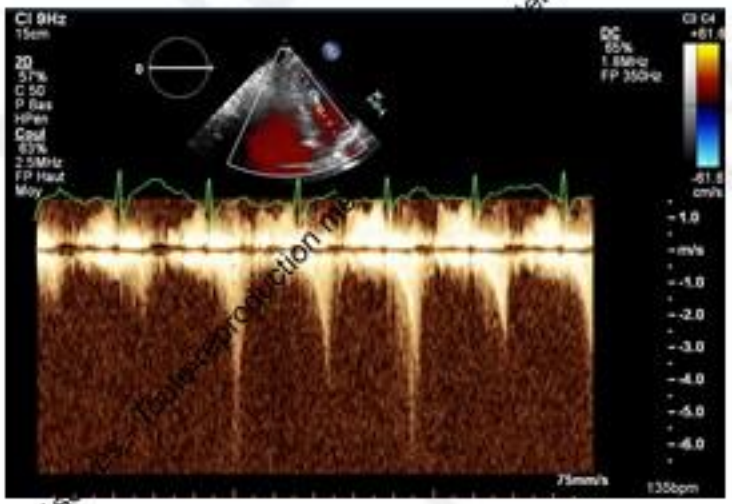
Echographie d'effort.



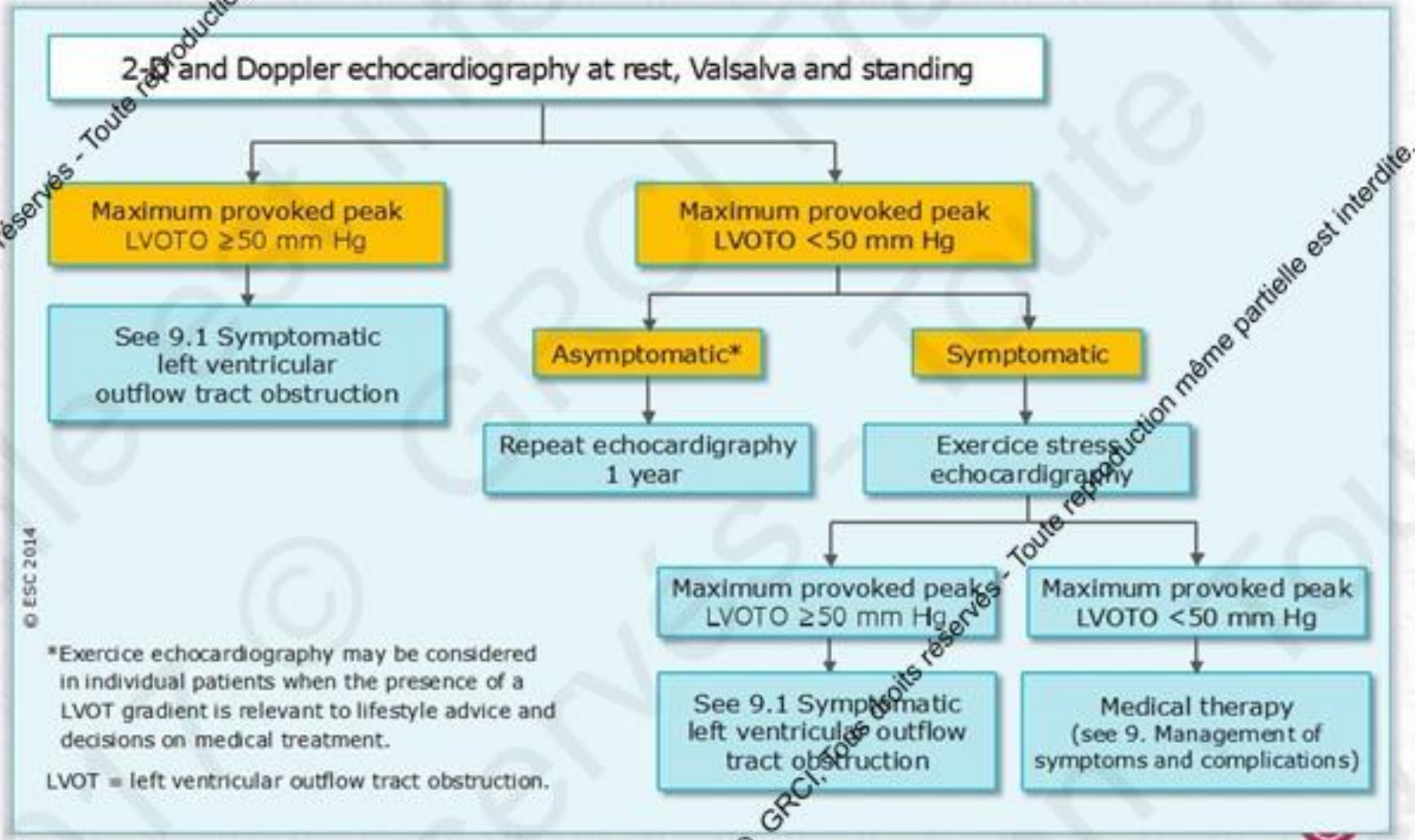


Echographie d'effort.





Protocol for the assessment and treatment of left ventricular outflow tract obstruction

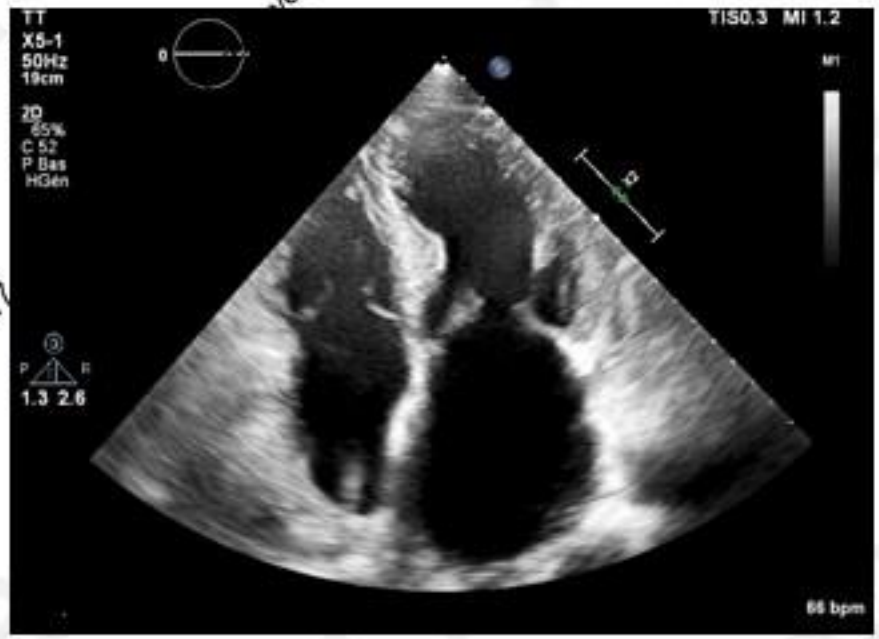


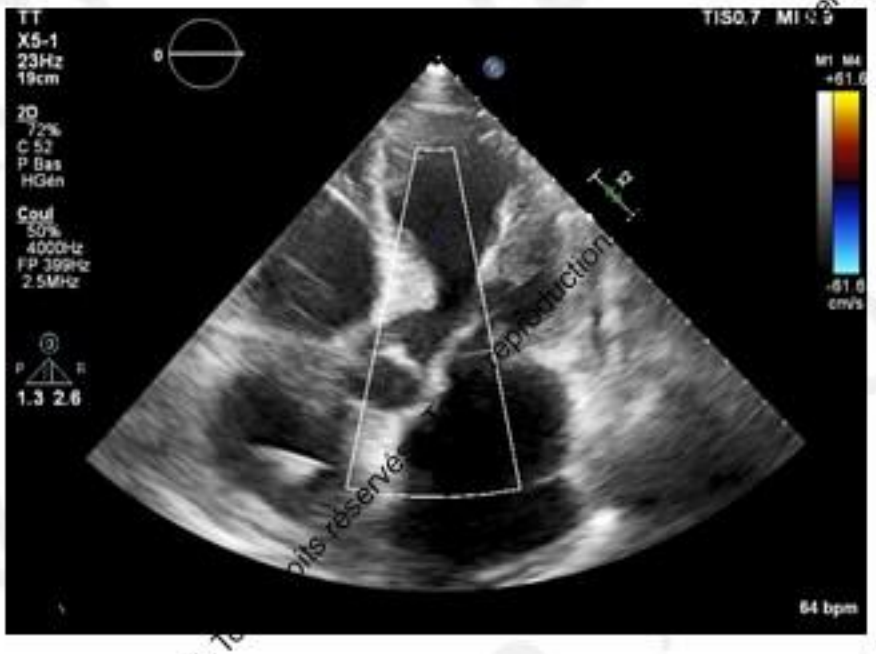
www.escardio.org/guidelines European Heart Journal (2014);35:2733-2779 - doi:10.1093/eurheartj/ehu284

Stimulation cardiaque et CMO

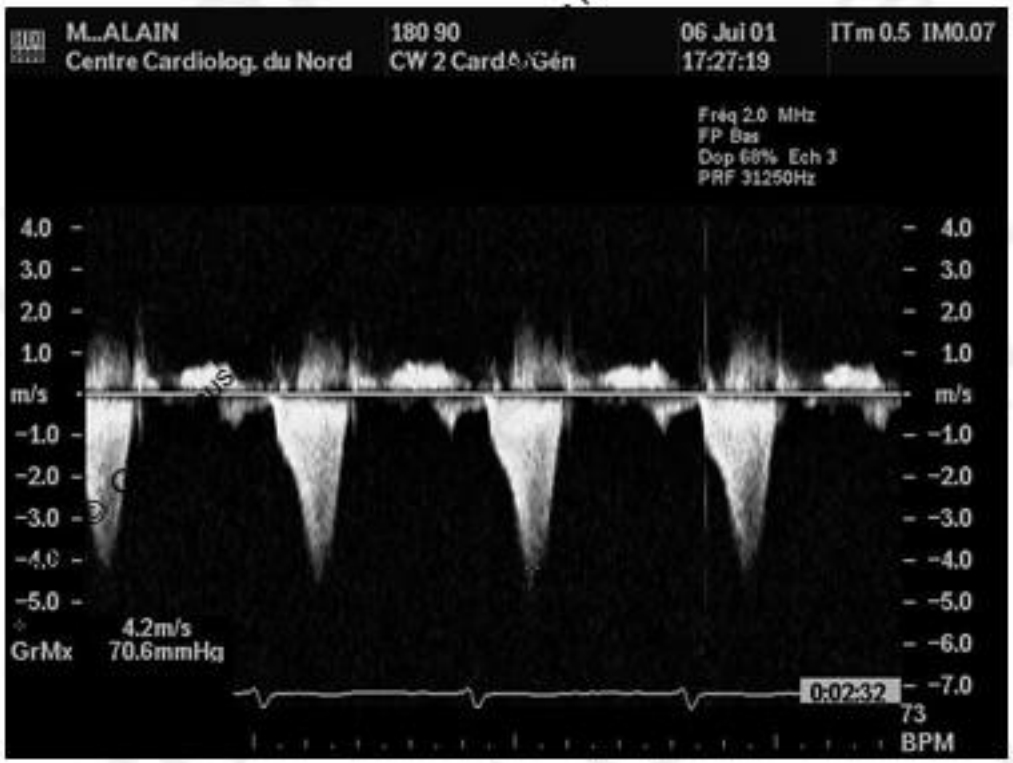
- Stimulation à l'apex du ventricule droit
- Inverser la séquence d'activation/contraction du ventricule gauche par primo-stimulation de l'apex, et entraîner un mouvement paradoxal du septum
- Fonction du DAV (= court pour anticiper la contraction spontanée du VD et la capturer). Mais ne pas le programmer trop court.
- Intérêt d'y associer des beta-bloquants pour allonger un peu le DAV
- Résultats contradictoires des études >> Classe IIb

CMH et stimulation VD

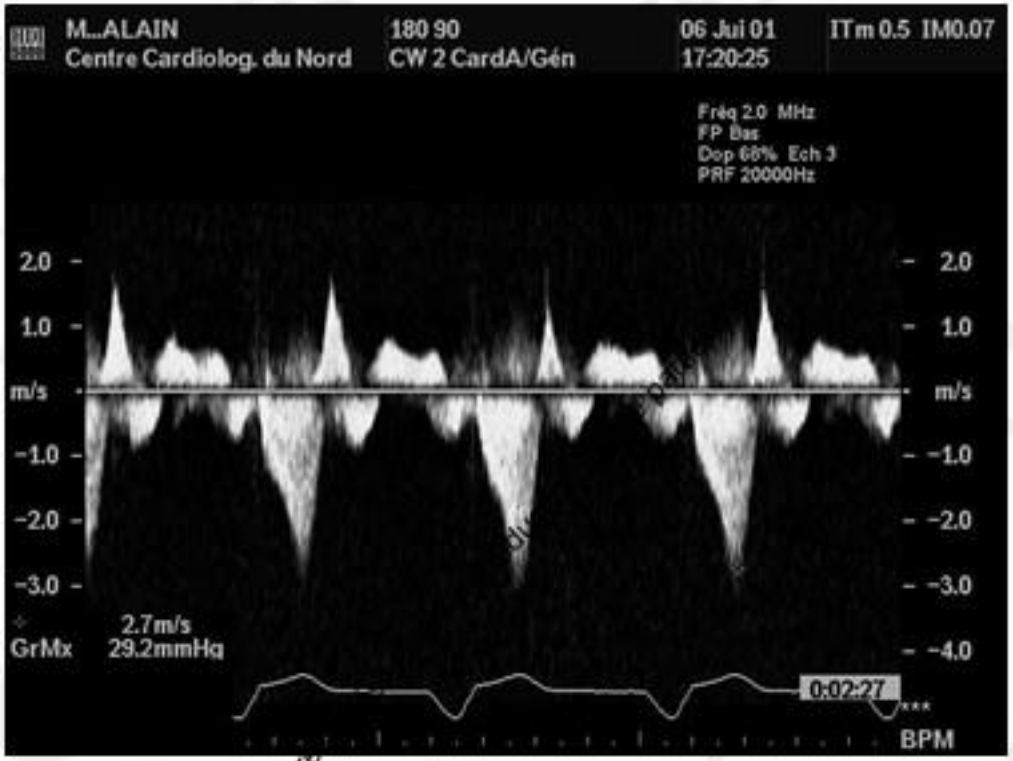




Gradient intraventriculaire gauche chez un patient ayant une CMH obstructive avant et après implantation d'un stimulateur cardiaque



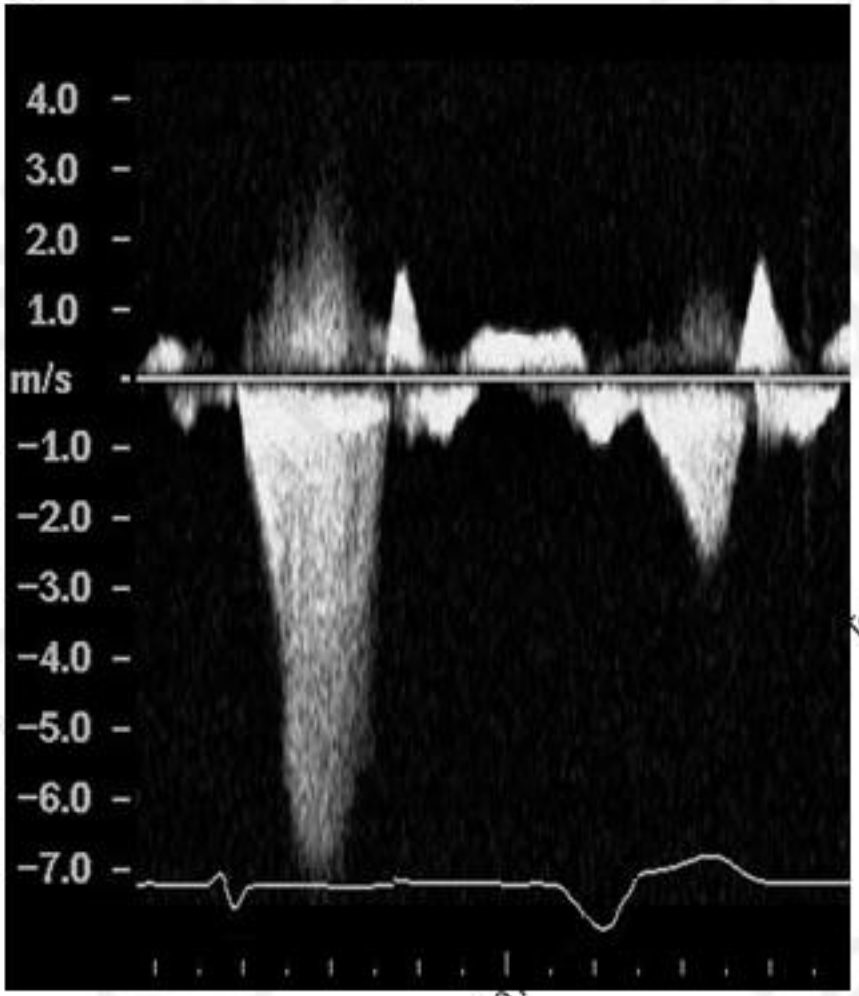
Gradient maximal de 70,6 mmHg



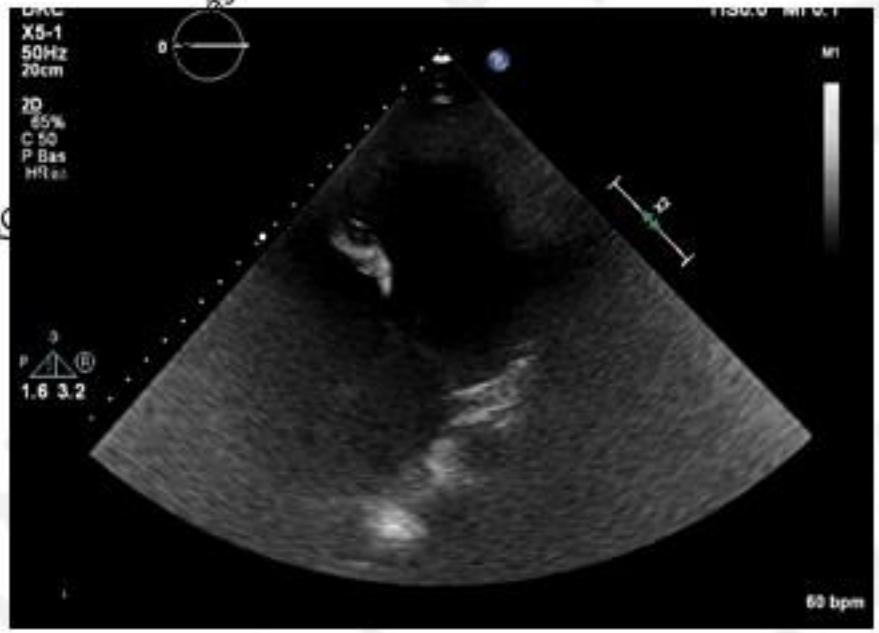
Gradient maximal de 29,2 mmHg

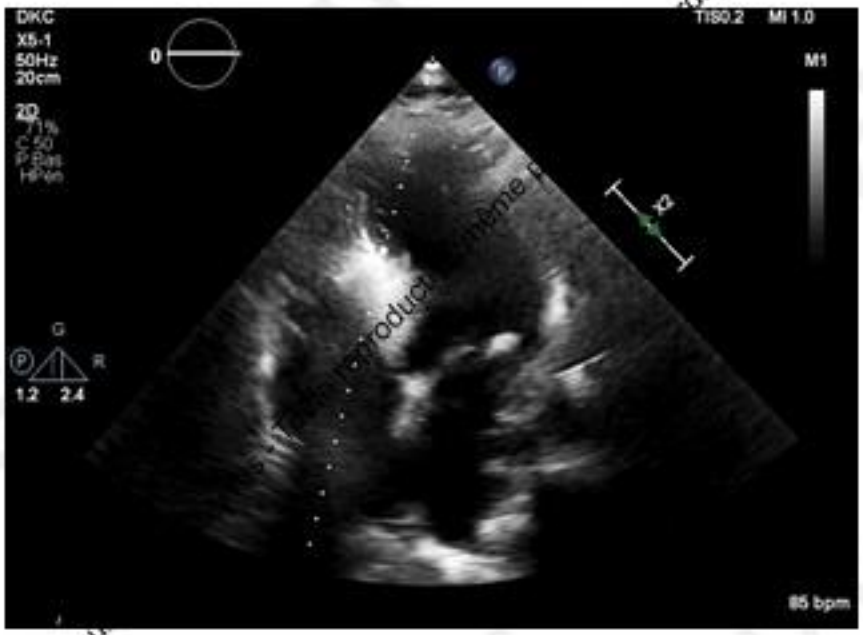
(échelle des vitesses différente entre les deux figures).

Flux d'insuffisance mitrale en rythme sinusal (premier complexe QRS) puis en stimulation à l'apex du VD (deuxième complexe QRS). Cette stimulation diminue le mouvement systolique antérieur de la grande valve mitrale (SAM) réduisant ainsi la fuite mitrale délétère.

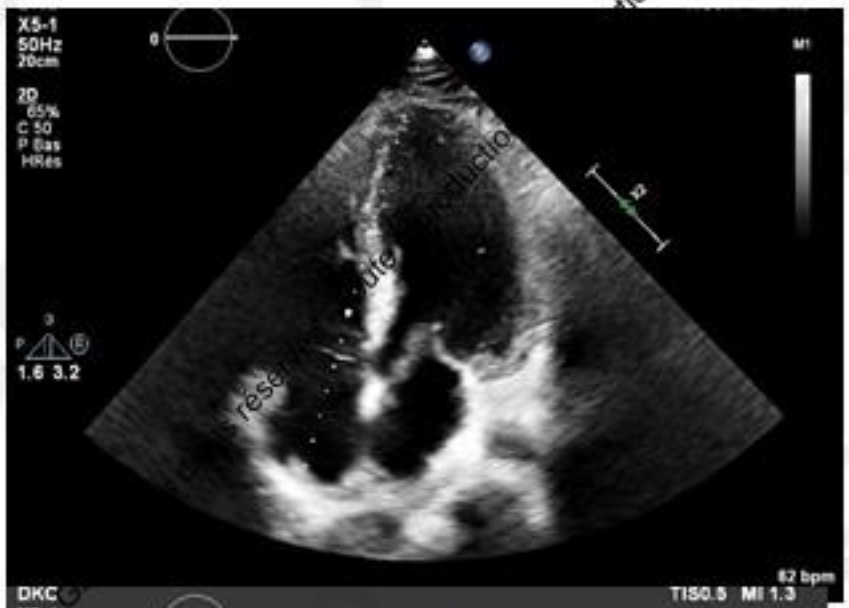


CMH. Alcoolisation septale

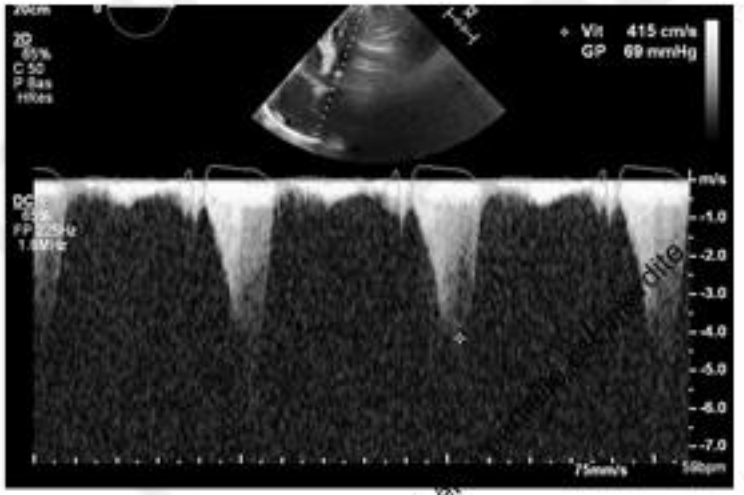


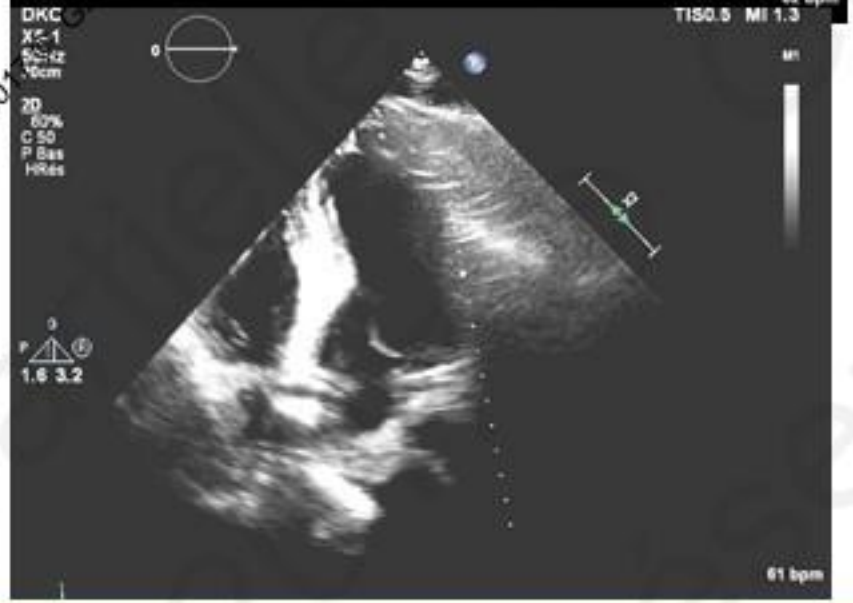


CMH. Alcoolisation septale

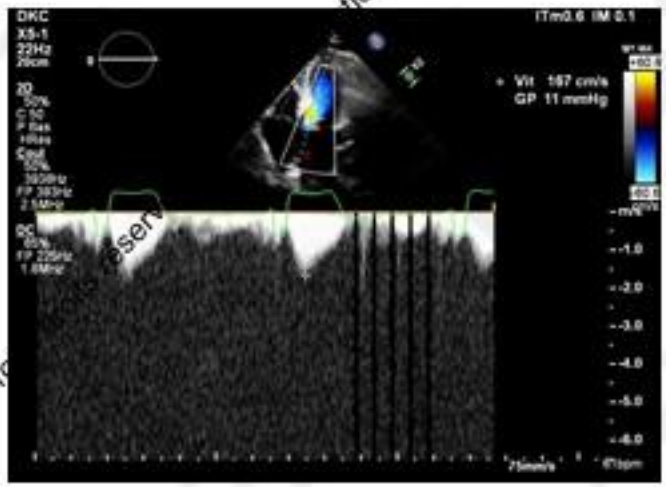


Avant

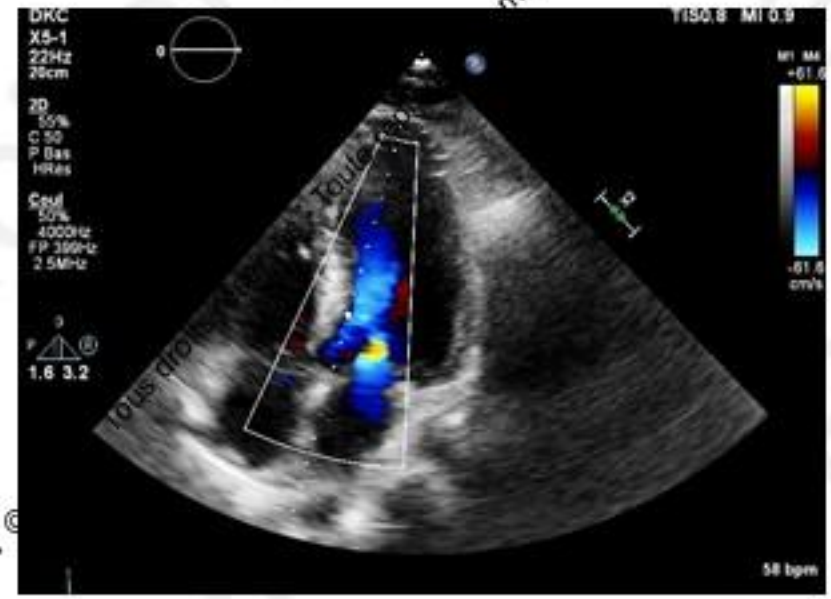


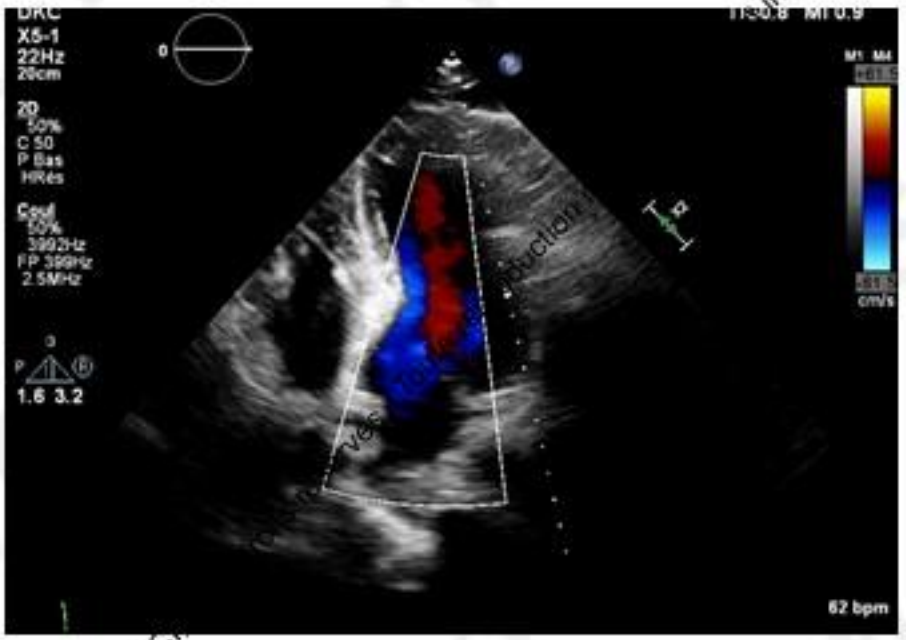


Après

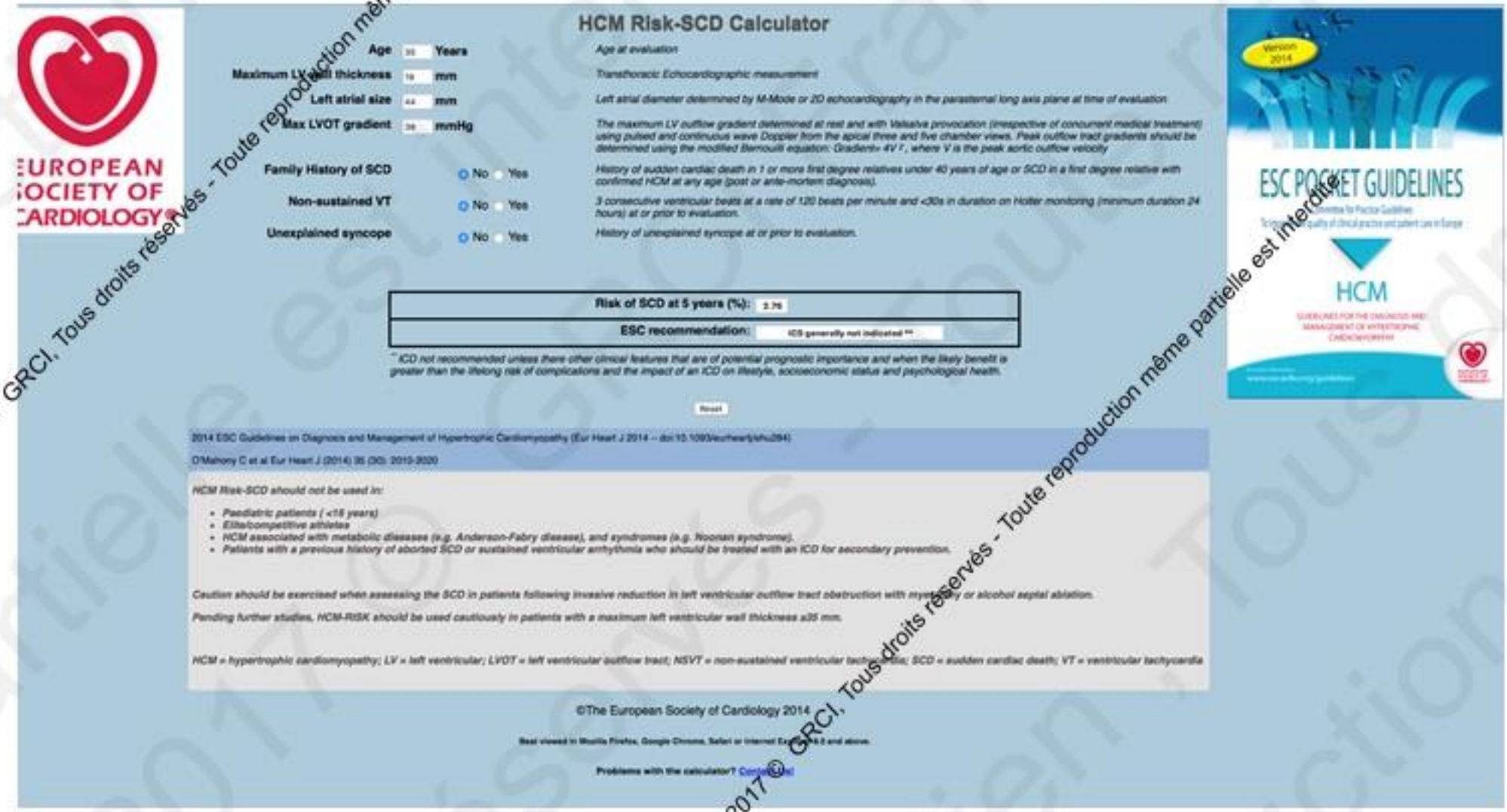


Evolution de l'IM après alcoolisation





Calculateur du risque de mort subite



HCM Risk-SCD Calculator

Age	35	Years
Maximum LV wall thickness	19	mm
Left atrial size	44	mm
Max LVOT gradient	38	mmHg

Family History of SCD	No / Yes
Non-sustained VT	No / Yes
Unexplained syncope	No / Yes

Age at evaluation

Transthoracic Echocardiographic measurement

Left atrial diameter determined by M-Mode or 2D echocardiography in the parasternal long axis plane at time of evaluation

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 Bernoulli equation: Gradient= 4V², where V is the peak aortic outflow velocity

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).

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.

History of unexplained syncope at or prior to evaluation.

Risk of SCD at 5 years (%):	2.76
ESC recommendation:	ICD generally not indicated **

** *ICD not recommended unless there other clinical features that are of potential prognostic importance and when the likely benefit is greater than the lifelong risk of complications and the impact of an ICD on lifestyle, socioeconomic status and psychological health.*

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 (<18 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.

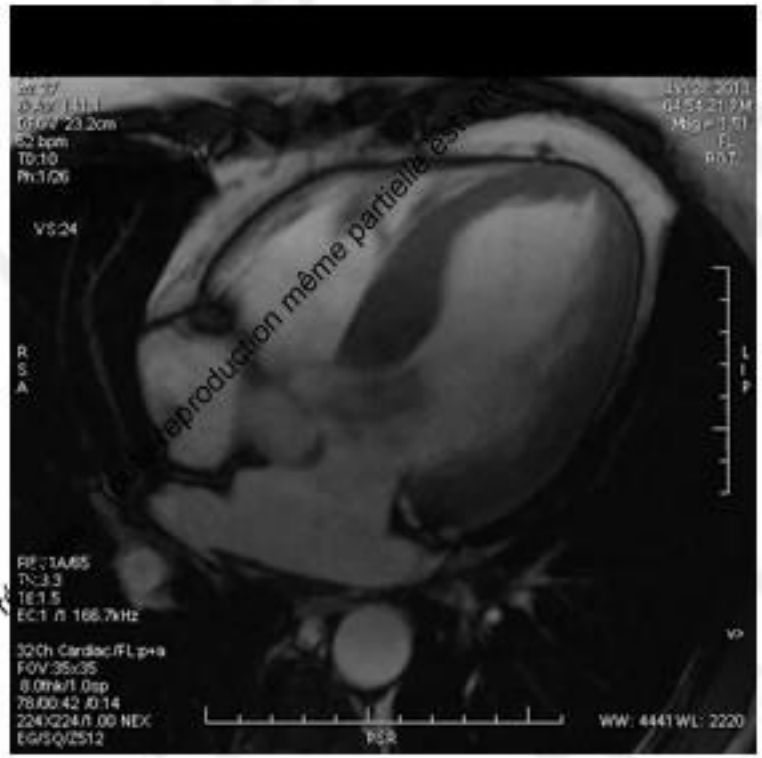
HCM = hypertrophic cardiomyopathy; LV = left ventricular; LVOT = left ventricular outflow tract; NSVT = non-sustained ventricular tachycardia; SCD = sudden cardiac death; VT = ventricular tachycardia

Best viewed in Mozilla Firefox, Google Chrome, Safari or Internet Explorer 8.0 and above.

Problems with the calculator? Contact Us!

IRM et CMH

- Supériorité/écho pour
  - les CMH apicales, les anévrysmes et les thrombi
  - Détection de la fibrose (réhaussement tardif)
- Moins performante pour l'évaluation du gradient et à l'effort



Conclusion

- Intérêt de l'échocardiographie pour
  - Détecter la CMH
  - Orienter le diagnostic étiologique
  - Aider à déterminer des critères de sévérité et le pronostic
  - Contrôler les diverses stratégies thérapeutiques
  - Surveiller les patients

Table 7 **Major clinical features associated with an increased risk of sudden cardiac death in adults**

Risk Factor	Comment
Age	• The effect of age on SCD has been examined in a number of studies[73,82,99,208,244,372–374] and two have shown a significant association, with an increased risk of SCD in younger patients.[73,99] • Some risk factors appear to be more important in younger patients, most notably, NSVT,[69] severe LVH[375] and unexplained syncope.[99]
Non-sustained ventricular tachycardia	• NSVT (defined as ≥3 consecutive ventricular beats at ≥120 BPM lasting <30 seconds) occurs in 20–30% of patients during ambulatory ECG monitoring and is an independent predictor of SCD.[69,73,83,246,248,374] • There is no evidence that the frequency, duration or rate of NSVT influences the risk of SCD.[69,374]
Maximum left ventricular wall thickness	• The severity and extent of LVH measured by TTE are associated with the risk of SCD.[69,120,121,373] • Several studies have shown the greatest risk of SCD in patients with a maximum wall thickness of ≥30 mm but there are few data in patients with extreme hypertrophy (≥35 mm).[69,73,120,247,248,371,377,378]
Family history of sudden cardiac death at a young age	• While definitions vary,[73,120,372,377] a family history of SCD is usually considered clinically significant when one or more first-degree relatives have died suddenly aged <40 years with or without a diagnosis of HCM, or when SCD has occurred in a first-degree relative at any age with an established diagnosis of HCM.
Syncope	• Syncope is common in patients with HCM but is challenging to assess as it has multiple causes.[379] • Non-neurocardiogenic syncope for which there is no explanation after investigation is associated with increased risk of SCD.[73,83,99,244,246–248] • Episodes within 6 months of evaluation may be more predictive of SCD.[99]
Left atrial diameter	• Two studies have reported a positive association between LA size and SCD.[73,99] There are no data on the association between SCD and LA area and volume. Measurement of LA size is also important for assessing the risk of AF (see section 9.4).
Left ventricular outflow tract obstruction	• A number of studies have reported a significant association with LVOTO and SCD.[73,82,83,246,372,380] Several unanswered questions remain, including the prognostic importance of provocable LVOTO and the impact of treatment (medical or invasive) on SCD.
Exercise blood pressure response	• Approximately one third of adult patients with HCM have an abnormal systolic blood pressure response to exercise characterised by progressive hypotension or a failure to augment the systolic blood pressure that is caused by an inappropriate drop in systemic vascular resistance and a low cardiac output reserve.[241,381] • Various definitions for abnormal blood pressure response in patients with HCM have been reported[69,83,246,377]; for the purposes of this guideline an abnormal blood pressure response is defined as a failure to increase systolic pressure by at least 20 mm Hg from rest to peak exercise or a fall of >20 mm Hg from peak pressure.[237] • Abnormal exercise blood pressure response is associated with a higher risk of SCD in patients aged ≤40 years,[237] but its prognostic significance in patients >40 years of age is unknown.

HCM = hypertrophic cardiomyopathy; LA = left atrium; LVH = left ventricular hypertrophy; LVOTO = left ventricular outflow tract obstruction; NSVT = non-sustained ventricular tachycardia; SCD = sudden cardiac death; TTE = transthoracic echocardiography.

Réhaussement tardif en IRM