Cardiomyopathies hypertrophiques:

Données de l'échocardiographie en faveur d'une obstruction

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Marseille - France

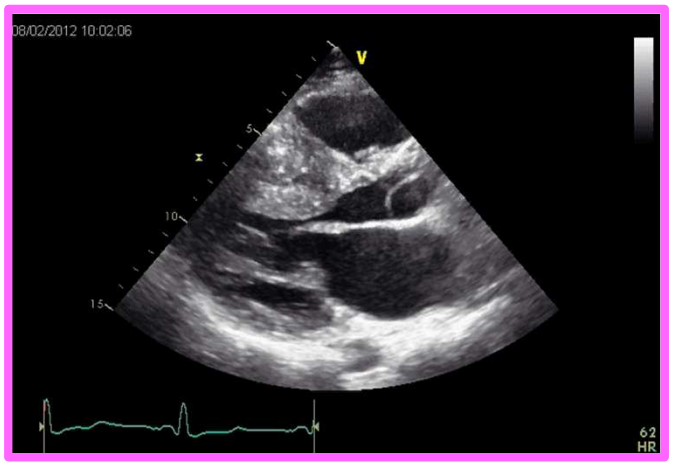
Avignon, le 24 Mars 2023

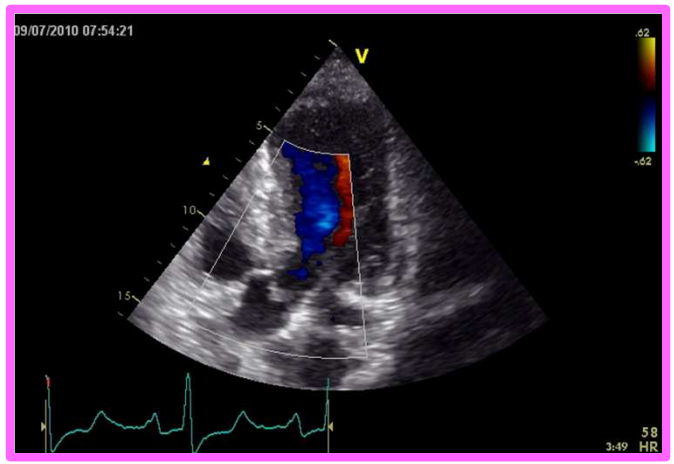
FACULTÉ DE MÉDECINE DE MARSEILLE

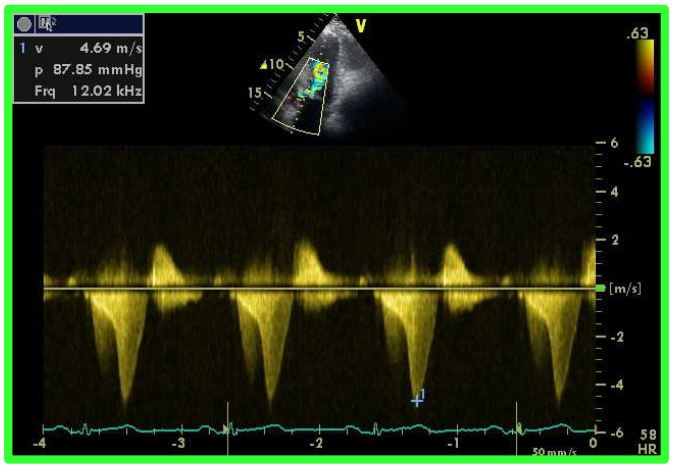
Centre de Compétence des Cardiomyopathies

Aix*Marseille université

L'obstruction dans les CMH

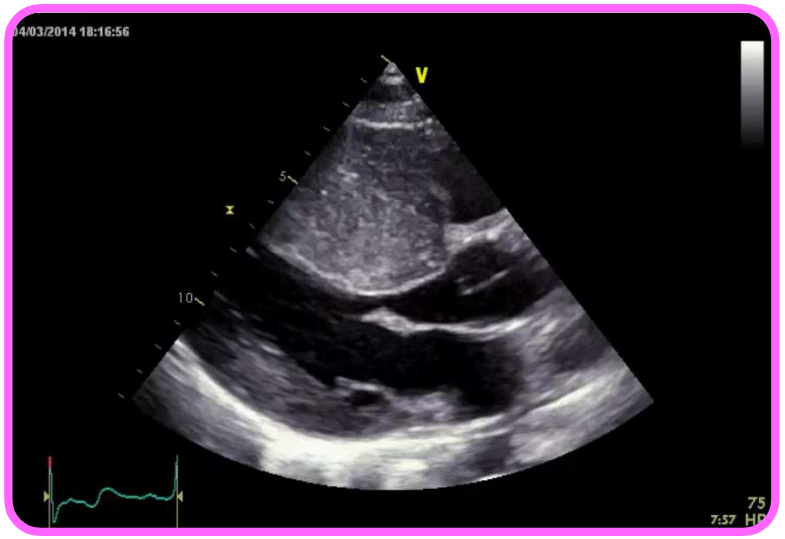


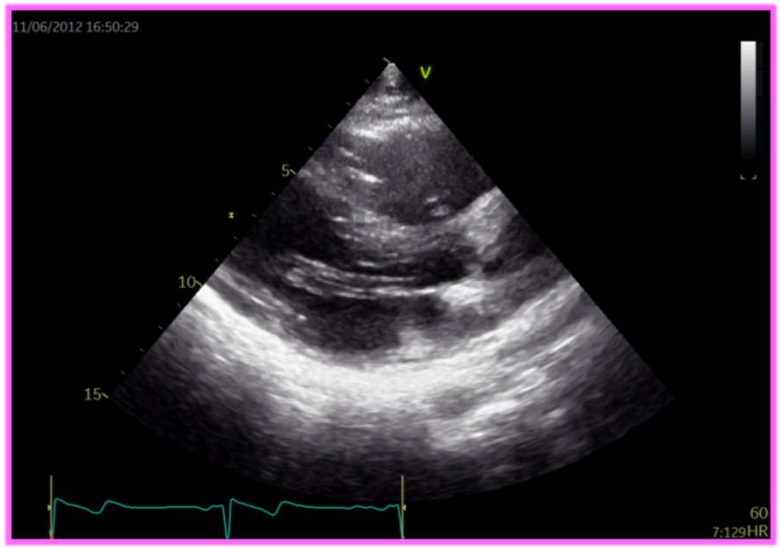




- Obstruction = gradient maximal > 30mmHg
- Indication thérapeutique = gradient maximal > 50 mmHg

L'obstruction dans les CMH





- Obstruction = gradient maximal > 30mmHg
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L'obstruction dans les CMH

1. **Mécanisme**
2. **Diagnostic**
3. **Valeur pronostique**
4. **Cible du traitement**

L'obstruction dans les CMH

1. *Mécanisme*
2. Diagnostic
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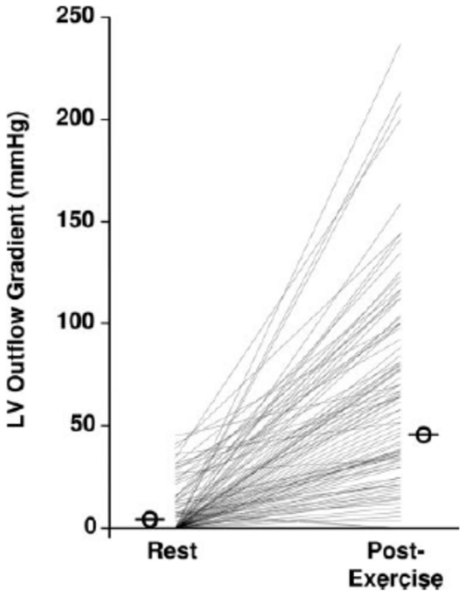
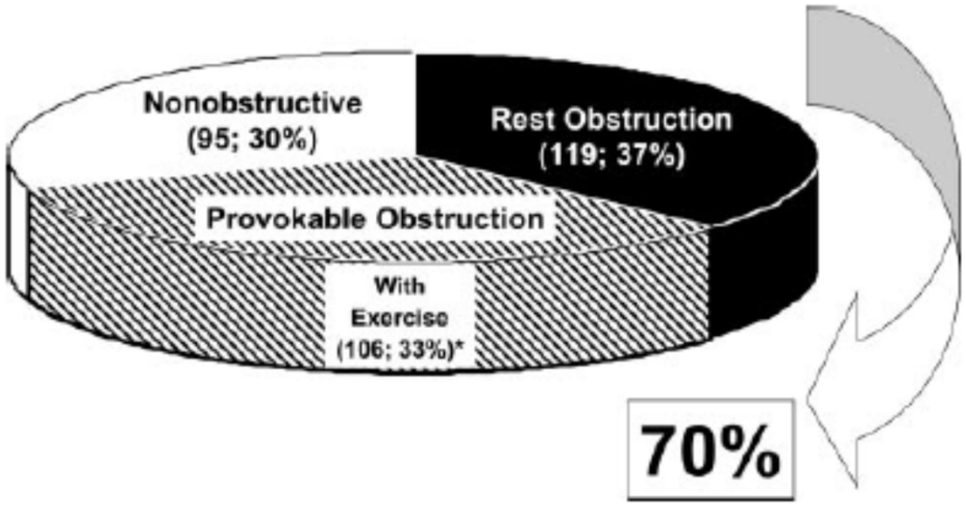
Obstruction intra-VG

Maron MS. Circulation 2006

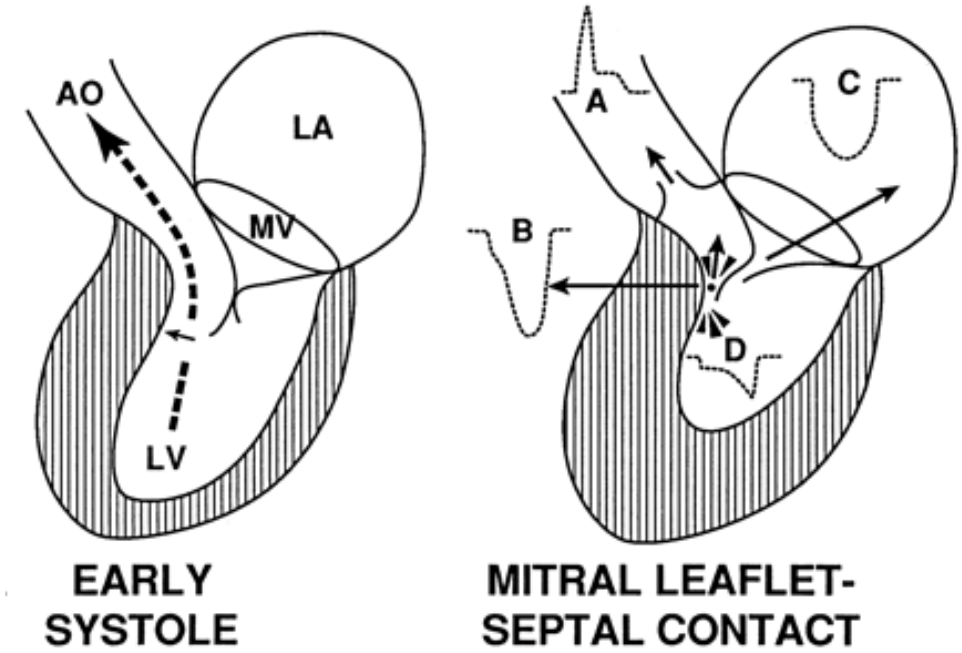
Phénomène dynamique

Incidence: 70% des pts avec une CMH ont une obstruction au repos ou à l'effort



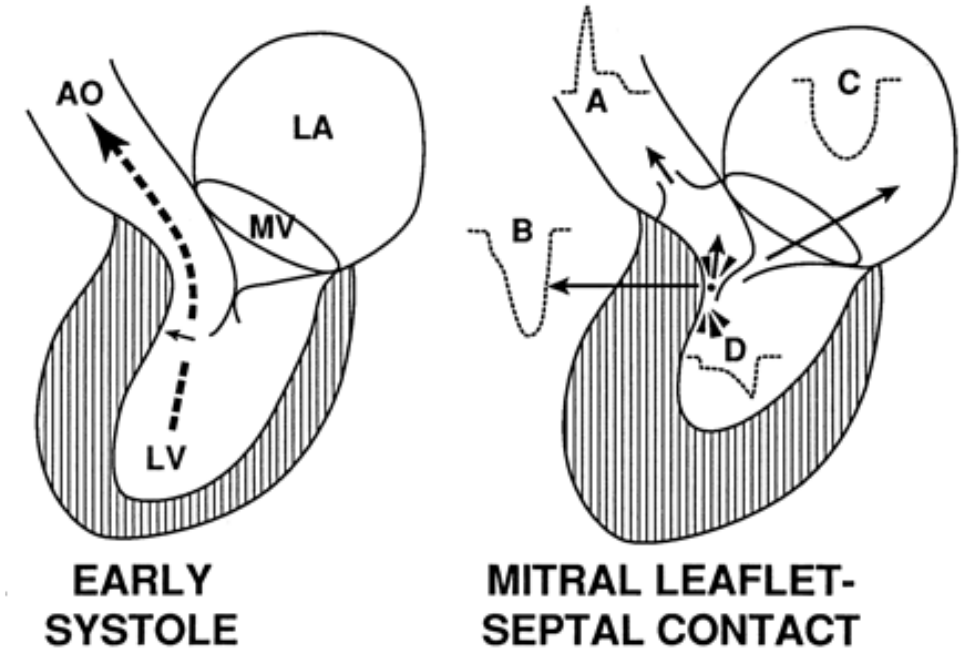


Facteurs favorisant le SAM



- hypertrophie septale sous aortique
- hypercontractilité VG
- déplacement antérieur des piliers
- élongation du feuillet antérieur

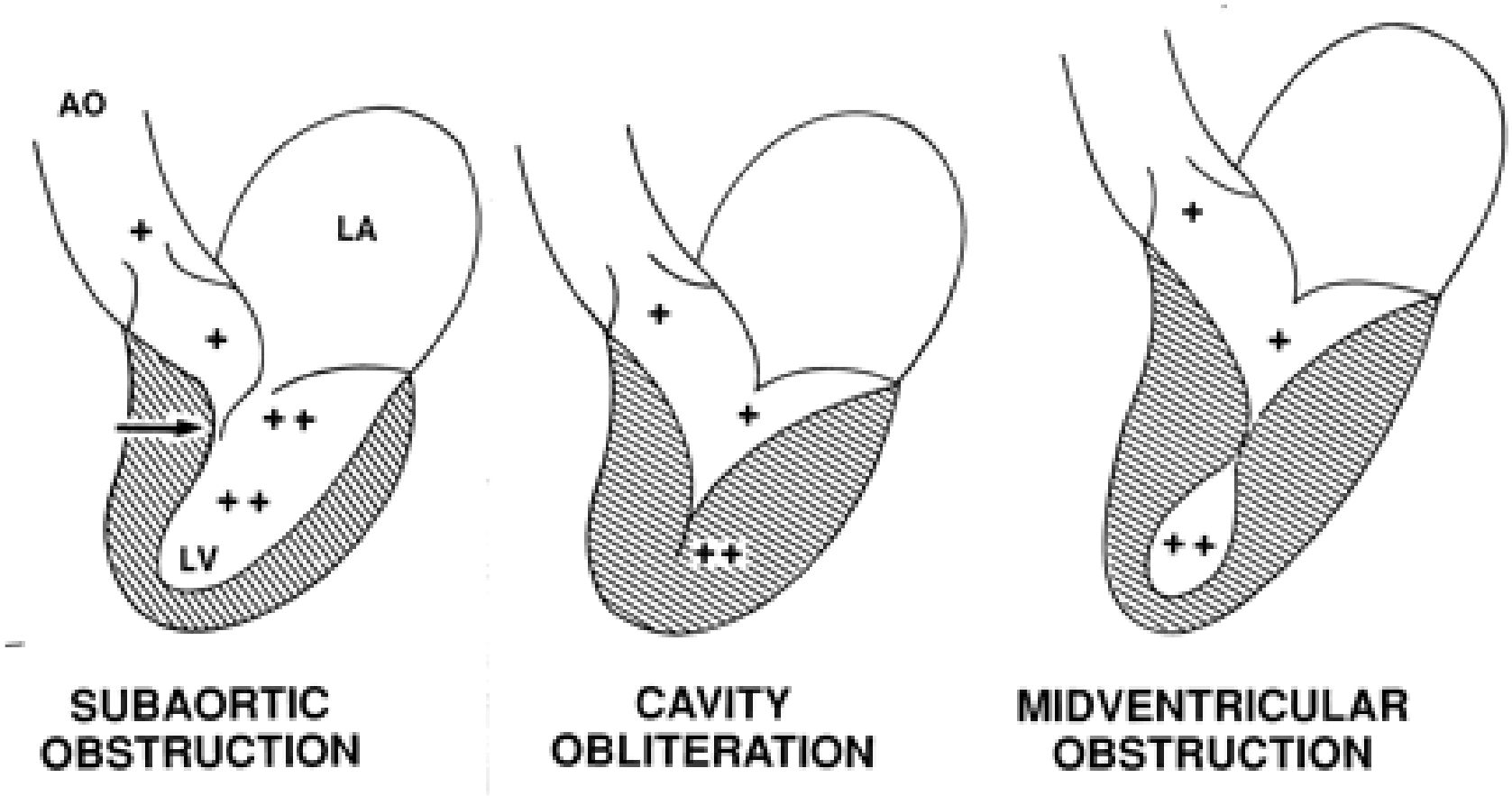
Mécanismes du SAM: effet Venturi



- Chambre de chasse de taille réduite
- L'hypercontractilité augmente la vitesse du sang au dessus de la mitrale
- Conservation de l'énergie: Si vitesse augmente (E cinétique), la pression diminue (E pression) au dessus de la valve
- Pression sous la valve

> Pression au dessus de la valve: SAM

Localisation de l'hypertrophie



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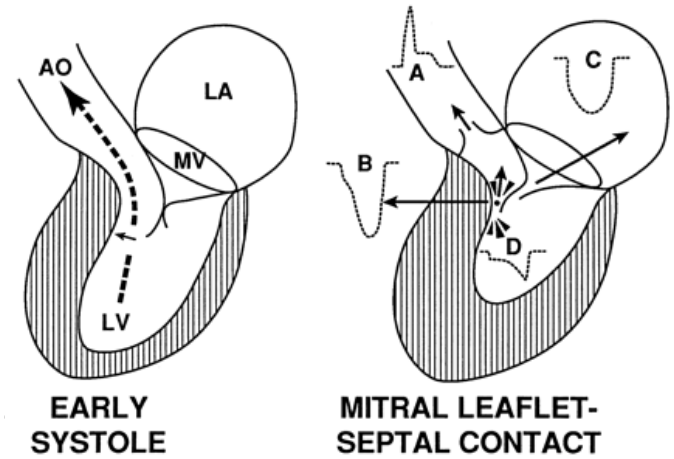
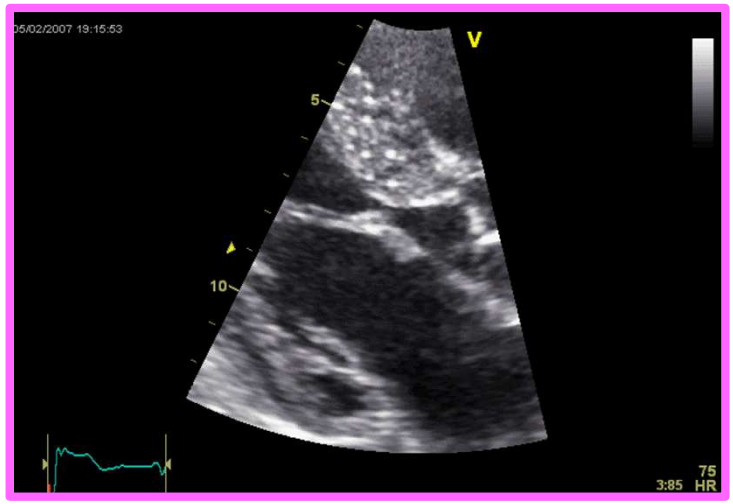
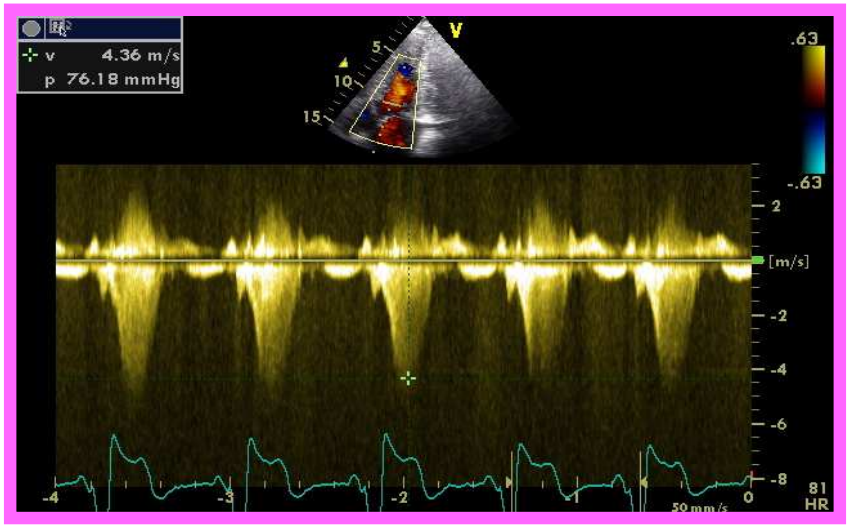
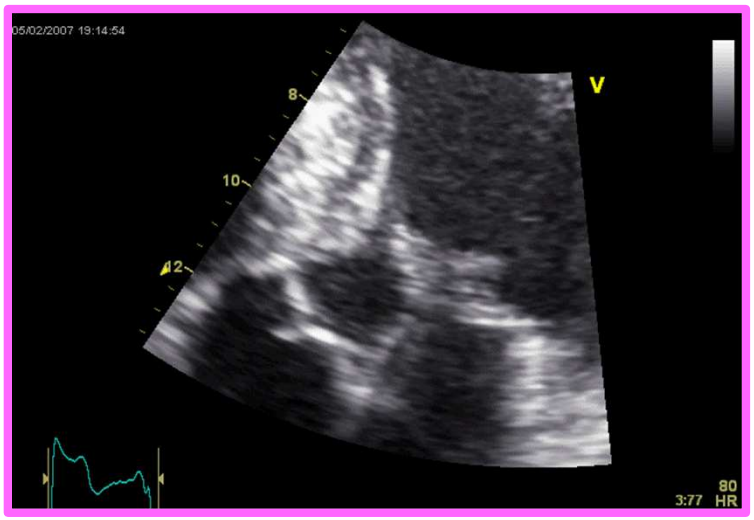
Obstruction intra-VG

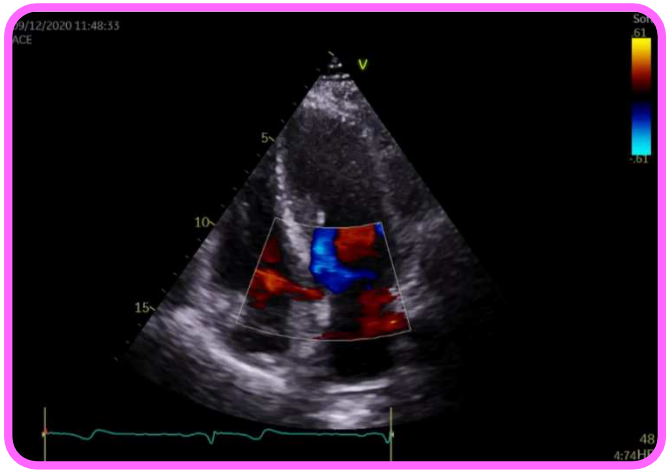


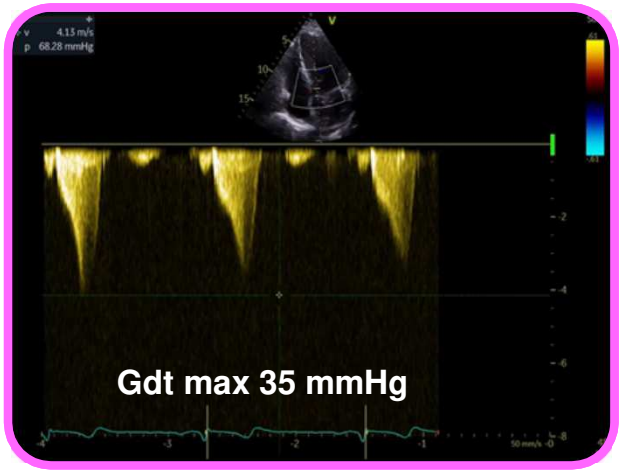


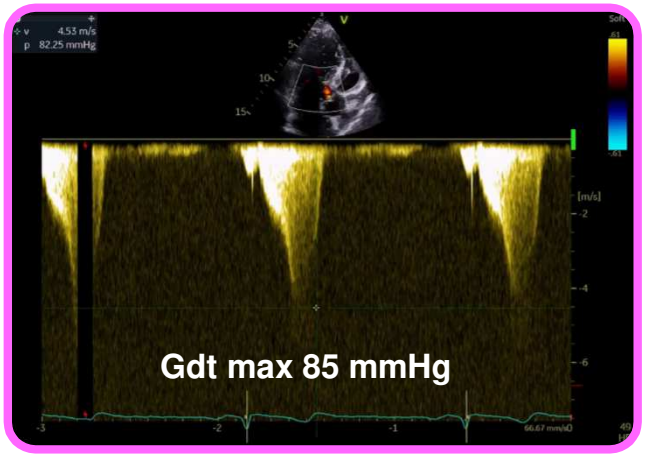




Obstruction intra-VG Valsalva





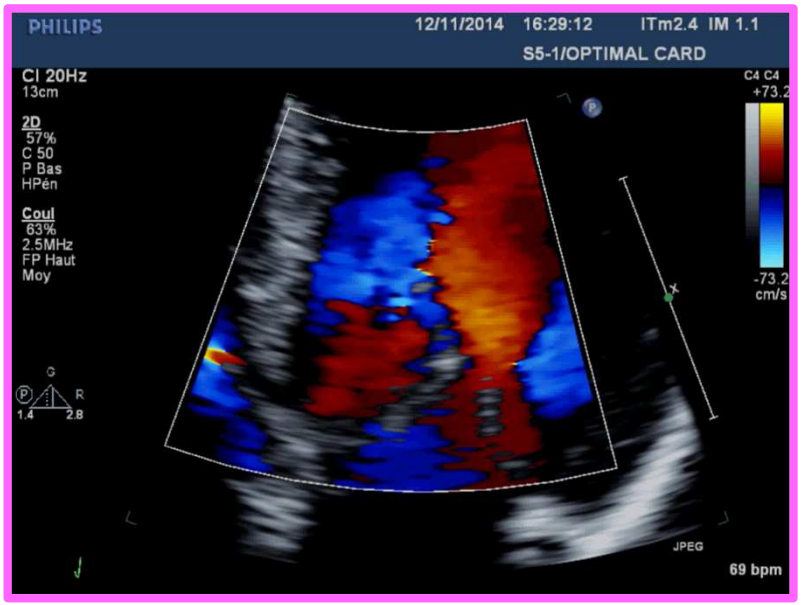


basal

Valsalva

Manœuvres provocatrices

- Manœuvres de Valsalva assis et penché en avant
- Puis debout
- Echocardiographie d'effort chez patients symptomatiques
- Echocardiographie dobutamine contre-indiquée



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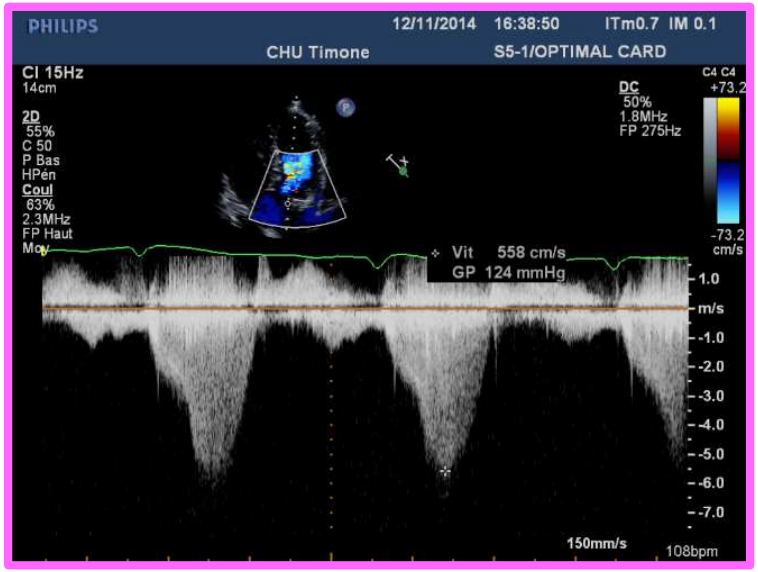

Transthoracic echocardiography evaluation in hypertrophic cardiomyopathy

Recommendations	Class	Level
In all patients with HCM at initial evaluation, transthoracic 2-D and Doppler echocardiography are recommended, at rest and during Valsalva manœuvre in the sitting and semi-supine positions—and then on standing if no gradient is provoked.	I	B
In *symptomatic* patients with a resting or provoked peak instantaneous LV outflow tract gradient <50 mm Hg, 2-D and Doppler echocardiography *during exercise* in the standing, sitting or semi-supine position is recommended to detect provocable LVOTO and exercise induced mitral regurgitation.	I	B
In *asymptomatic* patients with a resting or provoked peak instantaneous LV outflow tract gradient <50 mm Hg, 2-D and Doppler echocardiography *during exercise*—in the standing, sitting or semi-supine positions—may be considered when the presence of a LVOT gradient is relevant to lifestyle advice, and decisions on medical treatment.	IIb	C
In patients with sub-optimal images or with suspected LV apical hypertrophy or aneurysm, TTE with LV cavity opacification—using intravenous echocardiographic contrast agents—should be considered as an alternative to CMR imaging.	IIa	C

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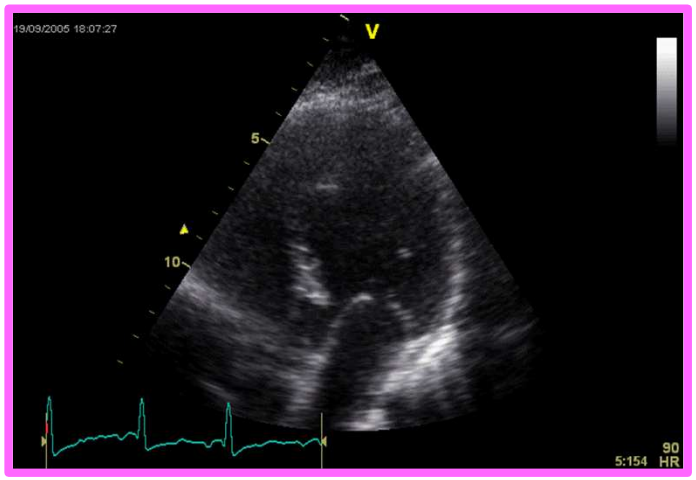
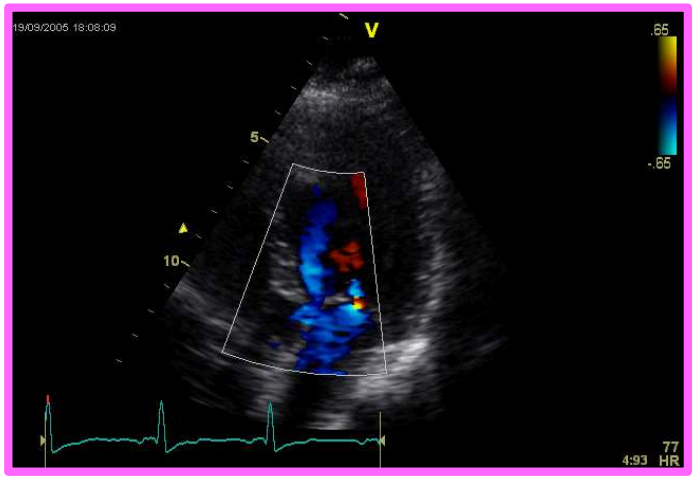
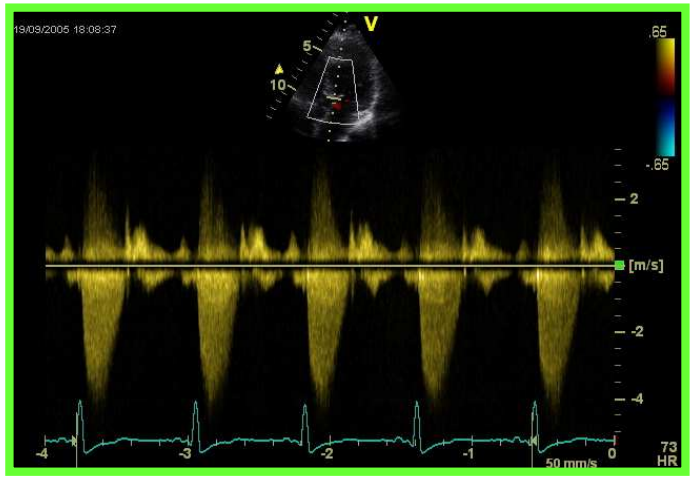
Diagnostic différentiel







membrane / diaphragme sous-aortique

L'obstruction dans les CMH

1. **Mécanisme**
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Les facteurs de risque de mort subite

Maron MS - N Engl J Med 2003;348:295-303.

1. TVNS (au moins 3 ESV consécutives) au Holter
2. Antécédents familiaux de mort subite
3. Syncope inexpliquée
4. Hypertrophie sévère (> 30 mm)
5. Obstruction intra-VG

6. + Anévrysme VG apical?
7. + Fibrose extensive à l'IRM?
8. + Baisse de la FEVG

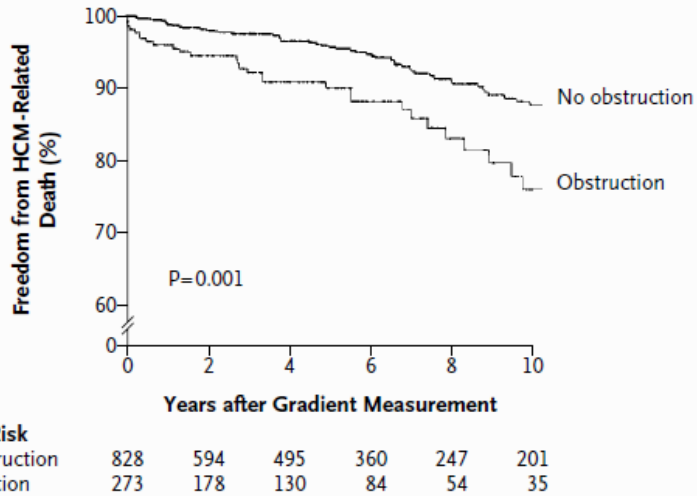


Figure 1. Probability of Hypertrophic Cardiomyopathy (HCM)–Related Death among 273 Patients with a Left Ventricular Outflow Gradient of at Least 30 mm Hg under Basal Conditions and 828 Patients without Obstruction at Entry.

Algorithme décisionnel « Européen »

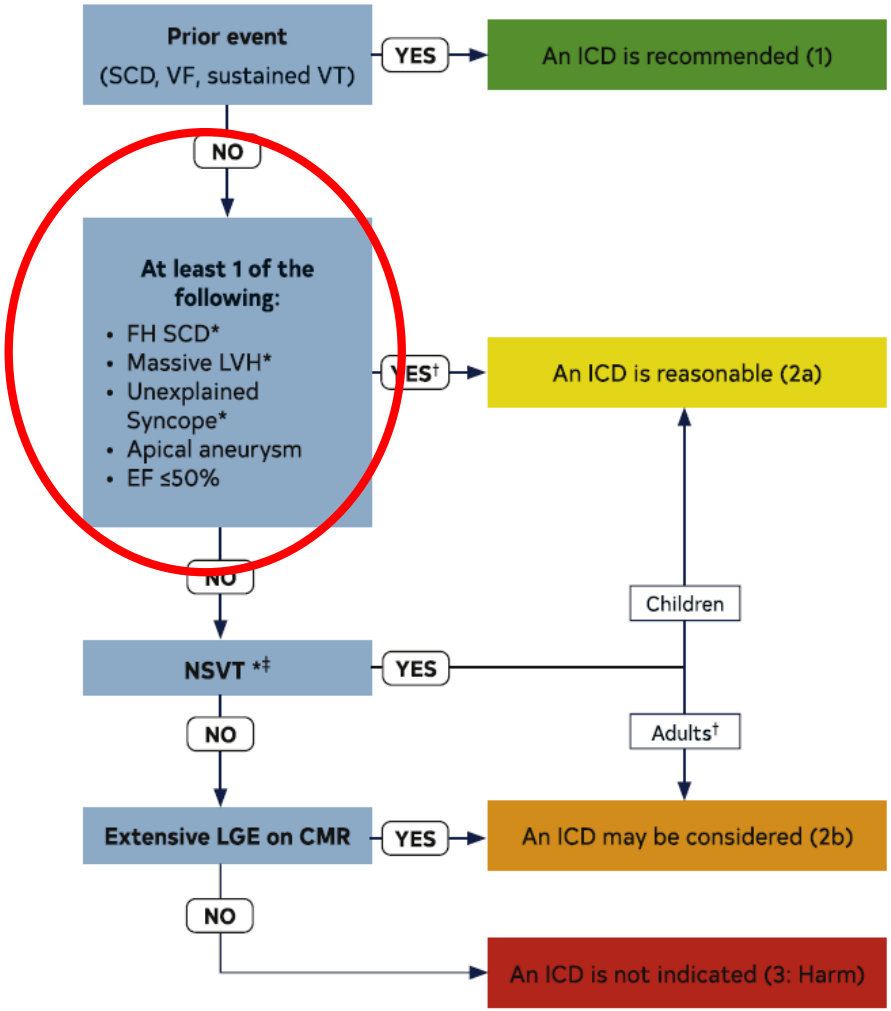
Table 7 Major clinical features associated with an increased risk of sudden cardiac death in adults

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.[69,376]
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).[69,73,120,247,248,373,377,378]
Family history of sudden cardiac death at a young age	• While definitions vary,[73,120,373,377] 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 in 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.[73,80,83,246,372,380] 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.

Algorithme décisionnel « américain »

FIGURE 3 ICD Patient Selection



Mme D – 15ans

ATCD Familiaux

CMH chez le père – MYH7

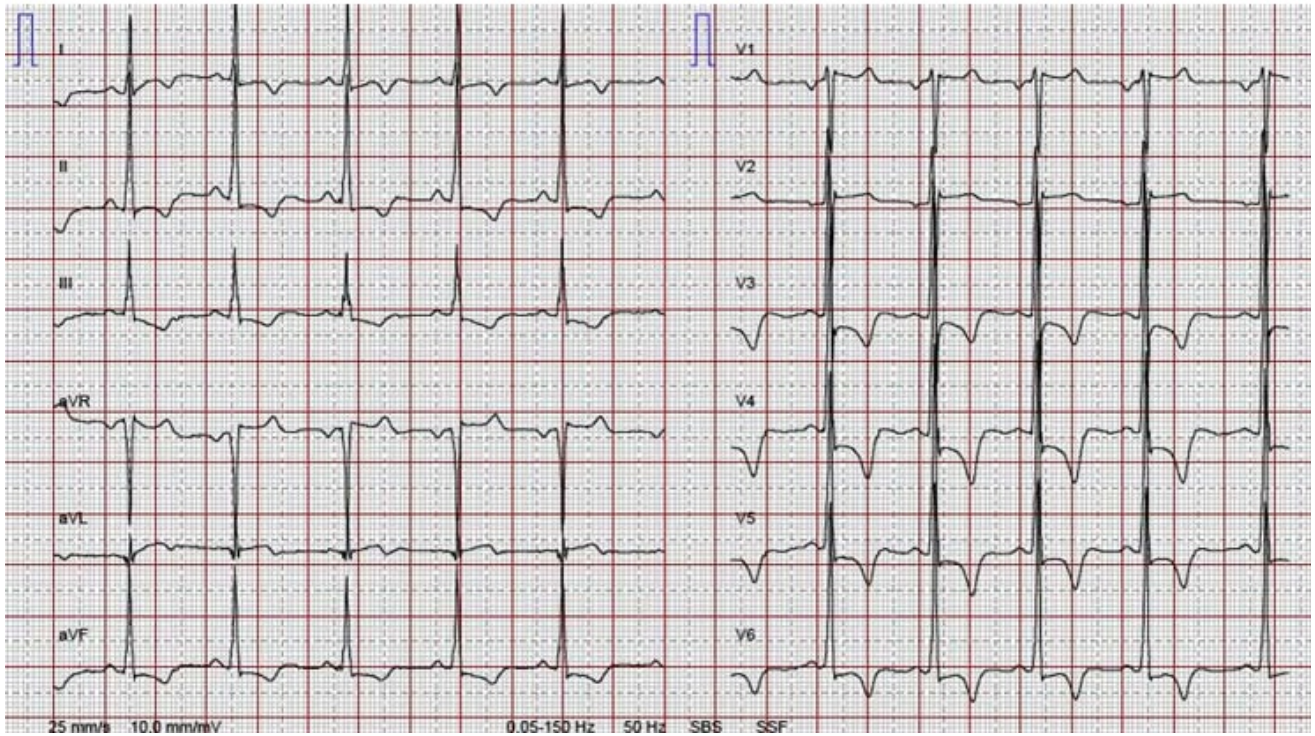
Mort Subite chez le grand père

ATCD Personnel

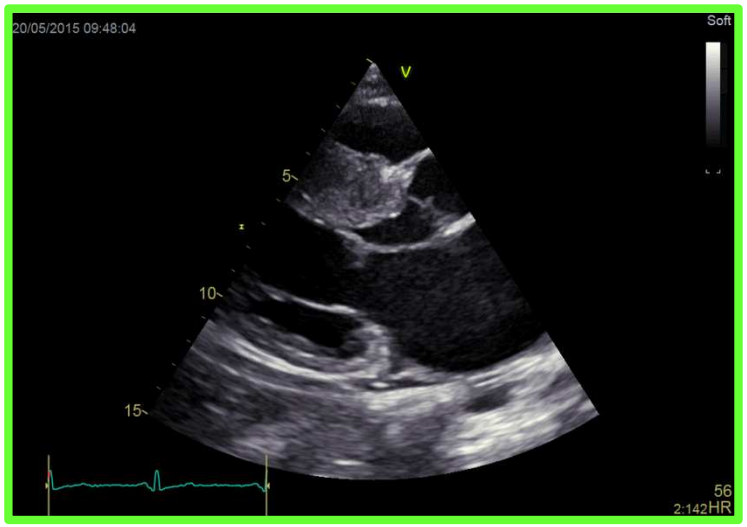
CMHO découverte à l'âge de 12ans

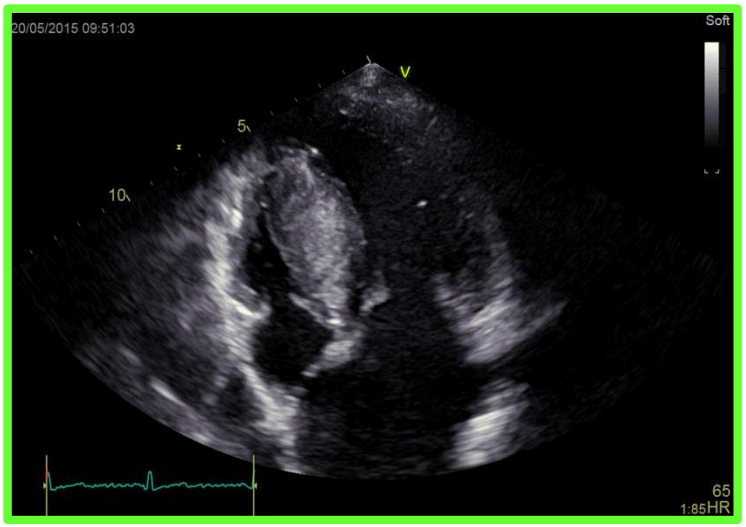
Dyspnée NYHA 2
Episode de lipothymie

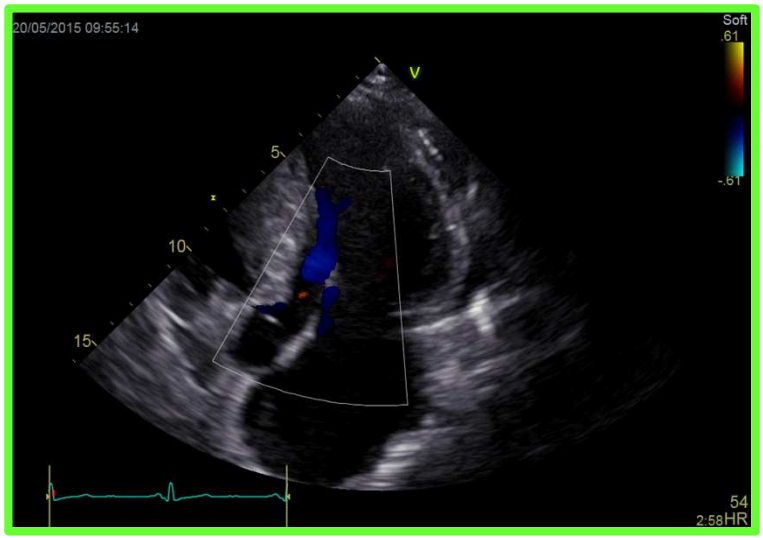
Souffle systolique mésocardiaque 4/6
Absence de signes congestifs

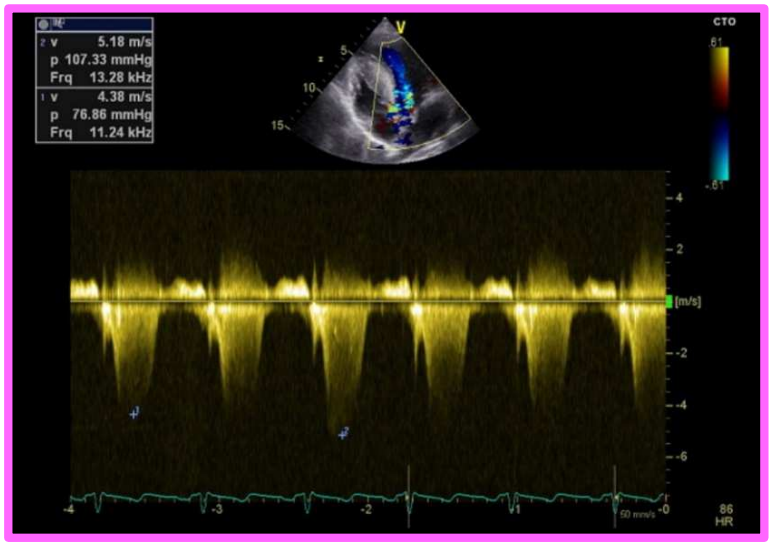


ETT 2015









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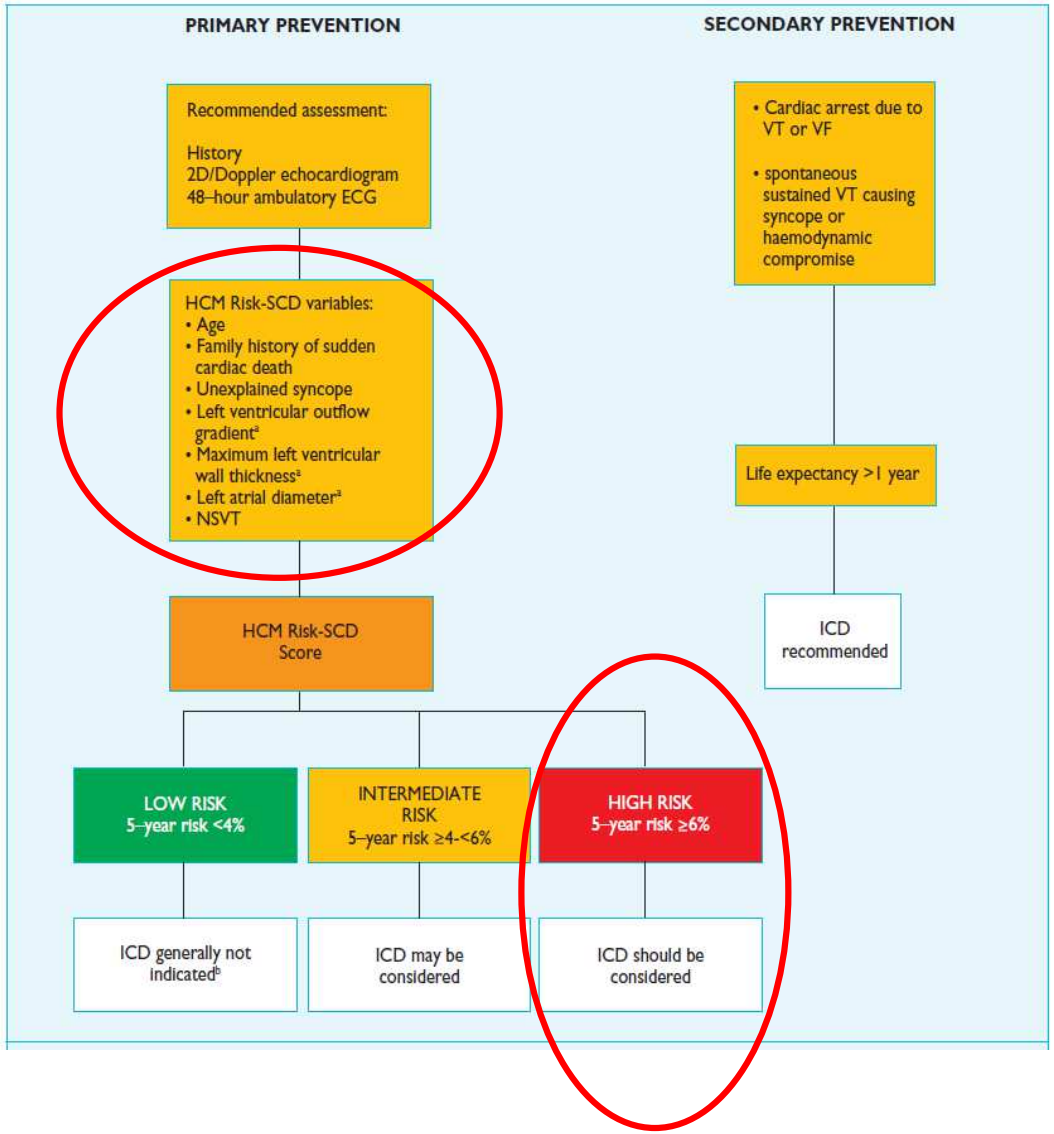

- Score à 12,9%
- Implantation d'un DAI Sous Cutané en 2015
- Hospitalisation en 2016 pour un CEI approprié sur FV
- NYHA 3 sous traitement BB

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Possibilités thérapeutiques

1. Traitement medical: Betabloquants, Verapamil, Rythmodan
2. Alcoolisation septale
3. Chirurgie – myomectomie
4. Nouveaux traitements: inkibiteurs de la myosine
5. Transplantation cardiaque

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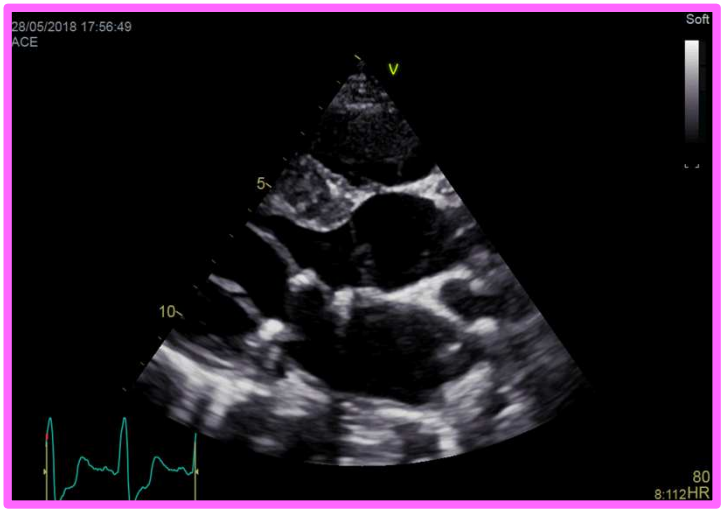
→ **Myomectomie + plastie mitrale en 2016**

ETT 2018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ACE
Soft
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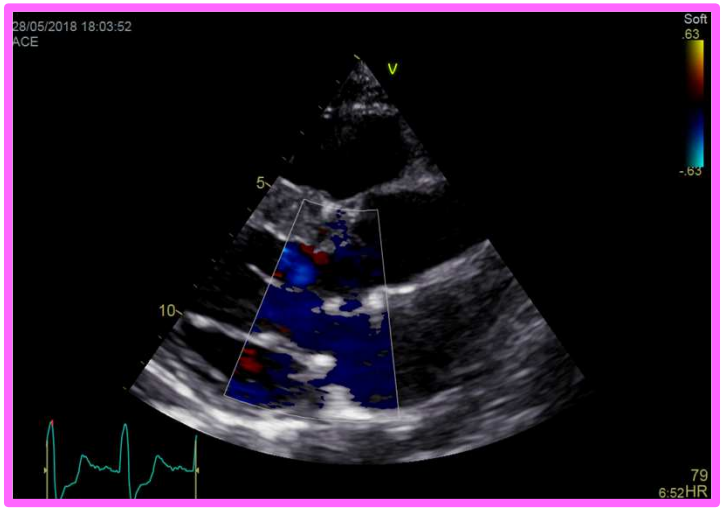
8:112HR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.63
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79
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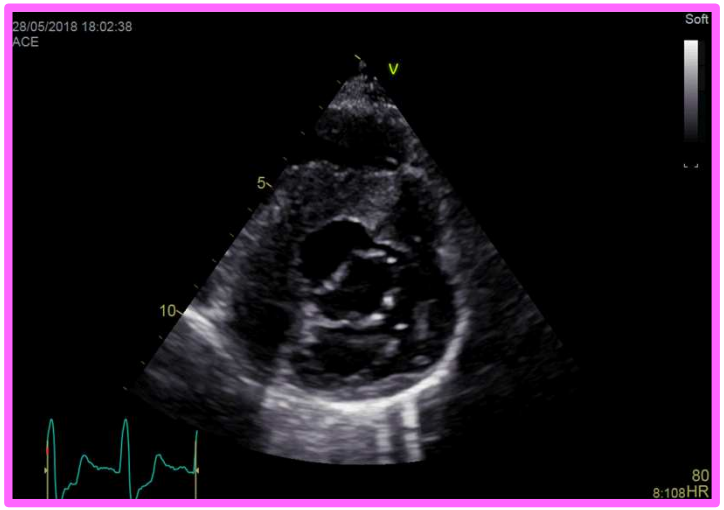
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ACE
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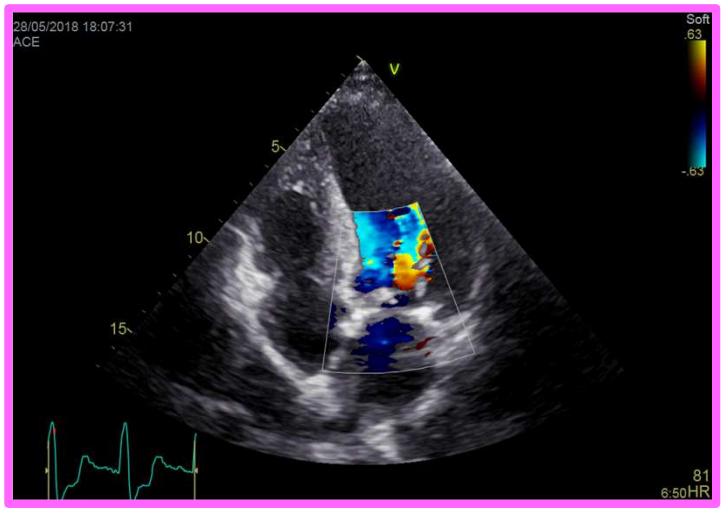
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Soft
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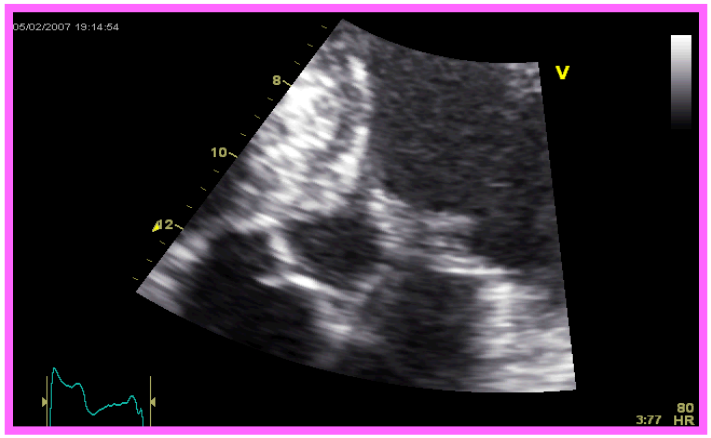
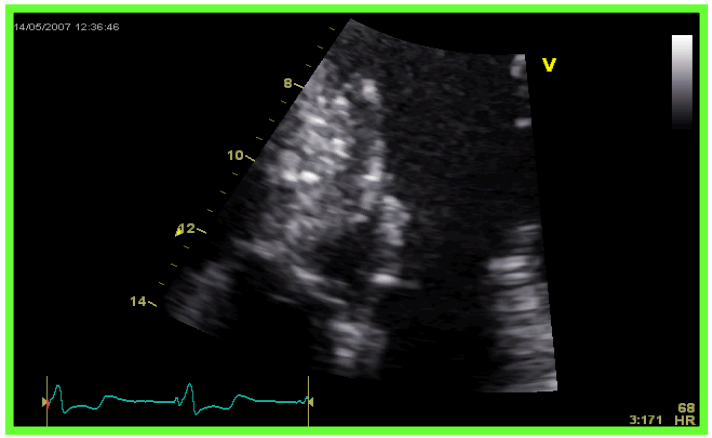
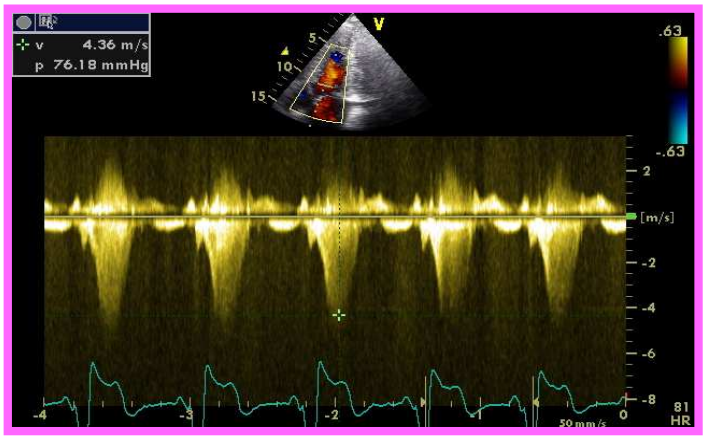
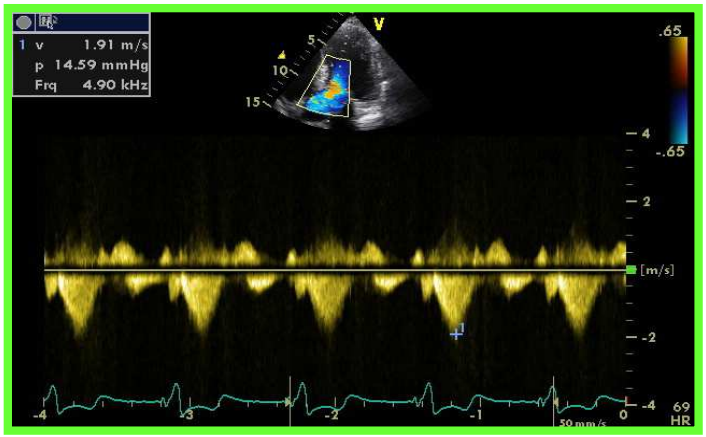
Alcoolisation septale









Avant alcoolisation septale

Après alcoolisation septale

Conclusion: l'obstruction dans les CMH

1. Présente spontanément ou provoquée dans 2/3 des CMH
2. Méthodologie diagnostique rigoureuse
3. Valeur pronostique discutée
4. Cible des différentes thérapeutiques médicamenteuses ou interventionnelles visant à améliorer les symptômes