Echocardiographic workup for HCM – Making the diagnosis and evaluation of cardiac function

Thor Edvardsen, MD, PhD – Oslo, Norway

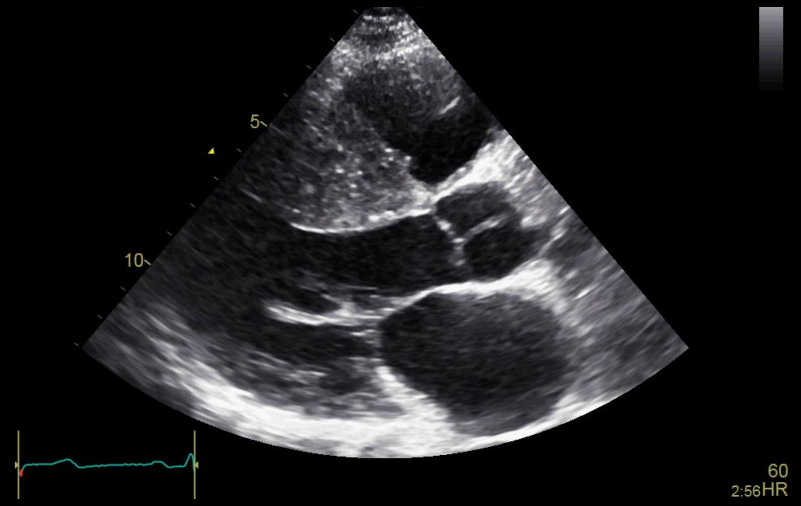
Disclosures

Speaker fees:

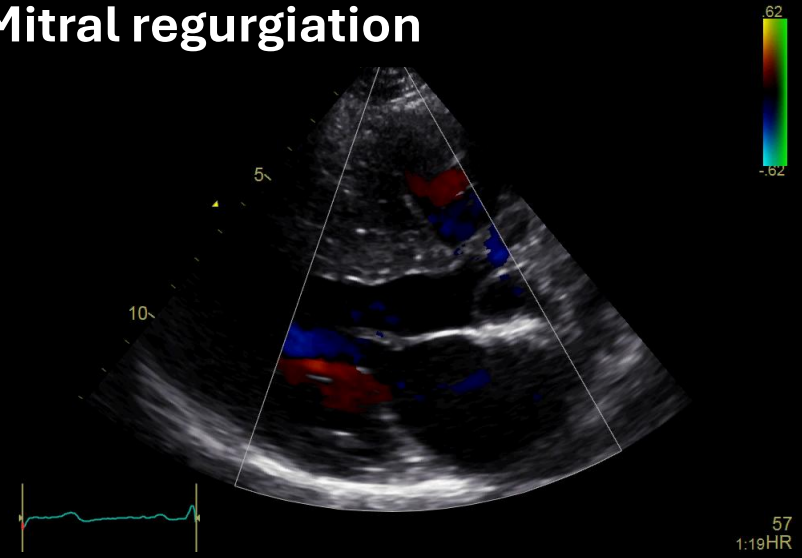
Bristol Myers Squibb

Typical echocardiographic findings in severe HCM

Systolic anterior motion (SAM) of the mitral valve

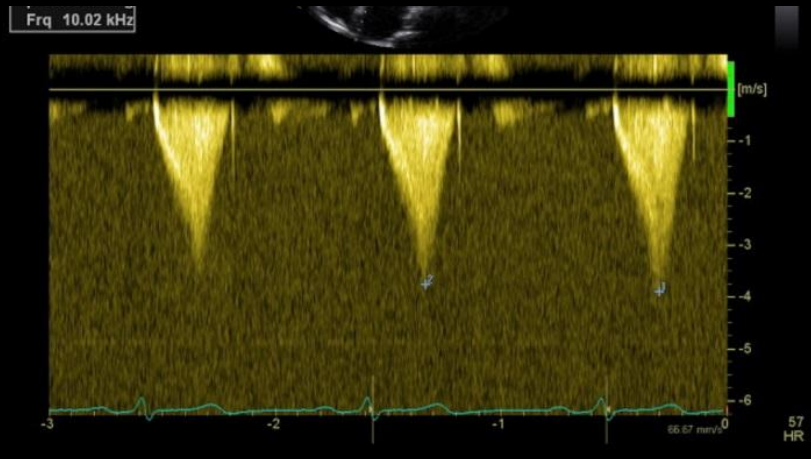


Mitral regurgiation

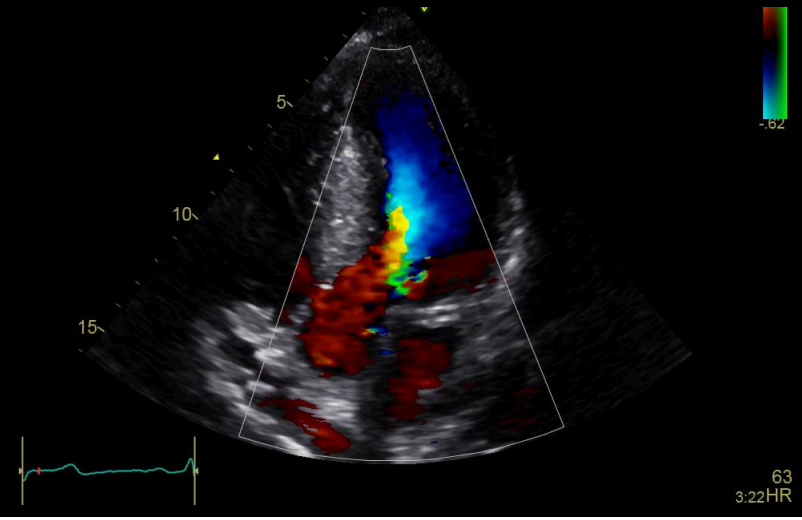


LVOT obstruction – CW doppler

typical dagger-shaped envelope



LVOT obstruction – Color doppler



Diagnostic Criteria

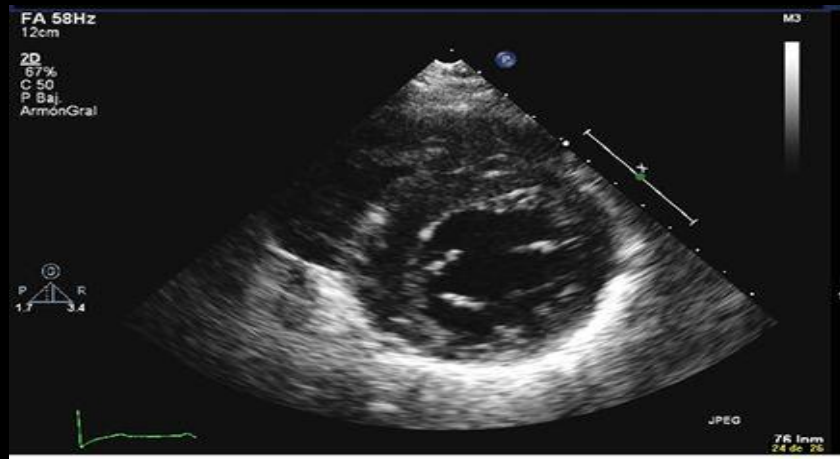
- **Adults**: LV wall thickness ≥15 mm in any myocardial segment
  - not explained by loading conditions

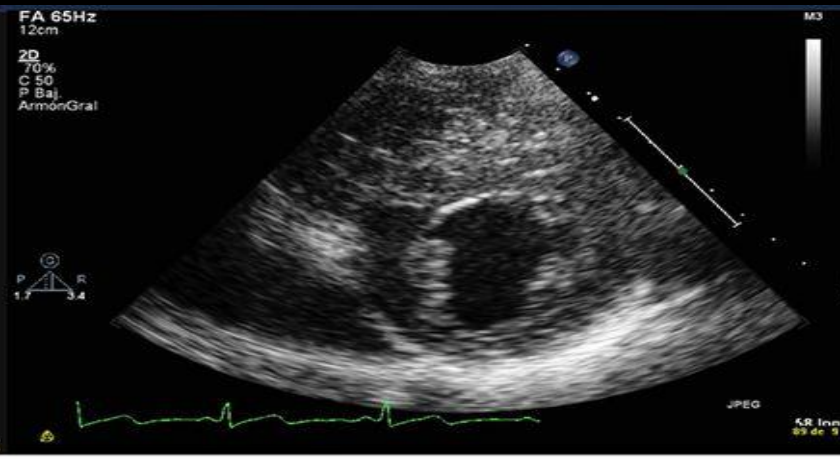
- **Relatives**: LV Wall thickness ≥13 mm

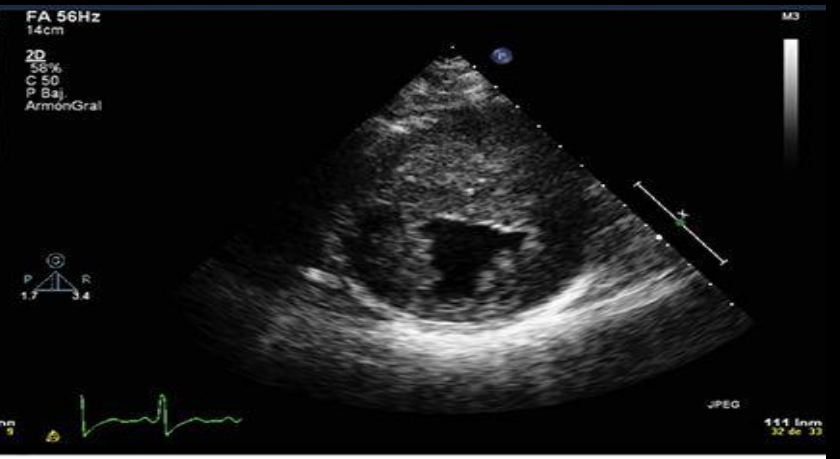
- **Children:** LV wall thickness > than 2 SD > predicted mean (z-score >2)

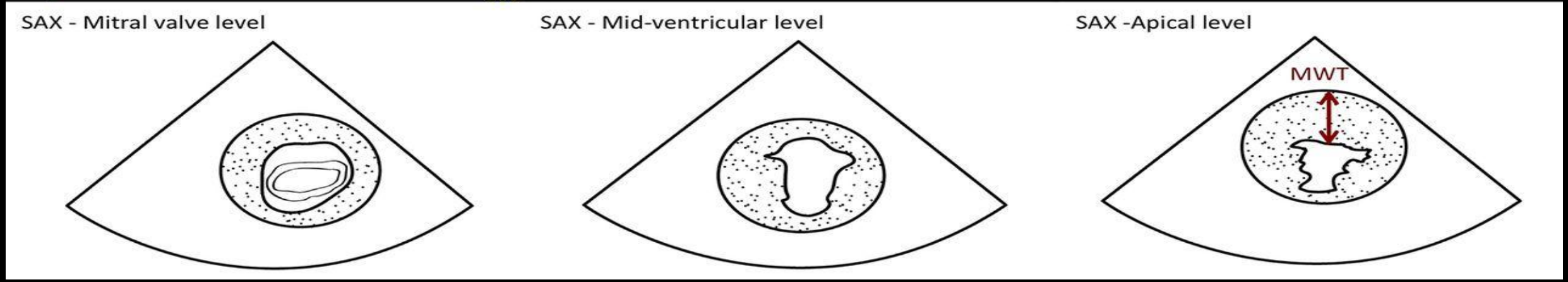
Left Ventricular Hypertrophy Evaluation in HCM

Assess MWT at different levels





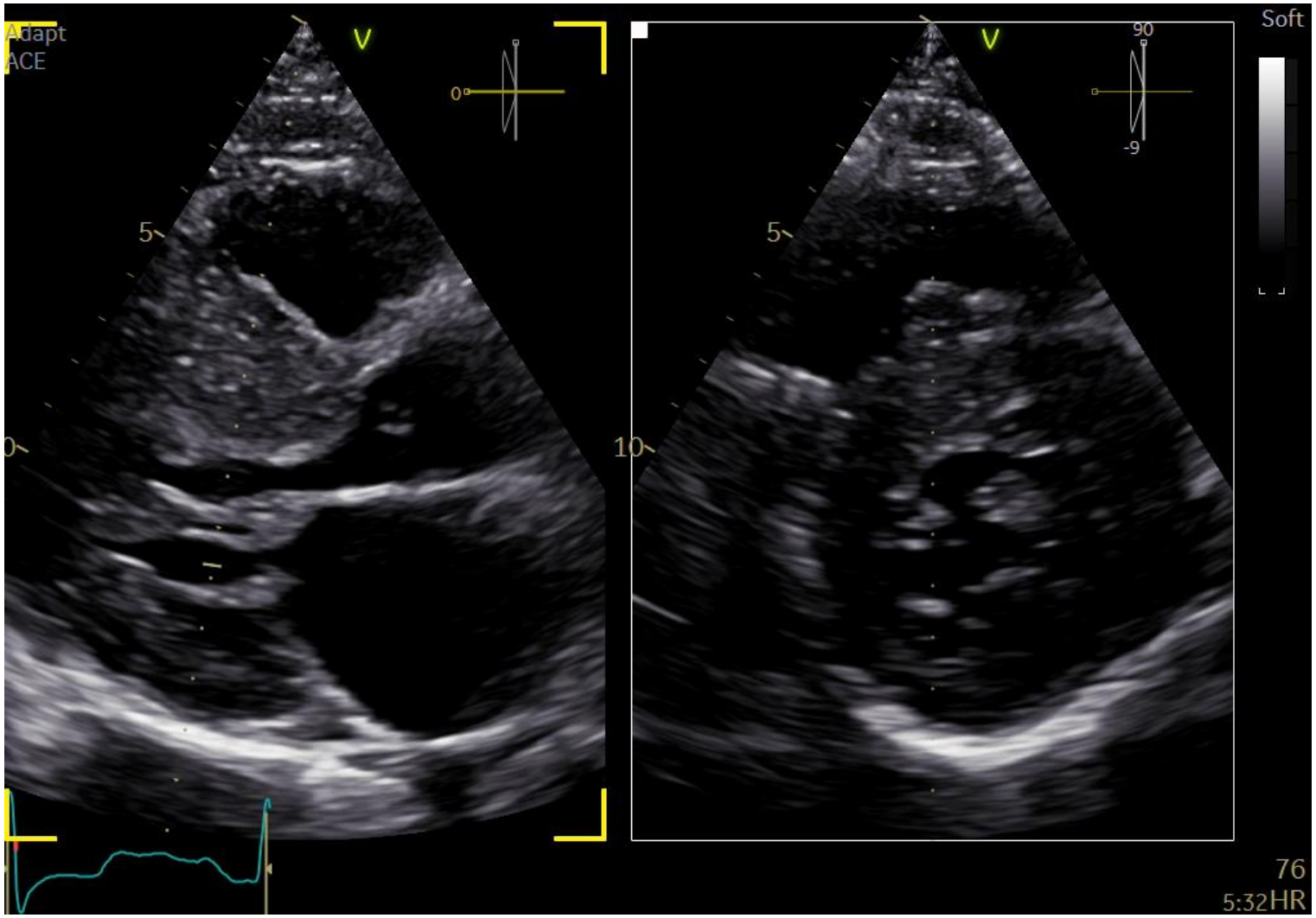




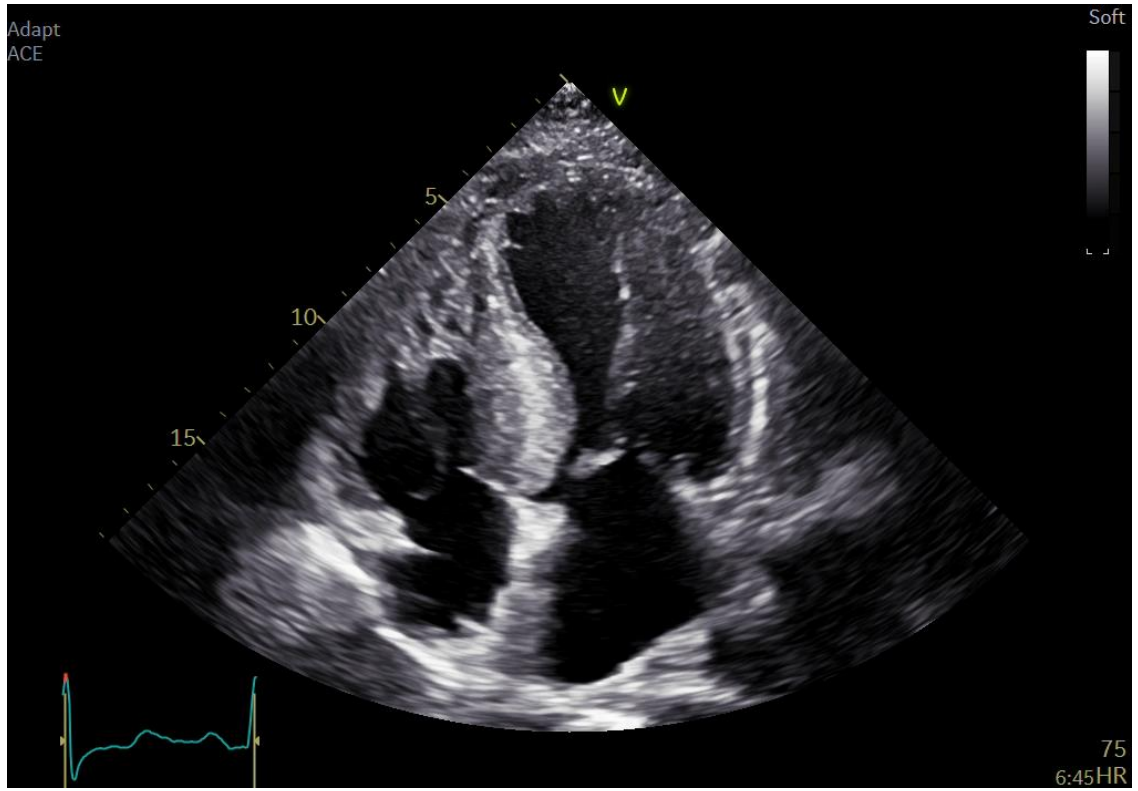
Dominguez, et al. *Heart*. 2018.

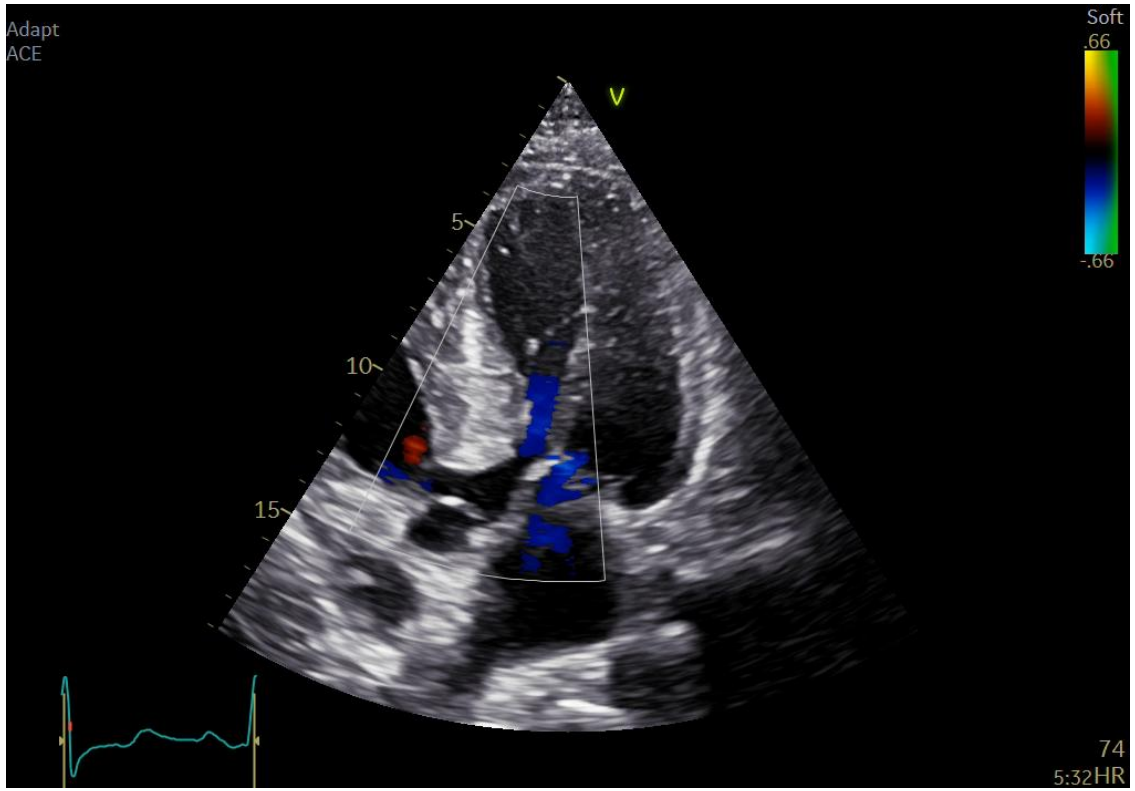
Man 37 years

- Dyspnoe
- NYHA class II

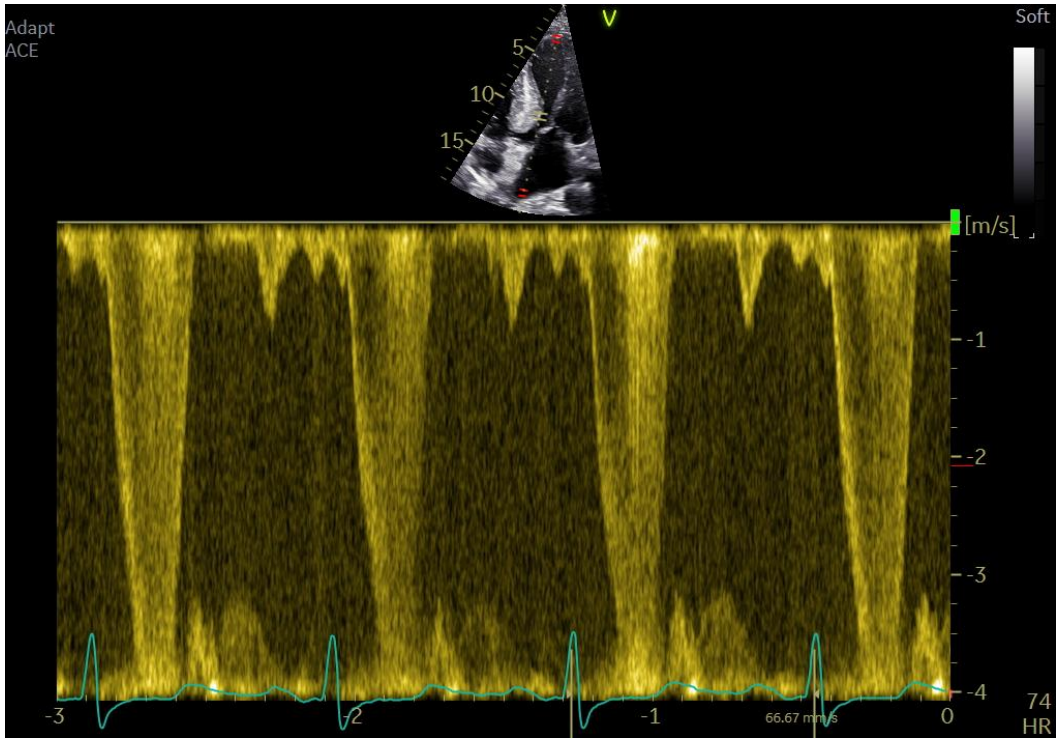

Adapt
ACE
V
0
5
0
V
90
-9
5
10
Soft
76
5:32HR

HCM


Adapt
ACE
Soft
V
5
10
15
75
6:45HR


Adapt
ACE
Soft
.66
-.66
V
5
10
15
74
5:32HR

Obstruction



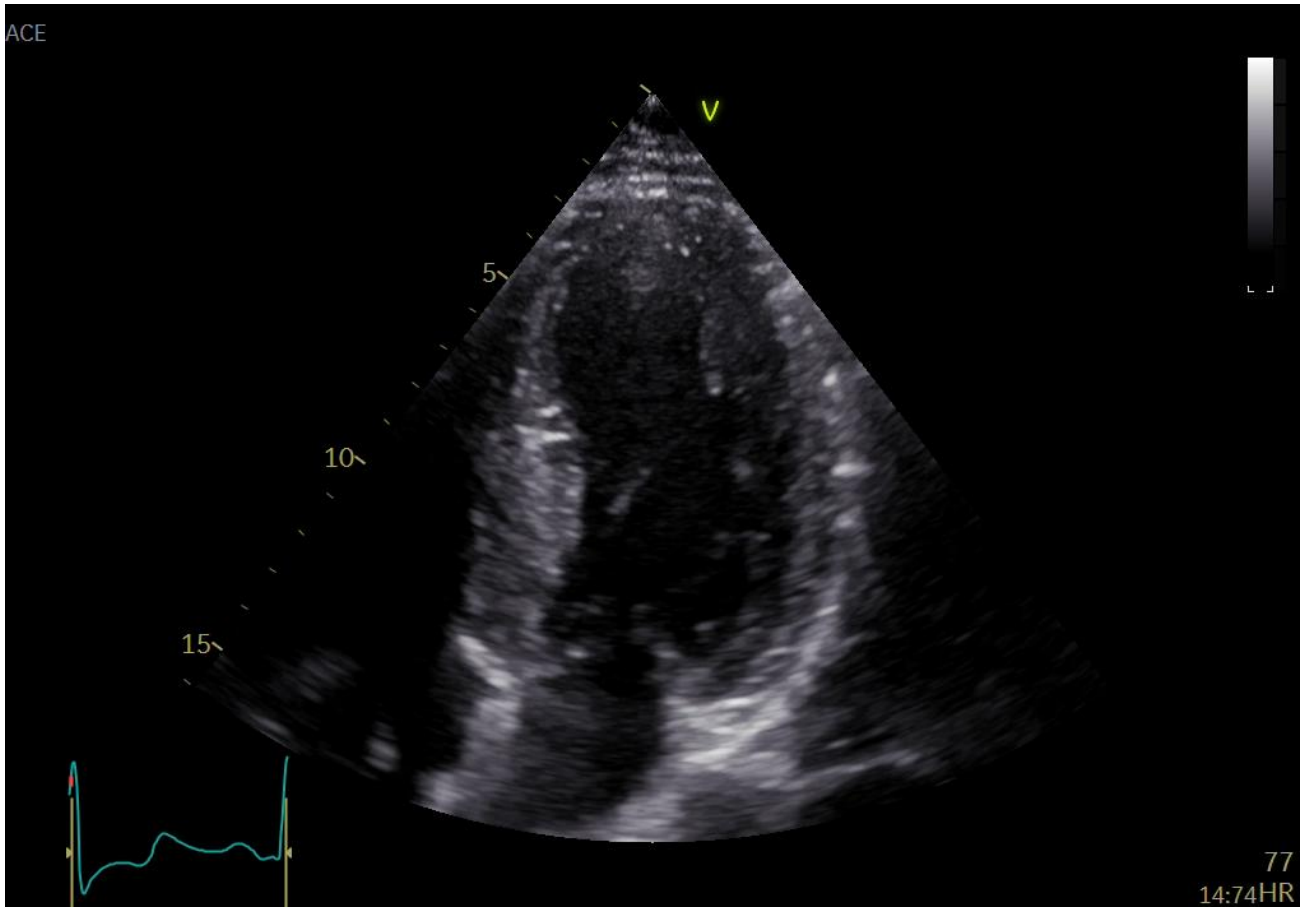
>5 m/s

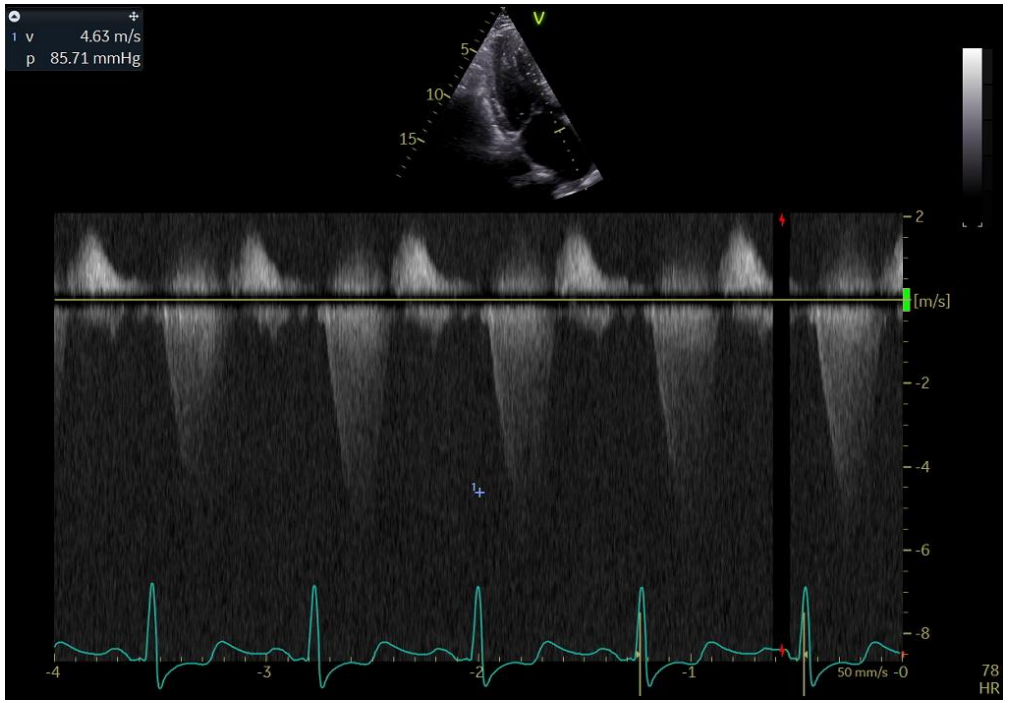
>100 mmHg

Man 37 years

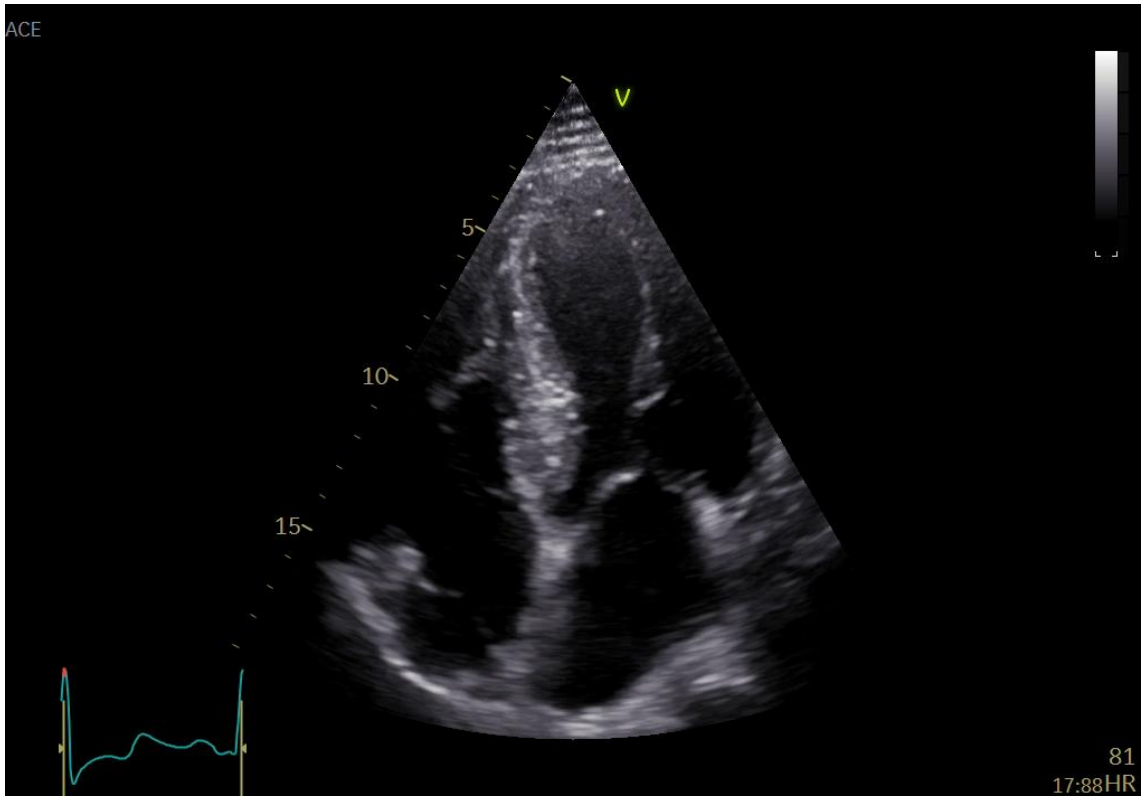
- Dyspnoe
- NYHA class II
- Genotype positive (MYBPC3)
- Risk calculator 5 year = 8.3% - received ICD
- Betablocker
  - minor effect
- Referred for possible Septal Reduction Therapy (May -24)
  - septal myectomy or alcohol septal ablation

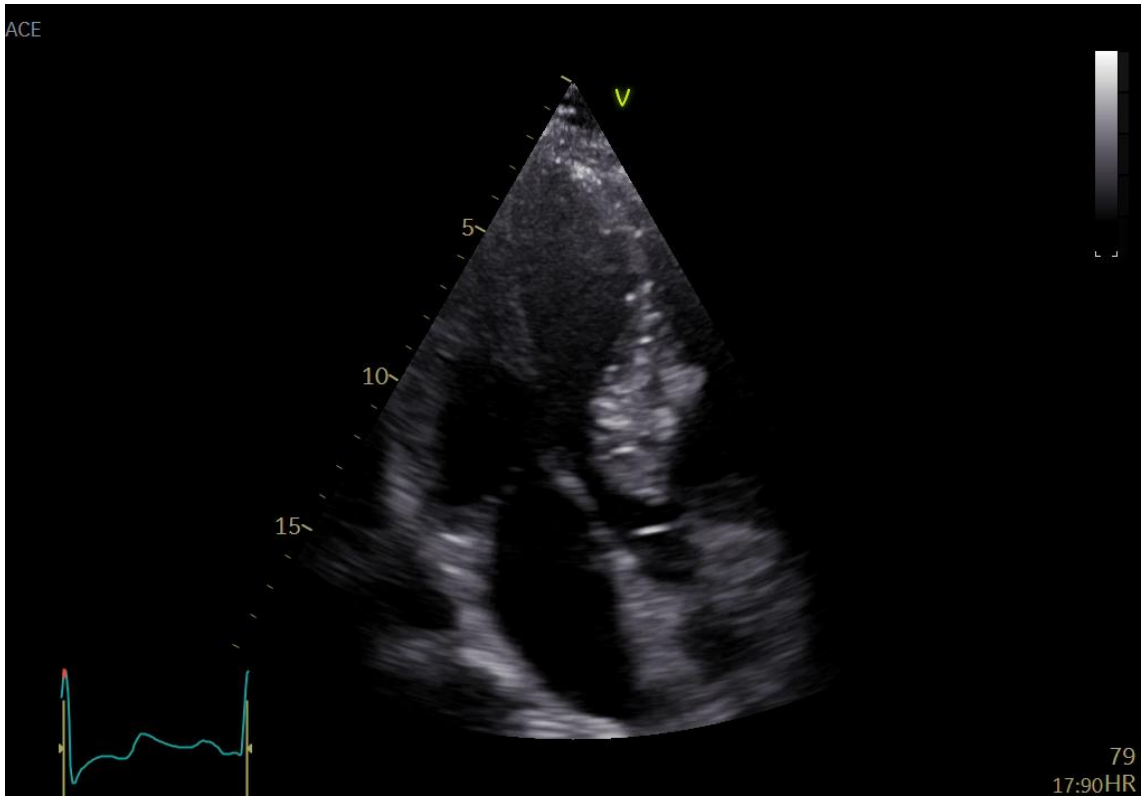
Before alcohol septal ablation





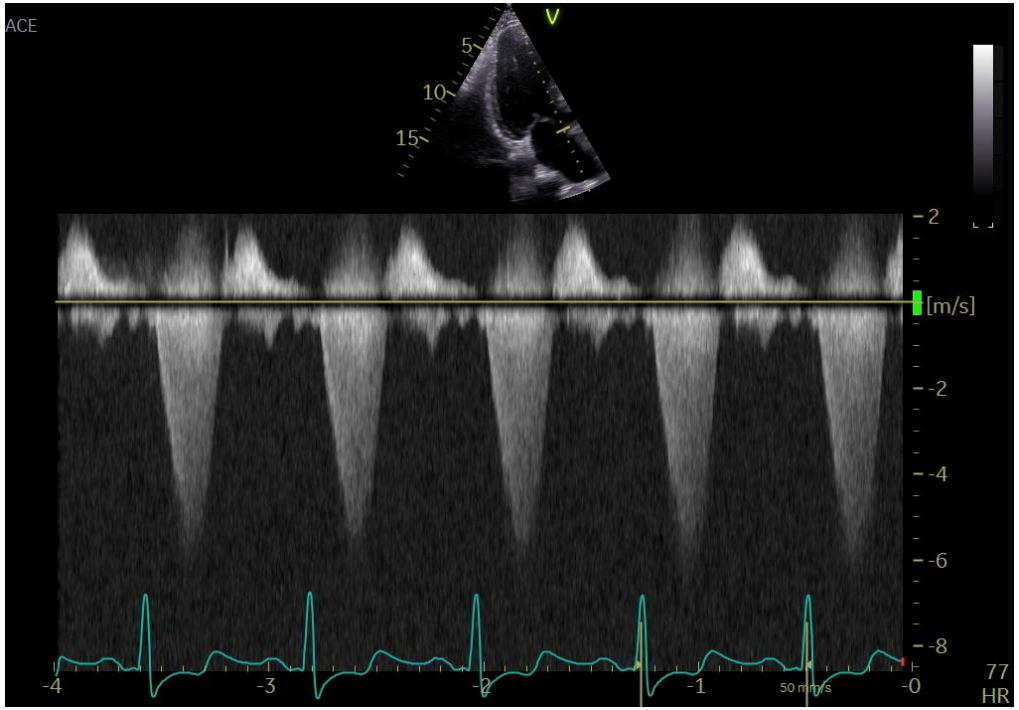
During alcohol septal ablation



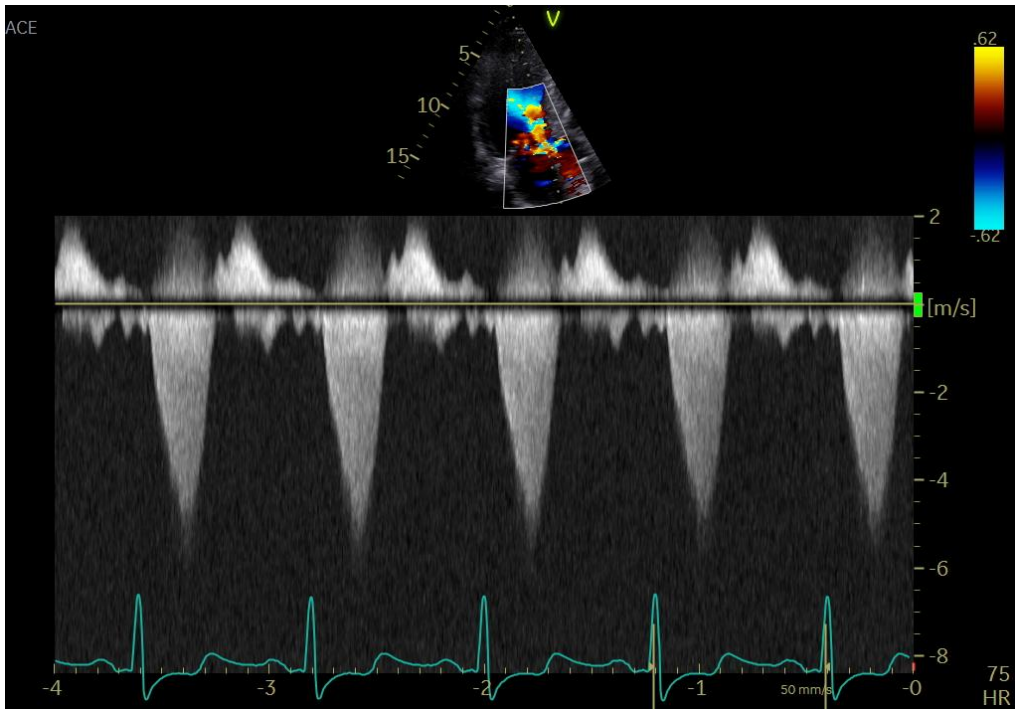


Comparison before and after alcohol septal ablation

Before



After



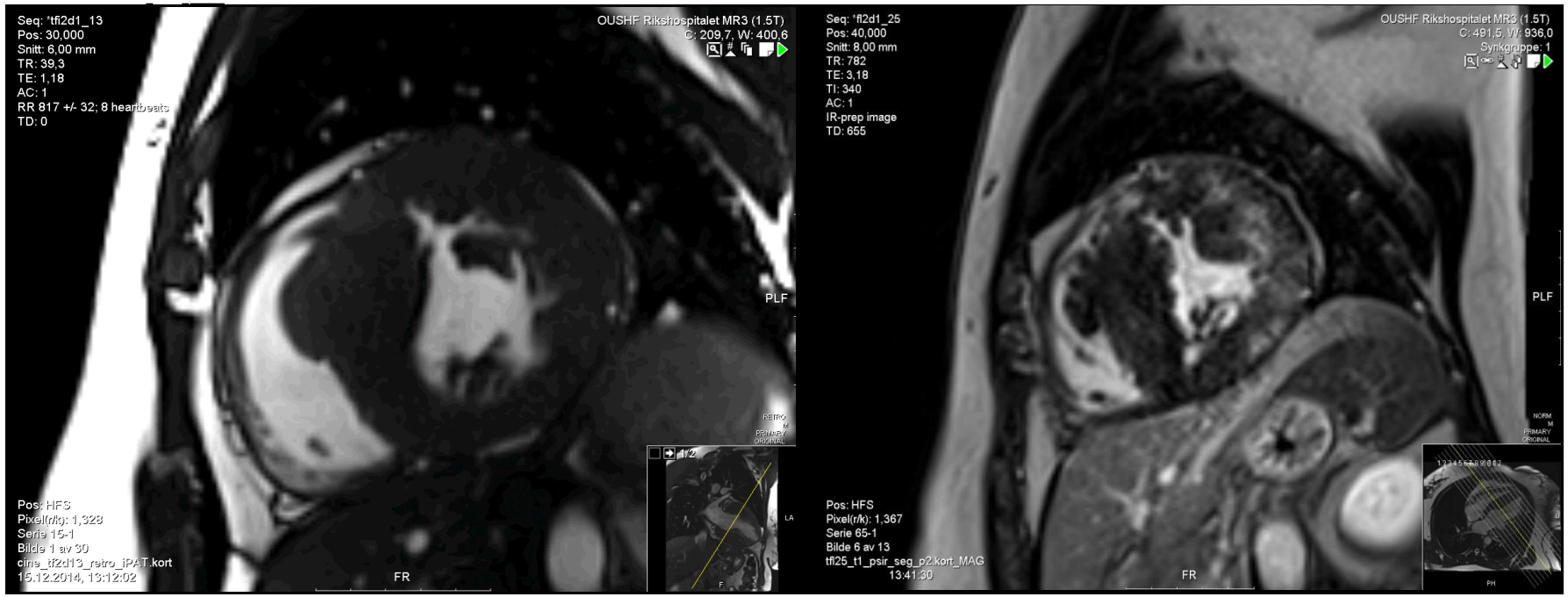
Status after ASA

- No complications during or after procedure
- Max Troponine T = 1700
- Max CK-MB = 118

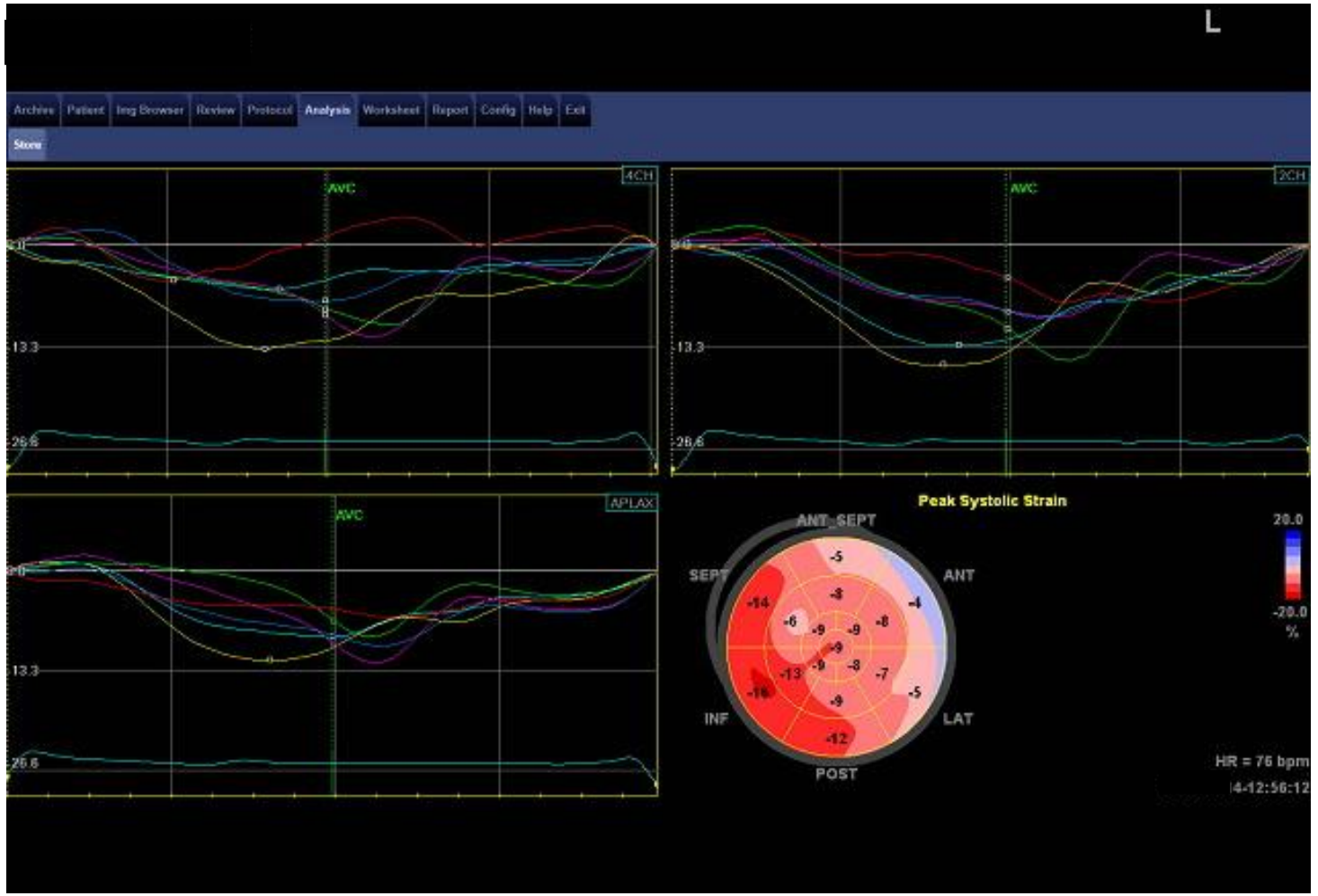
- Successful treatment?

CMR

35 y man, 3 SCD family



Strain - GLS



35 y man, 3 SCD family

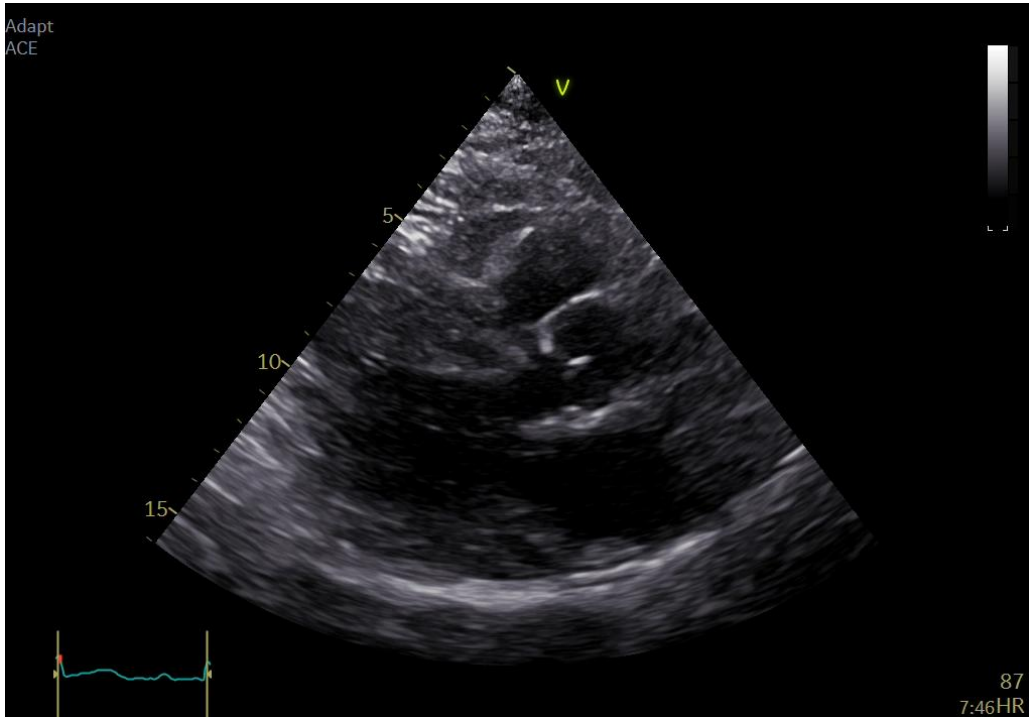
EF = 65%

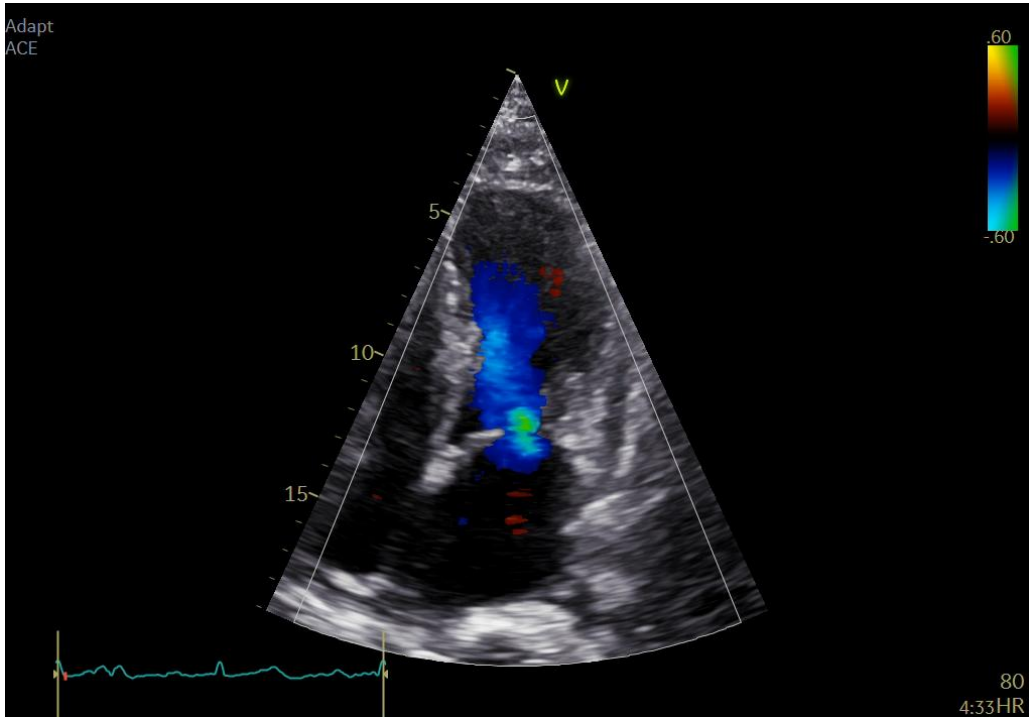
GLS = -9%

Woman 72 years

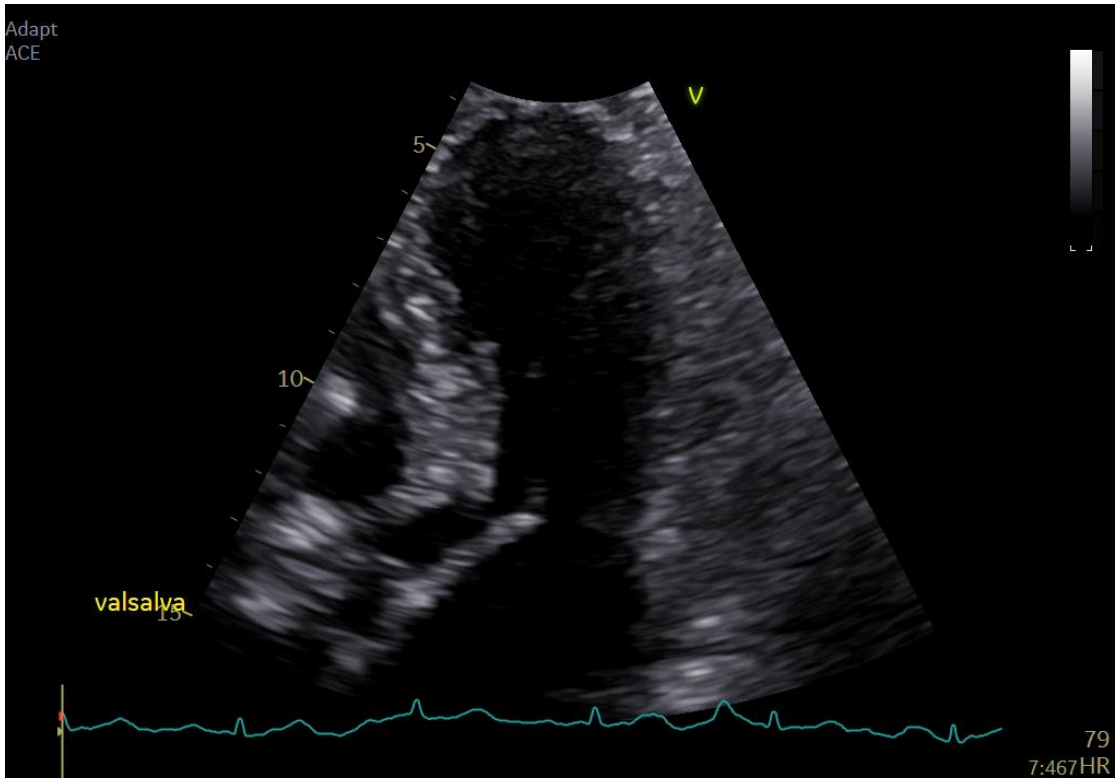
- Palpitations and dyspnoe
- NYHA class III

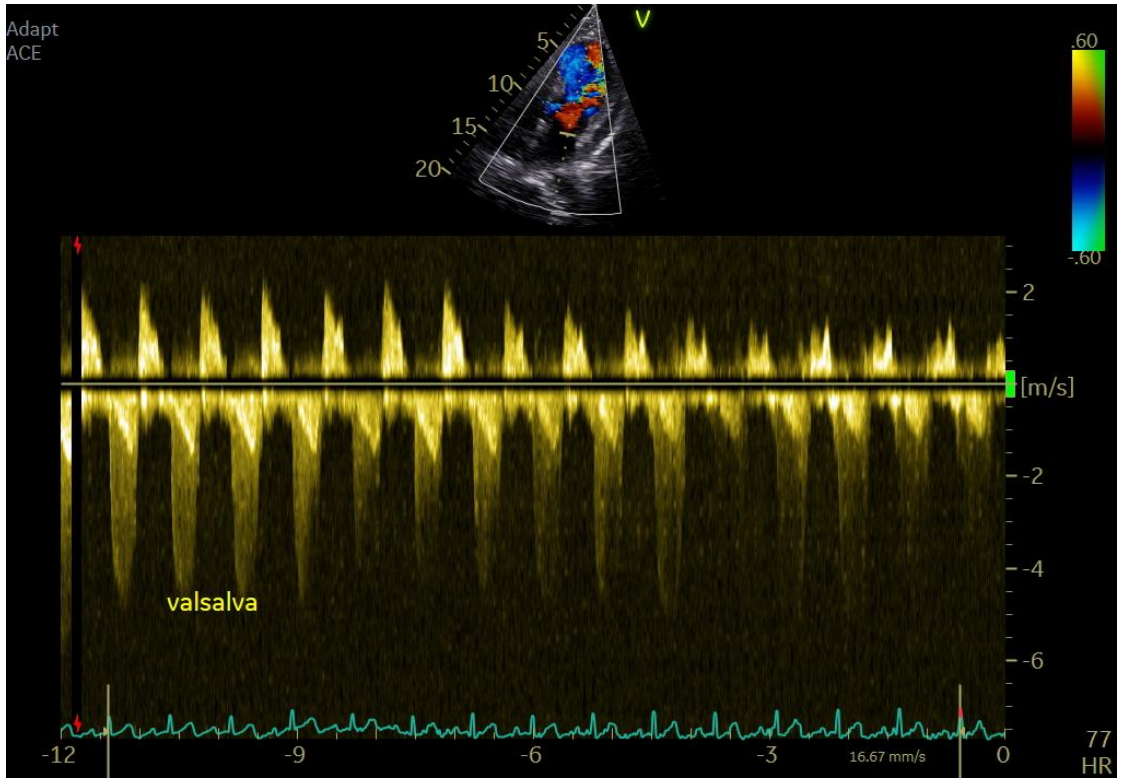
Midventricular obstruction



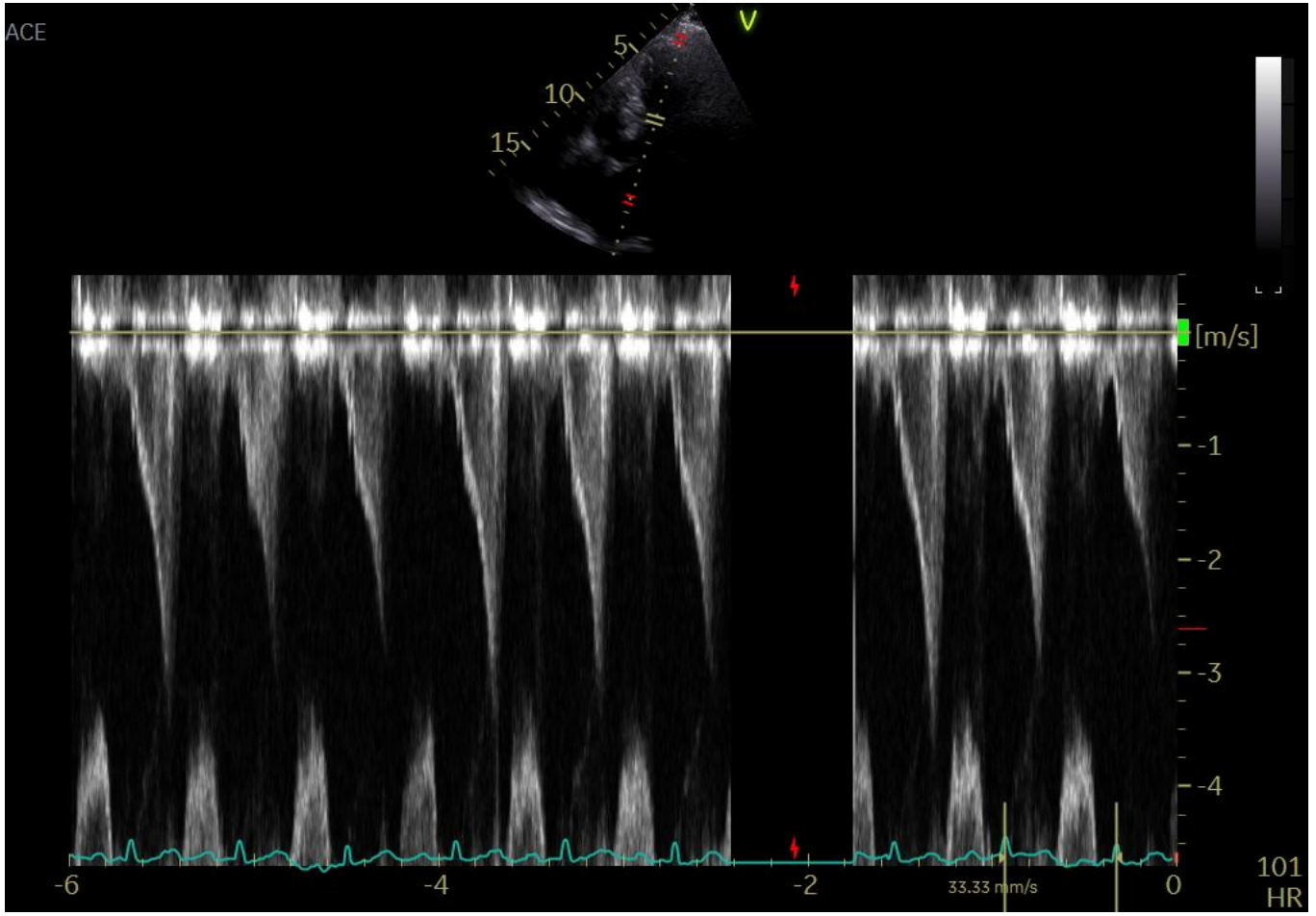


Valsalva


Adapt
ACE
V
5
10
15
valsalva
79
7:467 HR


Adapt
ACE
V
5
10
15
20
.60
-.60
2
[m/s]
-2
-4
-6
valsalva
-12
-9
-6
-3
16.67 mm/s
0
77
HR

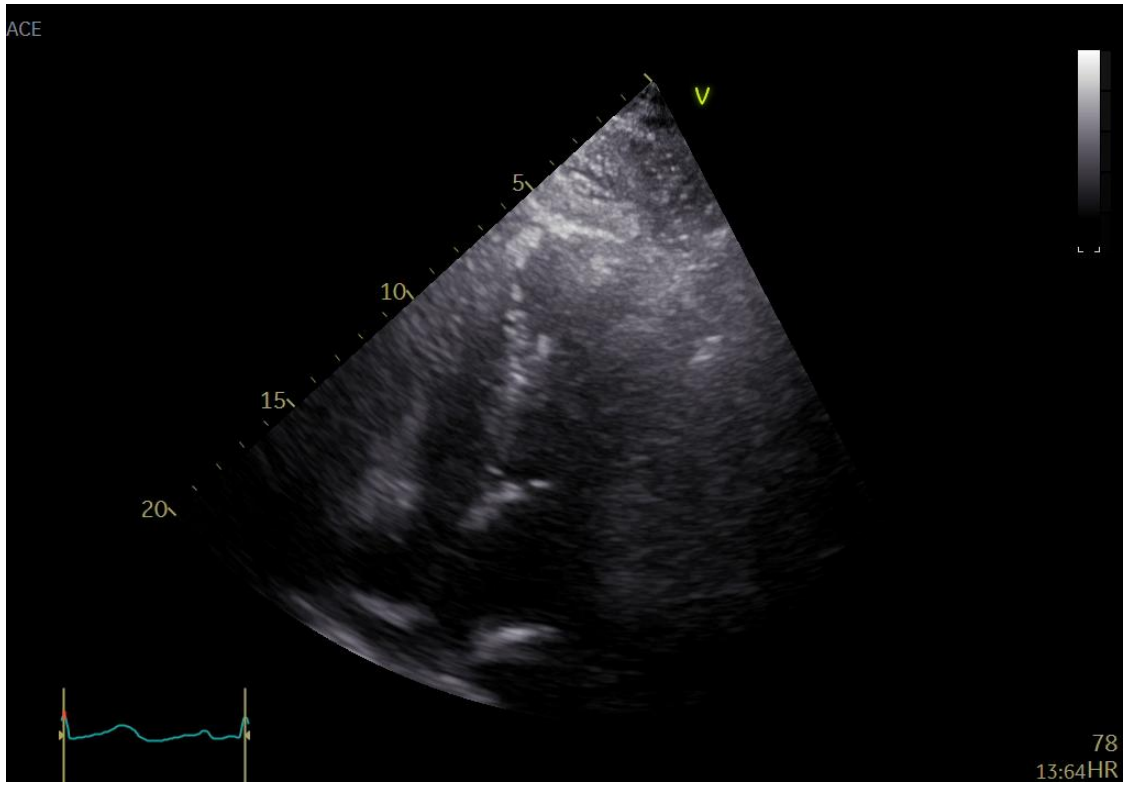
Squats

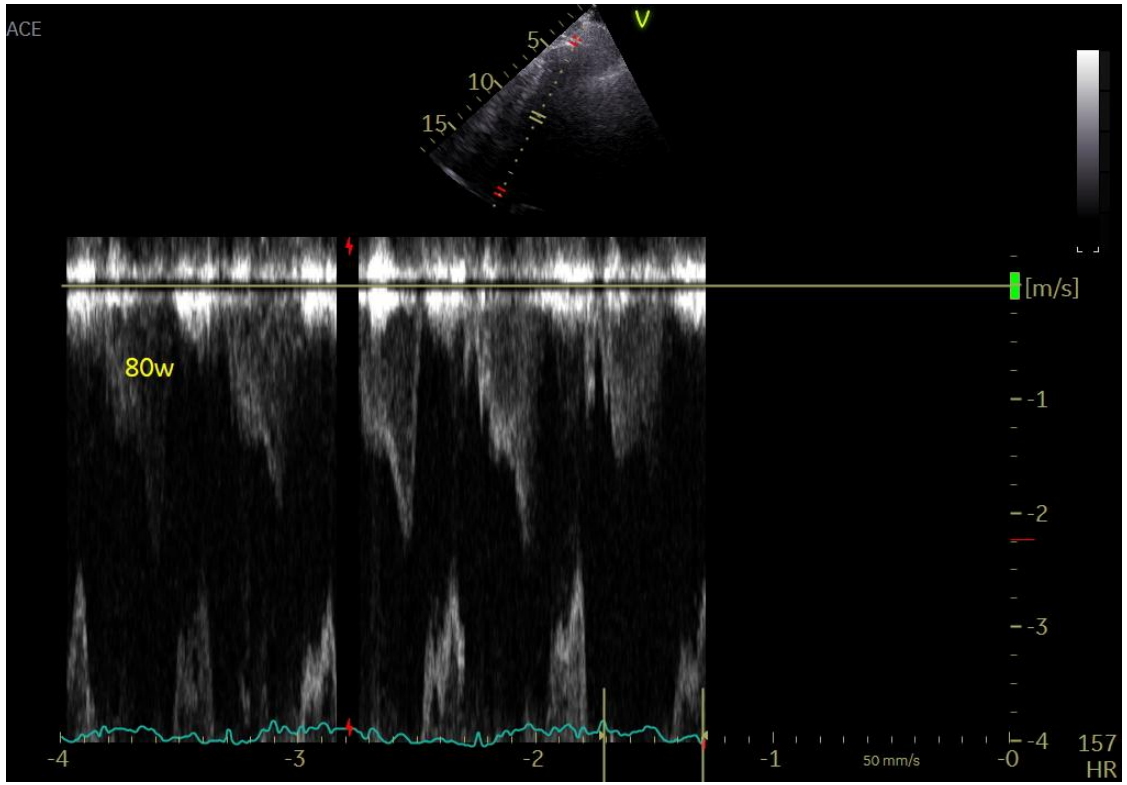


3.7 m/s

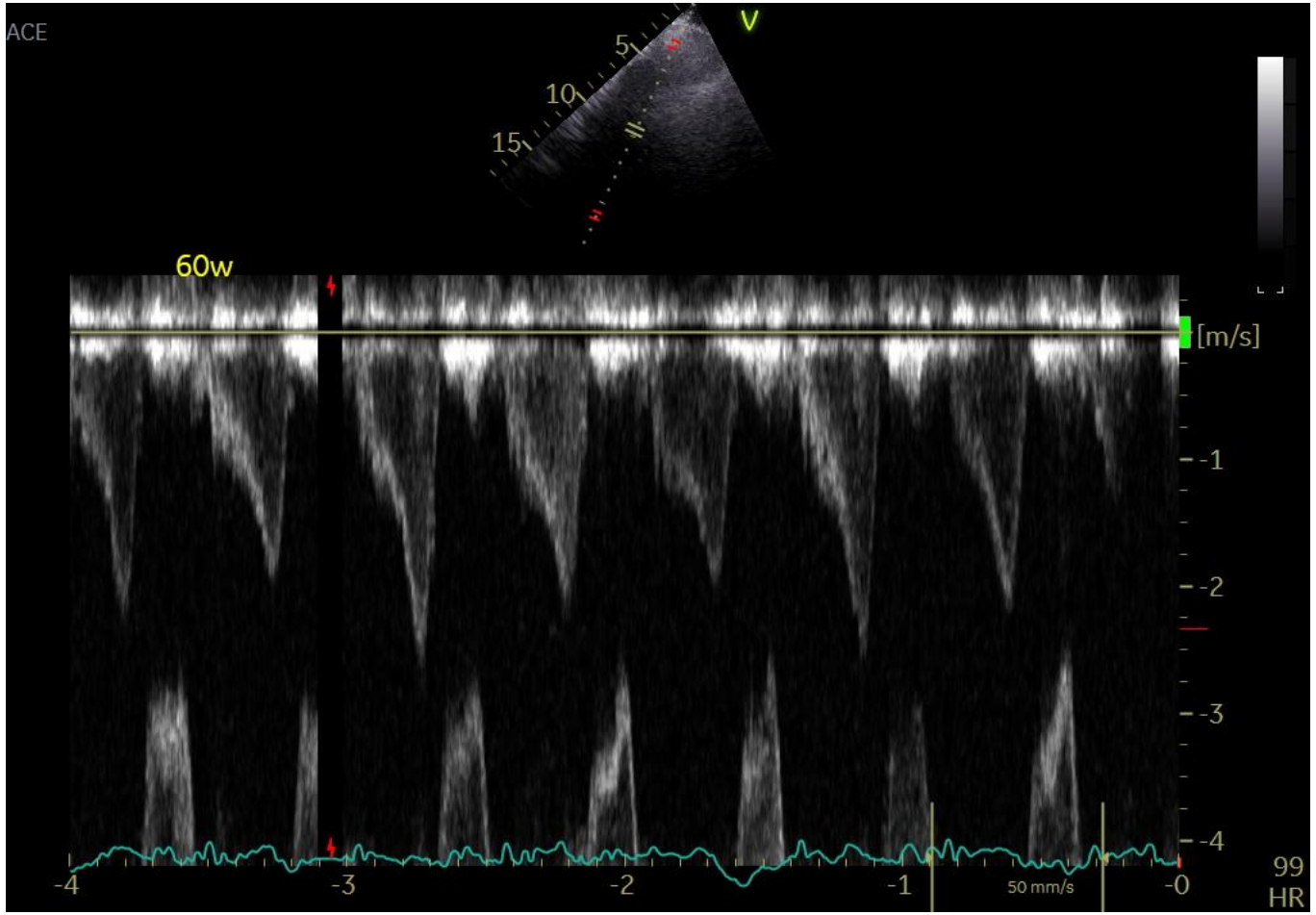
55 mmHg

Exercise echocardiography

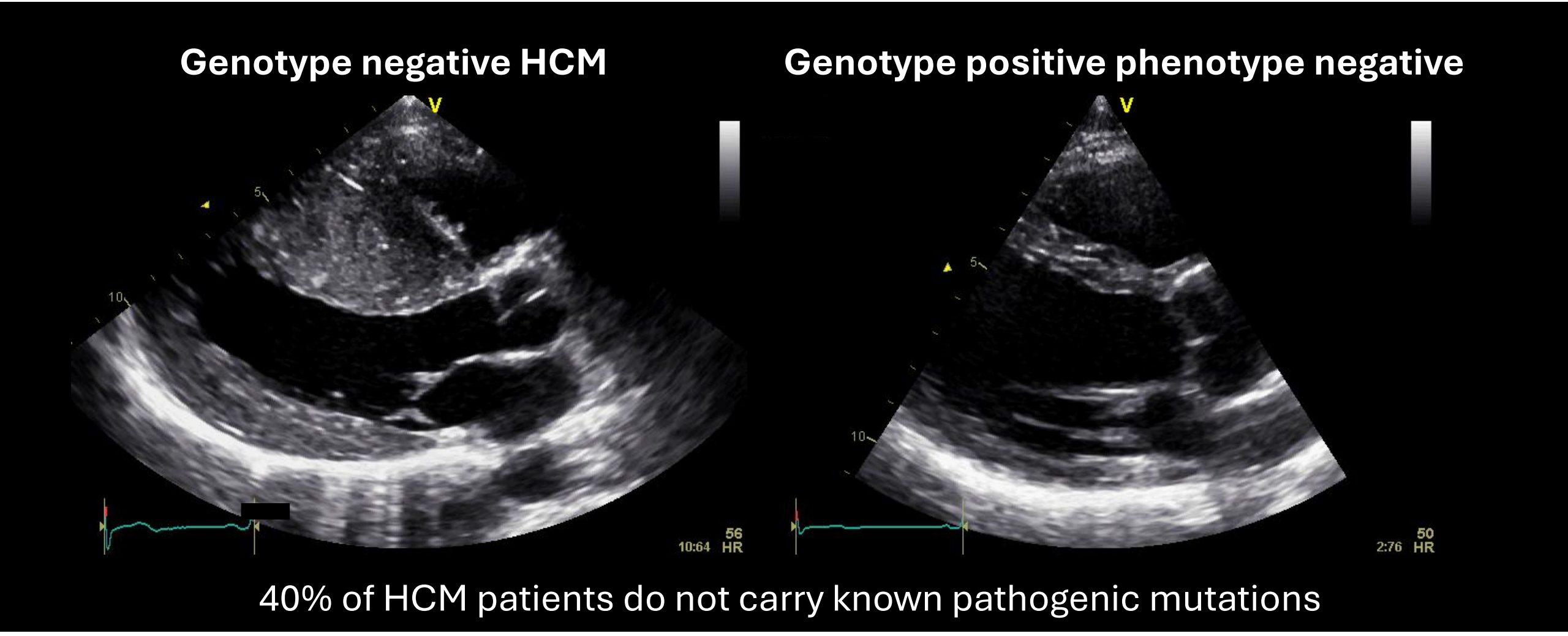




Exercise echocardiography



Genetic tests have limitations

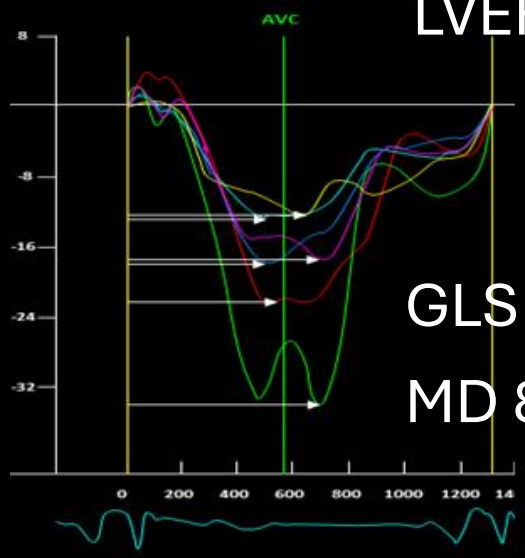


40% of HCM patients do not carry known pathogenic mutations

Strain imaging in HCM

Genotype positive HCM with mild phenotype (MWT 13 mm) and signs of systolic dysfunction

LVEF 60 %



GLS – 16 %

MD 88 ms

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

ESC GUIDELINES

2014 ESC Guidelines on diagnosis and management of hypertrophic cardiomyopathy

The Task Force for the Diagnosis and Management of Hypertrophic Cardiomyopathy of the European Society of Cardiology (ESC)

Authors/Task Force members: Perry M. Elliott* (Chairperson) (UK) Aris Anastasakis (Greece), Michael A. Borger (Germany), Martin Borggrefe (Germany), Franco Cecchi (Italy), Philippe Charron (France), Albert Alain Hagege (France), Antoine Lafont

European Heart Journal – Cardiovascular Imaging (2016) 17, 613–621
doi:10.1093/ehjci/jew005

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

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

- Strain imaging may facilitate diagnosis and management in early HCM disease
- Strain may be abnormal in mutation positive family members yet to develop typical HCM phenotype
- Link between abnormal strain, ventricular arrhythmias and fibrosis

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

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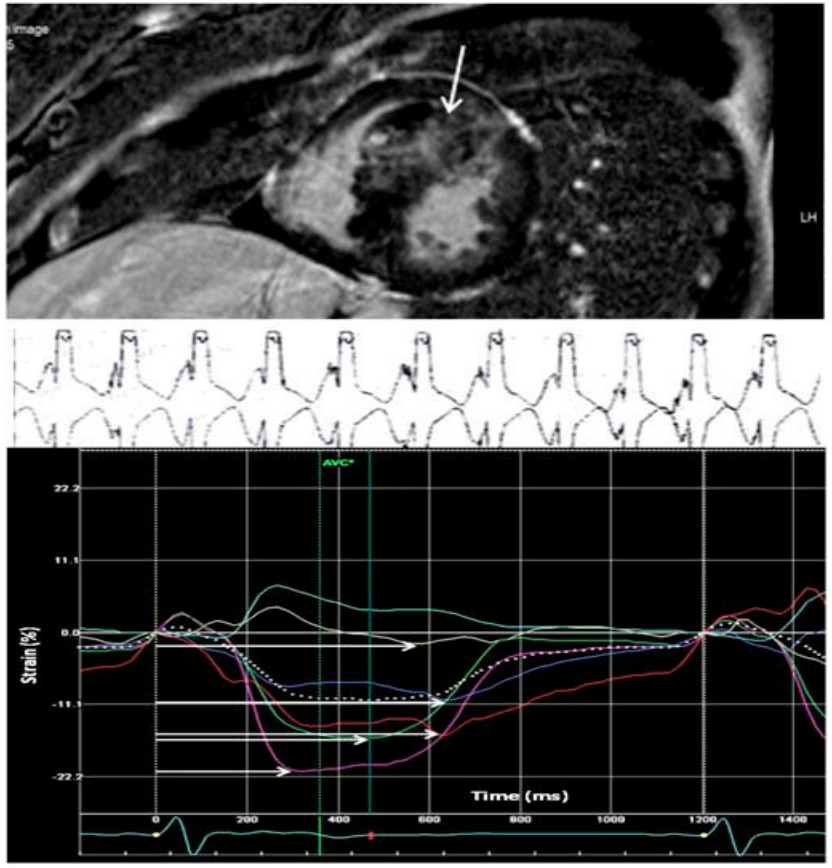

European Heart Journal – Cardiovascular Imaging
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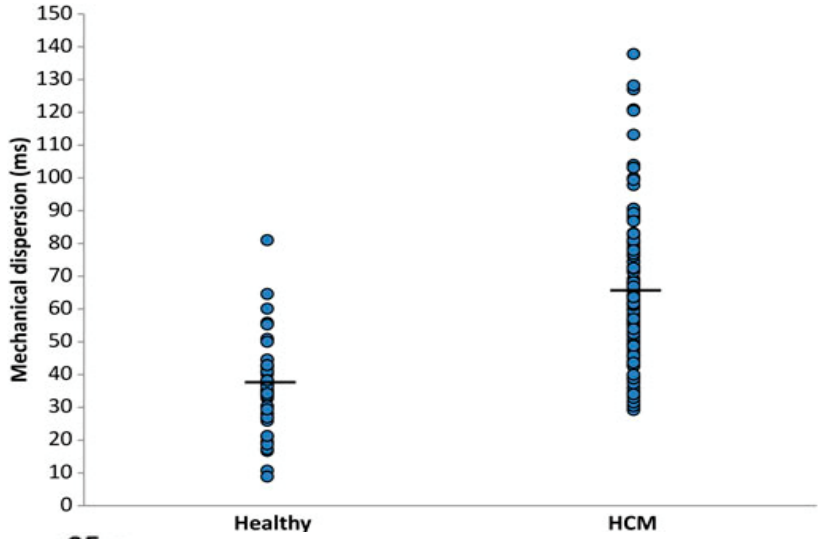
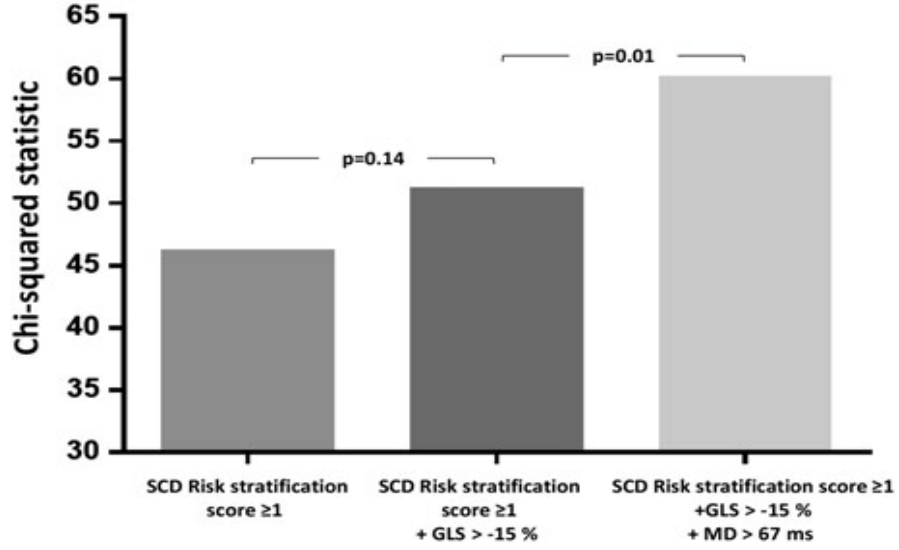
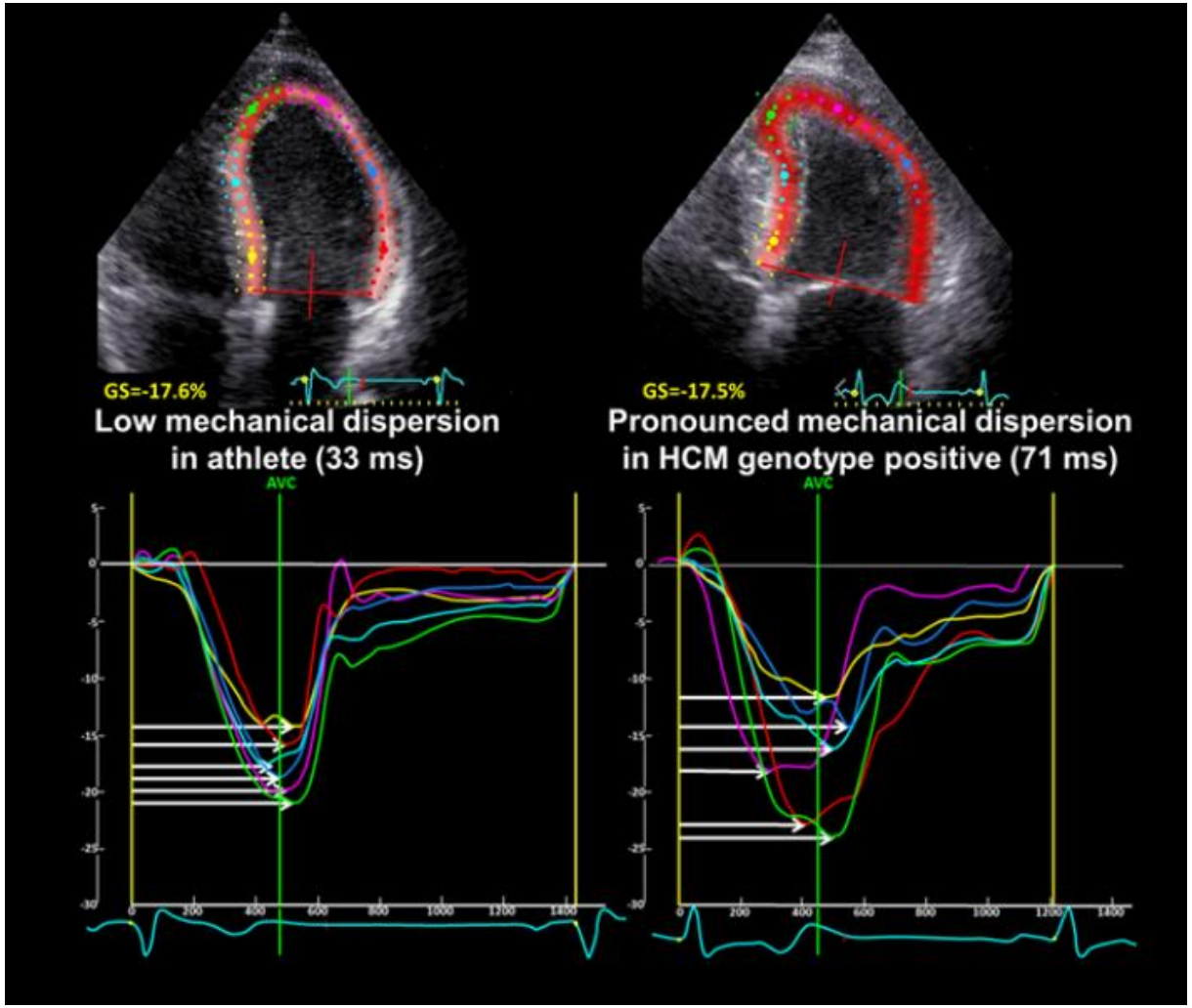
2016





Mechanical dispersion



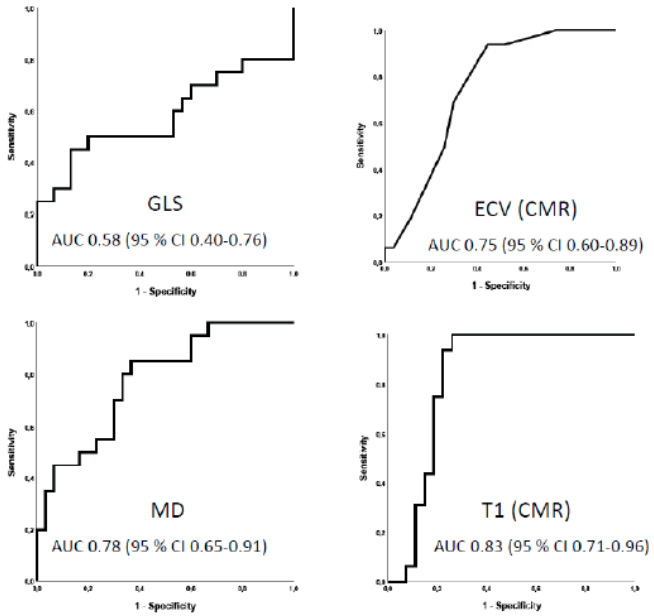


Figure 3. ROC curves of indices from strain echocardiography (**left** column) and T1 mapping techniques (**right** column). GLS > −17.7%, MD > 44 ms, T1 > 1230 ms and ECV > 22.5% were the optimal cut off values in the current dataset. CI = confidence interval. CMR = cardiac magnetic resonance imaging. ECV = extracellular volume. GLS = global longitudinal strain. MD = mechanical dispersion. T1 = native T1 time.

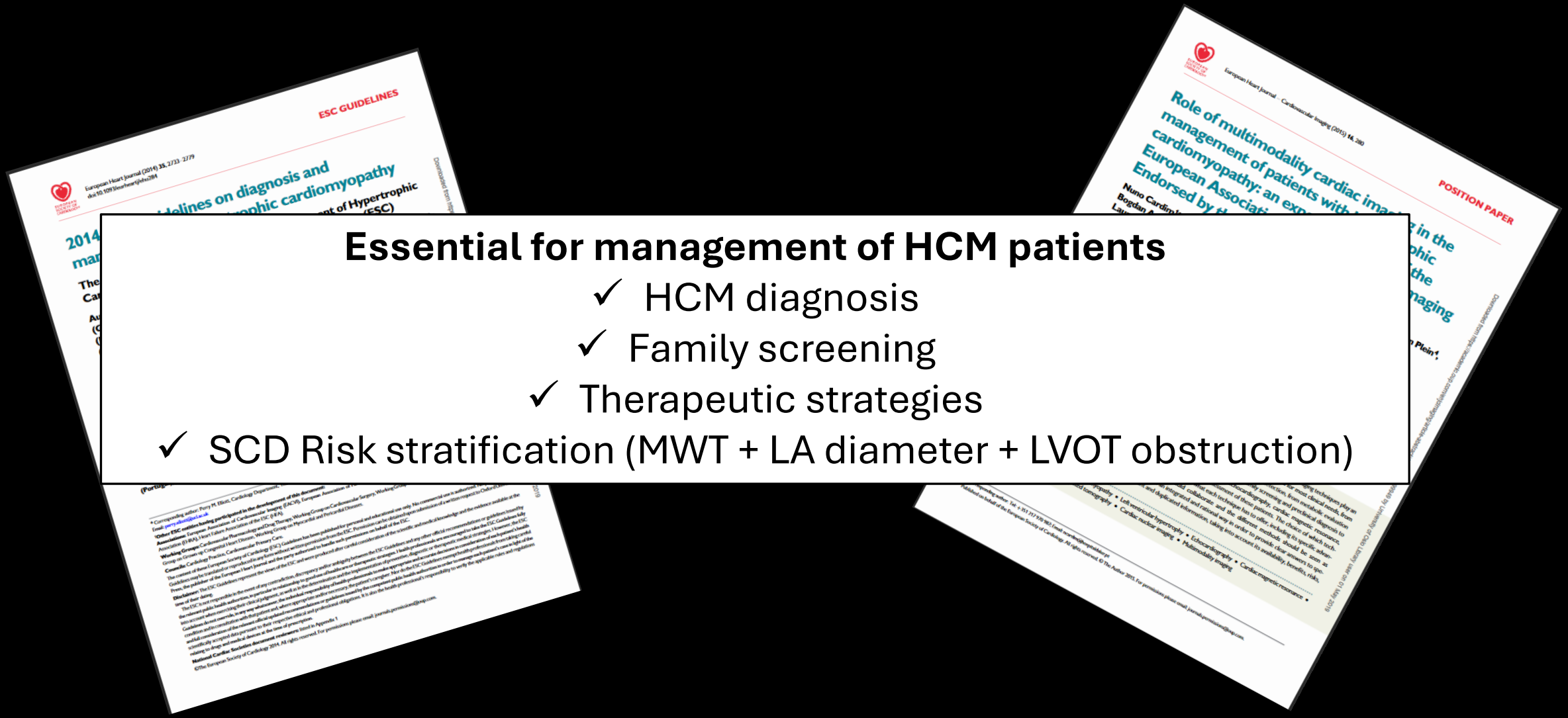
Table 2. Summary of results.

	Athletes	HCM	*p*	Optimal Cut Off	AUC	95% CI
Echocardiography						
Mechanical dispersion, ms	40 ± 11	54 ± 16	0.001	>44	0.78	0.65–0.91
Global longitudinal strain, %	−18.9 ± 1.8	−18.1 ± 3.7	0.28	>−17.7	0.58	0.40–0.76
CMR						
Extracellular volume, %	22.7 ± 3.2	25.6 ± 4.1	0.013	>22.5	0.75	0.60–0.89
Native T1 time, ms	1204 (1191, 1234)	1265 (1255, 1312)	<0.001	>1230	0.83	0.71–0.96

Values are mean ± SD or median with the IQR and were compared using Student's *t*-test or the Mann–Whitney U-test as appropriate. Indices from strain echocardiography and CMR and their ability to identify HCM mutation carriers from athletes in the study population were evaluated by means of ROC analysis. AUC = area under the curve. CI = confidence interval. CMR = cardiac magnetic resonance imaging. HCM = hypertrophic cardiomyopathy.

LG Klaeboe et al, 2024 Biomedicines

Echocardiography



Essential for management of HCM patients

- ✓ HCM diagnosis
- ✓ Family screening
- ✓ Therapeutic strategies
- ✓ SCD Risk stratification (MWT + LA diameter + LVOT obstruction)

Imaging strategy & follow-up

- Echocardiography
  - Every 1-2 years in clinical stable patients
- CMR
  - At least once (at the initial evaluation)
  - Repeated according to potential changes in clinical status
  - Specific clinical questions and problems.

Summary

- Cardiac Imaging – central role in cardiac phenotyping and disease monitoring in HCM
- Echocardiography – main imaging tool
  - from initial diagnosis to follow-up / widespread availability
  - Exercise echo – challenging
  - EF is preserved in HCM although systolic function is severely depressed
  - advanced techniques – GLS and MD
- CMR
  - allows tissue characterization
- Combining echo and CMR
  - facilitates risk stratification, particularly sudden cardiac death