Imaging Patients with Hypertrophic Cardiomyopathy

Sherif F. Nagueh, MD, FACC, FASE, FAHA

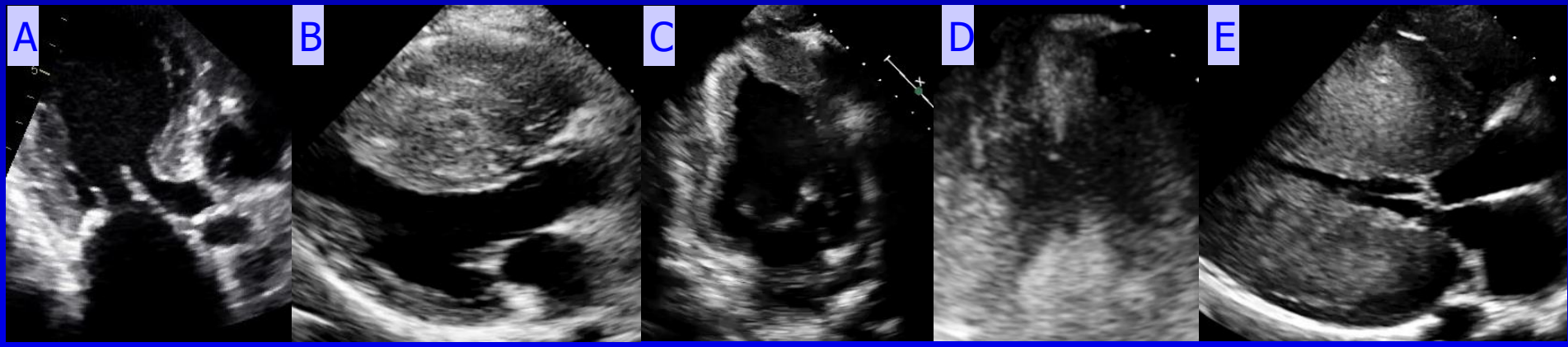
Methodist DeBakey Heart and Vascular Center

The Methodist Hospital

Houston, Texas

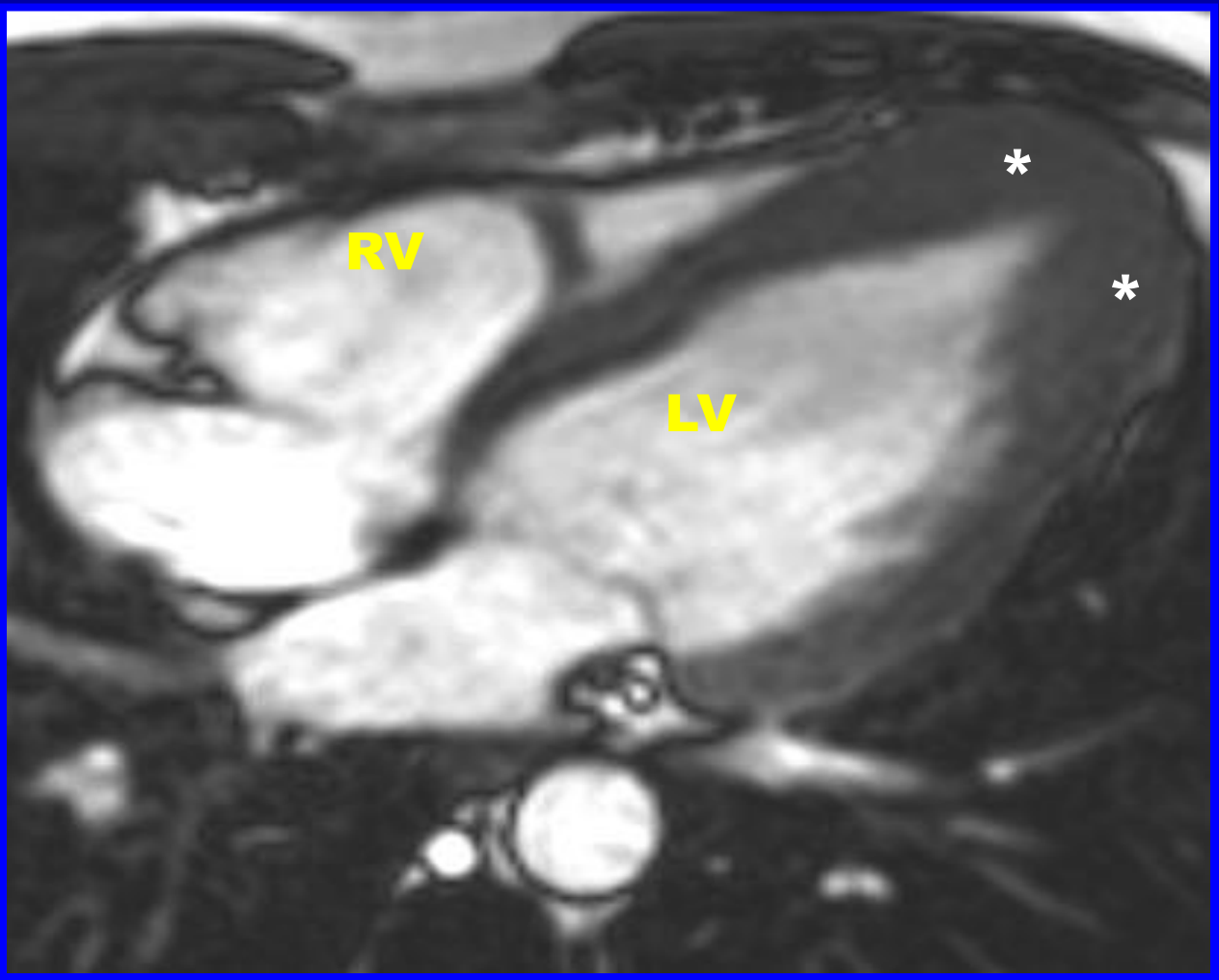
Morphology

Hypertrophy Patterns in HCM

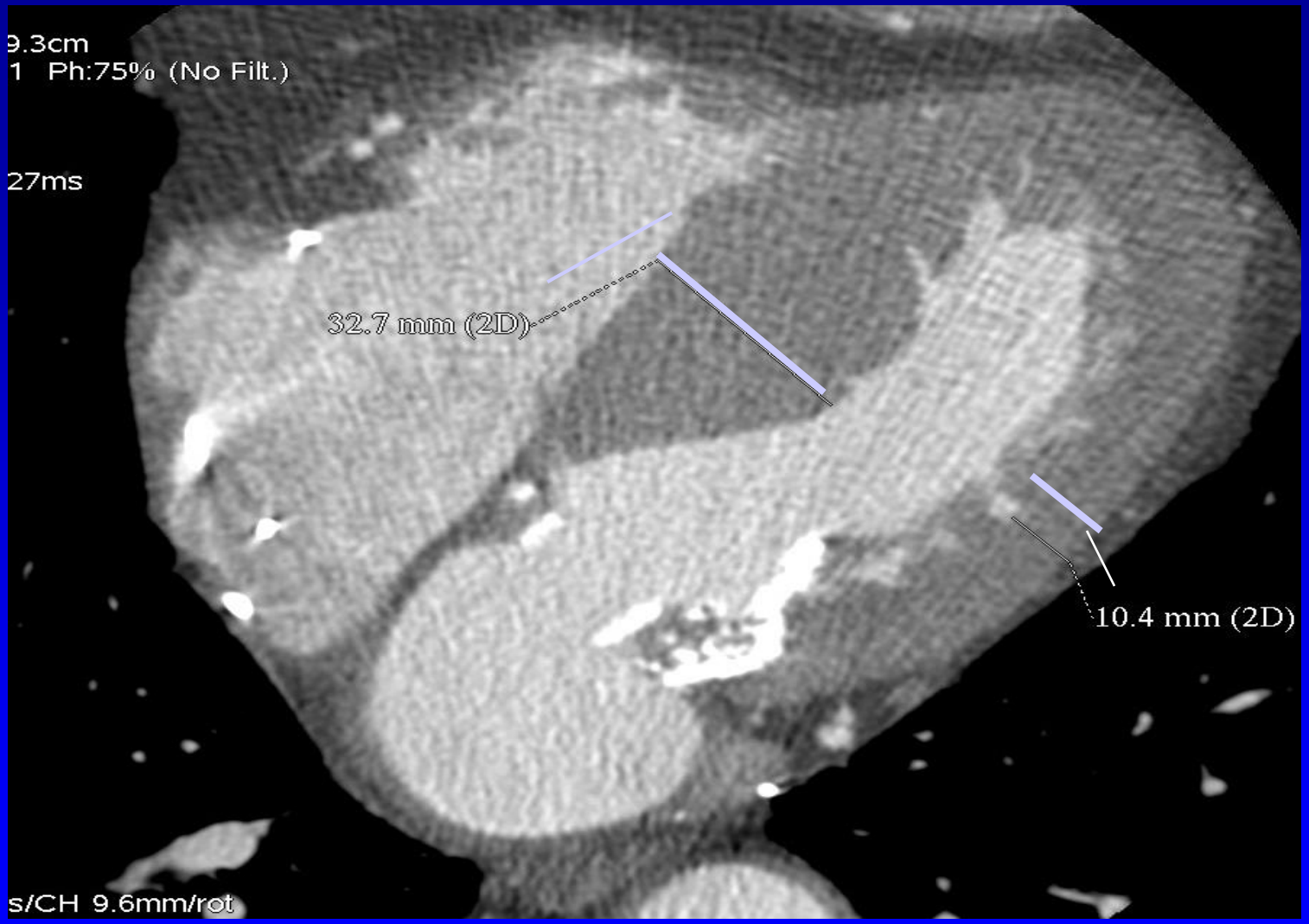


Nagueh et al *JASE* 2022;35:533 - 569

Apical HCM Visualized by CMR



Asymmetric Hypertrophy by Cardiac CT

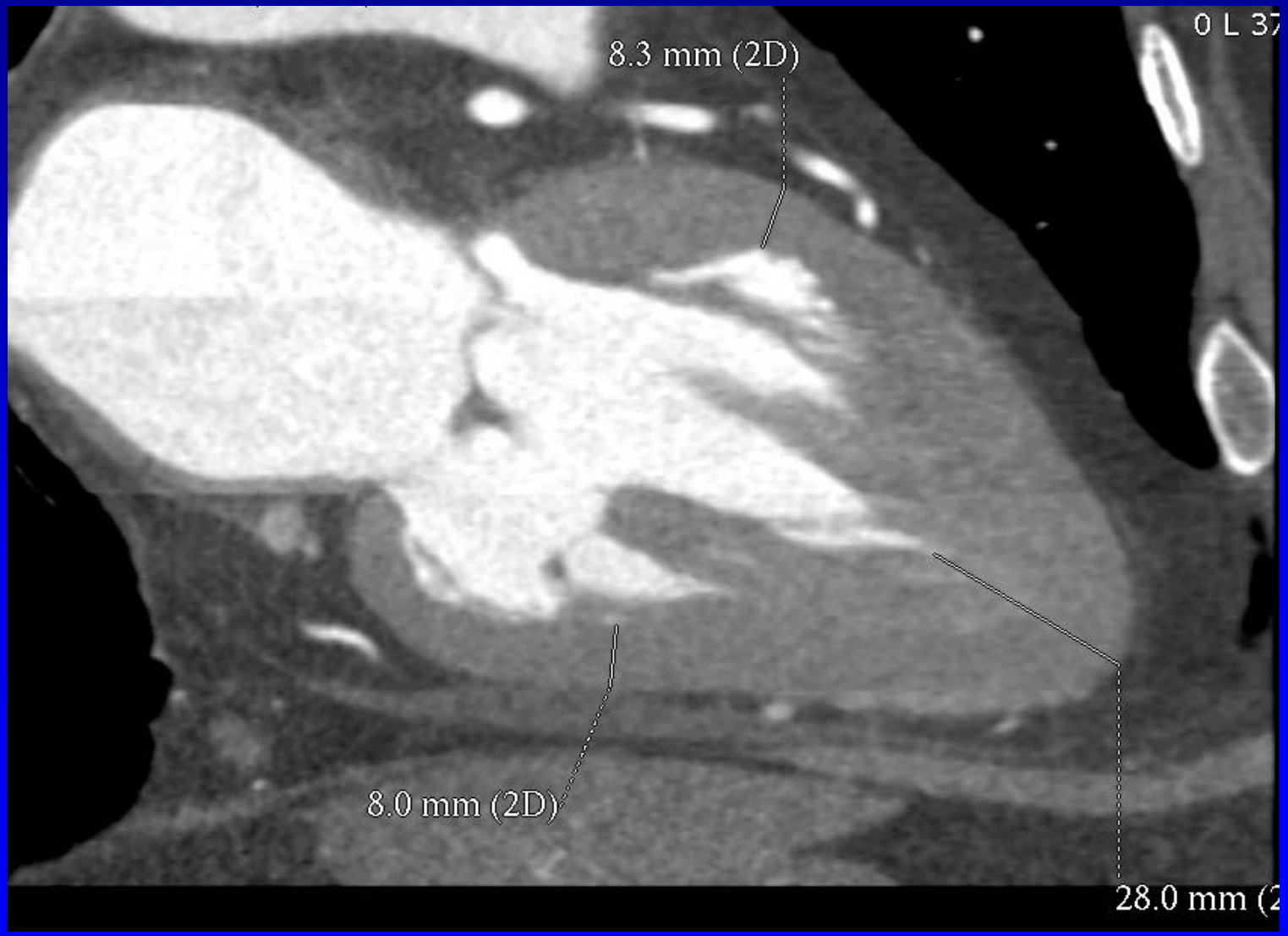


Nagueh et al *JASE* 2011;24:473 - 498

Asymmetric Hypertrophy by Cardiac CT

Nagueh et al *JASE* 2011;24:473 - 498

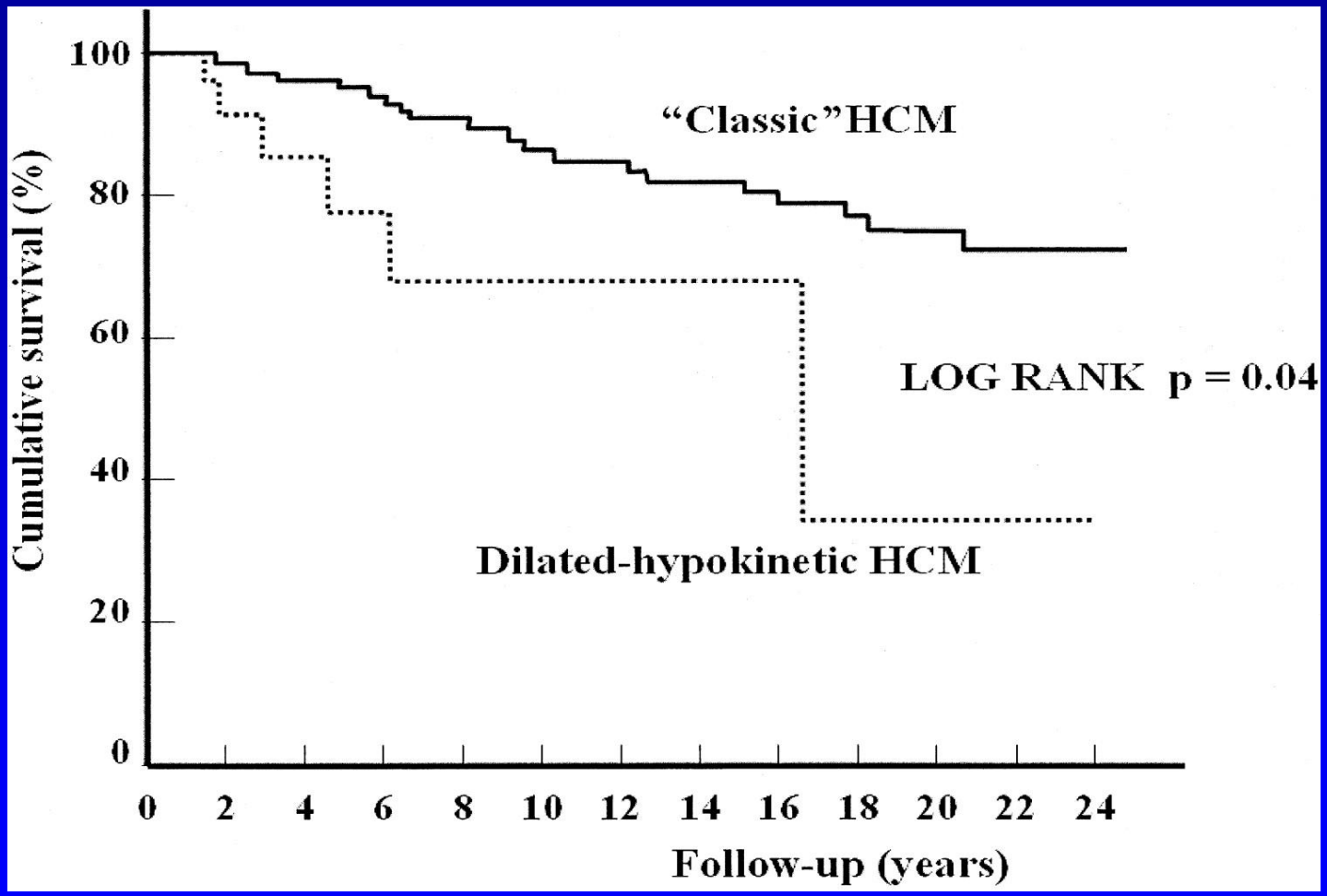
Apical Hypertrophy by Cardiac CT



Nagueh et al *JASE* 2011;24:473 - 498

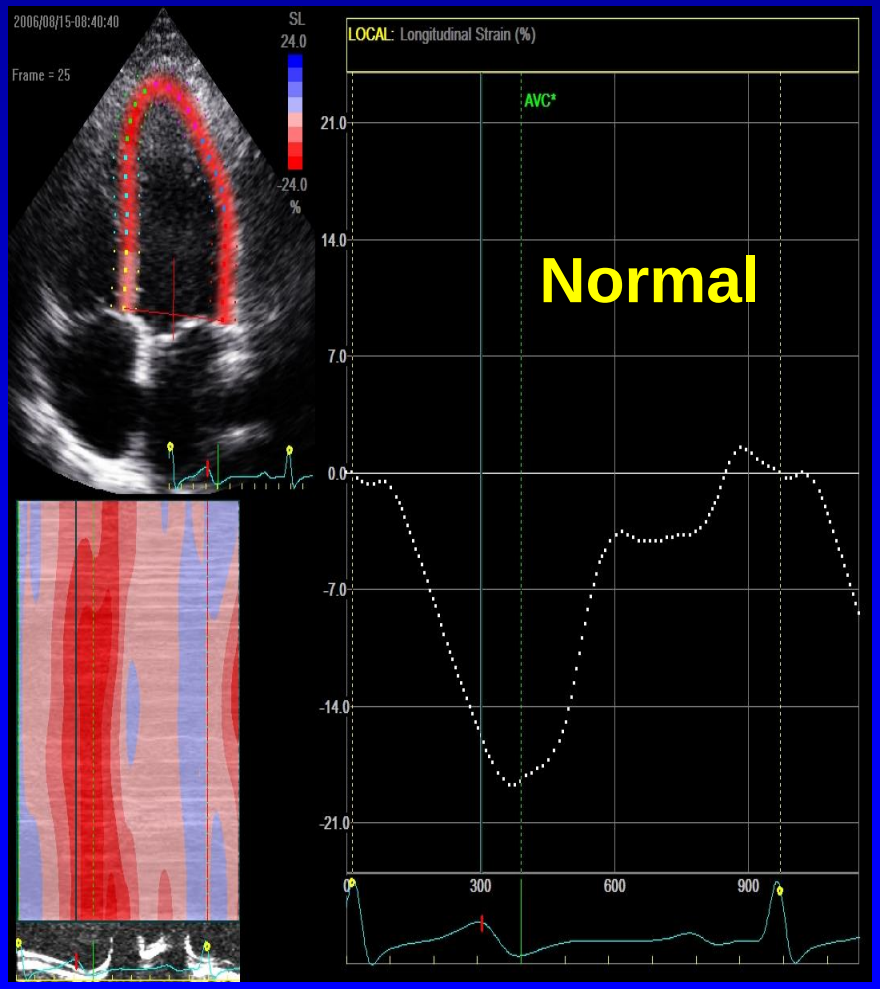
Systolic Function

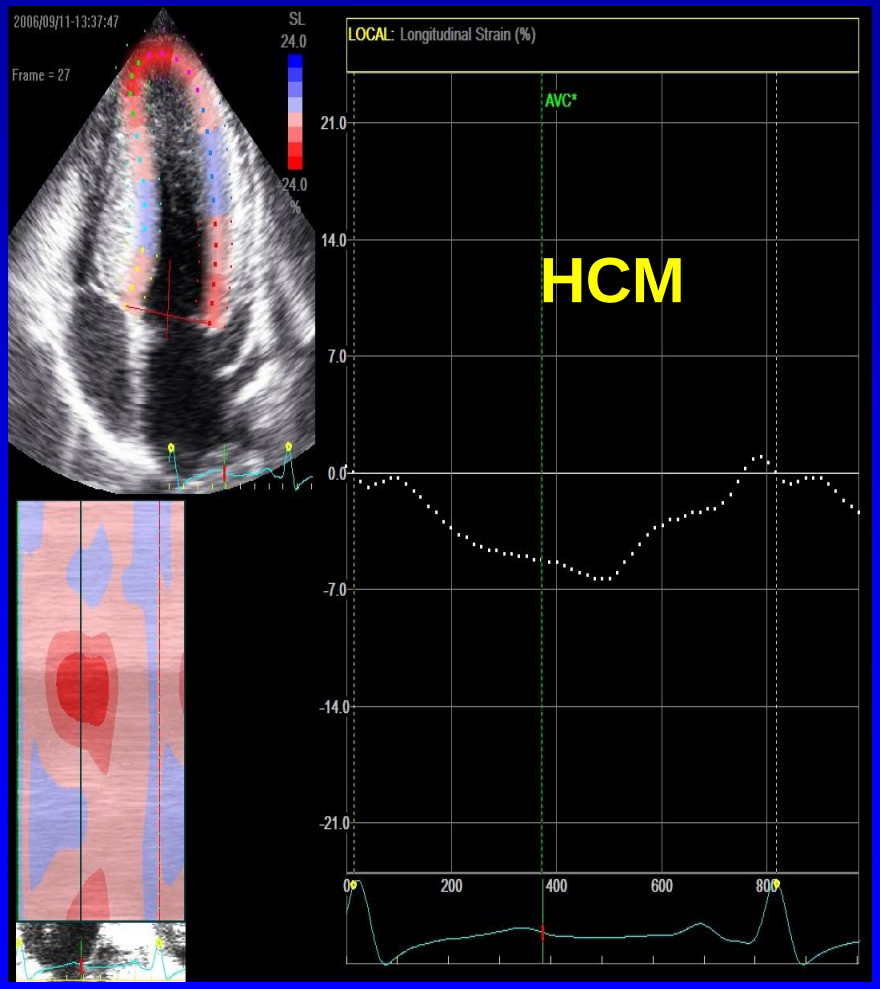
LV EF and Survival in HCM



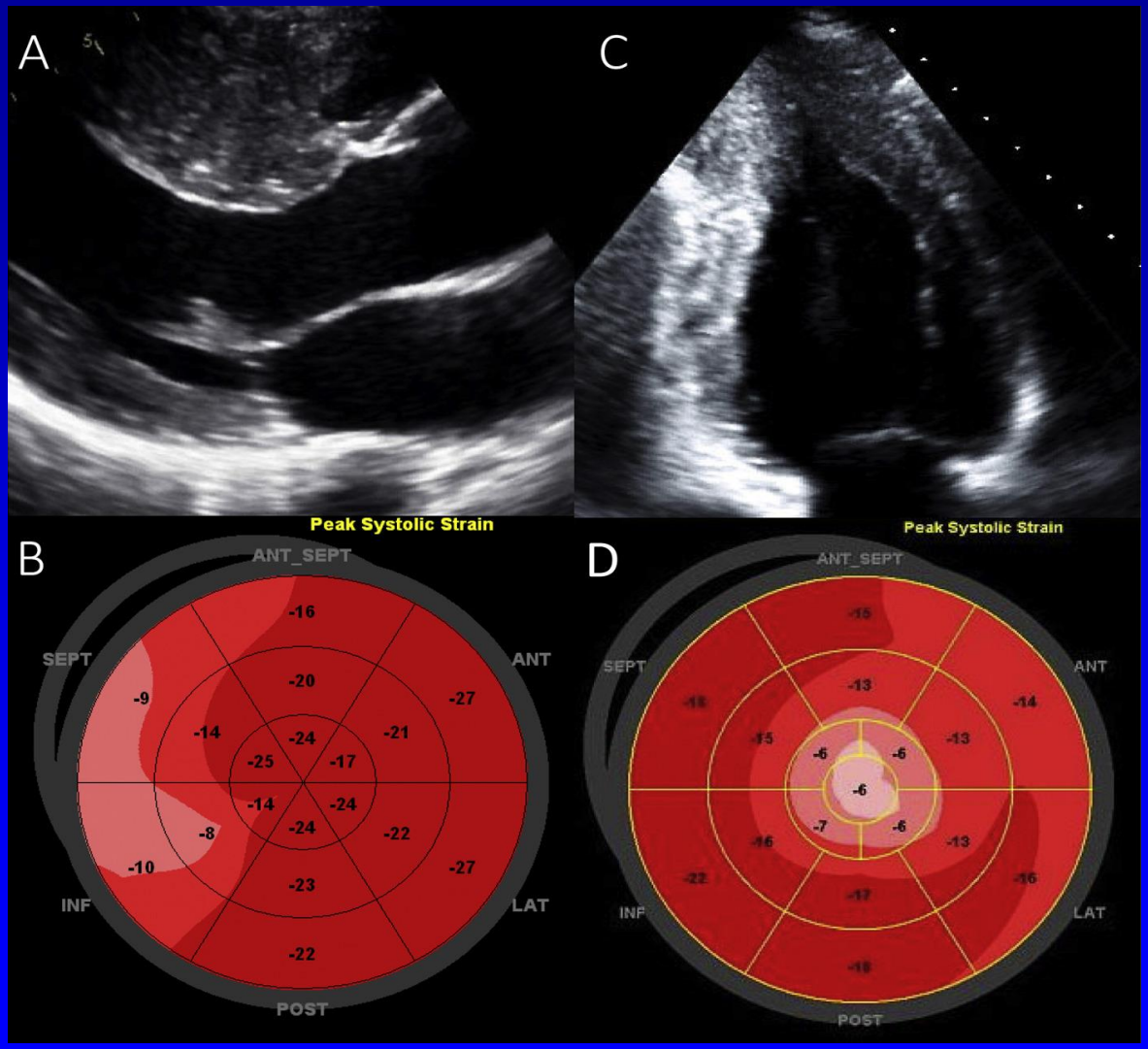
Biagini et al. J Am Coll Cardiol 2005;46:1543

Global Longitudinal Strain in HCM





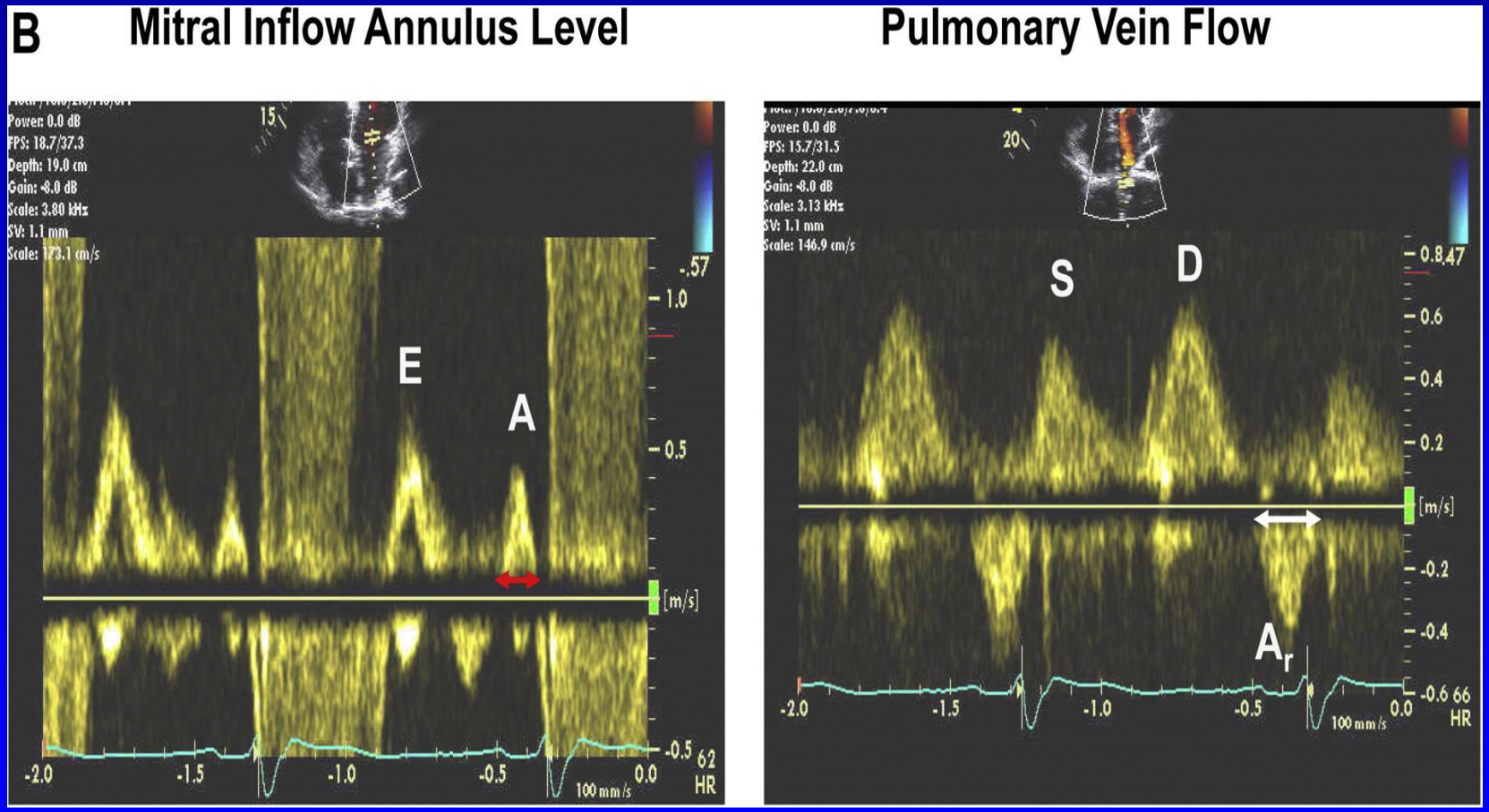
Strain Polar Map in HCM



Nagueh et al *JASE* 2022;35:533 - 569

Diastolic Function

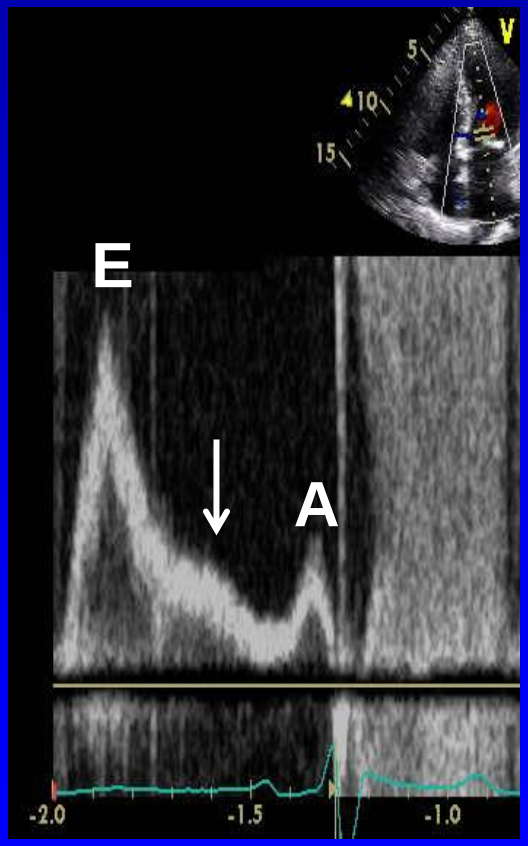
Pulmonary Venous Flow with Increased LV EDP in HCM



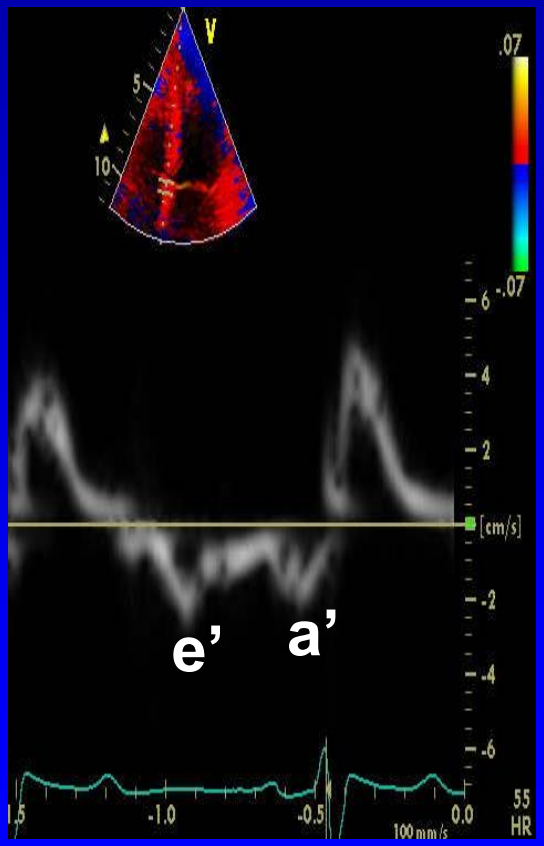
Nagueh et al *JASE* 2022;35:533 - 569

HCM with Restrictive LV Filling

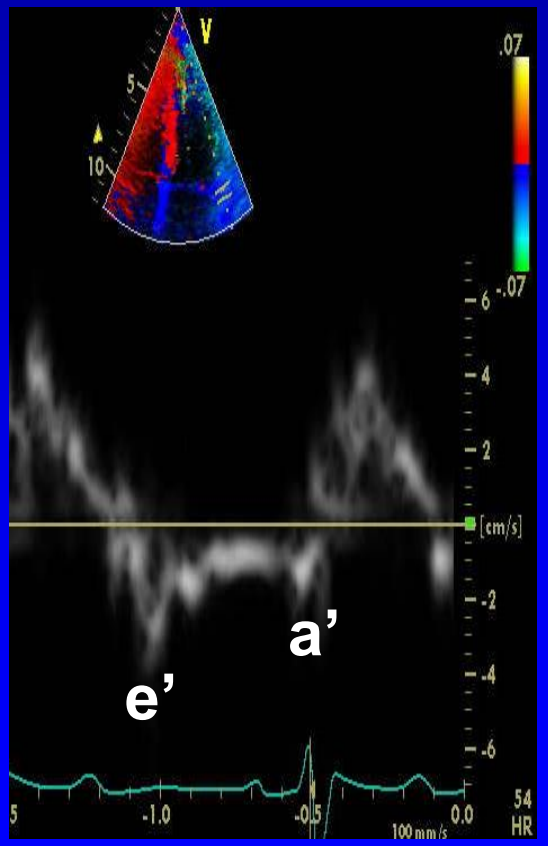
Mitral Inflow



Septal TD

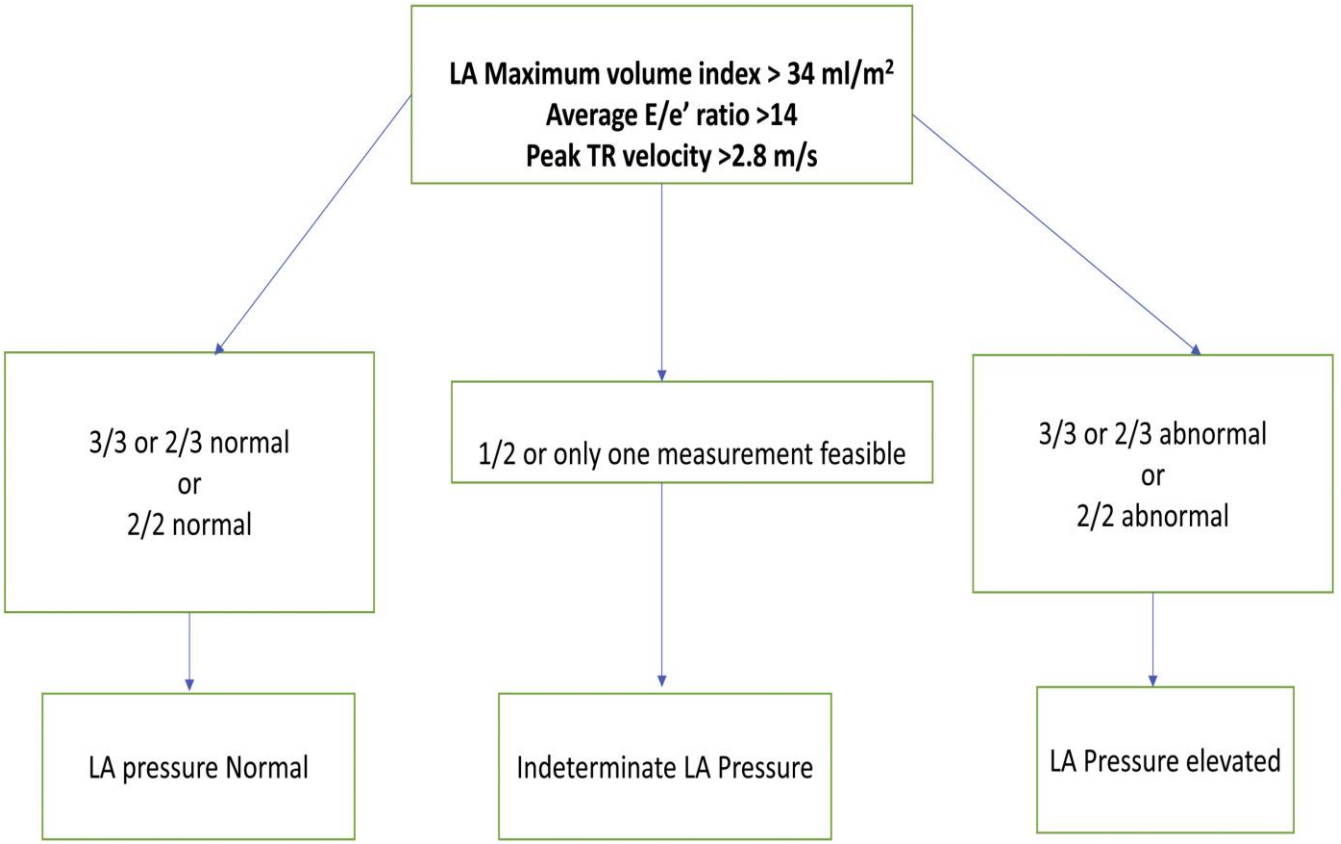


Lateral TD



Nagueh et al *JASE* 2011;24:473 - 498

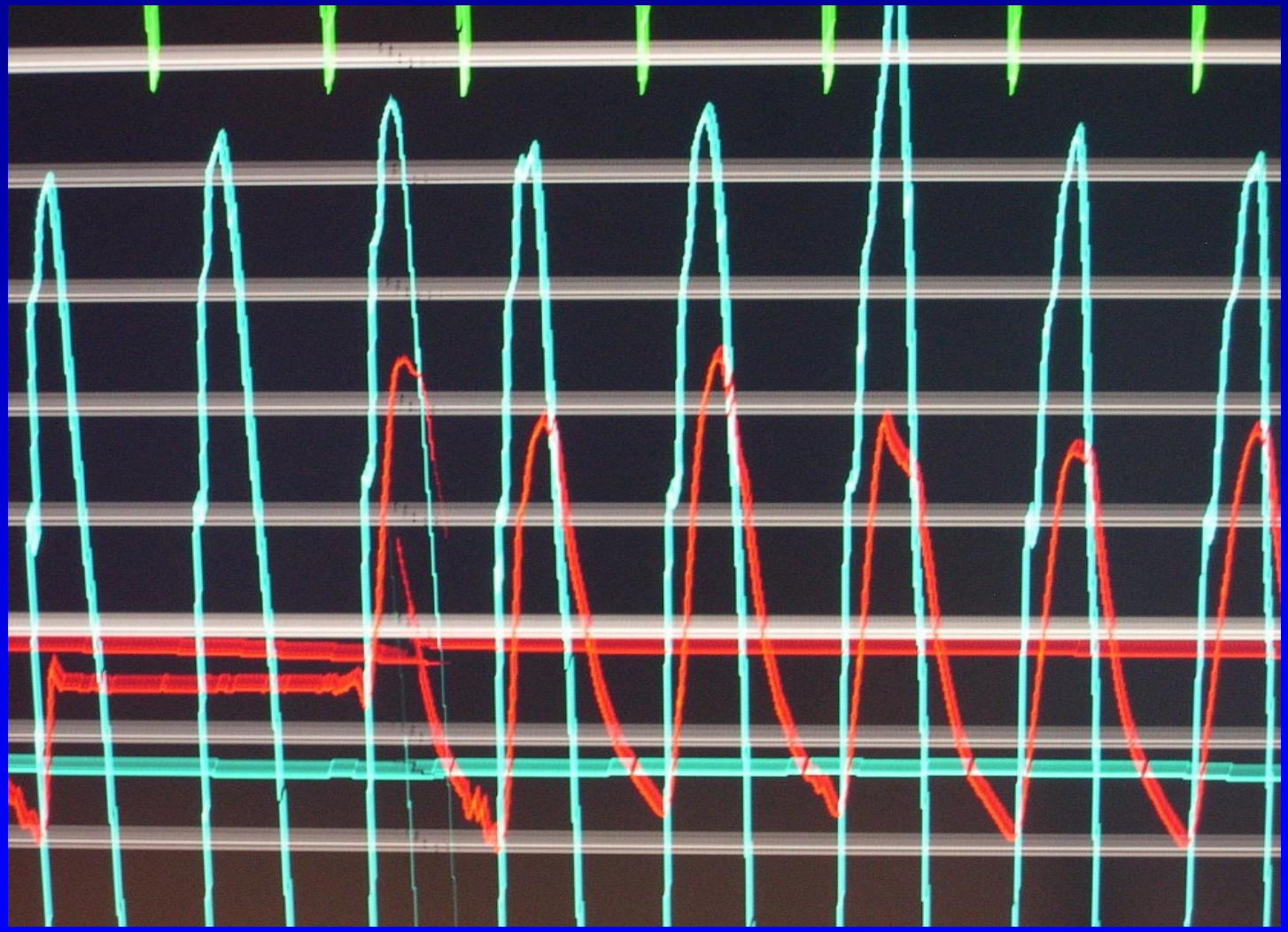
Algorithm for LAP Estimation in HCM



Nagueh et al *JASE* 2022;35:533 - 569

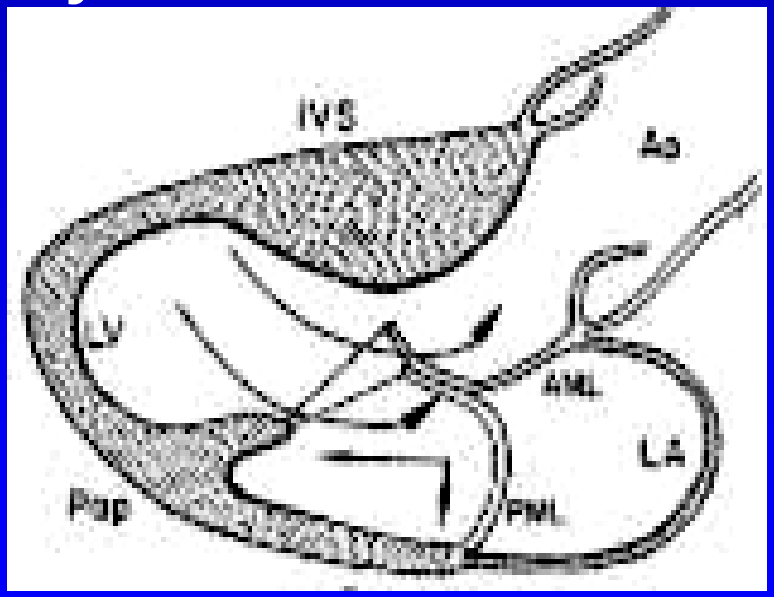
Dynamic Obstruction

LVOT Obstruction



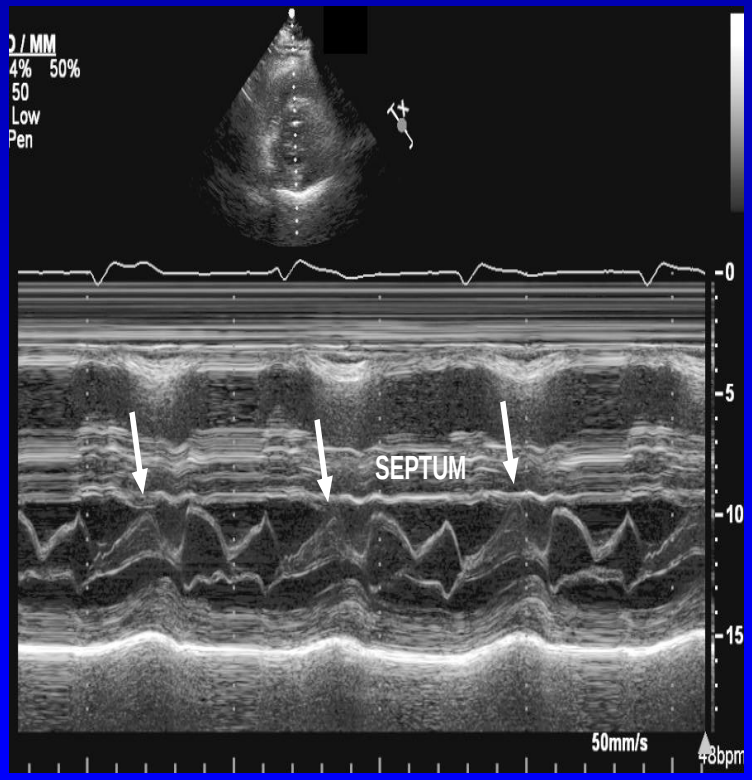
HYPERTROPHIC CARDIOMYOPATHY
MECHANISM OF SAM

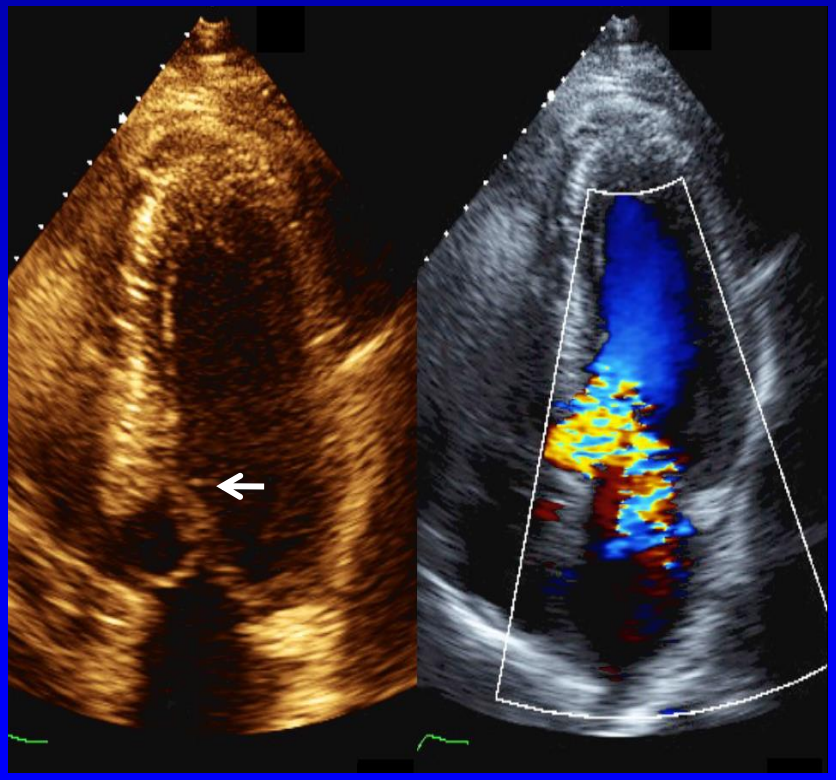
1. Anterior displacement of papillary muscles
2. Mitral leaflet elongation (relative to LV size)
3. Reduced posterior leaflet mobility
4. Curvature of septum
5. Hyperdynamic LV contraction



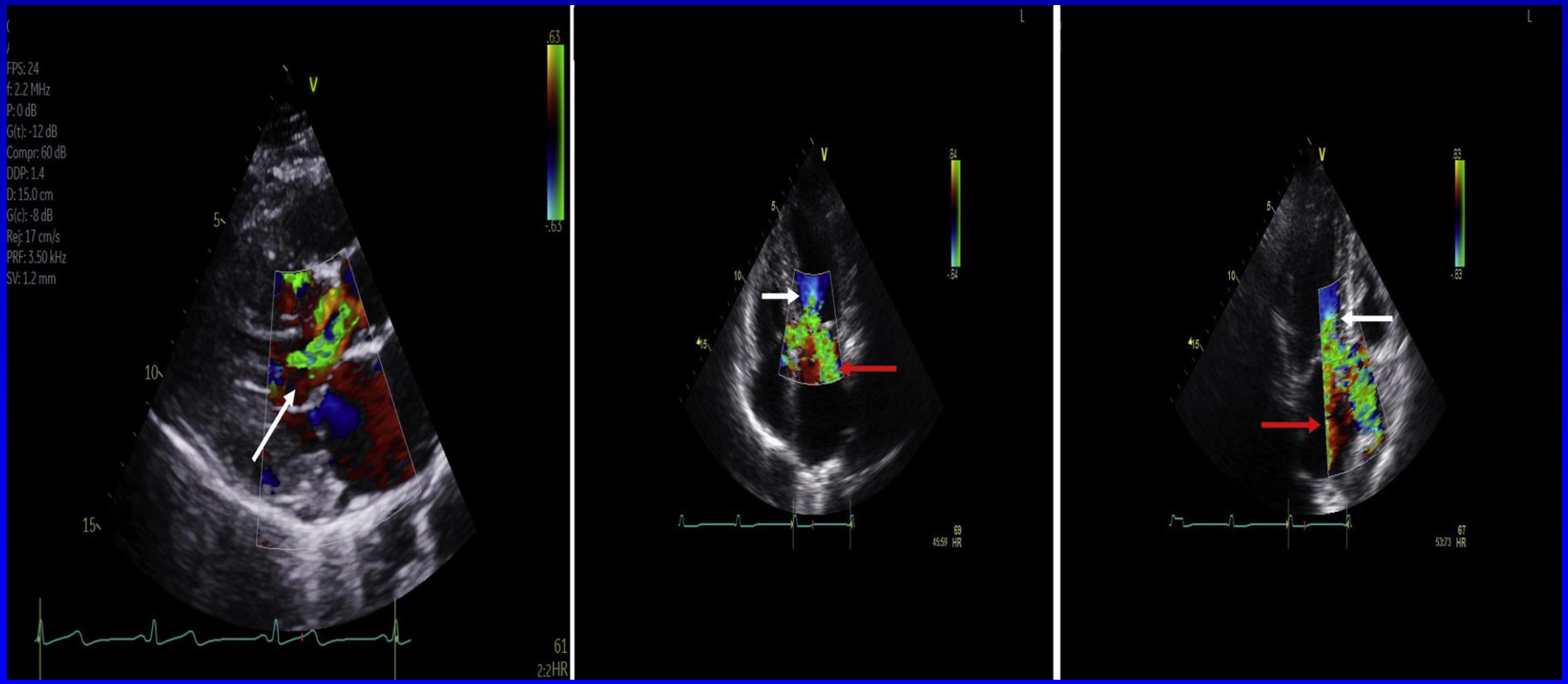
- Degree of SAM does not relate with severity of hypertrophy

HYPERTROPHIC CARDIOMYOPATHY
DYNAMIC LVOT OBSTRUCTION



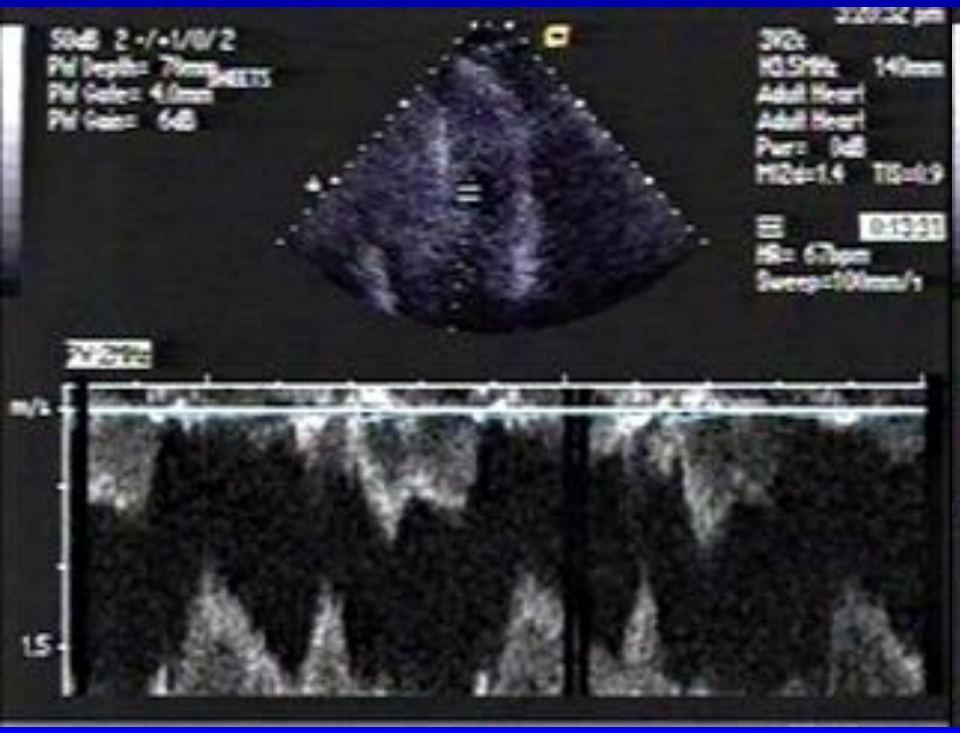


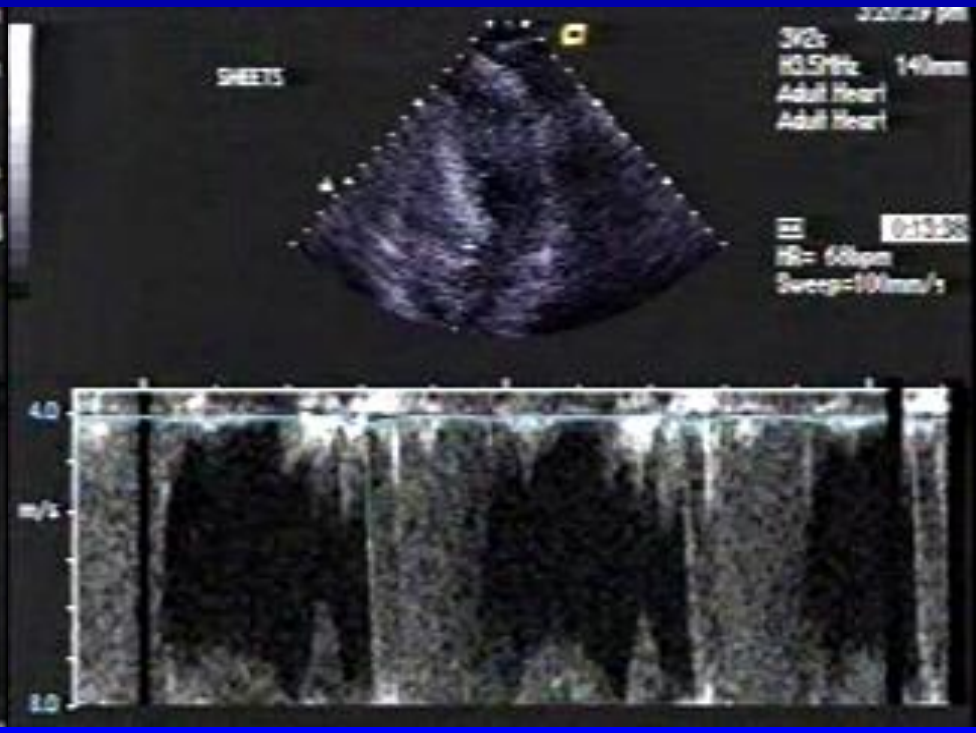
Color Doppler Showing Dynamic Obstruction in HCM



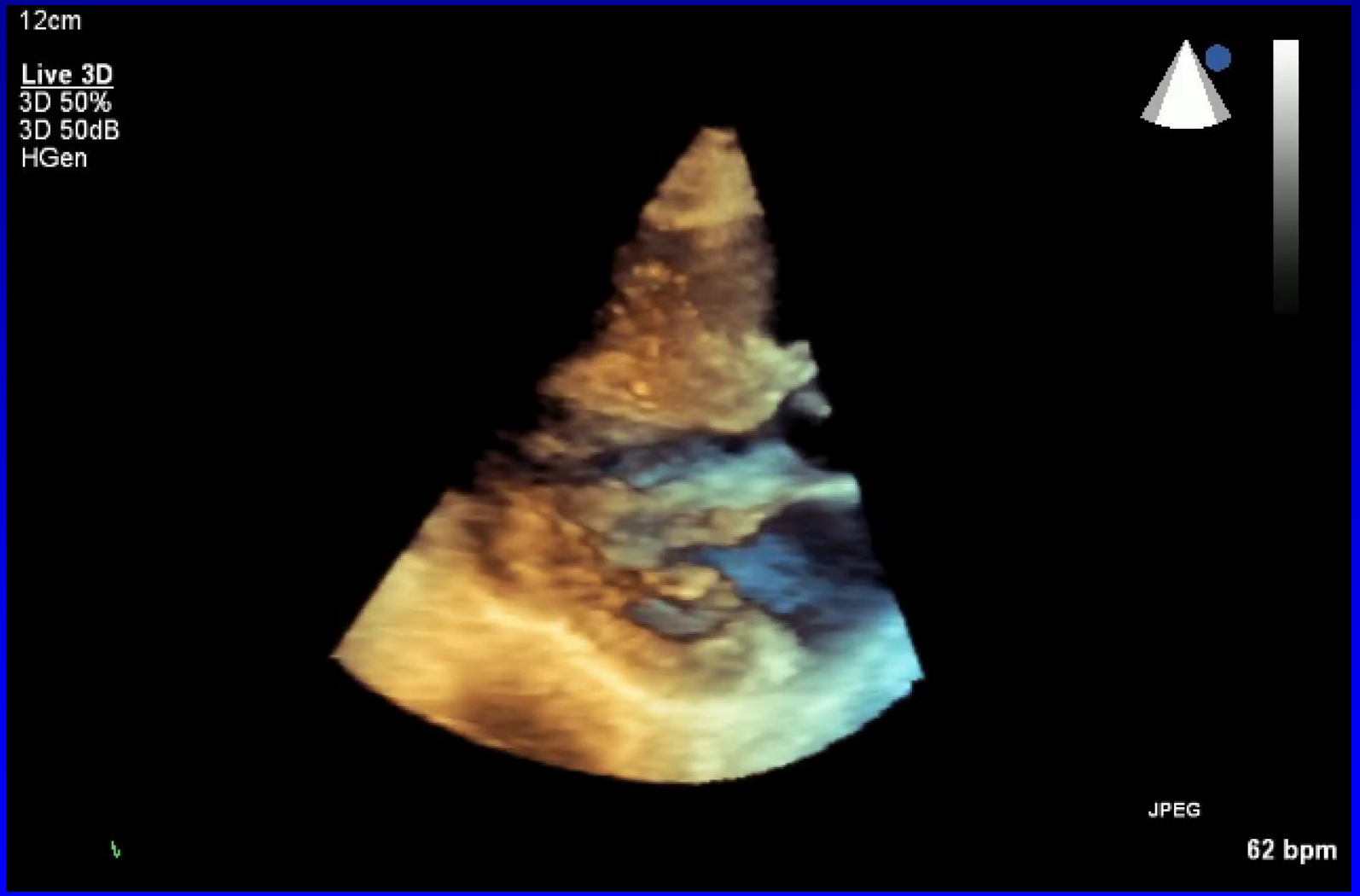
Nagueh et al *JASE* 2022;35:533 - 569

HYPERTROPHIC CARDIOMYOPATHY
DYNAMIC LVOT OBSTRUCTION





Dynamic Obstruction by 3 D in HCM

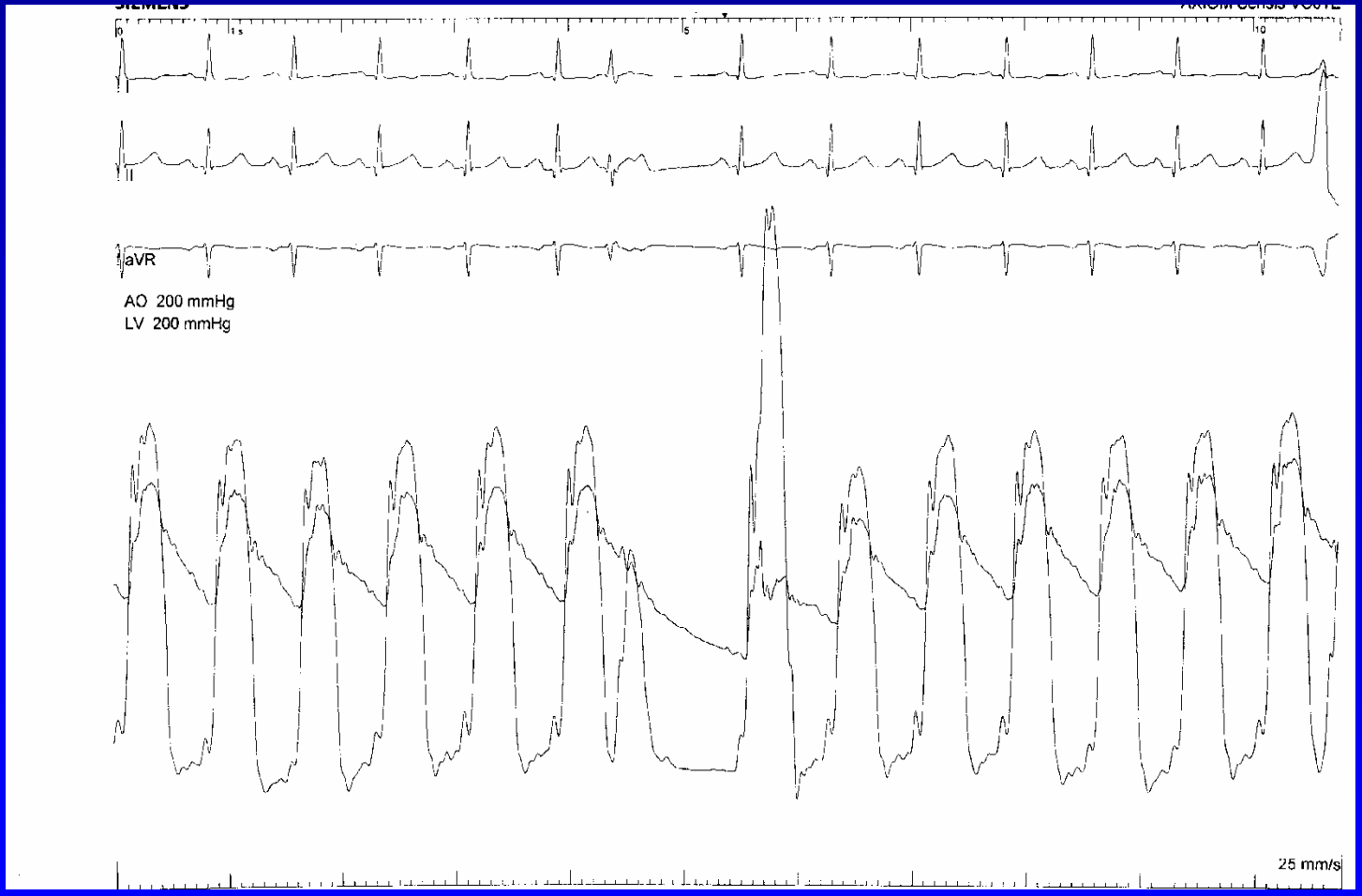


Nagueh et al *JASE* 2022;35:533 - 569

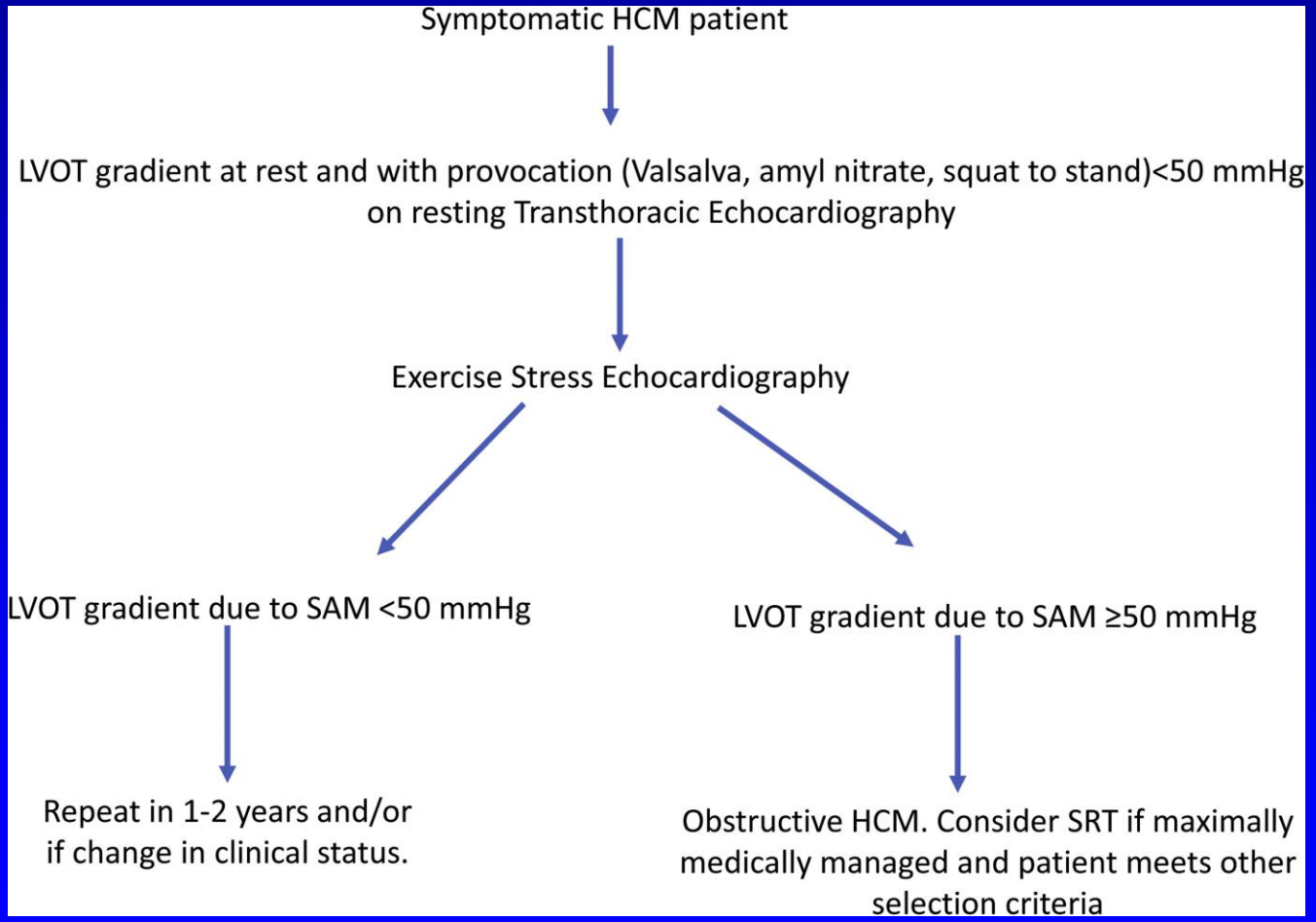
PROVOCABLE LVOT OBSTRUCTION

- Exercise
- Amyl nitrite
- Valsalva
- Squat to stand
- Isoproternol / dobutamine
- PVC

Post PVC Gradient



Algorithm for Provoking Dynamic Obstruction in HCM Patients



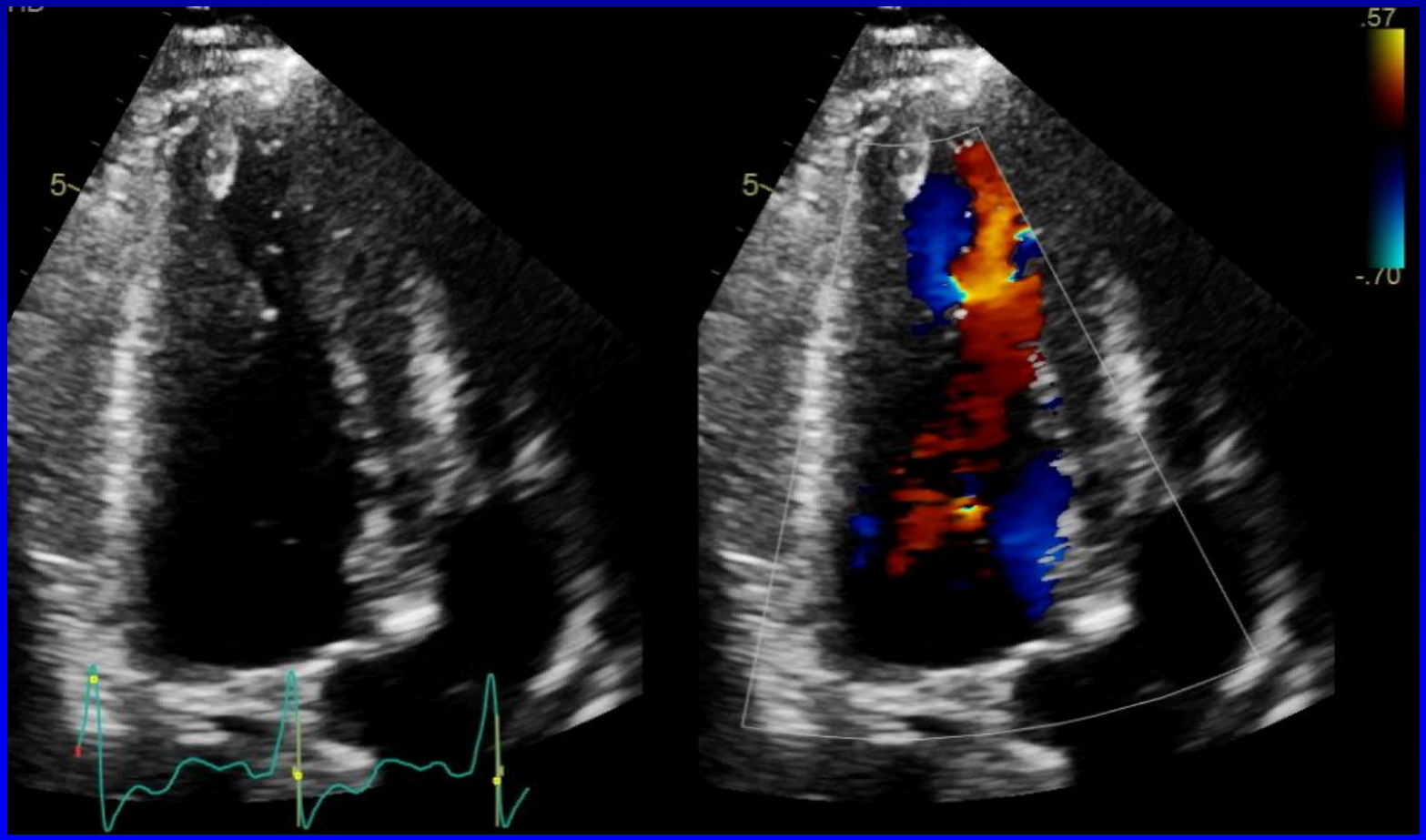
Nagueh et al *JASE* 2022;35:533 - 569

Mid-Ventricular Obstruction and Apical Aneurysm

MVO and Apical Aneurysms :Key Points

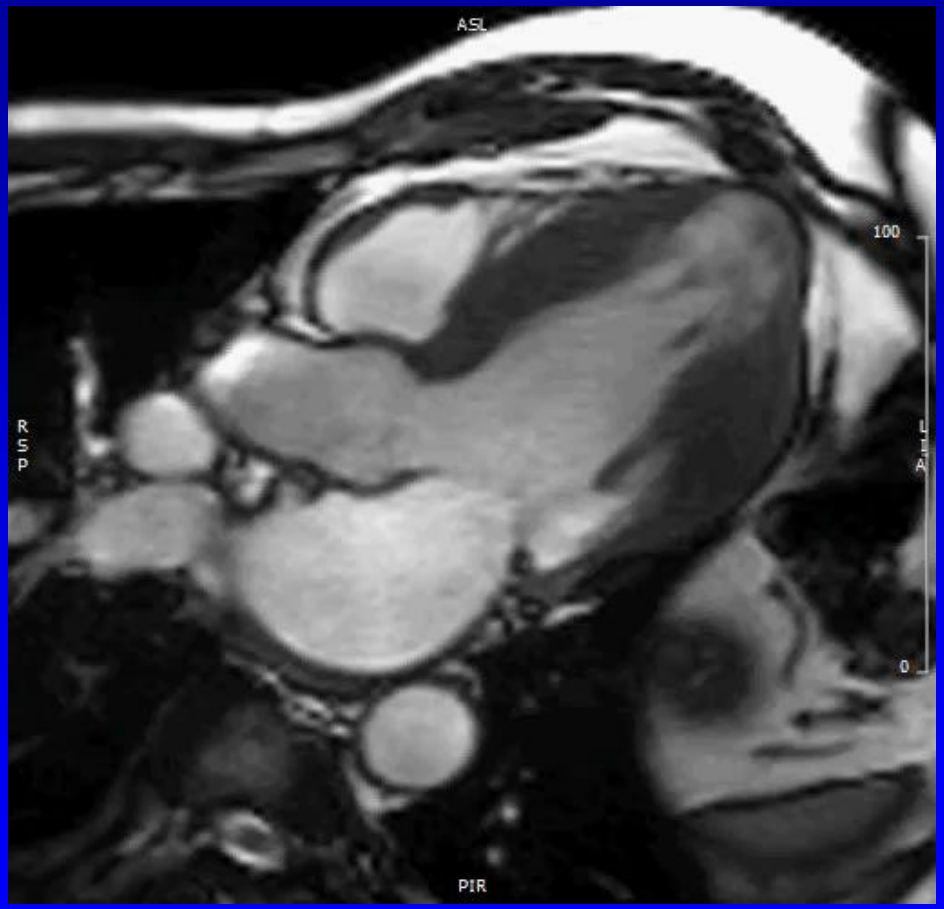
- Noticed in 3% of patients in recent registry
- Mid cavity obliteration with gradient ≥30 mmHg
- Echo Doppler with UEA and CMR helpful in diagnosis
- Portends higher risk for mortality and ventricular arrhythmias

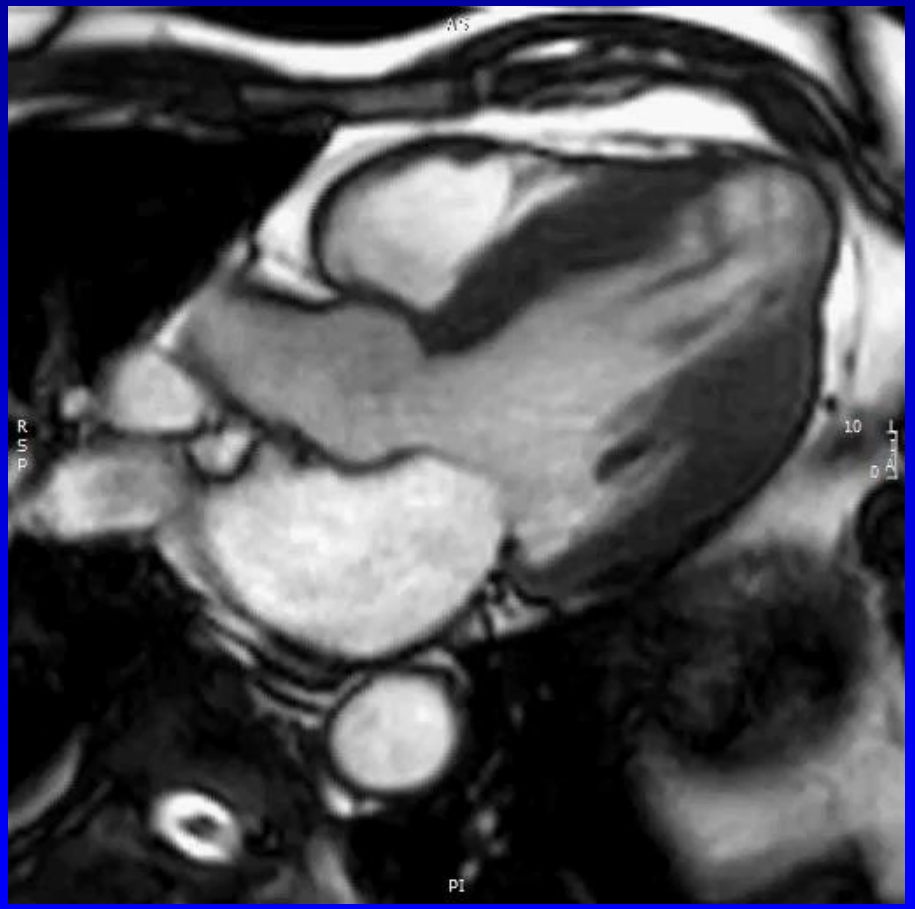
Echocardiography for MVO and Apical Aneurysm



Nagueh et al *JASE* 2022;35:533 - 569

CMR for MVO and Apical Aneurysm





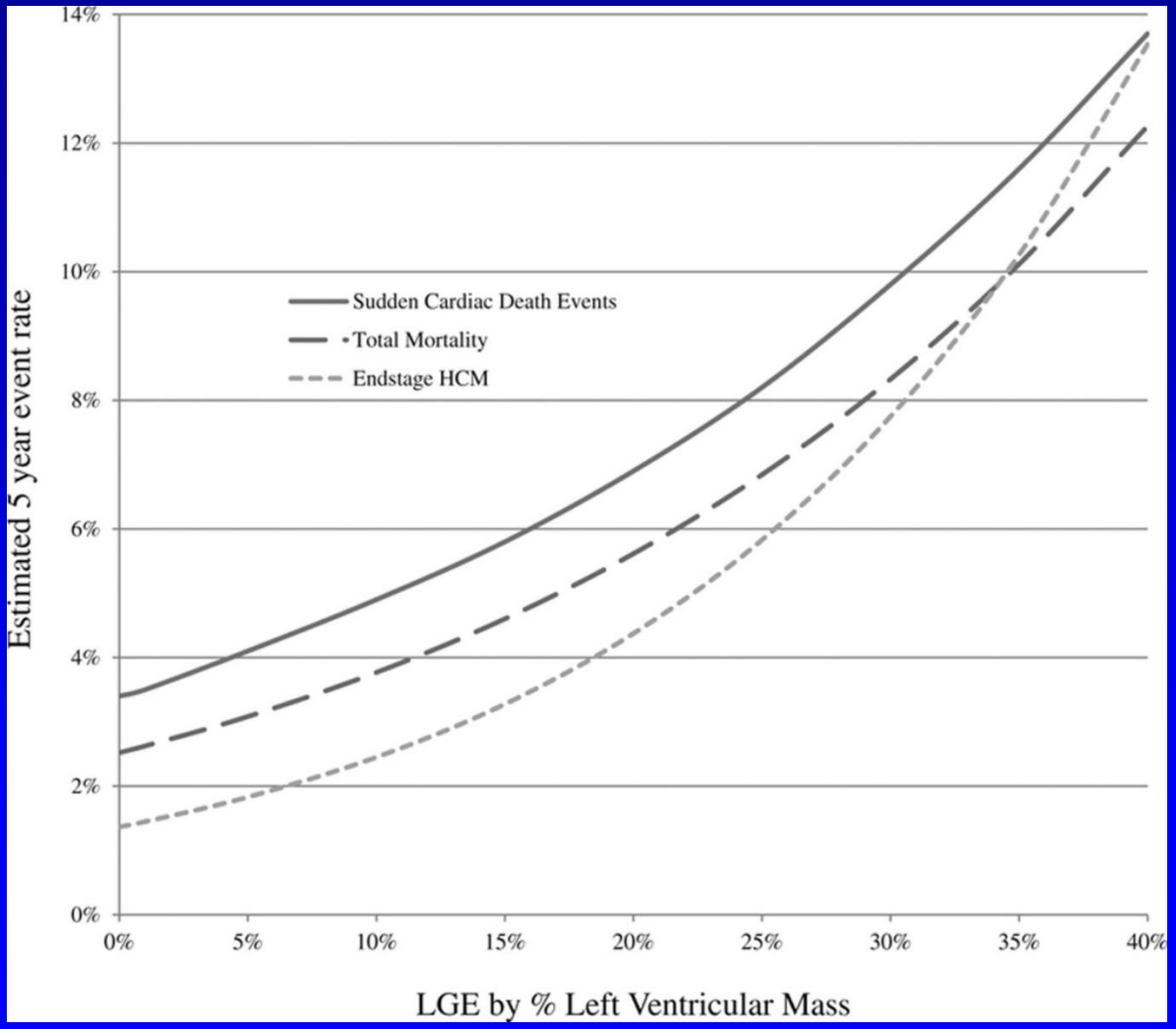
Nagueh et al *JASE* 2022;35:533 - 569

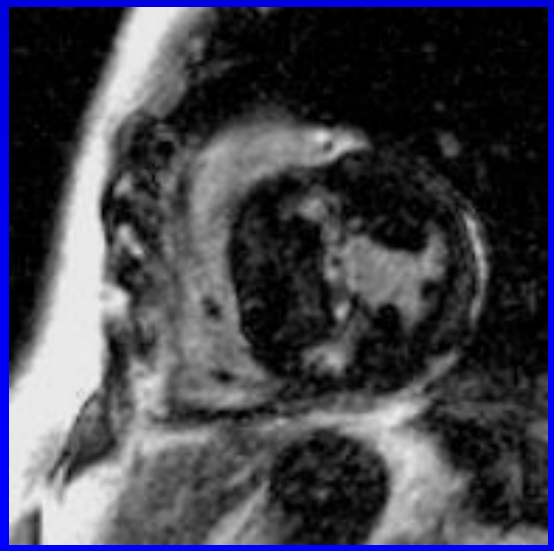
Imaging Role in Risk Stratification for SCD in HCM

Imaging Markers of Higher SCD Risk

- Maximum wall thickness ≥30 mm (consider ≥ 28 mm)
- Apical aneurysm (event rate at 5-15%/ year)
- LGE >15% of LV mass
- LA AP diameter in HCM Risk-SCD calculator
- LVOT obstruction (rest or Valsalva) in HCM-Risk-SCD calculator
- LV EF <50%
- Ischemia + blood flow reserve and stress myocardial blood flow heterogeneity by PET

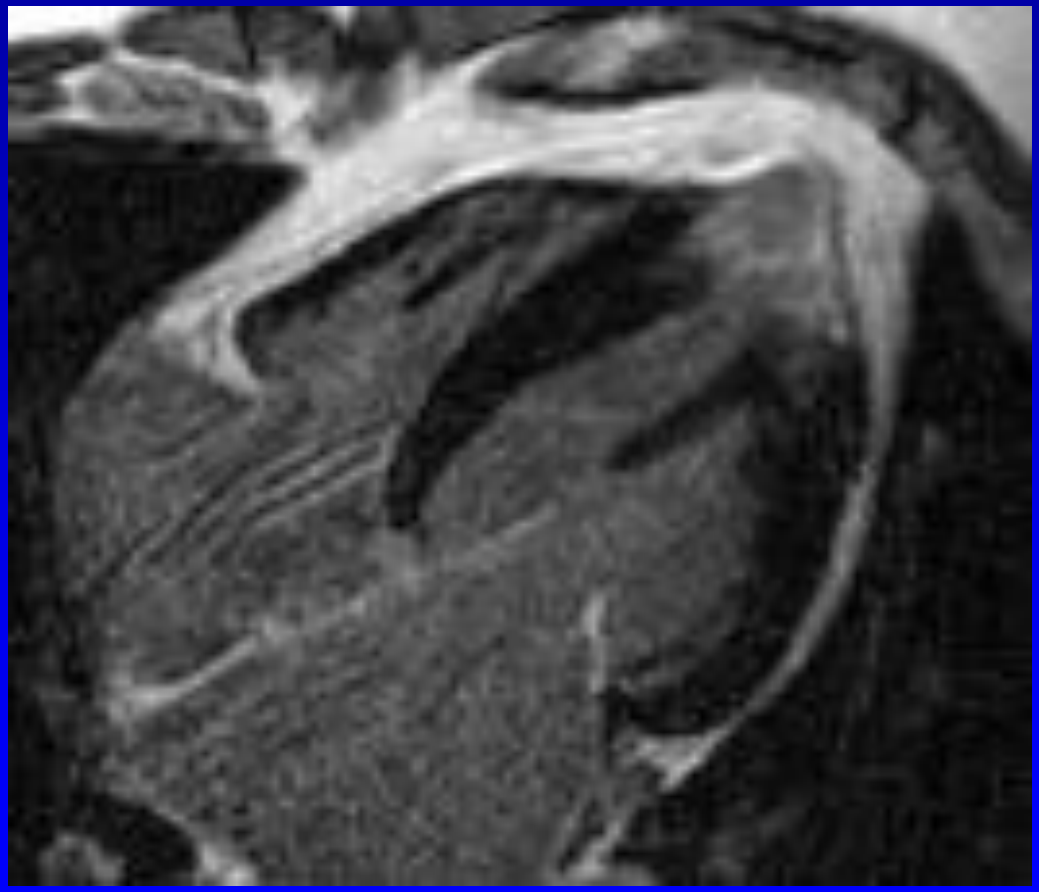
LGE in HCM and Outcomes





Chan et al. Circulation 2014;130:484-495

Apical HCM with Apical Infarction



Treatment of Dynamic Obstruction

Medical Therapy for HCM

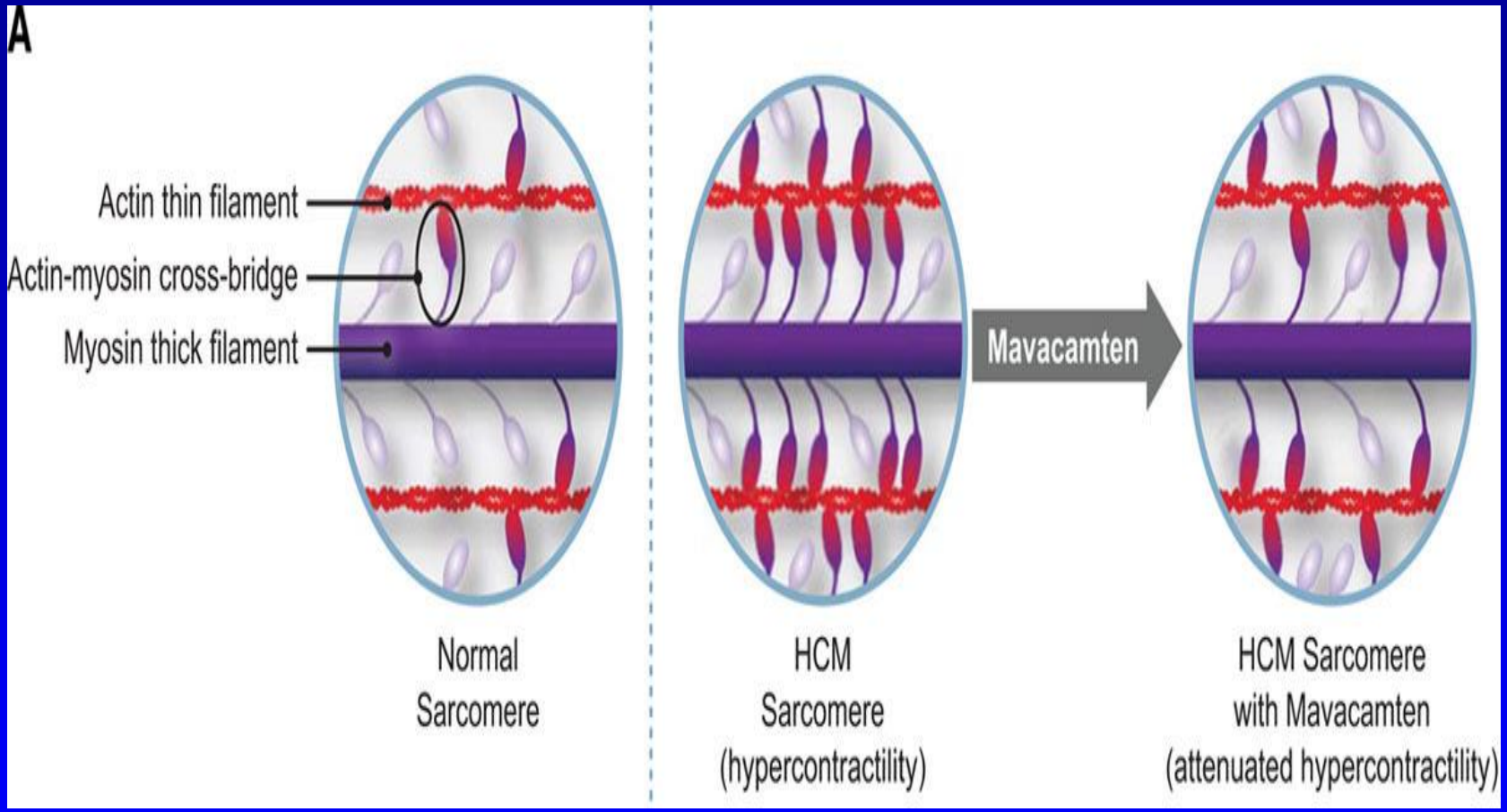
- Interval imaging with echo at 1-2 years interval, or earlier with symptomatic changes

- Objective of imaging: LVOT gradient, changes in ventricular size and function

- Detect changes in LV EF to identify dilated-hypokinetic LV to institute GDMT

- On myosin inhibitors main goals are LVOT gradients and quantitative LV EF to avoid heart failure due to reduced LV EF

Mechanism of Action of Mavacamten



Ho et al. Circulation Heart Failure. 2020;13:e006853

Response in Explorer- I

- End point: decrease in NYHA class by at least one level +1.5 ml/kg/min increase in MVO_2 or 3 ml/kg/min increase in MVO_2 without worsening of NYHA class

- **Patients on mavacamten had**

a-greater reductions than placebo in post-exercise LVOT gradient (−36 mm Hg, p<0·0001)

b-greater increase in pVO_2 (+1·4 mL/kg per min, p=0·0006)

c-improved symptom scores (p<0.001)

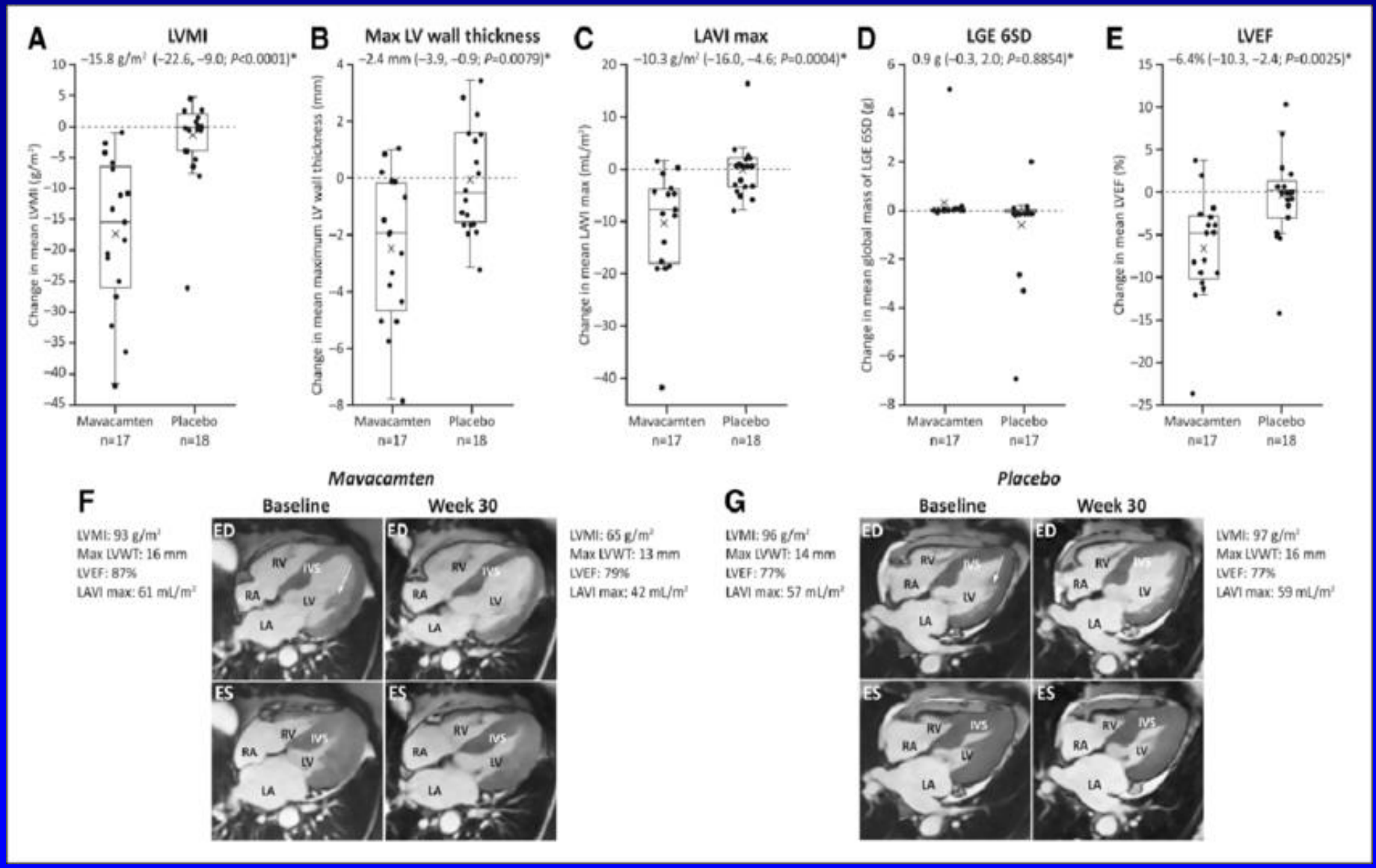
d-34% more patients improved by at least one class

e-Safety and tolerability similar to placebo

Response in Explorer-II

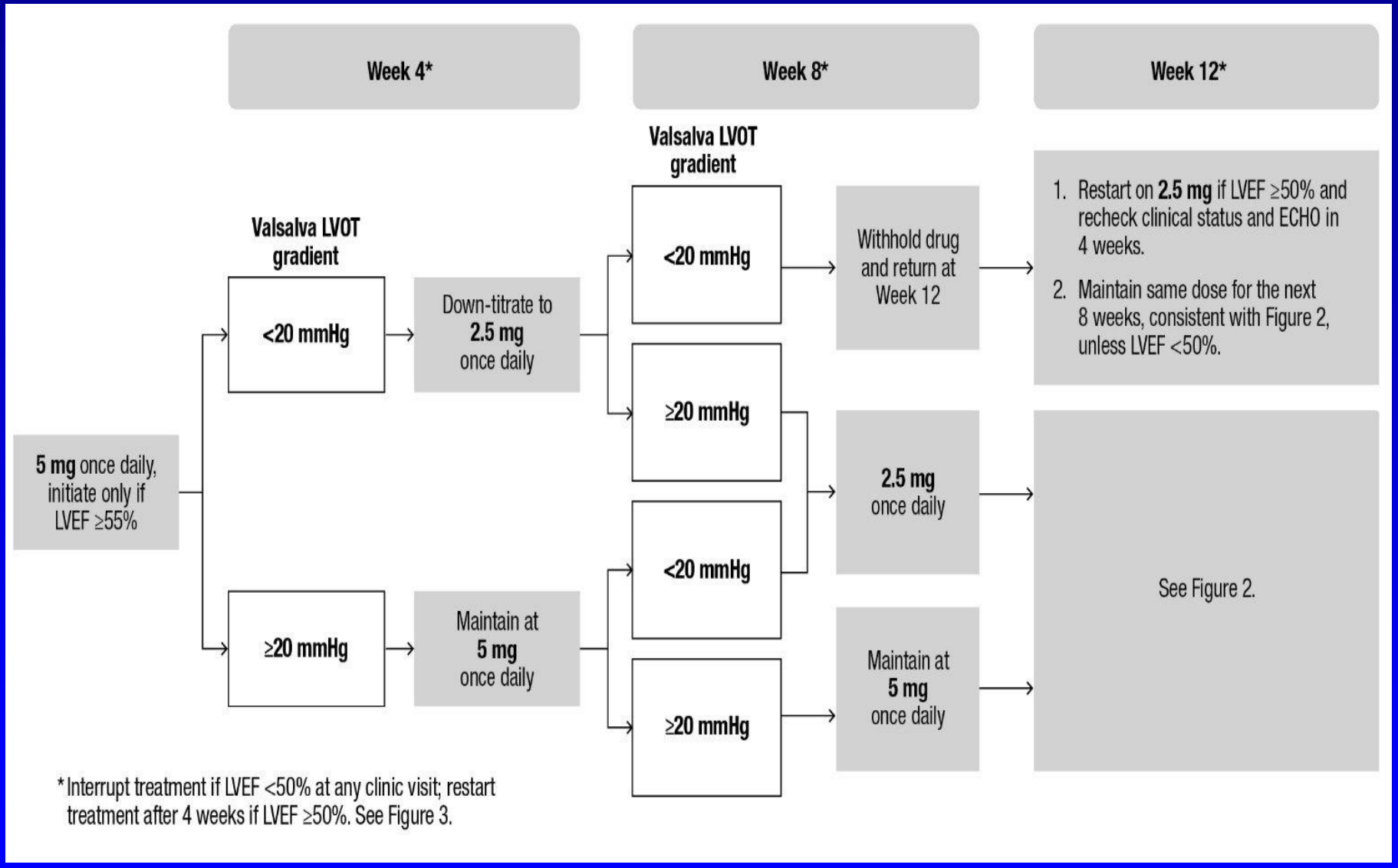
- End point met in 37% of drug arm (n=123) vs 17% on placebo (n=128) at 30 weeks
- NYHA class unchanged in 35% on the drug
- 26% of patients on the drug left with LVOT gradient >50 mmHg with exercise
- 43% patients on the drug left with LVOT gradient >30 mmHg

Mavacamten Decreases LA Volume, LV Mass, and LV EF

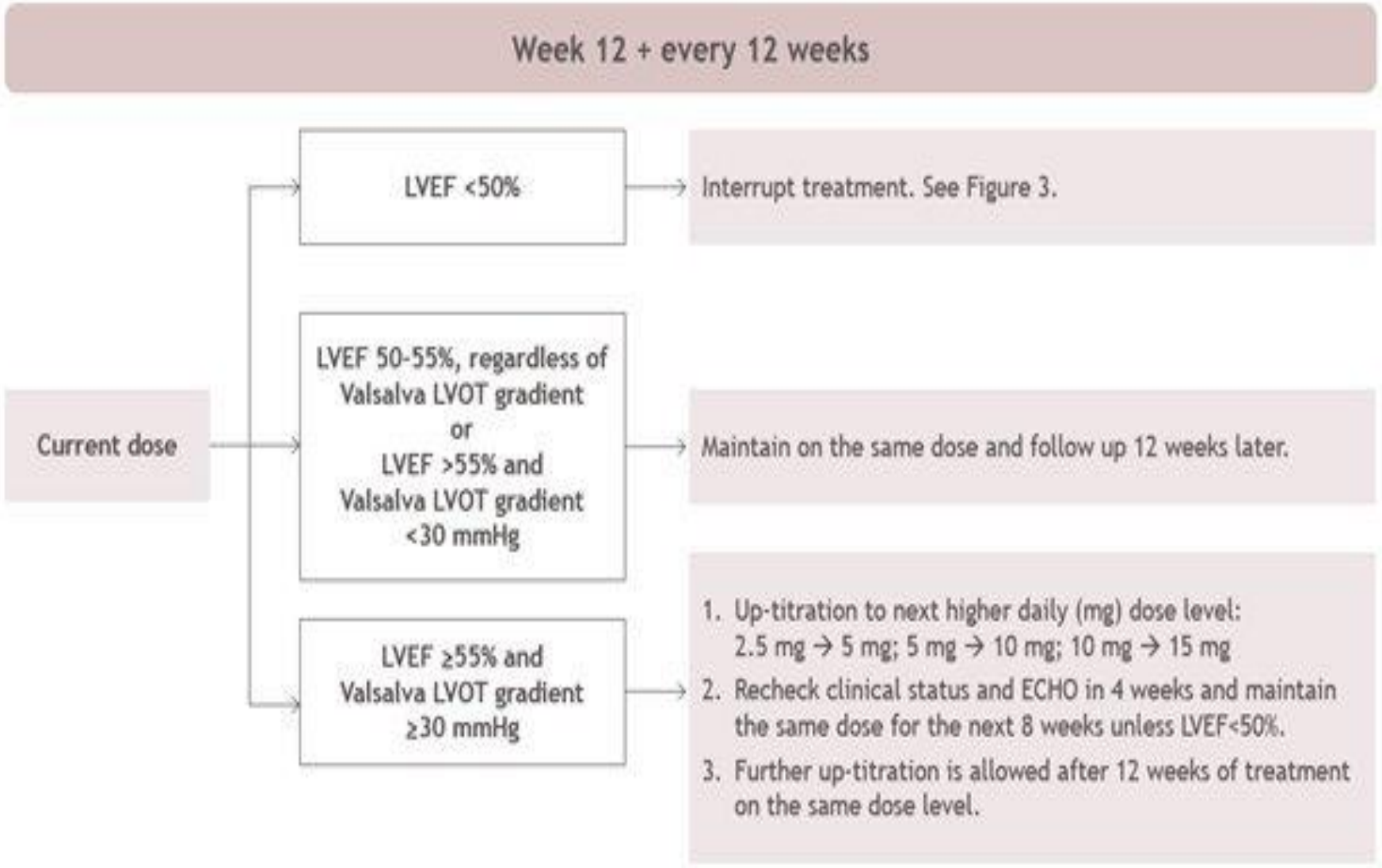


Saberi et al. Circulation 2021;143: 606-608

Mavacamten Initiation Phase

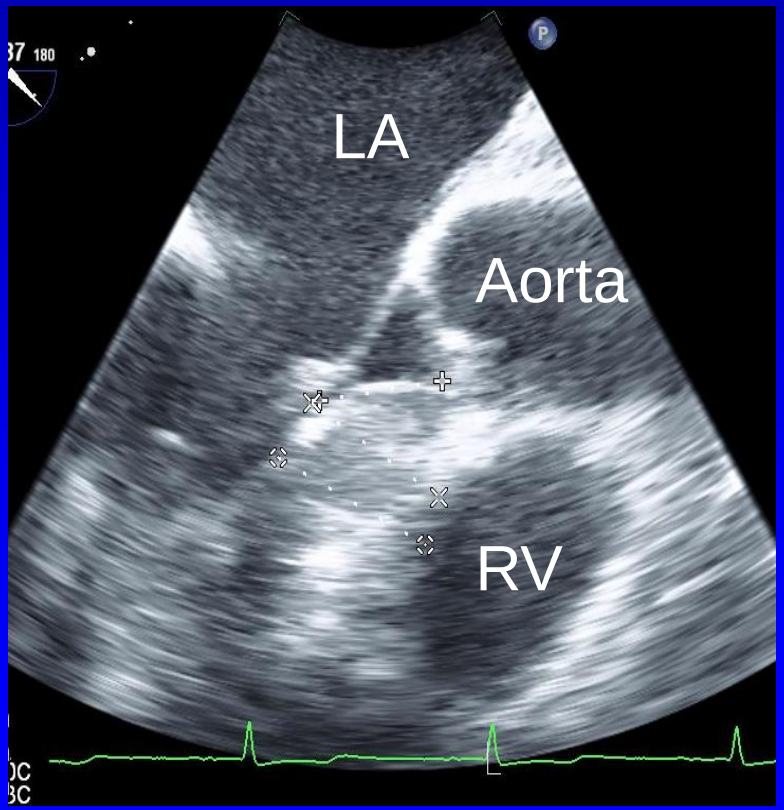


Mavacamten Maintenance Phase

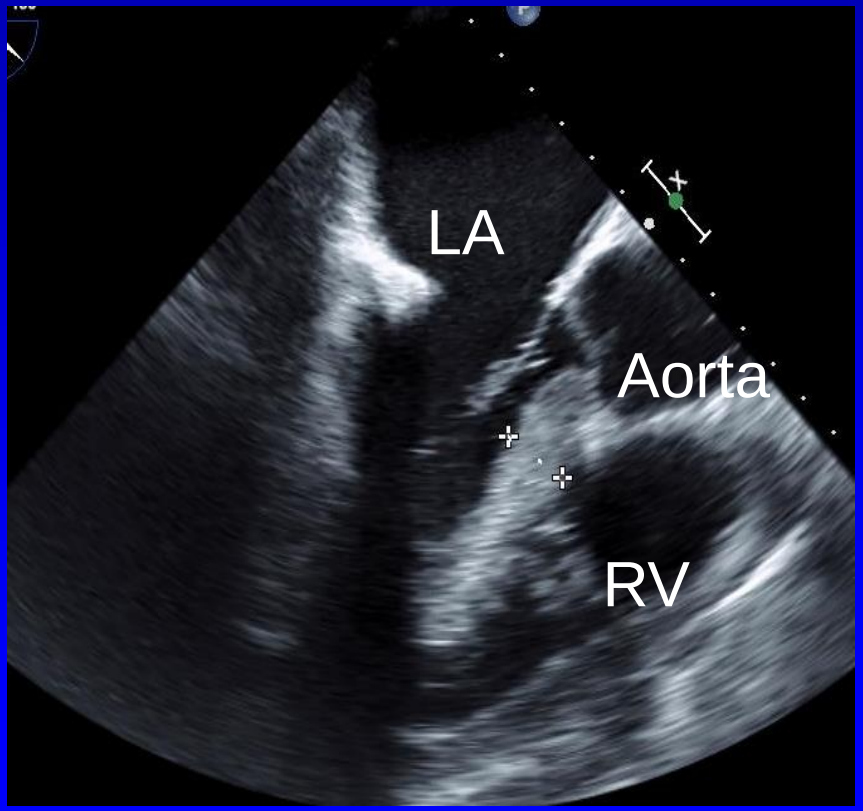


Acute Changes in Septal Thickness After Myectomy

Pre - Myectomy

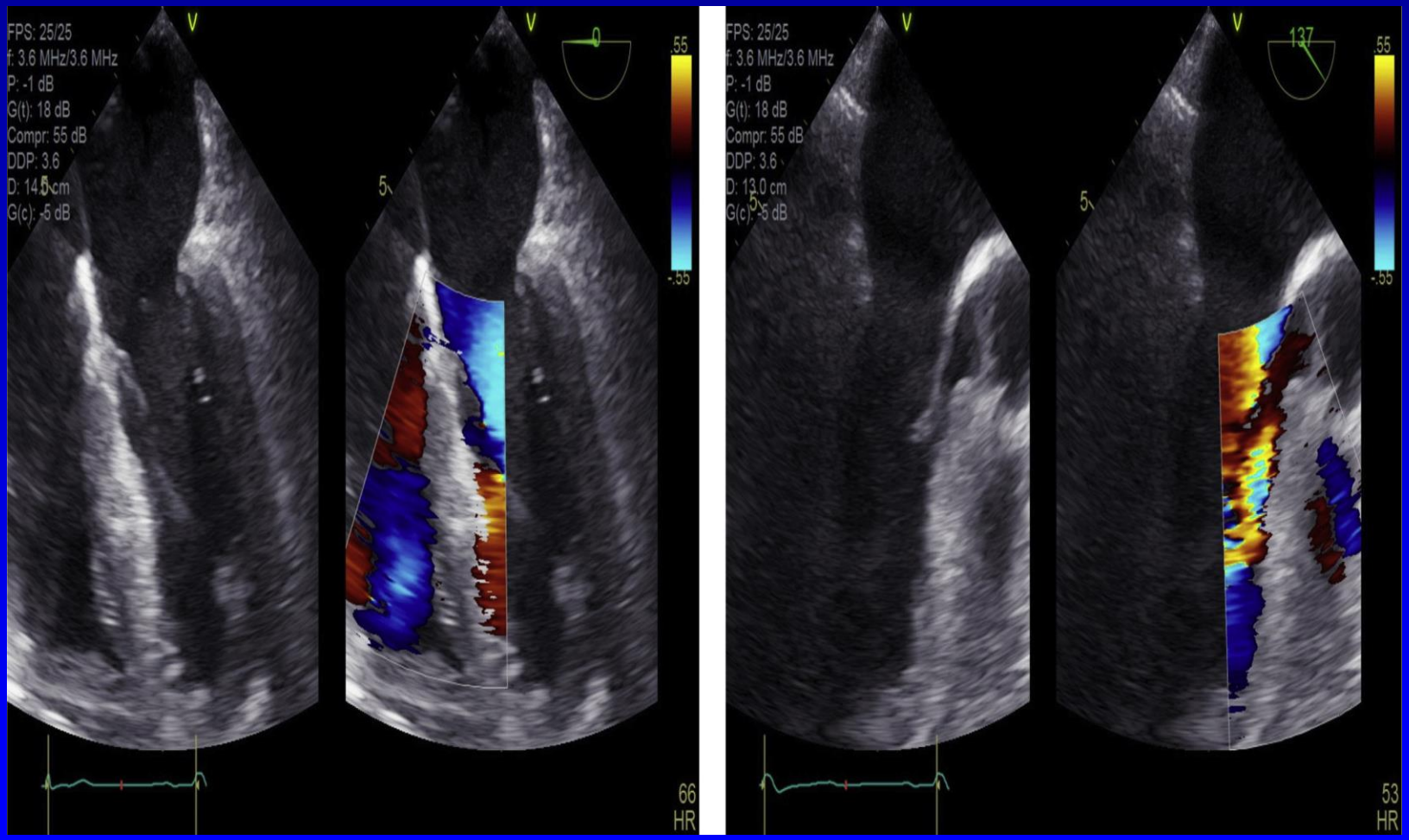


Post - Myectomy



Nagueh et al *JASE* 2011;24:473 - 498

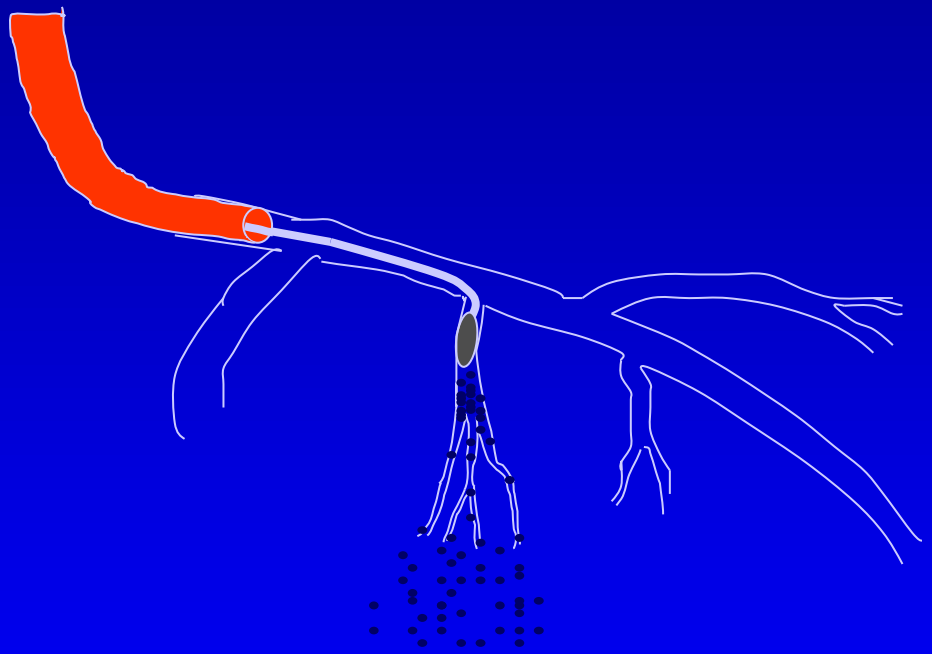
Screening for VSD after Myectomy



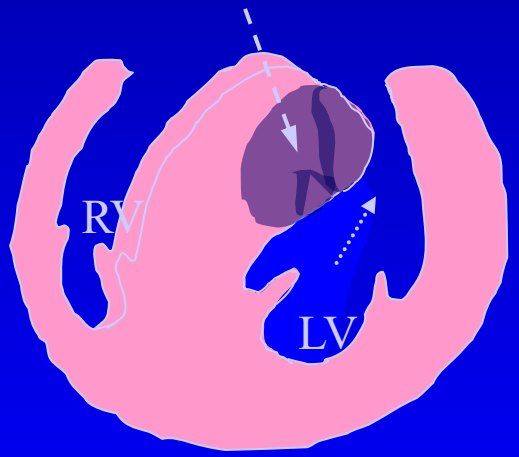
Nagueh et al *JASE* 2022;35:533 - 569

NONSURGICAL SEPTAL REDUCTION THERAPY (NSRT)-Alcohol Septal Ablation (ASA)

Confirmation Step With Contrast Injection and Echocardiography

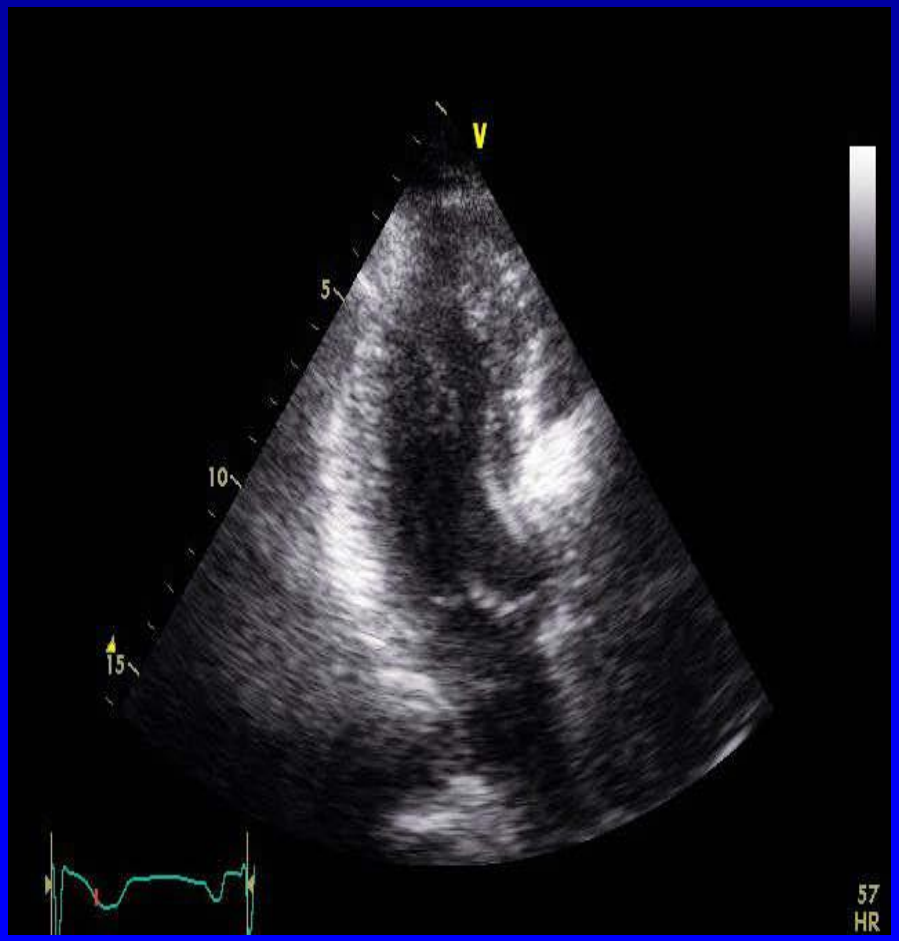


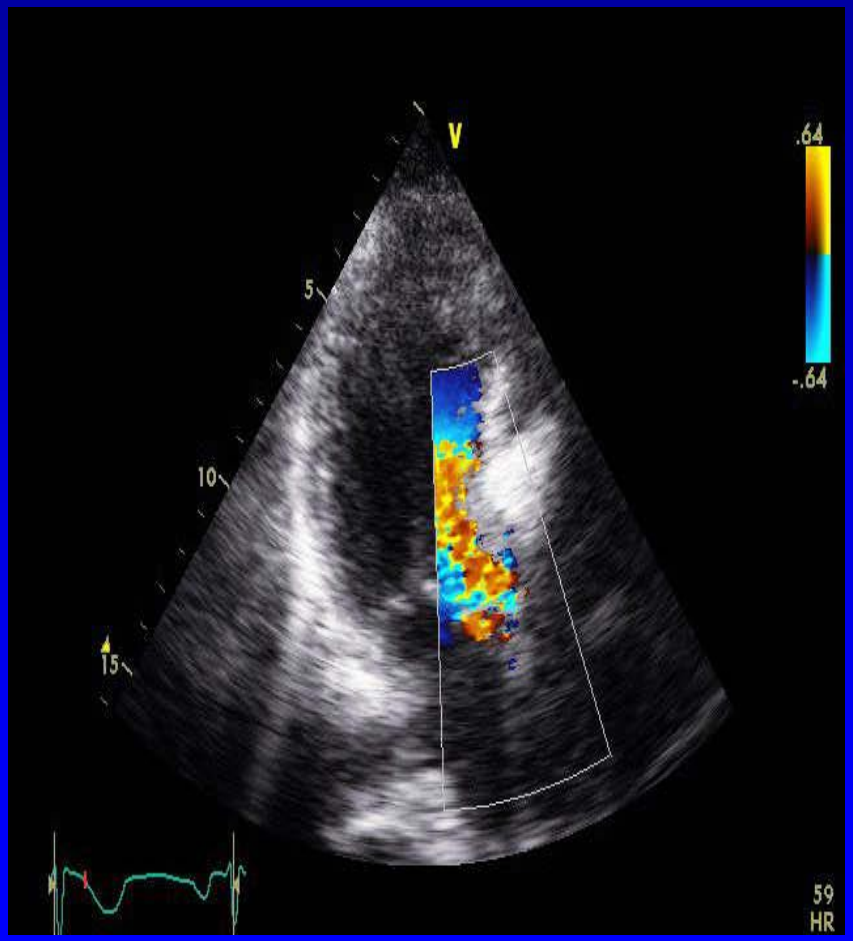
Hypertrophied septum by contrast echo



Contrast (Albunex ,Optison, or Levovist) is injected through the balloon lumen into the septum.

Septal Opacification by MCE in Relation to Dynamic Obstruction



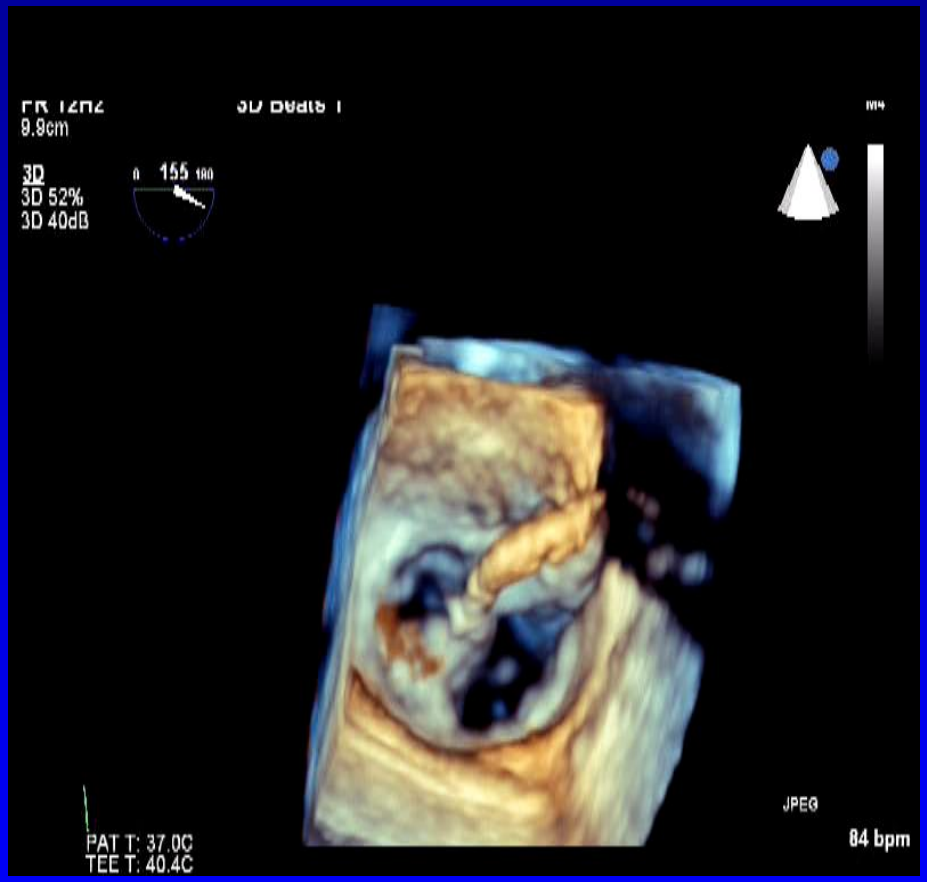


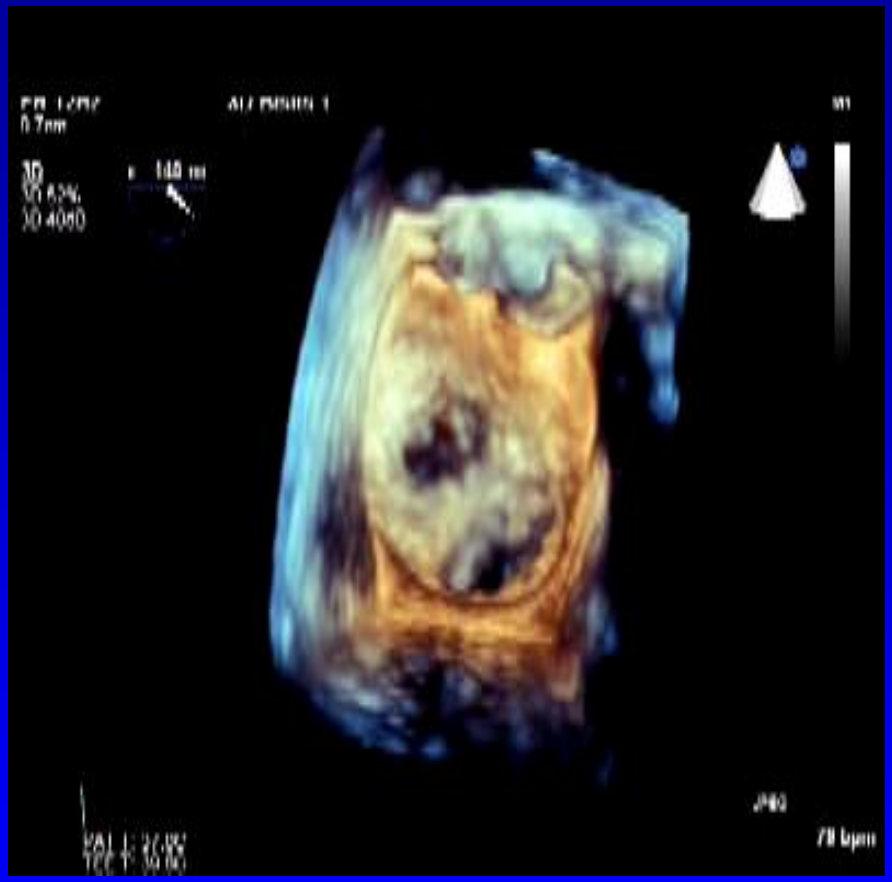
Nagueh and Mahmarian J Am Coll Cardiol 2006;48:2410

TEER for Dynamic Obstruction

Nagueh et al *JASE* 2022;35:533 - 569

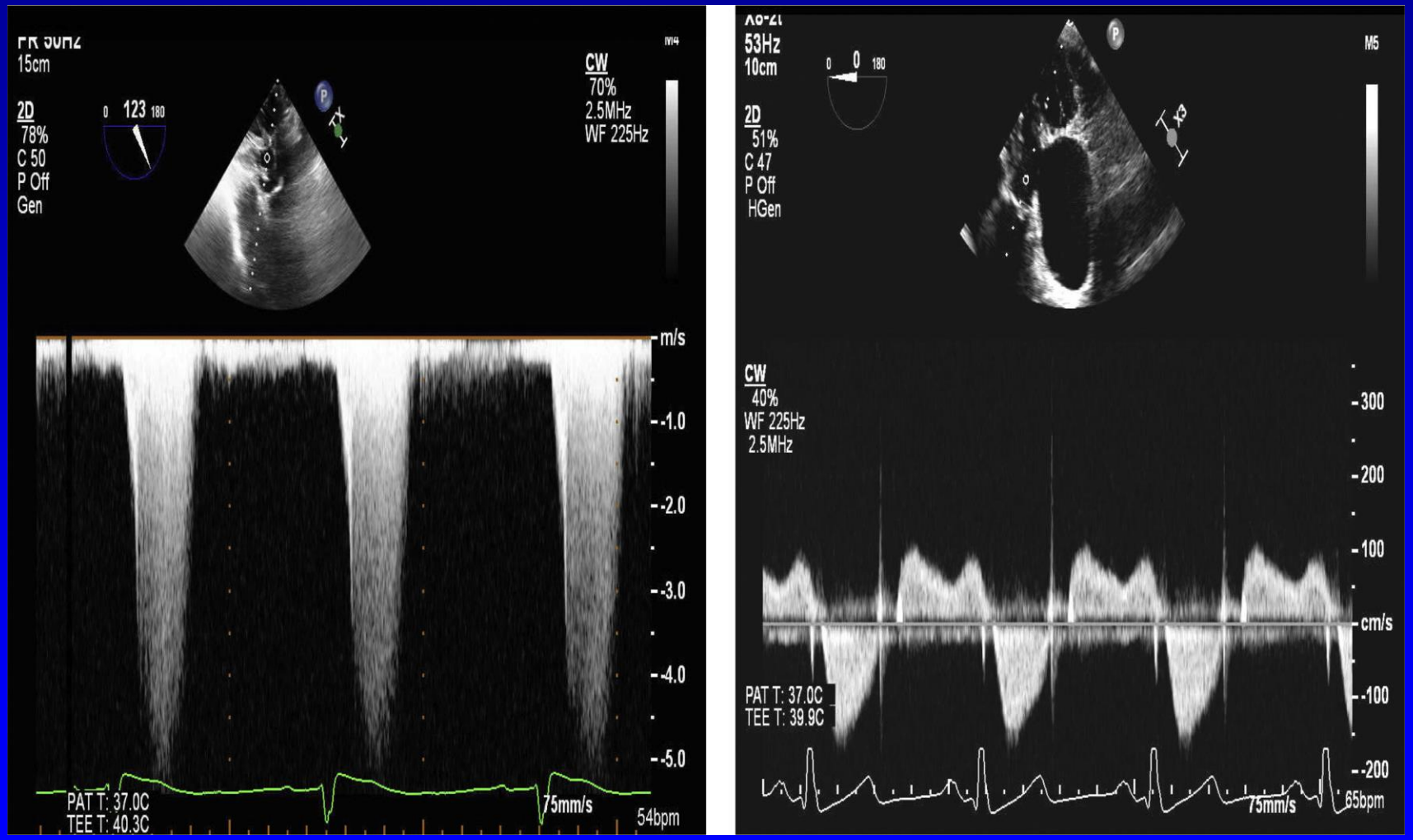
TEER for Dynamic Obstruction





Nagueh et al *JASE* 2022;35:533 - 569

LVOT Gradient Pre and Post TEER



Nagueh et al *JASE* 2022;35:533 - 569

Conclusions

- Imaging is essential for evaluation of patients with known or suspected HCM in conjunction with clinical findings

- Imaging plays critical role in informing risk stratification for SCD and evaluation of patients with chest pain and possible CAD

- Guide treatment with negative inotropic drugs, SRT, TEER

- Staff should have clear understanding of strengths and limitations of different imaging modalities and clinical implications of findings ascertained by imaging. Results should be communicated clearly with clinical team taking care of the patient.