HYPERTROPHIC CARDIOMYOPATHY

DR PAVAN ROY

INTERVENTIONAL CARDIOLOGIST

BANKERS HEART INSTITUTE

Outline

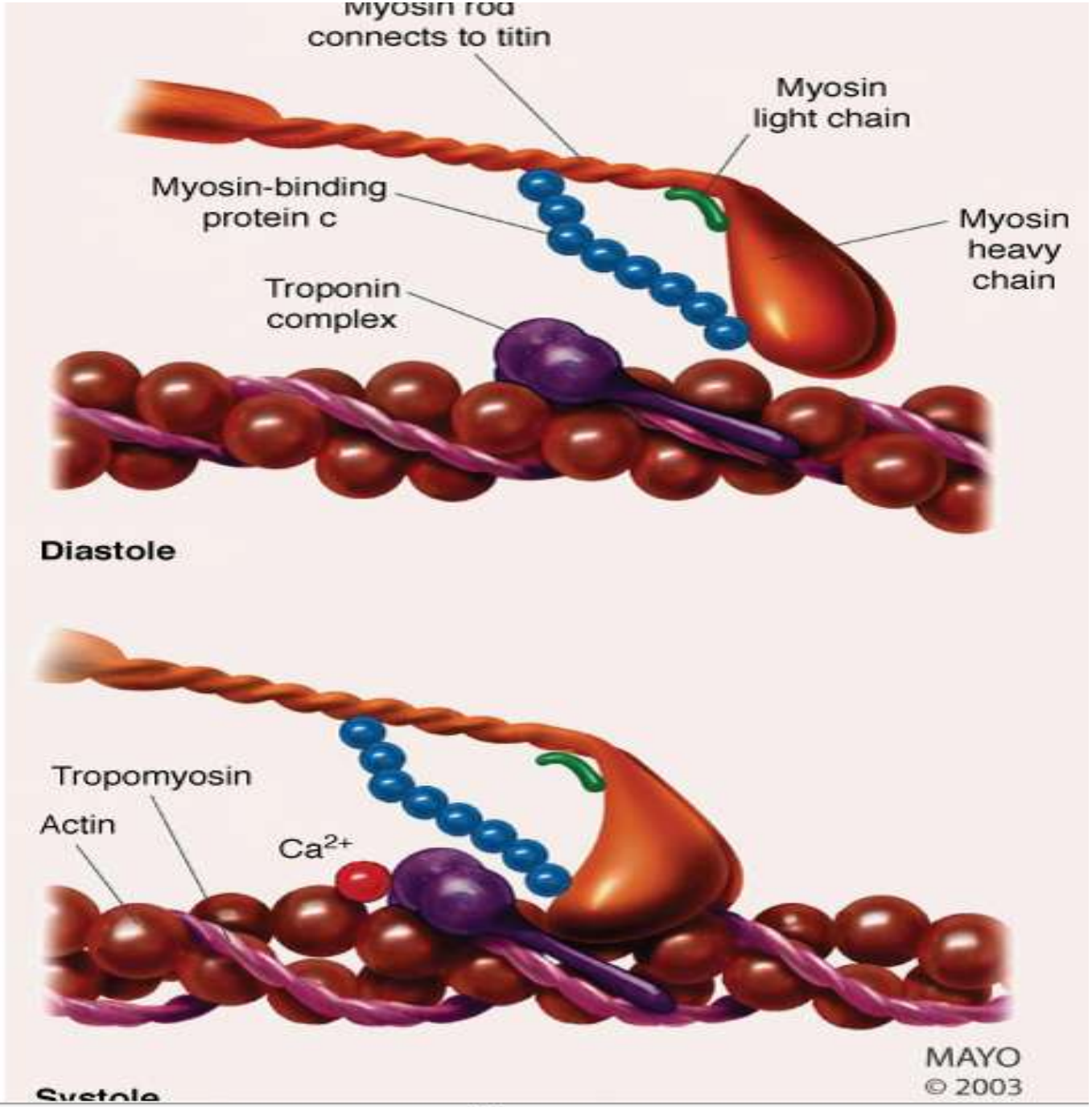
- Definition
- Genetics
- Pathophysiology
- Clinical features
- Evaluation
- Treatment

DEFINITION

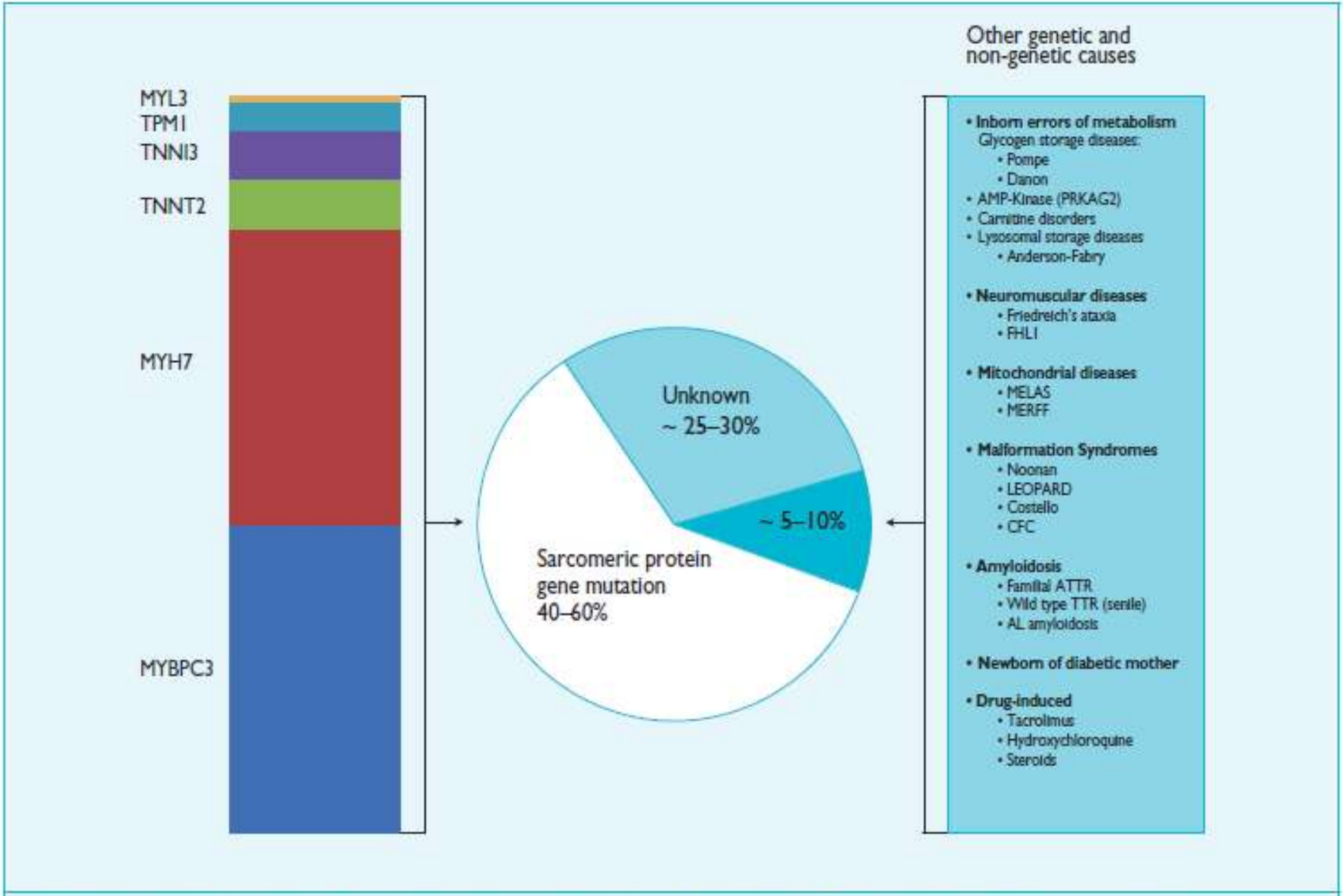
HCM is a genetic disease state characterized by unexplained LV hypertrophy associated with nondilated ventricular chambers in the absence of another cardiac or systemic disease that itself would be capable of producing the magnitude of hypertrophy evident in given patient.

It's prevalence estimated to be 1:500

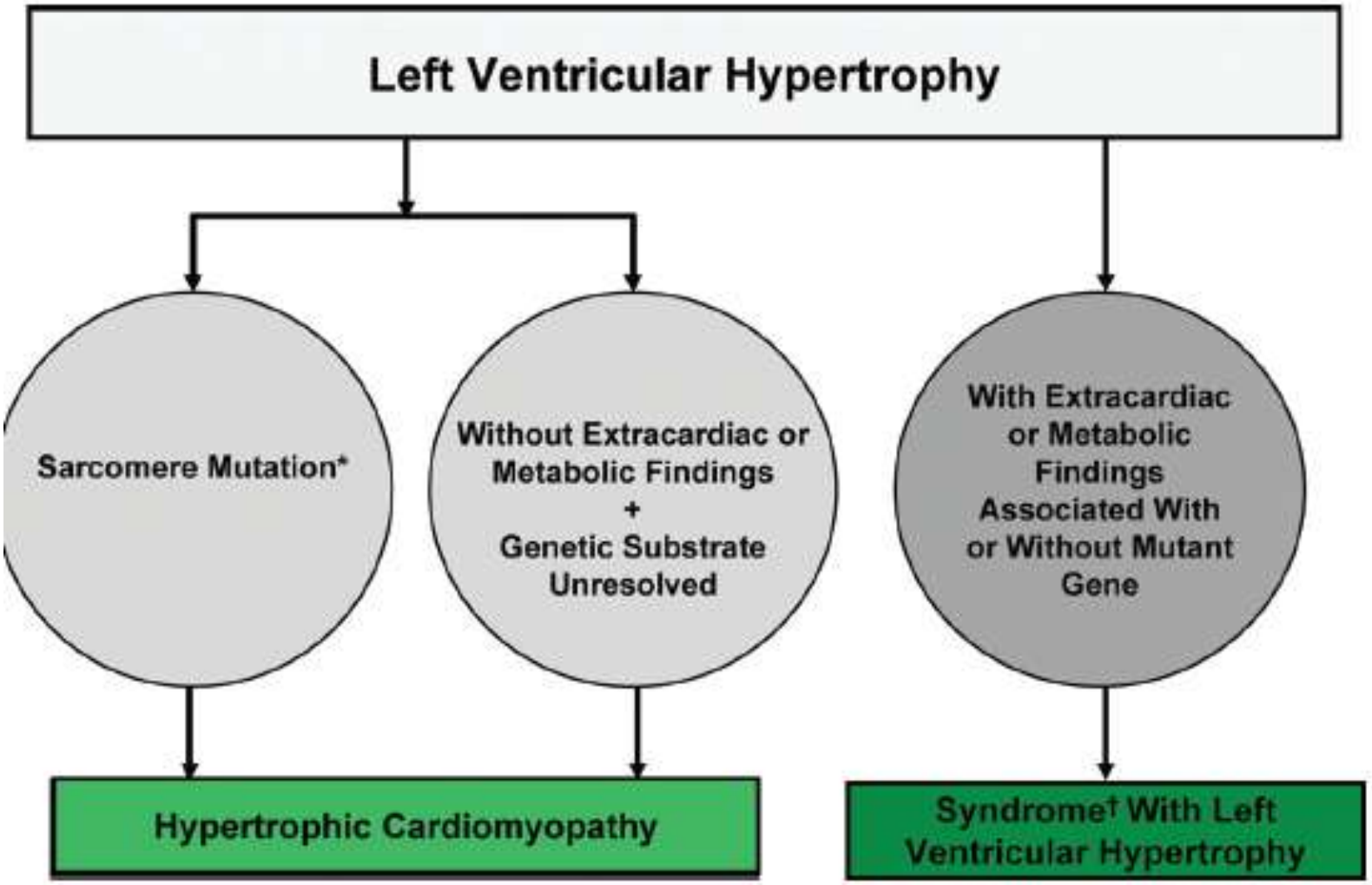
IHSS, HOCM are other terms


Myosin rod
connects to titin
Myosin
light chain
Myosin-binding
protein c
Myosin
heavy
chain
Troponin
complex
Diastole
Tropomyosin
Actin
Ca2+
MAYO
© 2003
Systole

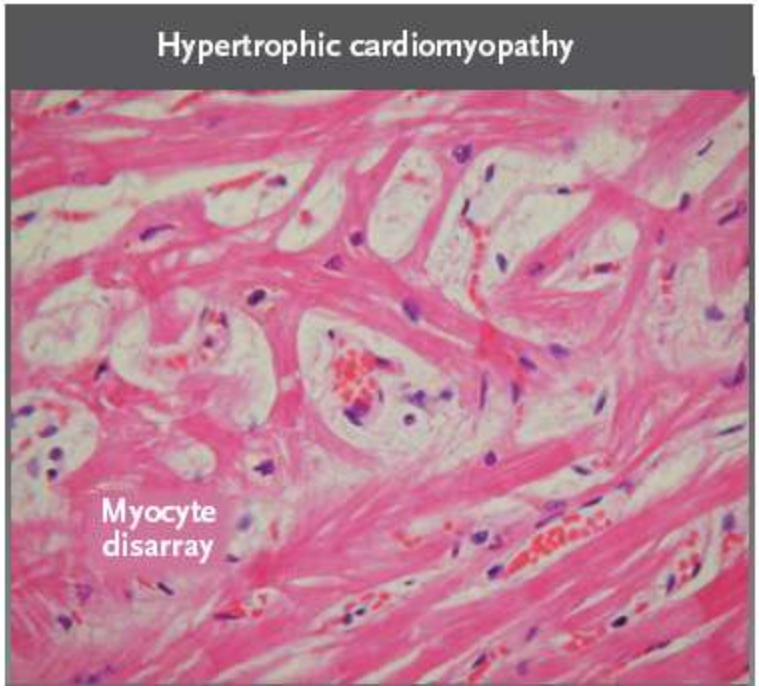
- Beta MHC mutations-clinical presentation apparent by late adolescents and develop substantial hypertrophy and more severe diseases.
- MyBPC mutations can have delayed clinical presentation until age 50 or older. Less severe symptoms.
- cTnT mutations-modest hypertrophy, increased risk of sudden death
- cTnI mutations- Greater predisposition of apical hypertrophy
- Alpha tropomyosin-relatively good survival. Variable degree of hypertrophy

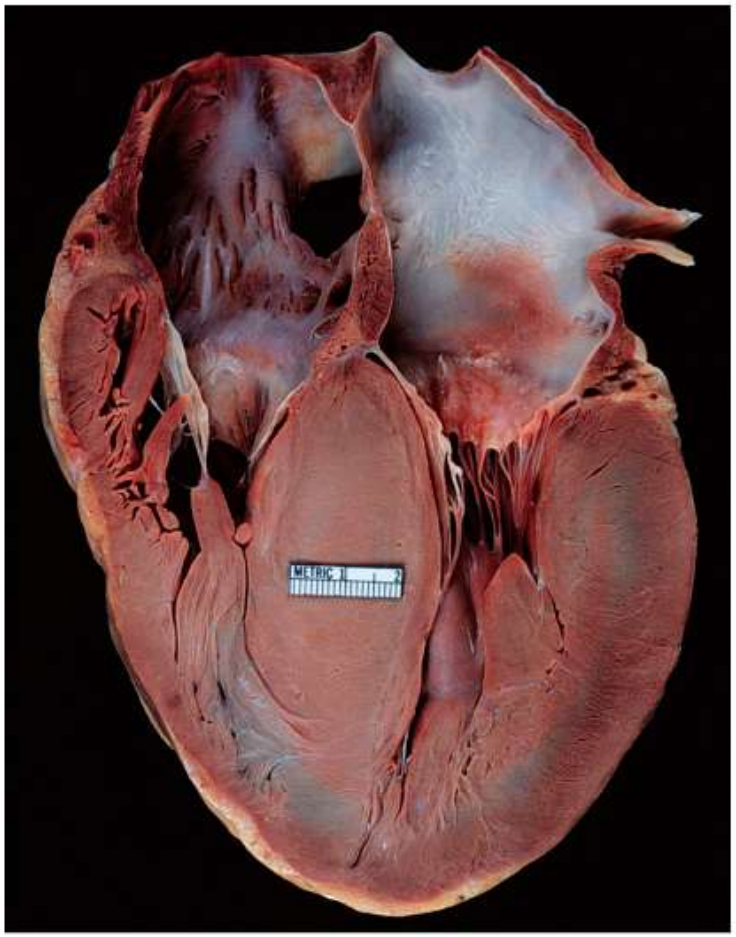


The majority of cases in adolescents and adults are caused by mutations in sarcomere protein genes. AL = amyloid light chain; ATTR=amyloidosis, transthyretin type.
CFC = cardiofaciocutaneous; FHL-1=Four and a half LIM domains protein 1; LEOPARD = lentigines, ECG abnormalities, ocular hypertelorism, pulmonary stenosis, abnormal genitalia, retardation of growth, and sensorineural deafness; MELAS = mitochondrial encephalomyopathy, lactic acidosis, and stroke-like episodes; MERFF = myoclonic epilepsy with ragged red fibres; MYL3 = myosin light chain 3; MYBPC3 = myosin-binding protein C, cardiac-type; MYH7 = myosin, heavy chain 7; TNNI3 = troponin I, cardiac; TNNT2 = troponin T, cardiac; TPM1 = tropomyosin 1 alpha chain; TTR = transthyretin.


Left Ventricular Hypertrophy
Sarcomere Mutation*
Without Extracardiac or Metabolic Findings + Genetic Substrate Unresolved
With Extracardiac or Metabolic Findings Associated With or Without Mutant Gene
Hypertrophic Cardiomyopathy
Syndrome† With Left Ventricular Hypertrophy

HISTOPATHOLOGY





Whorling and fibrosis

PATHOPHYSIOLOGY

- LV outflow tract obstruction
- Diastolic dysfunction
- Myocardial ischemia
- Mitral regurgitation
- Arrhythmias
- End stage/ burned out

LV OUTFLOW OBSTRUCTION

- Produced by SAM of mitral valve
- Explanations for the SAM of the mitral valve

1. Mitral valve is drawn toward the septum because of the lower pressure that occurs as blood is ejected at high velocity through a narrowed outflow tract (Venturi effect)
2. Mitral valve is pulled against the septum by contraction of the papillary muscles, which occurs because of the valve's abnormal location and septal hypertrophy altering the orientation of the papillary muscles
3. Hydrodynamic “drag” or the “pushing” force of flow

DYNAMIC OBSTRUCTION IS WORSENED BY

- Increase in contractility
  - VPC
  - Dobutamine,Isoproterenol
  - Exercise
- Decrease in afterload/volume
  - Valsalva maneuver
  - Standing
  - Nitroglycerine/amylnitrite inhalation
  - Blood loss
  - Dehydration

DEFINITIONS OF DYNAMIC LEFT VENTRICULAR OUTFLOW TRACT OBSTRUCTION

Hemodynamic state	Conditions	Outflow gradient
Basal obstruction	Rest	>30mmHg
Non obstructive	Rest	<30mmHg
	Physiologically provoked	<30mmHg
Dynamic obstruction	Rest	<30mmHg
	Physiologically provoked	>30mmHg

DYNAMIC LVOT OBSTRUCTION - DDS

- Hypercontractile states
- Anomalous papillary muscle insertion
- Anteroapical infarction with hyperkinetic basal segments
- Elderly women with LVH/sigmoid septum and hyperdynamic ventricular function
- After mitral valve repair

DIASTOLIC DYSFUNCTION

- Impaired relaxation
- Decreased compliance
  - Hypertrophy
  - Disorganised cellular architecture
  - Replacement scarring
  - Interstitial fibrosis
- Accounts for symptoms of exertional dyspnea
  - Increased filling pressures → increased pulmonary venous pressure

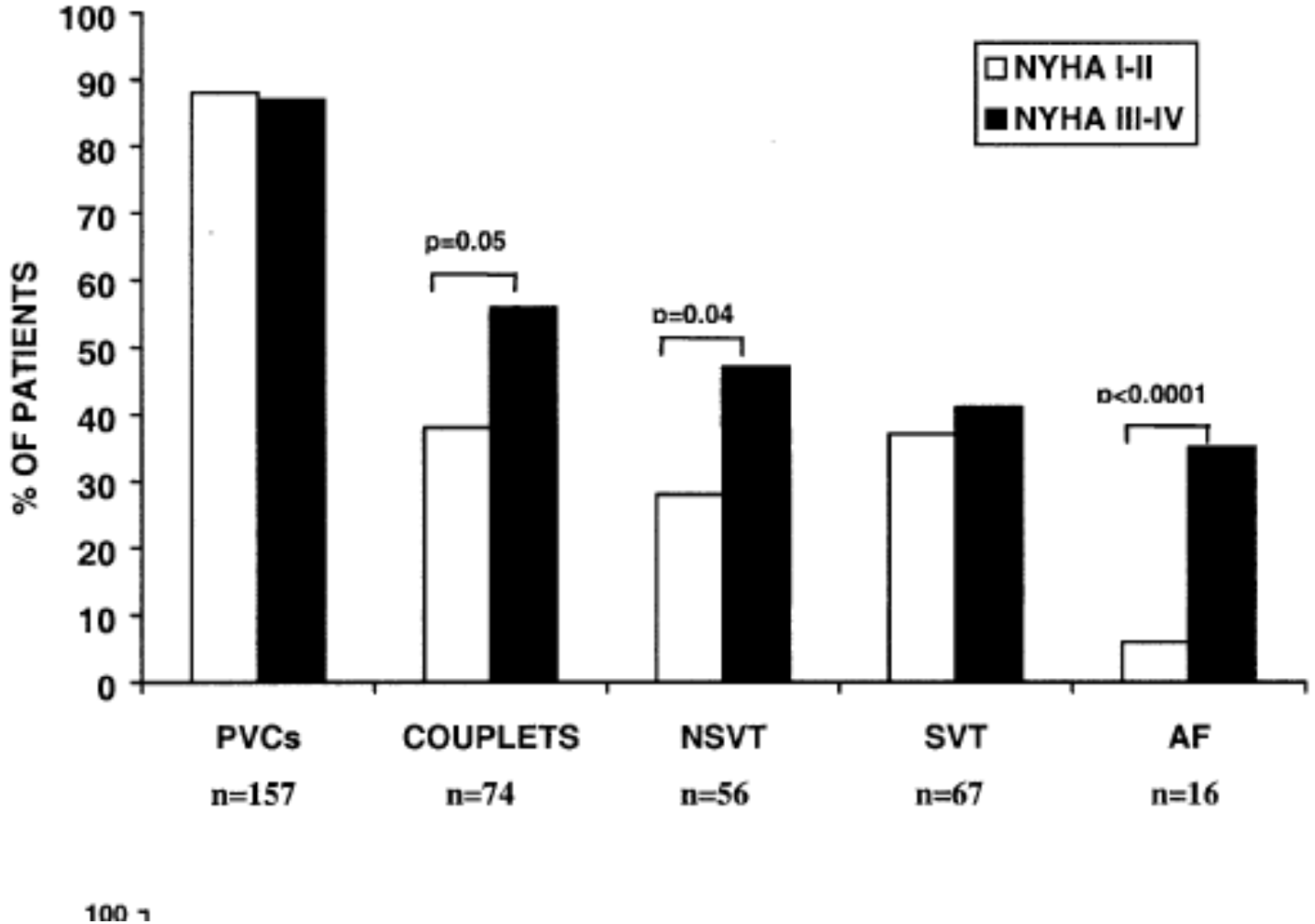
MYOCARDIAL ISCHEMIA

- Often occurs without atherosclerotic coronary artery disease
- Postulated mechanisms
  - Abnormally small and partially obliterated intramural coronary arteries as a result of hypertrophy
  - Inadequate number of capillaries for the degree of LV mass and increased myocardial oxygen consumption-supply demand mismatch
  - Increased filling pressures resulting in subendocardial ischemia

MITRAL VALVE APPARATUS

- Twice the normal size due to elongation of both leaflets or segmental enlargement of only anterior leaflet or mid portion of posteror leaflet
- Congenital and anomalous anterolateral papillary muscle insertion into the anterior leaflet without interposition of chordae tendineae and produce muscular midcavity outflow obstruction >>SAM>>LVOTO
- Variations in leaflet length (posterior/anterior leaflet length mismatch) – restrict the ability of the posterior leaflet to follow the anterior leaflet and to coapt effectively resulting in MR
- Severity of MR directly proportional to LV outflow obstruction
- Results in symptoms of dyspnea, orthopnea

ARRHYTHMIAS



CLINICAL PRESENTATION

- Majority are asymptomatic
- Dyspnea on exertion (90%), orthopnea, PND
- Palpitations (PAC, PVC, sinus pauses, AF, A flutter, SVT and VT)
- Congestive heart failure (2^{o} to increased filling pressures and myocardial ischemia)
- Sudden cardiac death (<1%)
- Angina (70-80%)
- Syncope (20%), Presyncope (50%)
  - Outflow obstruction worsens with increased contractility during exertional activities resulting in decrease in cardiac output
  - Secondary to arrhythmias

PHYSICAL

- Jugular venous pulse: prominent a- wave
- Double carotid arterial pulse: declines in mid systole as gradient develop
- Double apical impulse:
  - Forceful left atrial contraction against non-compliant ventricle
- Triple apical impulse:
  - Late systolic bulge near isometric contraction
- S1: normal
- S2: normal or paradoxical split
- S3 gallop: decompensated Lt. ventricle
- S4: atrial systole against hypertrophic ventricle

ESM-BETWEEN APEX AND LLSB

Effect of selected maneuvers		
Maneuver	Mechanism	Effect on gradient and murmur
Valsalva (strain)	Decreased LV cavity, pre and after load	Increased
Standing	Decreased LV cavity, preload	Increased
Post PVC	Increased contractility	Increased
Squatting	Increased LV cavity, pre and after load	Decreased
Isometric hand grip	Increased afterload	Decreased

PHYSICAL

- Holosystolic Murmur of MR:
  - Retrograde ejection of blood flow into low pressure left atrium
  - Best heard at apex and axilla
  - Pt. with SAM* and significant LV outflow gradients

- Diastolic Decrescendo Murmur of AR: 10% of Pt.

* Systolic anterior motion

MIMICKING HYPERTROPHIC CARDIOMYOPATHY

- Chronic hypertension
- RV hypertrophy
- Cardiac amyloidosis
- Athlete's heart
- Valvular AS

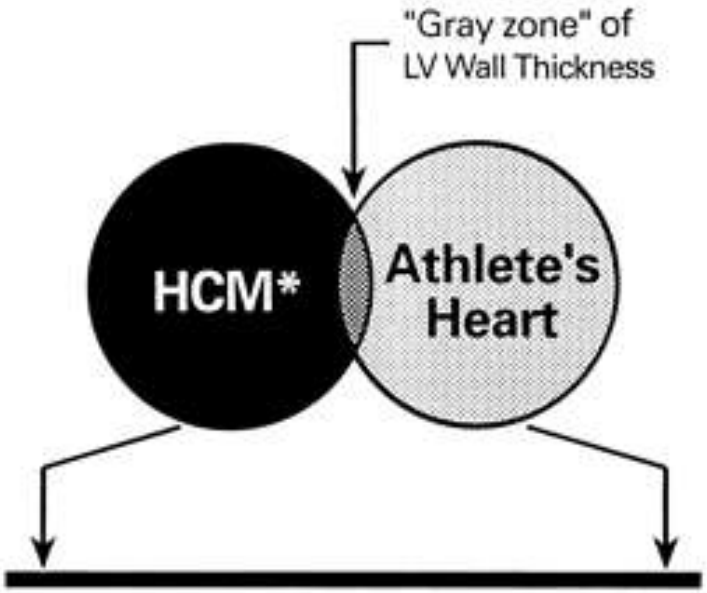
Apical hypertrophy - apical cavity obliteration caused by hypereosinophilic syndrome or noncompaction.

POINTS FAVOURING HCM IN HTN

- Family history of HCM
- Asymmetry
- Right ventricular hypertrophy
- Late gadolinium enhancement at the RV insertion points or localized to segments of maximum LV thickening on CMR
- Maximum LV wall thickness ≥15 mm (Caucasian); ≥20 mm (black)
- Severe diastolic dysfunction
- Marked repolarisation abnormalities, conduction disease or Q-waves on 12 lead ECG
- Regression of LVH

- ESC Guidelines2014

HCM VS. ATHLETE'S HEART



⊕	Unusual Patterns of LVH†	⊖
⊕	LV Cavity < 45 mm	⊖
⊖	LV Cavity > 55 mm	⊕
⊕	LA Enlargement	⊖
⊕	Bizarre ECG Patterns	⊖
⊕	Abnormal LV Filling	⊖
⊕	Female Gender	⊖
⊖	↓Thickness with Deconditioning	⊕
⊕	Family History HCM	⊖

Circulation 1995;91.

HCM VS AORTIC STENOSIS

	HCM	Fixed Obstruction
carotid pulse	spike and dome	parvus
murmur	↑ valsalva, standing ↓ squatting, handgrip ↓ passive leg elevation	radiate to carotids
systolic thrill	4th left ics	2nd right ics
systolic click	absent	present

DIAGNOSIS

- ECG
- IMAGING
    - ECHOCARDIOGRAPHY
    - CARDIAC MRI
    - OTHER MODALITIES
- CATH DATA
- TESTS TO RISK STRATIFY PATIENTS
- GENETIC TESTING
- FAMILY SCREENING

ECG

Abnormal - >90% of pts & >75% of asymptomatic relatives

- Increased voltages consistent with LV hypertrophy
- ST-T changes - marked T wave inversion in the lateral precordial leads
- Left atrial enlargement
- Deep and narrow Q waves lateral precardial leads
- Diminished R waves in the lateral precordial leads.

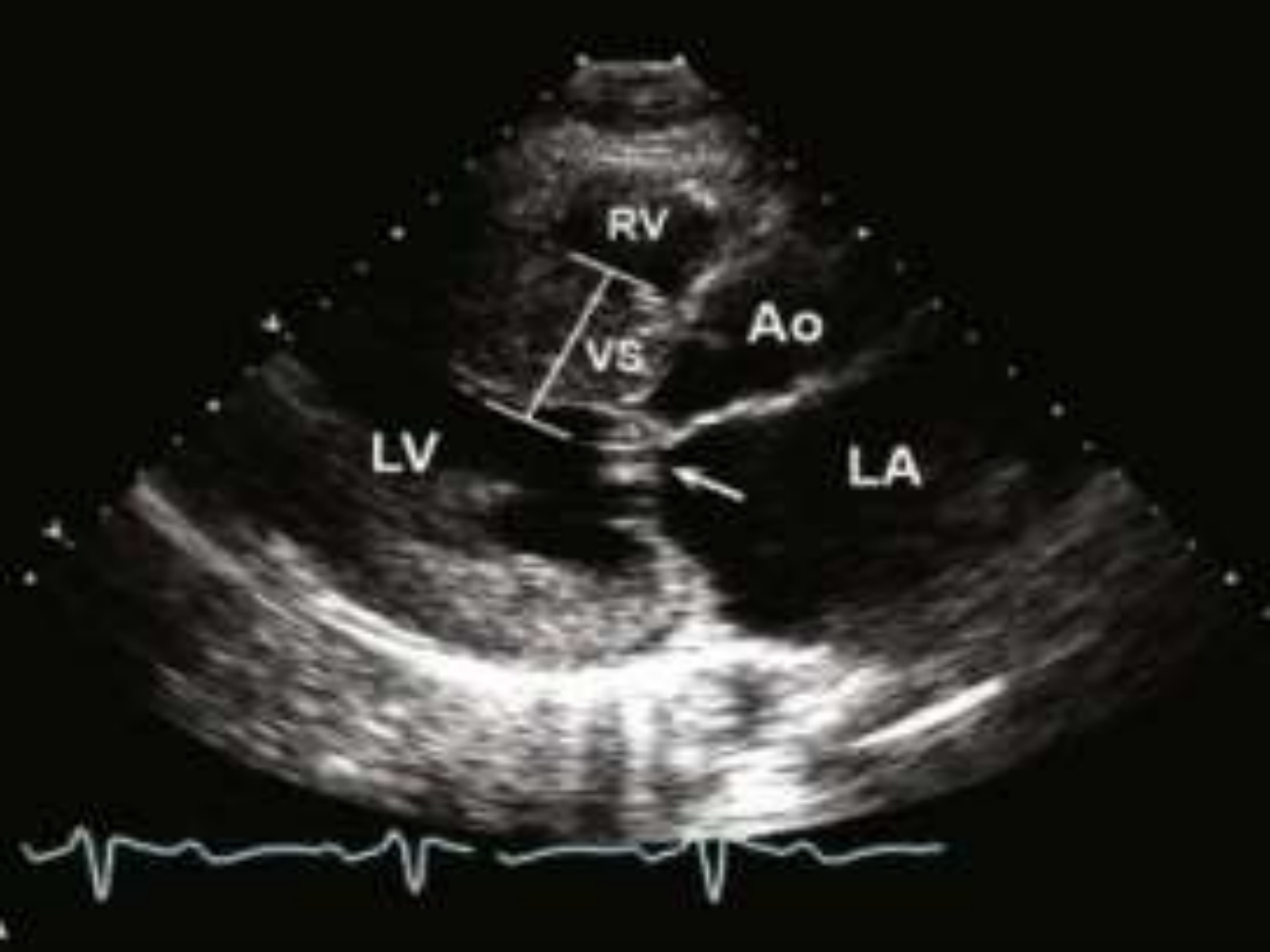
Normal ECG - 5% of pts

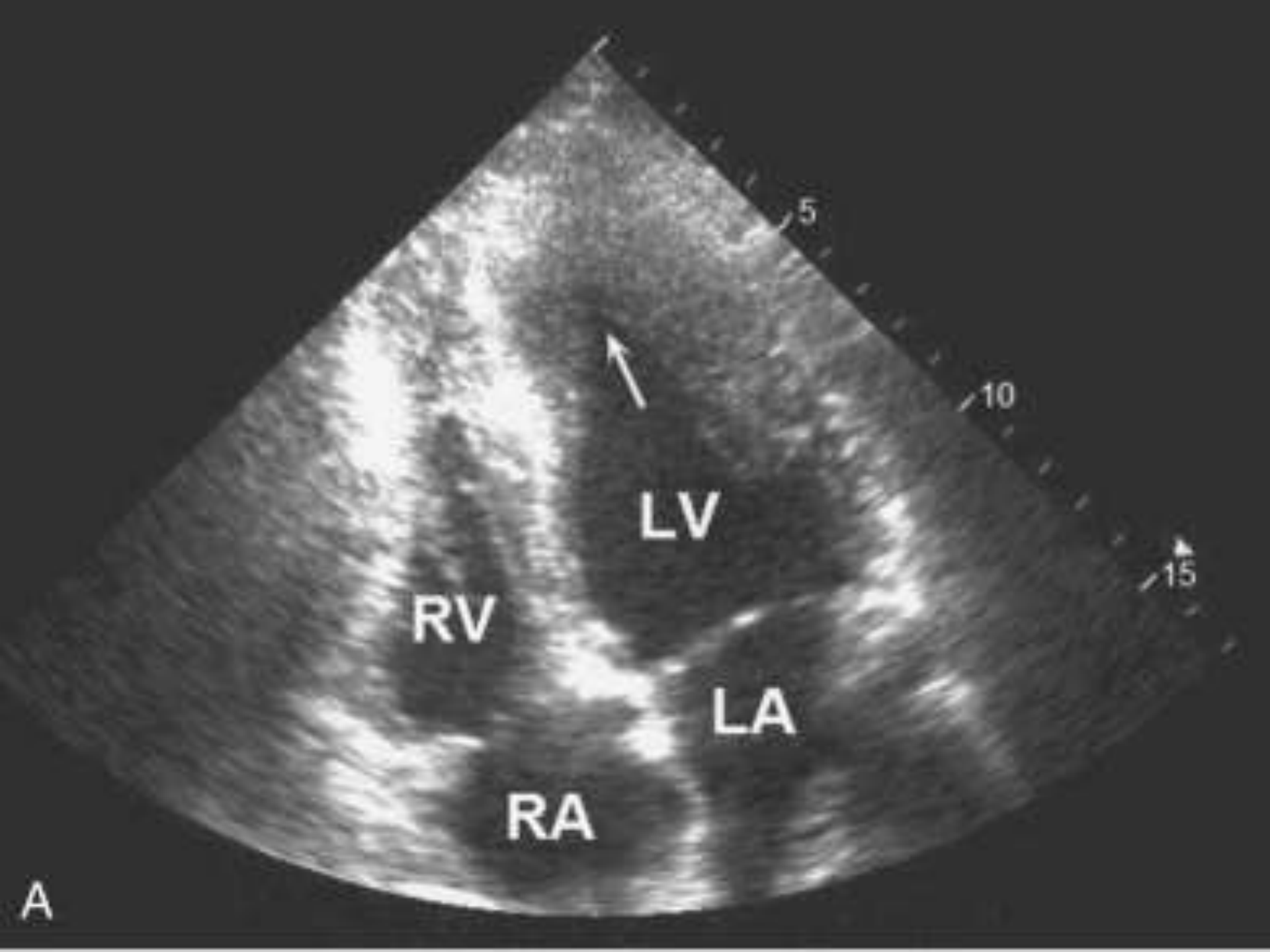
- Less severe phenotype and favorable course
- Not predictive of future sudden death

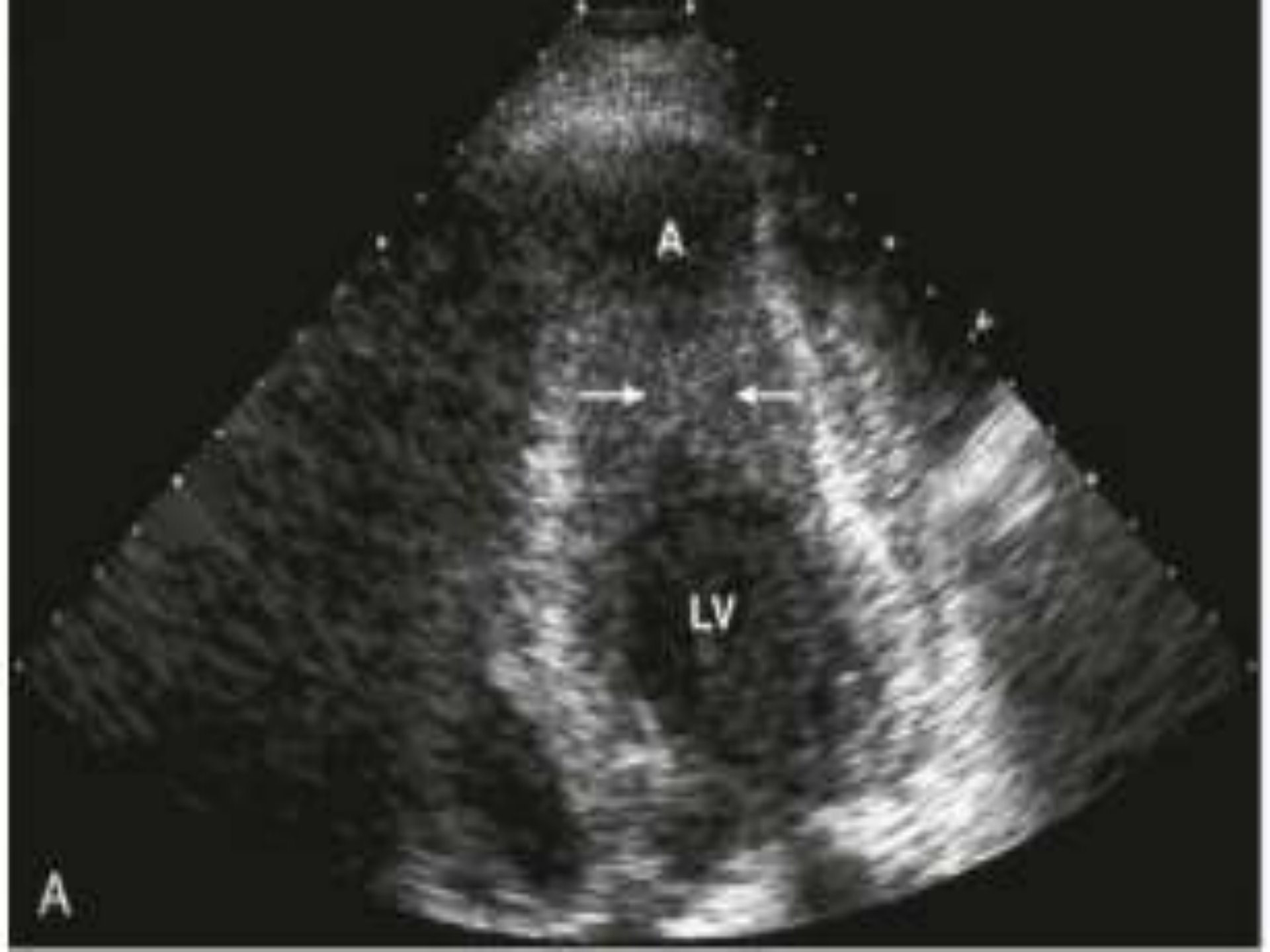
ECHOCARDIOGRAPHY

A COMPREHENSIVE ECHO EVALUATION REPORT

1. Presence of hypertrophy and its distribution; report should include measurements of LV dimensions and wall thickness (septal, posterior, and maximum)
2. LV EF
3. RV hypertrophy and whether RV dynamic obstruction is present
4. LA volume indexed to body surface area
5. LV diastolic function (comments on LV relaxation and filling pressures)
6. Pulmonary artery systolic pressure
7. Dynamic obstruction at rest and with Valsalva maneuver; report should identify the site of obstruction and the gradient
8. Mitral valve and papillary muscle evaluation, including the direction, mechanism, and severity of mitral regurgitation; if needed, TEE should be performed to satisfactorily answer these questions

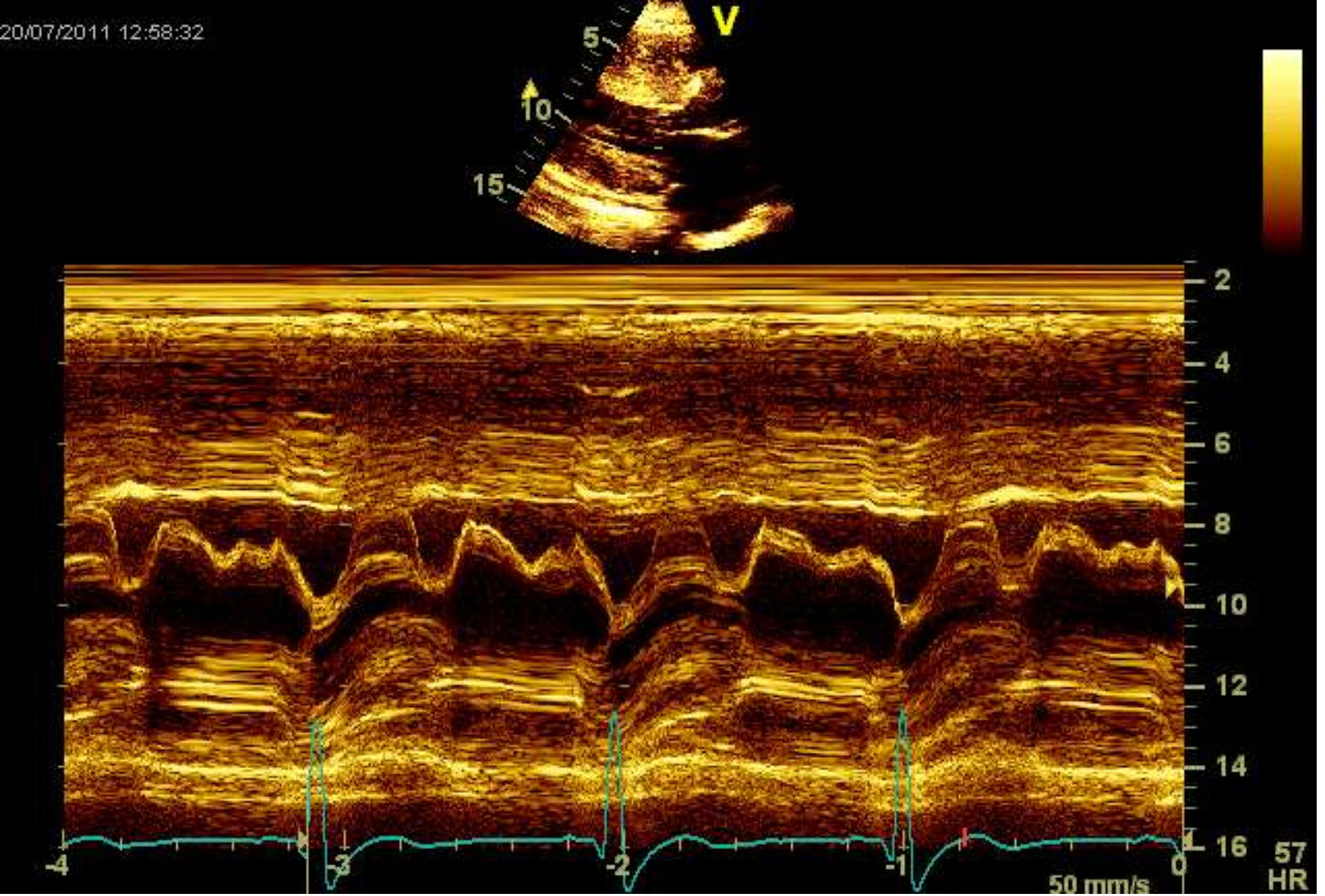

RV
VS
Ao
LV
LA


5
10
15
LV
RV
LA
RA
A

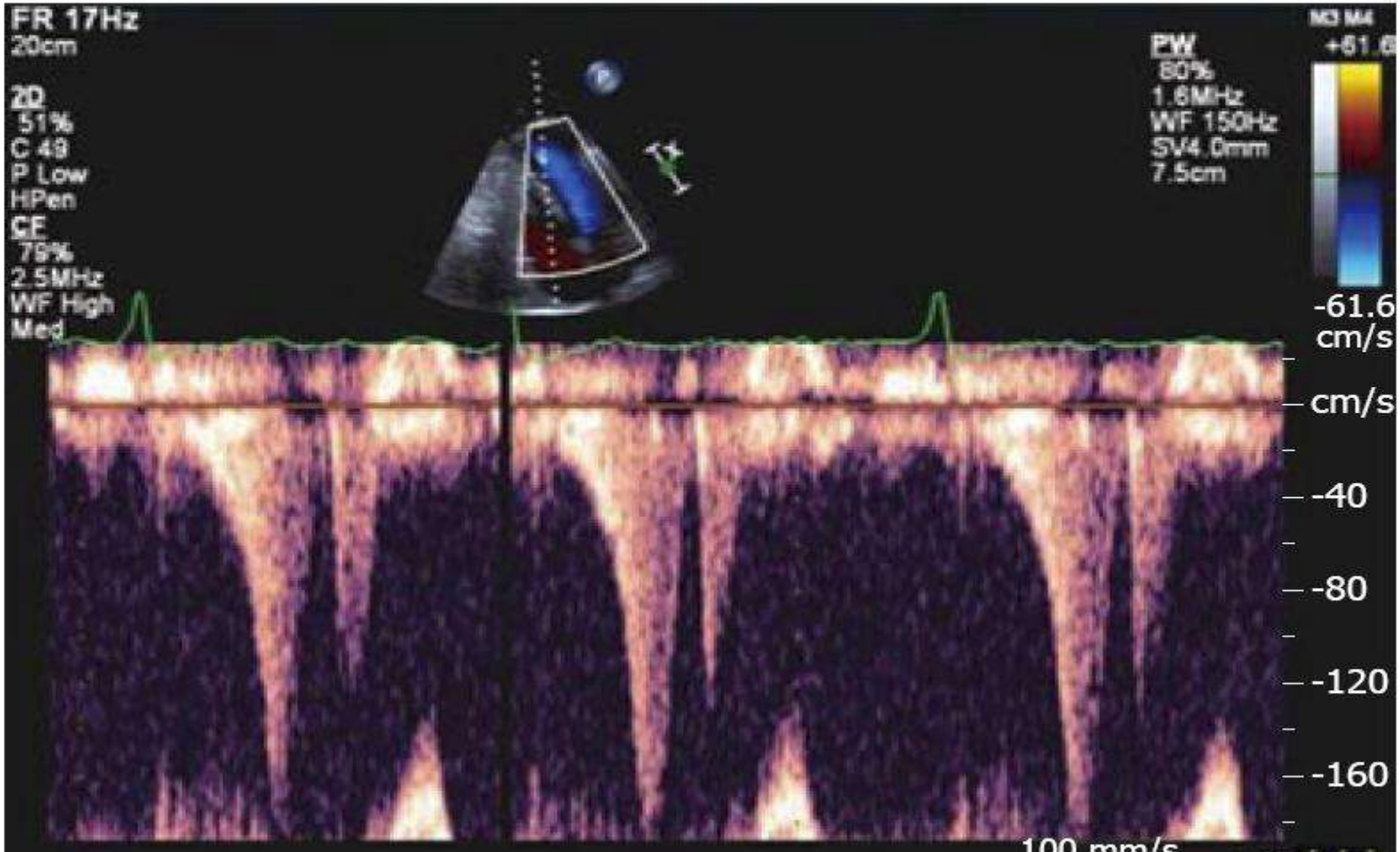

A
LV
A

LV hypertrophy

- Diffuse hypertrophy of the ventricular septum and anterolateral free wall (70% to 75%)
- Basal septal hypertrophy (10% to 15%)
- Concentric hypertrophy (5%)
- Apical hypertrophy (<5%)
- Hypertrophy of the lateral wall (1% to 2%).


20/07/2011 12:58:32
V
5
10
15
2
4
6
8
10
12
14
16
-4
-3
-2
-1
0
50 mm/s
57
HR

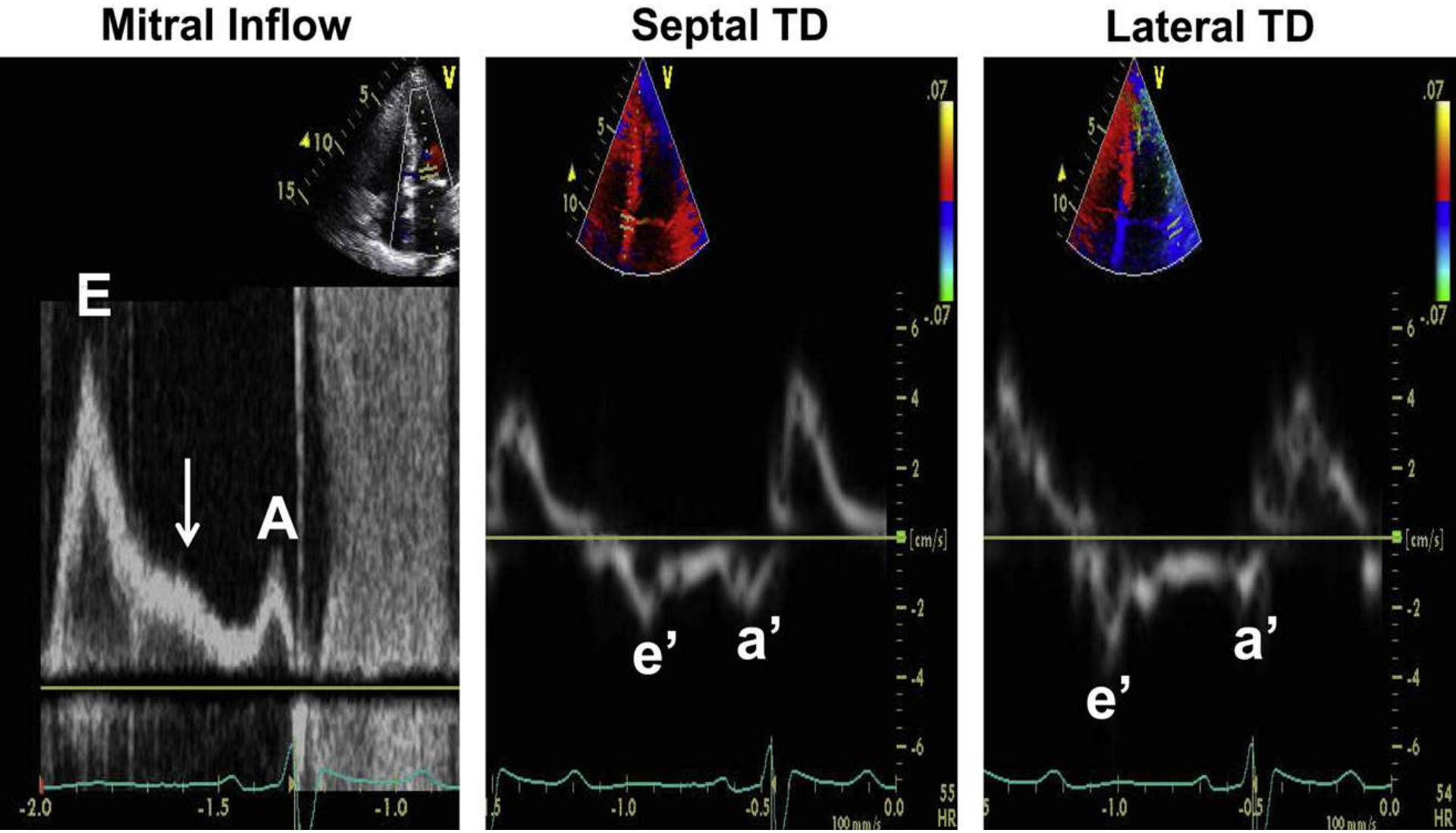
“THE DAGGER”


FR 17Hz
20cm
2D
51%
C 49
P Low
HPen
CE
75%
2.5MHz
WF High
Med
PW
80%
1.6MHz
WF 150Hz
SV4.0mm
7.5cm
-61.6
cm/s
cm/s
-40
-80
-120
-160
100 mm/s

LVEF

- Usually normal or increased
- Can have small LV end-diastolic volumes and therefore reduced stroke volumes despite having normal EFs
- Overt LV systolic dysfunction, termed the ‘‘dilated or progressive phase of HCM,’’ ‘‘end-stage HCM,’’ or ‘‘burnt-out HCM,’’ is usually defined as an LV EF < 50% and occurs in a minority (2%–5%) of patients
- Prognosis is worse in the presence of LV systolic dysfunction

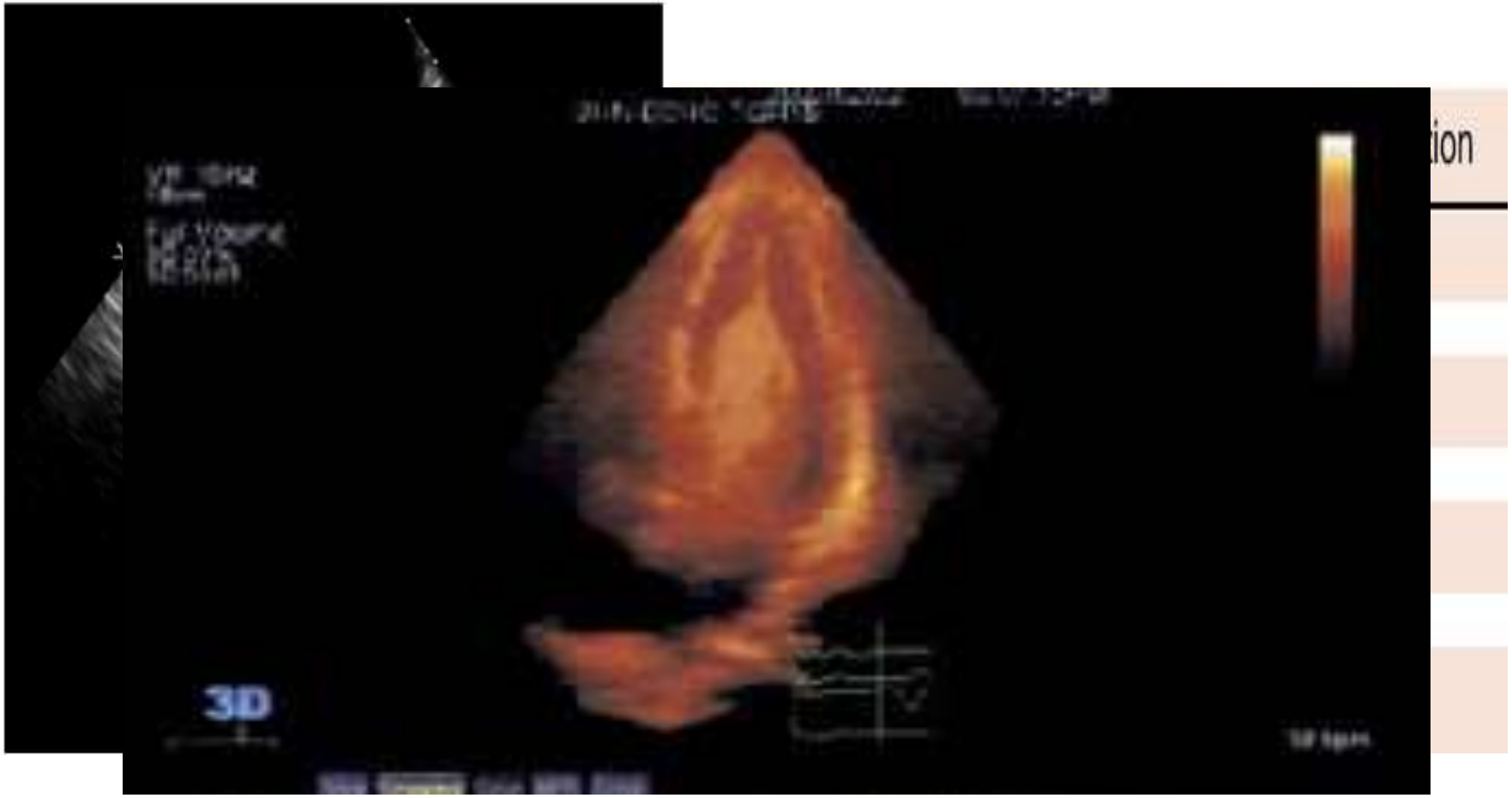
LV DIASTOLIC DYSFUNCTION



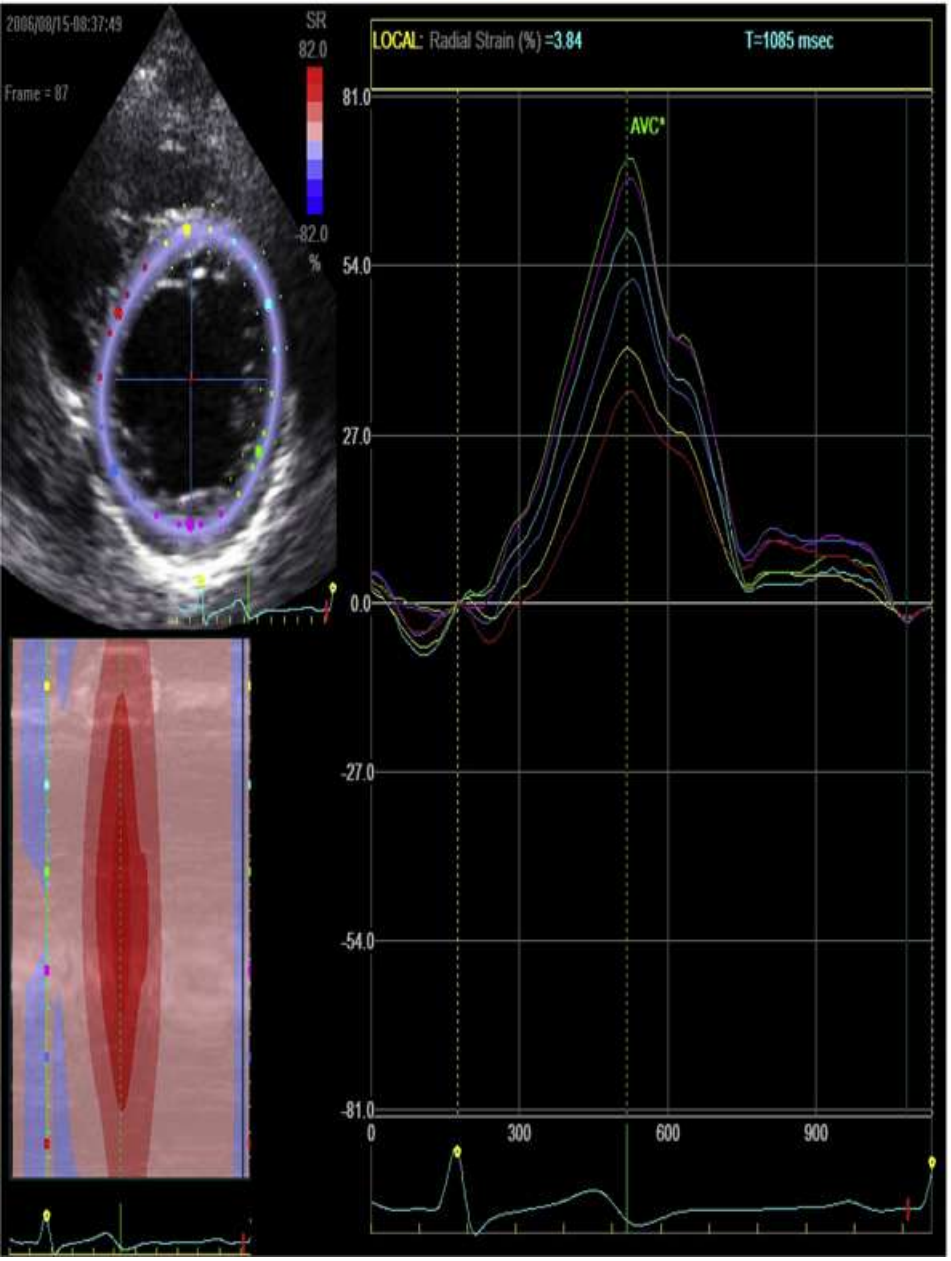
- Assessment of LV diastolic function in a patient with HCM with elevated LA pressure.
- Mitral inflow shows a restrictive inflow pattern (E velocity, 140 cm/sec). The arrow points to an L velocity in middiastole, which is observed in the presence of impaired relaxation and increased filling pressures.
- Lateral annular and septal annular tissue Doppler (TD) velocities (both e’ and a’) are markedly reduced consistent with severely impaired LV relaxation.
- The markedly increased E/e0 ratio is consistent with increased LA pressure > 20 mm Hg.
- The reduced mitral A velocity with its short deceleration time and the severely reduced a’ velocity are consistent with increased LV end-diastolic pressure.

MCE

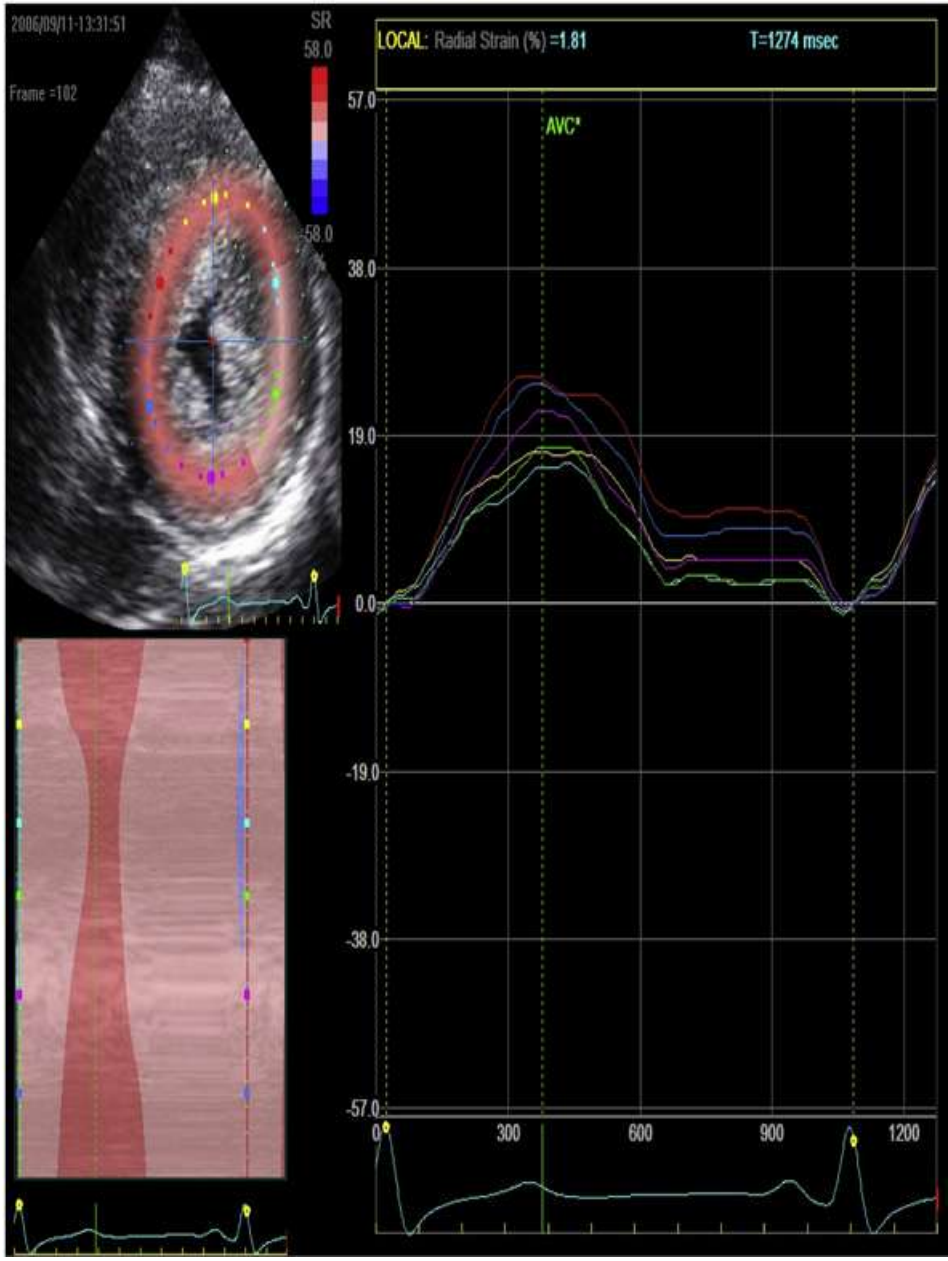
MCE opacifies LV side of septum

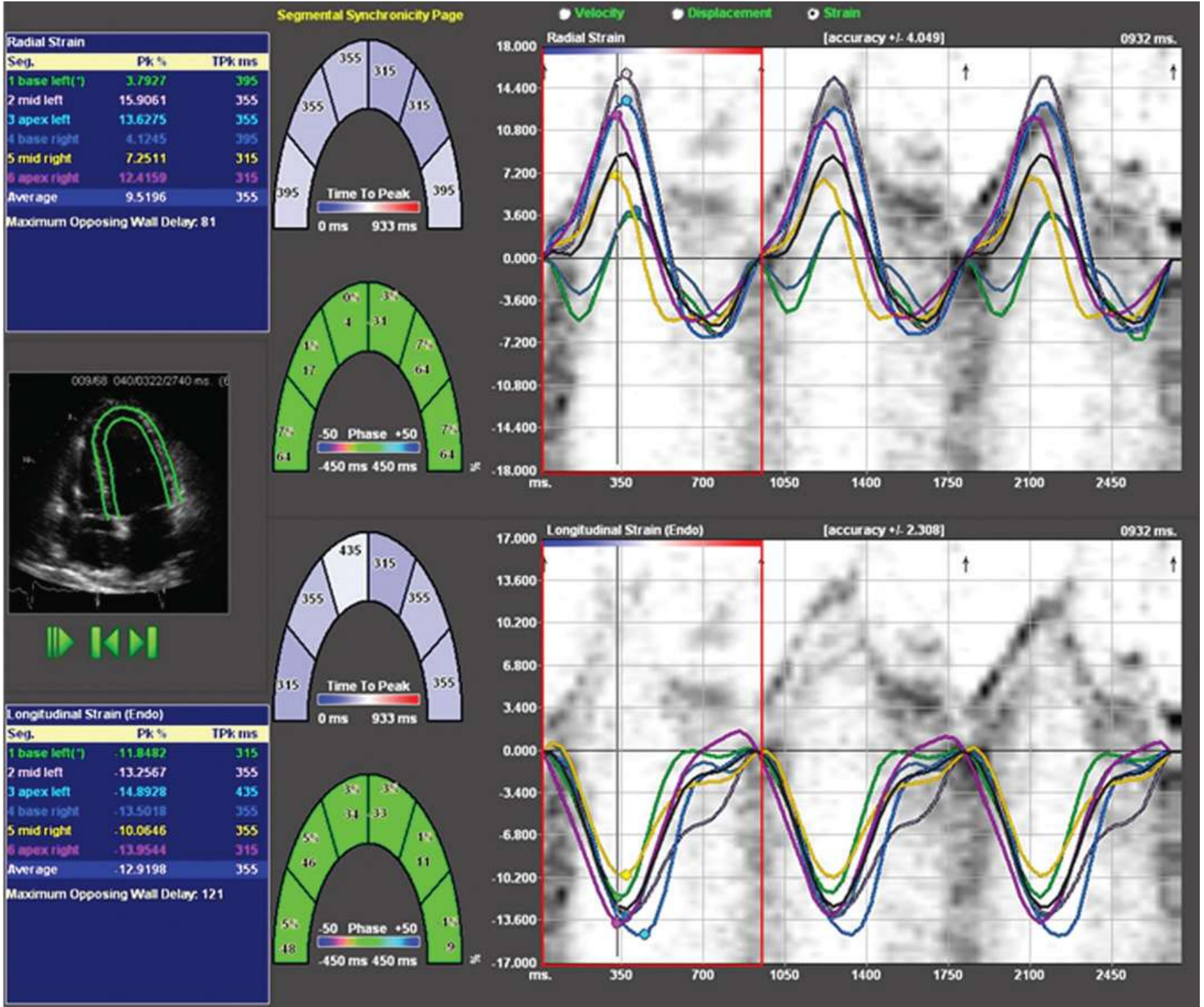


Normal


2006/08/15-08:37:49
Frame = 87
SR
82.0
-82.0
%
LOCAL: Radial Strain (%) =3.84
T=1085 msec
AVC'
81.0
54.0
27.0
0.0
-27.0
-54.0
-81.0
0
300
600
900

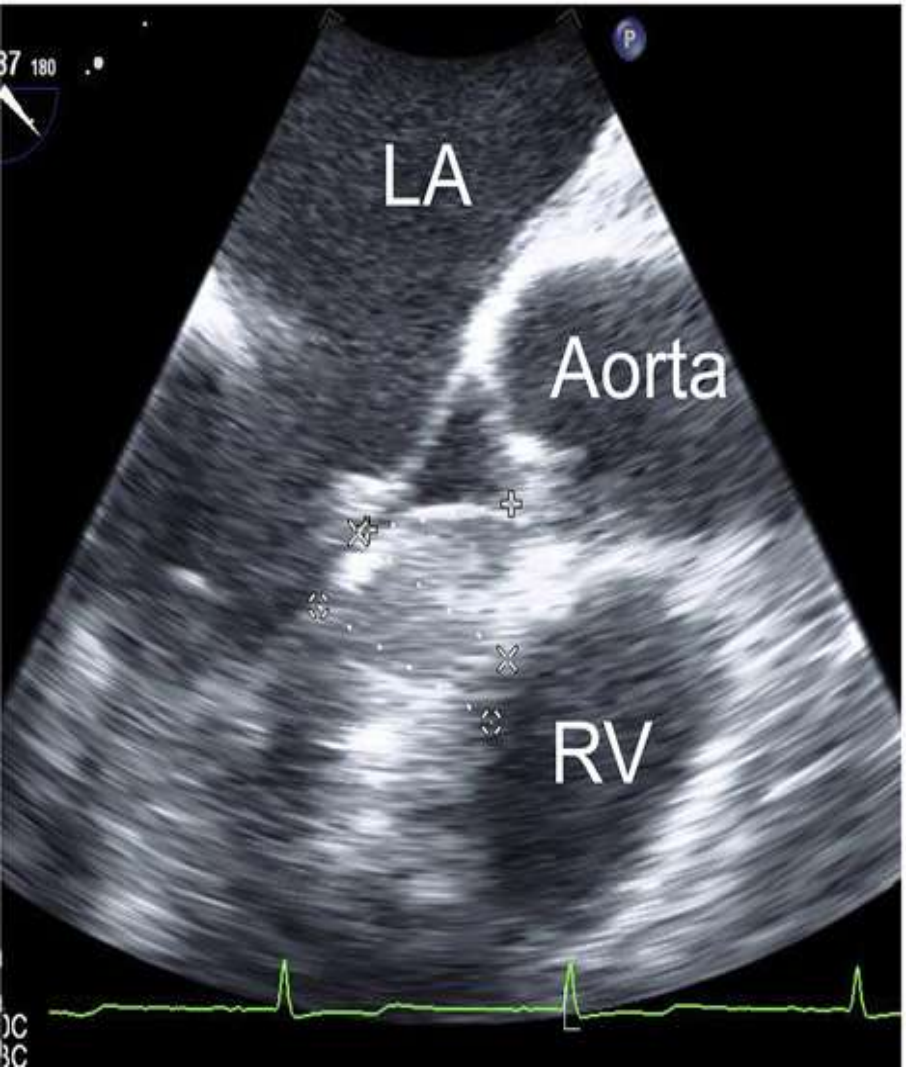
HCM


2006/09/11-13:31:51
Frame =102
SR
58.0
-58.0
%
LOCAL: Radial Strain (%) =1.81
T=1274 msec
AVC'
57.0
38.0
19.0
0.0
-19.0
-38.0
-57.0
0
300
600
900
1200

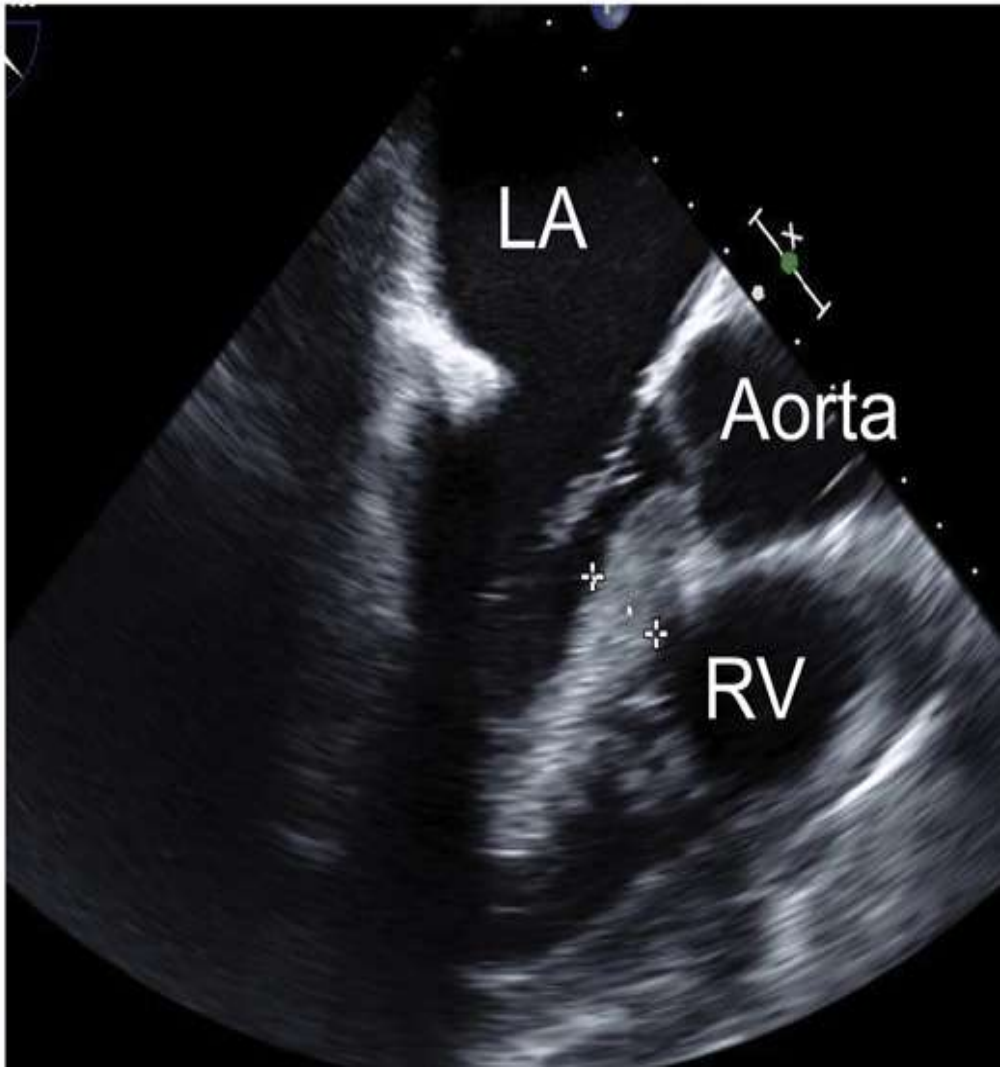

Segmental Synchronicity Page
Velocity
Displacement
Strain
Radial Strain
Seg. Pk % TPk ms
1 base left(*) 3.7927 395
2 mid left 15.9061 355
3 apex left 13.6275 355
4 base right 4.1245 395
5 mid right 7.2511 315
6 apex right 12.4159 315
Average 9.5196 355
Maximum Opposing Wall Delay: 81
Time To Peak
0 ms 933 ms
-50 Phase +50
-450 ms 450 ms
Radial Strain [accuracy +/- 4.049] 0932 ms.
18.000
14.400
10.800
7.200
3.600
0.000
-3.600
-7.200
-10.800
-14.400
-18.000
ms. 350 700 1050 1400 1750 2100 2450
Longitudinal Strain (Endo)
Seg. Pk % TPk ms
1 base left(*) -11.8482 315
2 mid left -13.2567 355
3 apex left -14.8928 435
4 base right -13.5018 355
5 mid right -10.0646 355
6 apex right -13.9544 315
Average -12.9198 355
Maximum Opposing Wall Delay: 121
Longitudinal Strain (Endo) [accuracy +/- 2.308] 0932 ms.
17.000
13.600
10.200
6.800
3.400
0.000
-3.400
-6.800
-10.200
-13.600
-17.000

TOE

Pre - Myectomy

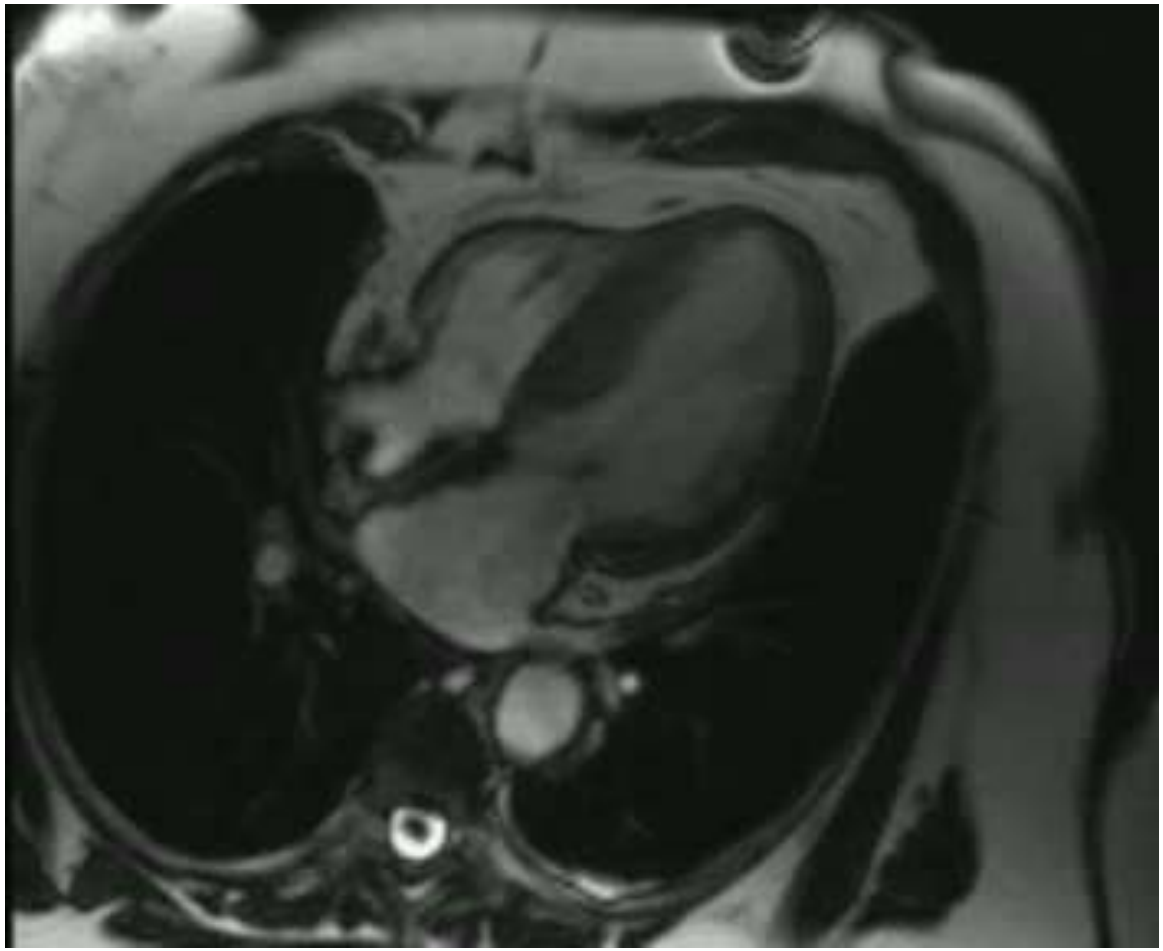


Post - Myectomy



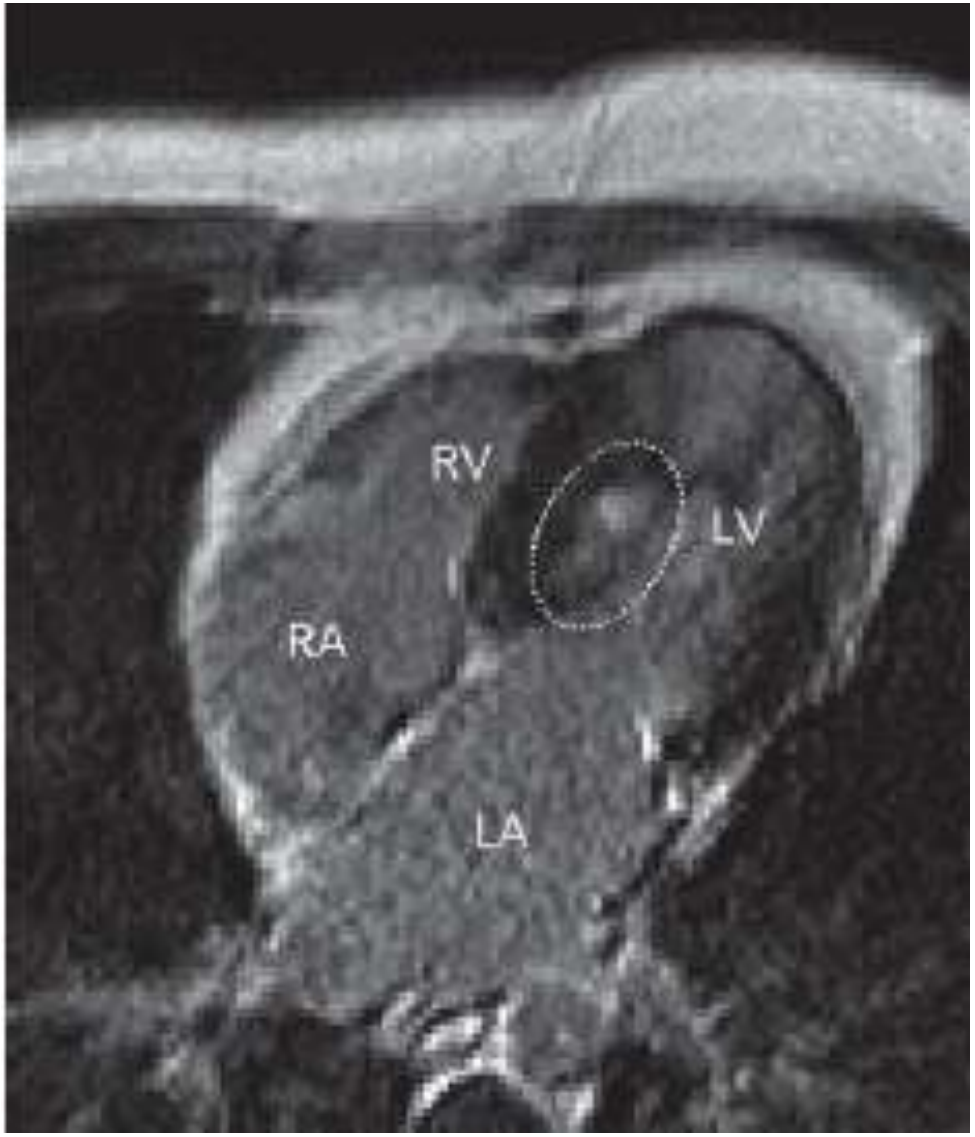
CARDIAC MRI

- Useful when echocardiography is questionable, particularly with apical hypertrophy
- Cines loops typically show obstruction and velocity mapping is useful in the assessment of peak velocities
- SAM of the mitral valve is clearly seen on cardiac MRI
- Improvement in obstruction after septal ablation or myomectomy can be demonstrated, as can the location and size of the associated infarction, which are useful for planning repeat procedures
- Cardiac MRI tagging identifies abnormal patterns of strain, shear, and torsion in cases of HCM, demonstrating significant dysfunction in hypertrophic areas of the ventricle



CARDIAC MRI

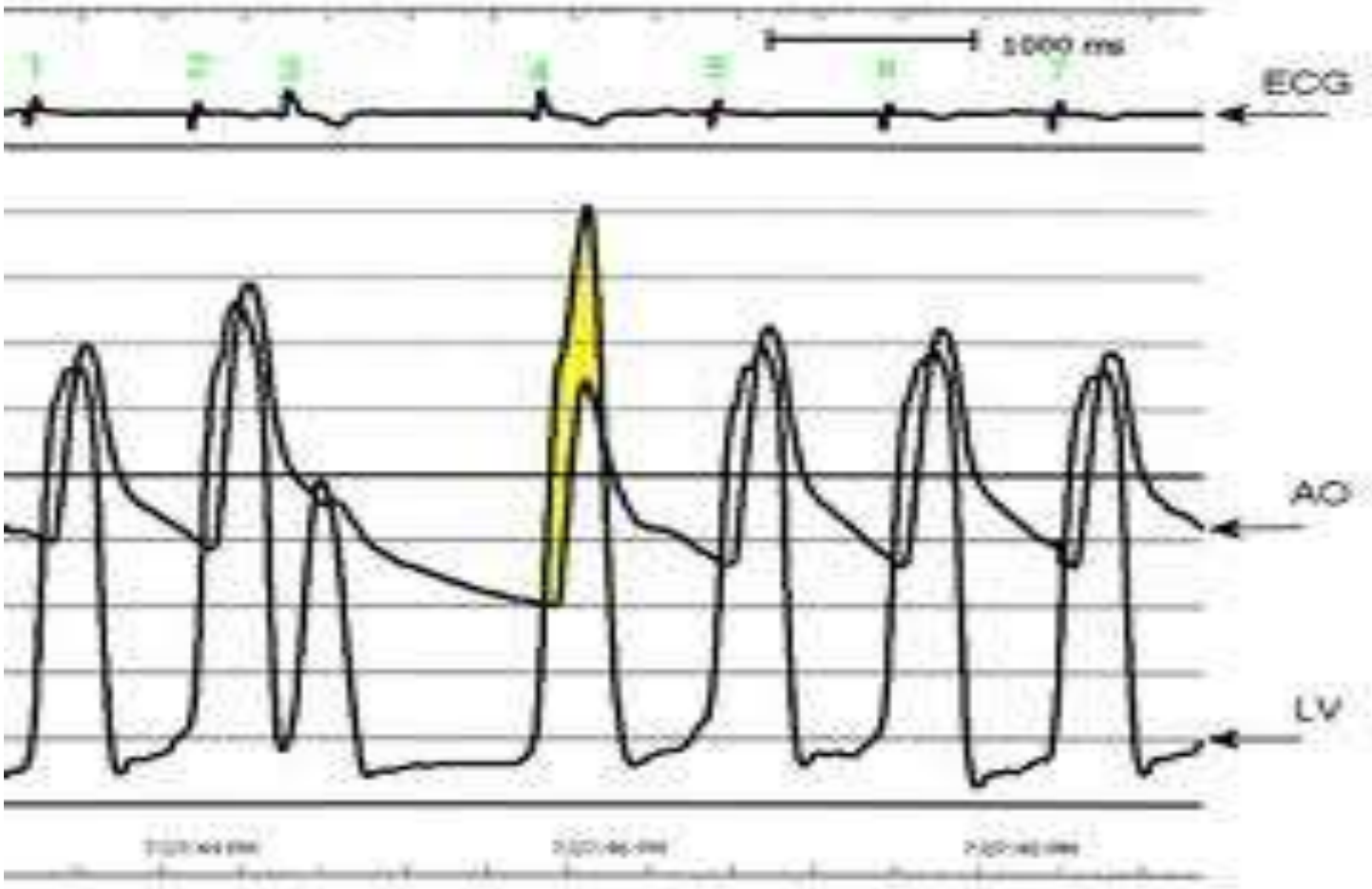
- Gadolinium contrast cardiac MRI - differentiating HCM from other causes of cardiac hypertrophy and other types of cardiomyopathy such as, amyloidosis, athletic heart, and Fabry's disease
- Late gadolinium enhancement occurring in HCM represents myocardial fibrosis
  - The greater the degree of late gadolinium enhancement, the more likely that the particular HCM patient has 2 or more risk factors for sudden death
  - More likely the patient has or will develop progression of ventricular dilation toward heart failure, thereby indicating a poorer prognosis
- Most patients with HCM have no gadolinium enhancement
  - Common benign pattern is 2 stripes running along the junction of the right ventricle insertion into the left ventricle


RV
LV
RA
LA

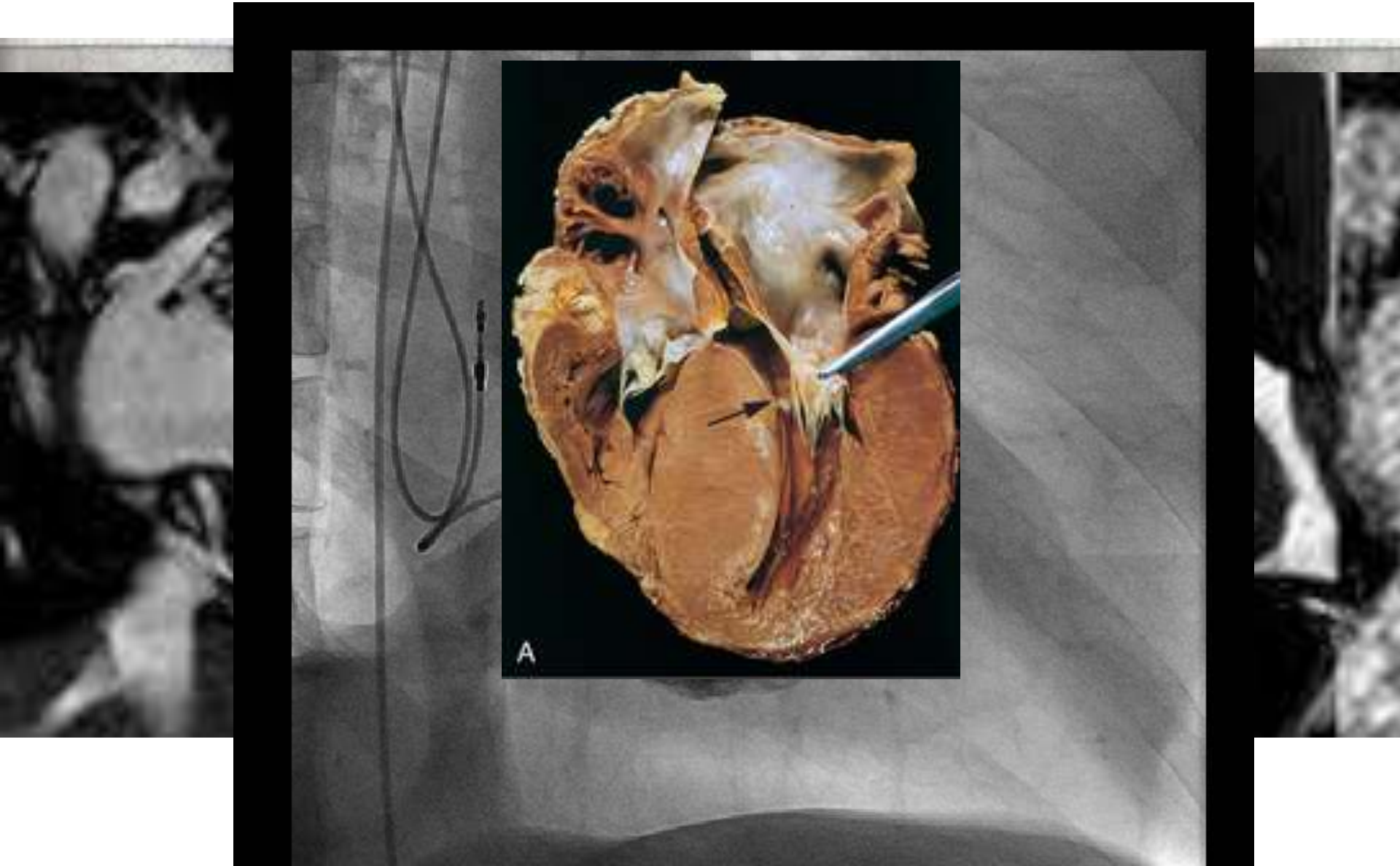
CARDIAC CATHETERIZATION

- Diagnostic cardiac catheterization is useful to determine the degree of LVOT obstruction, cardiac hemodynamics, the diastolic characteristics of the left ventricle, LV anatomy and coronary anatomy
- Reserved for situations when invasive modalities of therapy, such as a pacemaker or surgery, are being considered
- Therapeutic cardiac catheterization interventions, include transcatheter septal alcohol ablation
- The arterial pressure tracing found on cardiac catheterization may demonstrate a "spike and dome" configuration

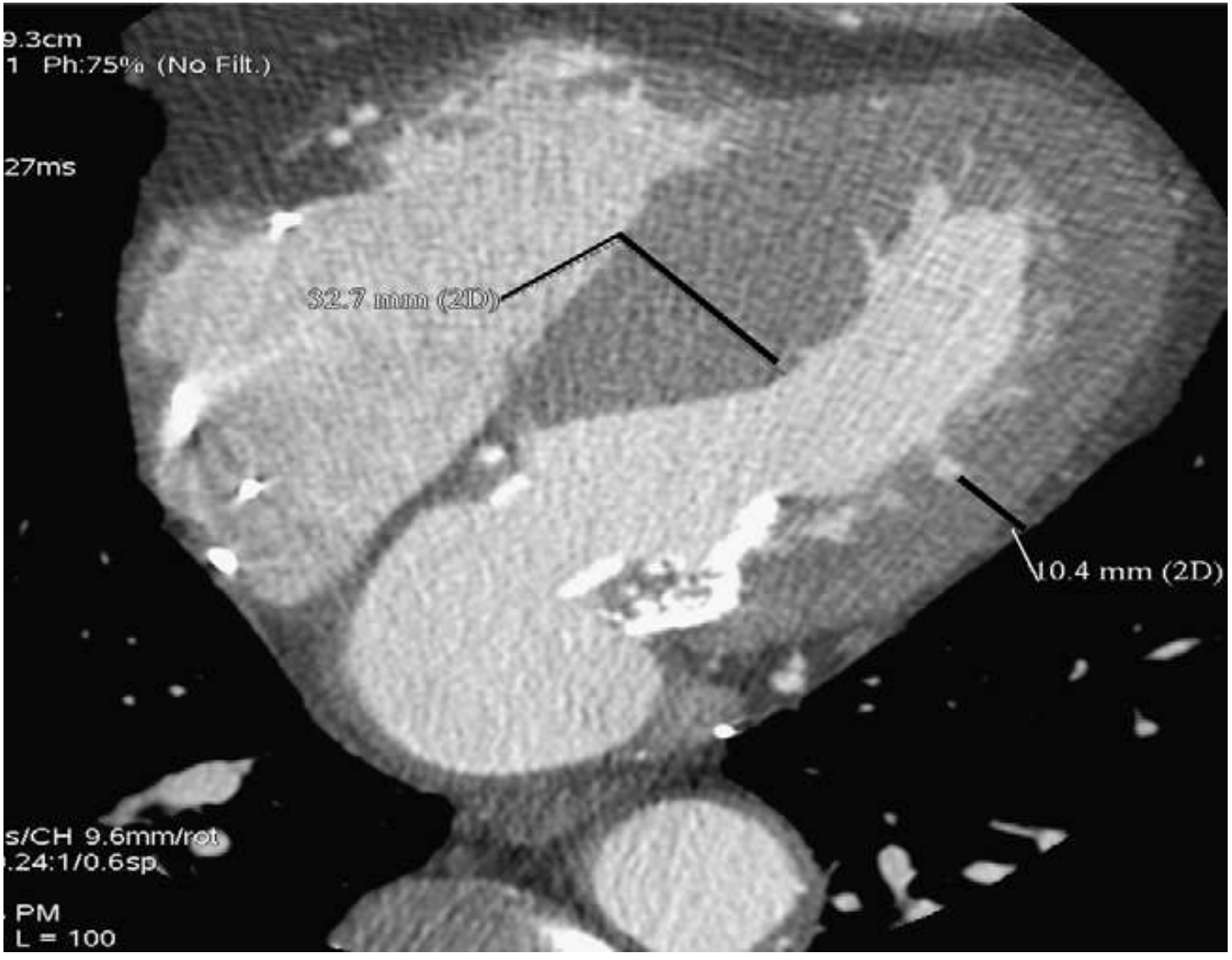
CARDIAC CATHETERIZATION



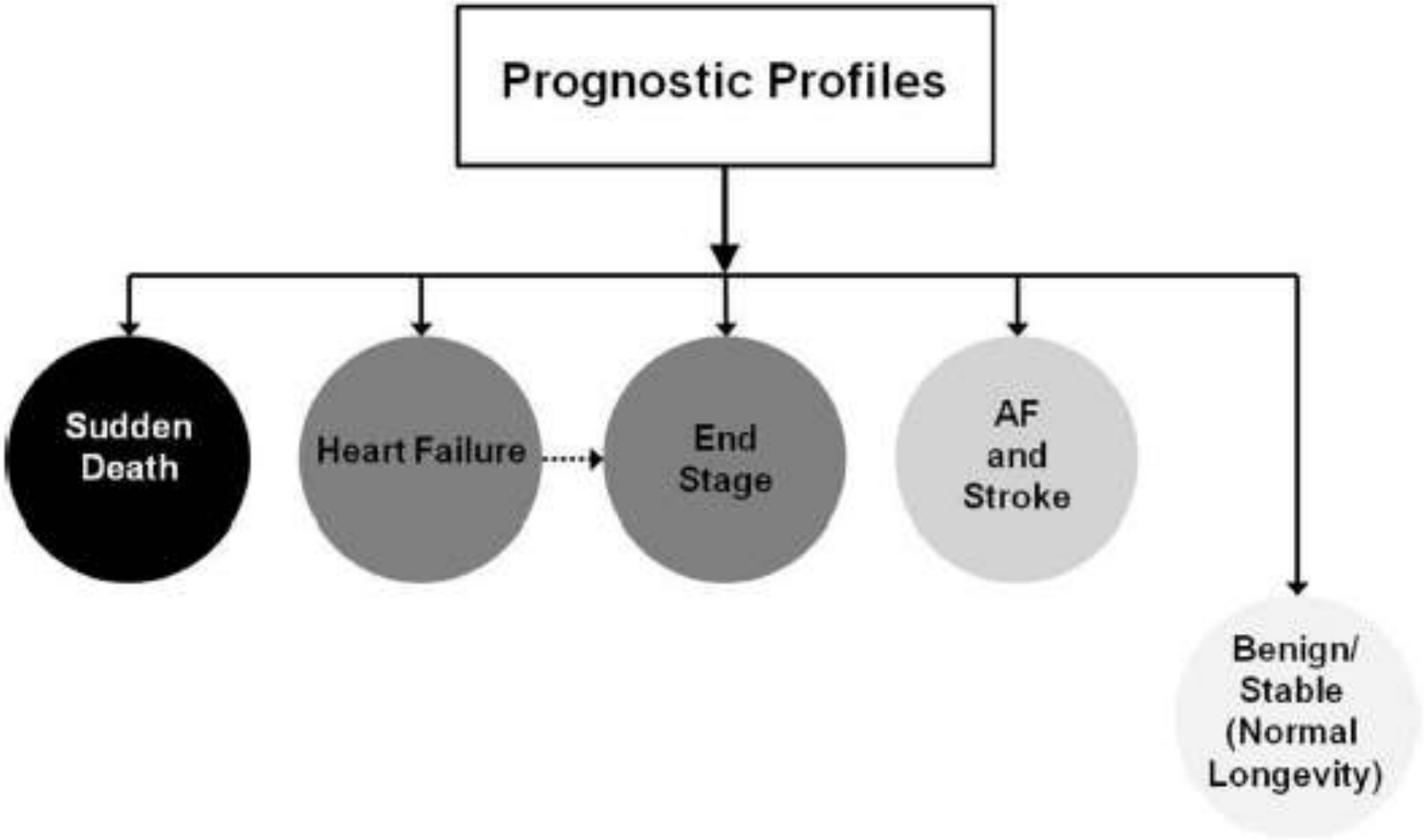
CARDIAC CATHETERIZATION



CARDIAC CT !


9.3cm
1 Ph:75% (No Filt.)
27ms
32.7 mm (2D)
10.4 mm (2D)
s/CH 9.6mm/rot
.24:1/0.6sp
PM
L = 100

NATURAL HISTORY



BURNT OUT HCM

- Burnt out HCM - 3%
- Systolic dysfunction EF <50%
- Associated with AF
- Wall thinning and cavity dilation
- Diffuse transmural scarring
- Progression to refractory heart failure or sudden death
- Mortality 10%/year
- Most reliable risk marker - a family history of the end stage

Medical Treatment

Beta blockers (Class 1)

- Slowing heart rate
- Reducing force of LV contraction
- Augmenting ventricular filling and relaxation
- Decreasing myocardial oxygen consumption
- Long-acting preparations - propranolol, atenolol, metoprolol or nadolol
- Blunt LV outflow gradient triggered by physiologic exercise.
- Target resting heart rate - 60 beats/min
- May require up to 400 mg equivalent of metoprolol

Verapamil (class I)

- Add on therapy to beta blockers if high doses of beta blockers are not tolerated
- First choice when beta blockers are contraindicated
- Maximal doses of 280 mg/day
- AVOID in NYHA class IV dyspnoea and hypotension
- When used as add-on therapy→to look for high grade AV block
- Diltiazem – when verapamil cannot be used

Disopyramide

- Negative inotropic effect decreases the gradient and improve symptoms.
- Concomitant beta blockade may be important to prevent rapid atrioventricular node conduction
- Between 300 and 600 mg/d
- The corrected QT interval must be monitored
- Anticholinergic side effects in older patients

Table. Agents That Modulate Myocardial Metabolic Efficiency and Their Major Mechanism(s) for Potentially Improving Energetics

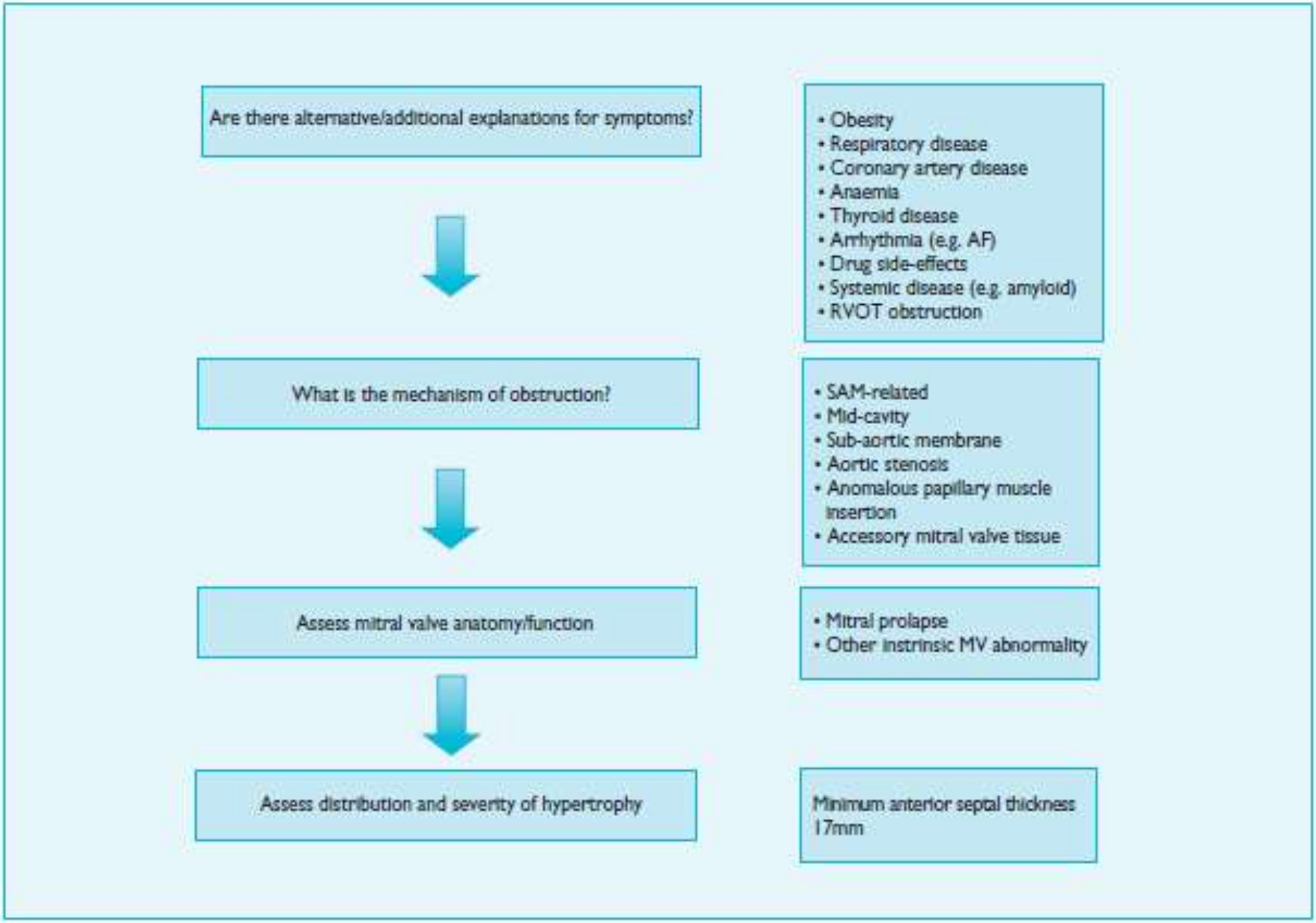
Agent	Mechanisms of Action
Perhexiline	CPT_1/CPT_2 inhibition
	NAD(P)H oxidase inhibition
	NO potentiation
Amiodarone	CPT_1 inhibition
	β-Adrenoceptor blockade
Trimetazidine	PFOX inhibition
	? CPT_1 inhibition
Ranolazine	PFOX inhibition
	? Late Na^+ current inhibition
Metformin	AMPK stimulation

DRUGS THAN CAN HARM(CLASS III)

- Nifedipine, Nitrates, Diuretics -potent vasodilator and hence avoided
- Digoxin, dobutamine, noradrenaline, dopamine-positive inotropes
- **MANAGEMENT OF ACUTE HYPOTENSION IN HOCM**
  - i.v fluids
  - phenylephrine
  - Iv beta blockers
- **MANAGEMENT OF HCM WITH LV SYSTOLIC DYSFUNCTION**
  - ACEI/ARBS, diuretics-standard HF treatment
  - Discontinue verapamil, diltiazem, disopyramide (class iii)

MANAGEMENT OF AF

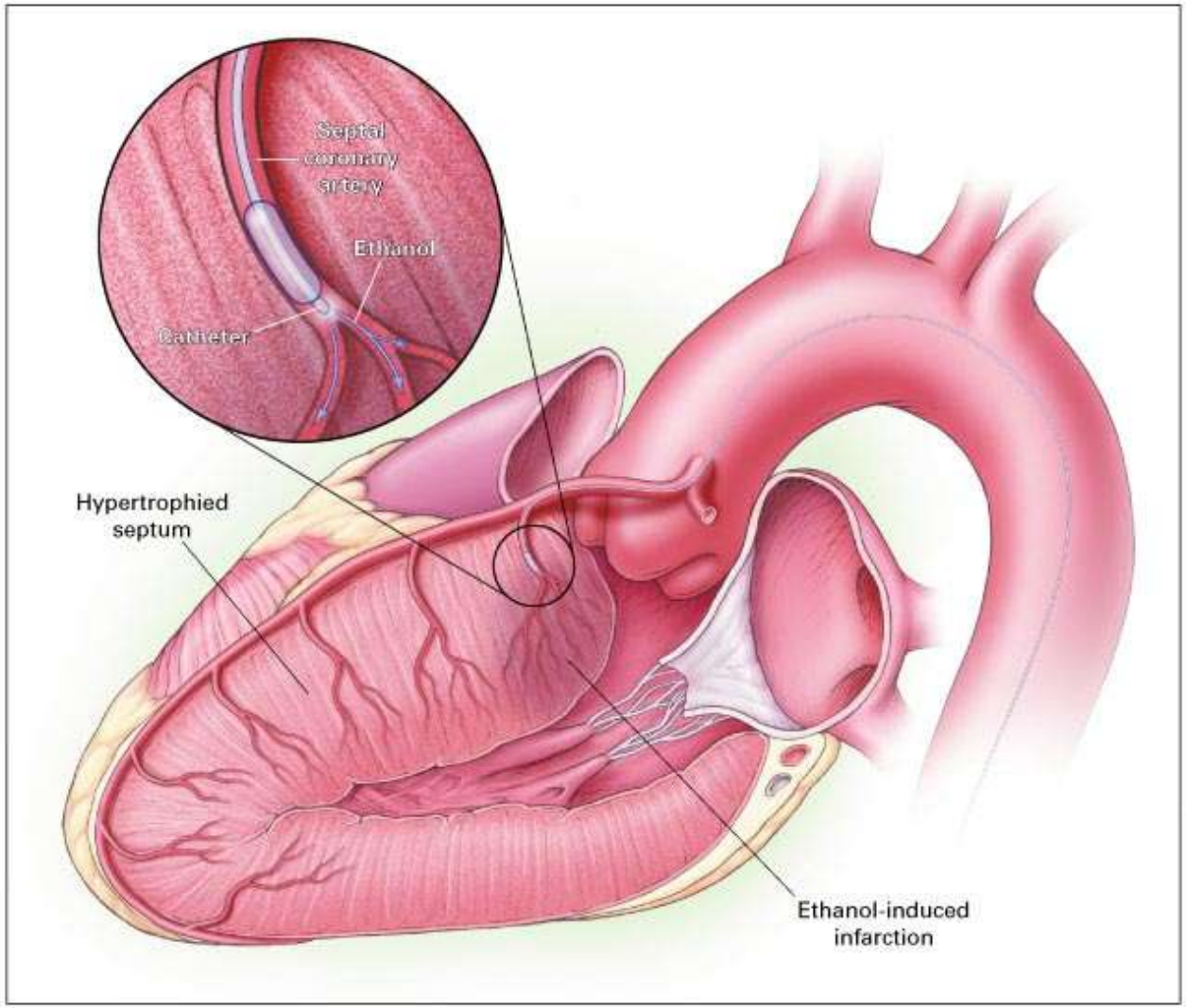
- A/c AF+ hemodynamically unstable= DC
- A/C AF + angina /pulmonary edema = BB/A–iv
- Hemodynamically stable= 3W OAC >>DC
- Rate control with oral BB/ CCB
- rhythm control with amiodarone
- Flecainide and propafenone, should be avoided as they may prolong QRS duration and the QT intervaL
- AV nodal ablation F/B DDD/VVIR/CRT IN drug refractory
- Radiofrequency ablation (II A)

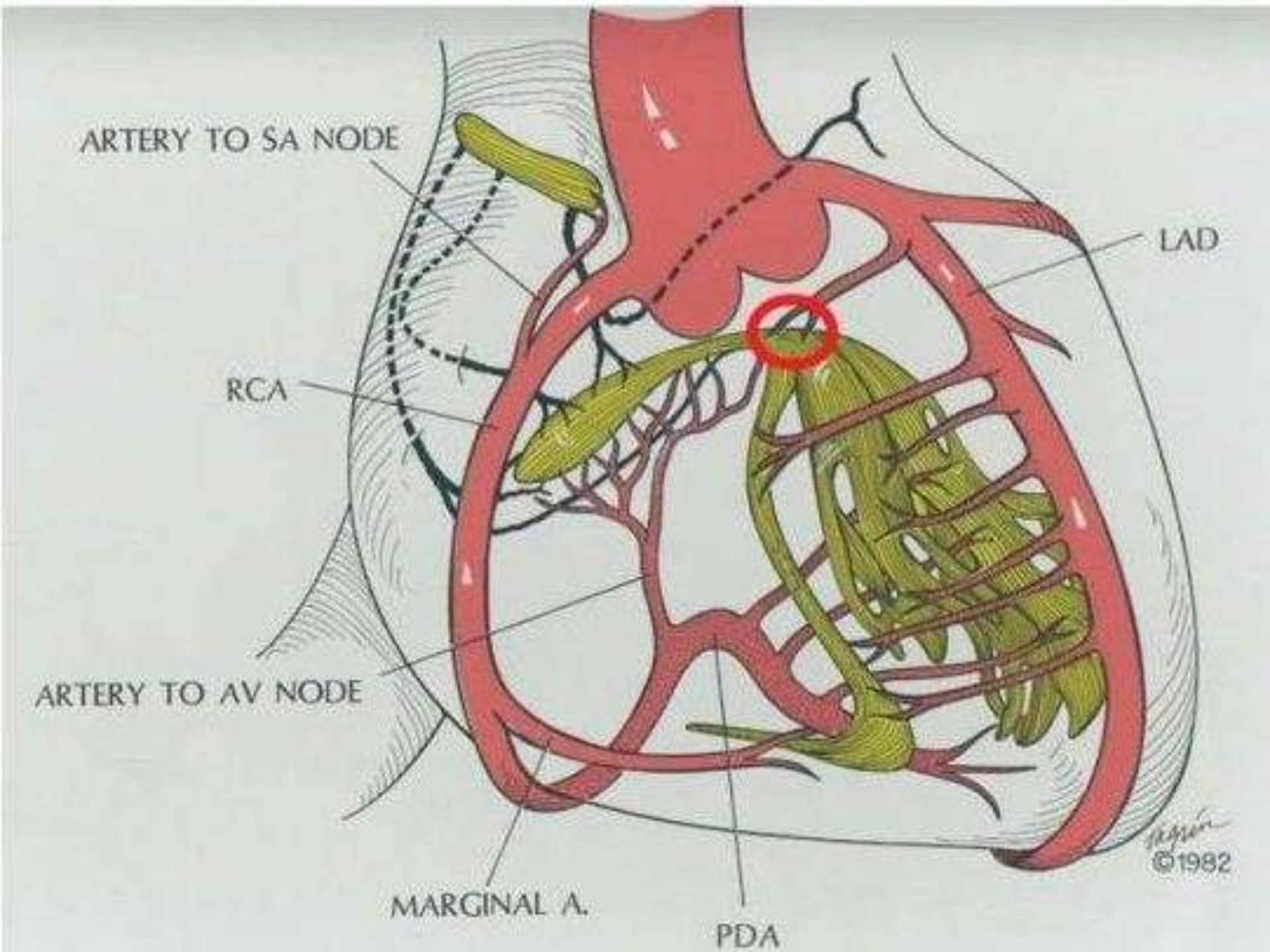

Are there alternative/additional explanations for symptoms?
• Obesity
• Respiratory disease
• Coronary artery disease
• Anaemia
• Thyroid disease
• Arrhythmia (e.g. AF)
• Drug side-effects
• Systemic disease (e.g. amyloid)
• RVOT obstruction
What is the mechanism of obstruction?
• SAM-related
• Mid-cavity
• Sub-aortic membrane
• Aortic stenosis
• Anomalous papillary muscle insertion
• Accessory mitral valve tissue
Assess mitral valve anatomy/function
• Mitral prolapse
• Other instrinsic MV abnormality
Assess distribution and severity of hypertrophy
Minimum anterior septal thickness 17mm

SEPTAL MYECTOMY (Morrow procedure)

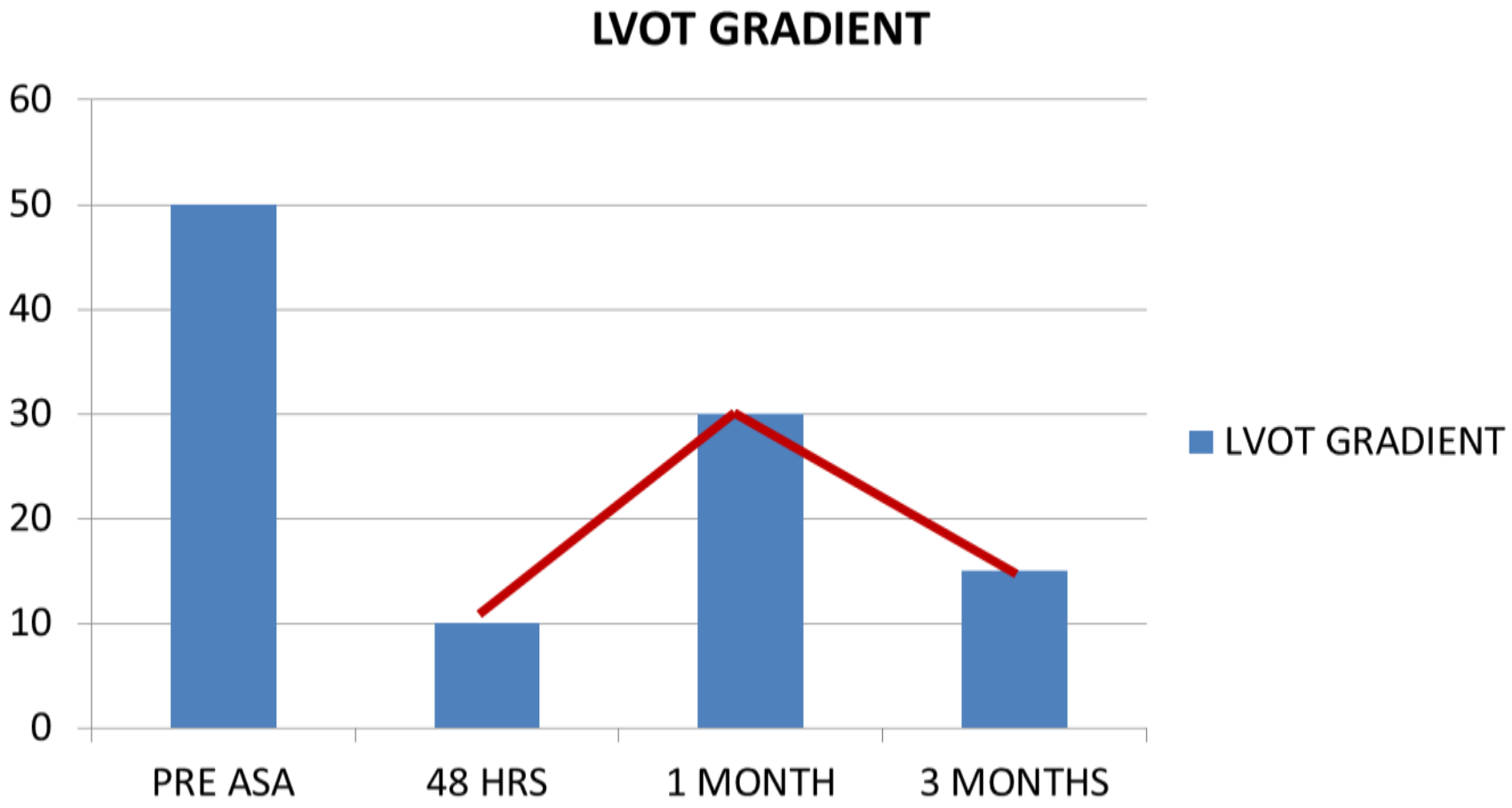
1. Drug-refractory heart failure symptoms
2. NYHA Classes III and IV or exertional syncope
3. LV outflow obstruction
   - gradient ≥ 50 mm Hg
4. Better surgical candidates
   1. Younger age
   2. Greater septal thickness (≥30 mm)
   3. Concomitant cardiac diseases –valvular/CAD
5. **Complications**
   1. Perioperative Mortality (1-5%)
   2. VSD - 3%
   3. CHB and PPI (5%)
   4. CVA 1-2%
   5. LBBB-40%

Alcohol-Induced Septal Ablation




ARTERY TO SA NODE
LAD
RCA
ARTERY TO AV NODE
MARGINAL A.
PDA
©1982

TRIPHASIC RESPONSE



OTHER THAN ALCOHOL!!!

- Polyvinyl alcohol foam particles,
- Microspheres,
- Absorbable gelatin sponges,
- Septal coils

- Gross CM, Schulz-Menger J, Kramer J, Siegel I, Pilz B, Waigand J, Friedrich MG, Uhlich F, Dietz R. Percutaneous transluminal septal artery ablation using polyvinyl alcohol foam particles for septal hypertrophy in patients with hypertrophic obstructive cardiomyopathy: acute and 3-year outcomes. *J Endovasc Ther*2004;11:705–711.
- Llamas-Esperon GA, Sandoval-Navarrete S. Percutaneous septal ablation with absorbable gelatin sponge in hypertrophic obstructive cardiomyopathy. *Catheter Cardiovasc Interv* 2007;69:231–235.
- Lafont A, Durand E, Brasselet C, Mousseaux E, Hagege A, Desnos M. Percutaneous transluminal septal coil embolisation as an alternative to alcohol septal ablation for hypertrophic obstructive cardiomyopathy. *Heart*2005;91:92

Updated Meta-Analysis of Septal Alcohol Ablation Versus Myectomy for Hypertrophic Cardiomyopathy

Shikhar Agarwal, MD, MPH,* E. Murat Tuzcu, MD,† Milind Y. Desai, MD,†
Nicholas Smedira, MD,‡ Harry M. Lever, MD,† Bruce W. Lytle, MD,‡ Samir R. Kapadia, MD,†
Cleveland, Ohio

Conclusion

SA does seem to show promise in treatment of HOCM owing to ***similar mortality rates as well as functional status*** compared with SM; however, the caveat is ***increased conduction abnormalities and a higher post-intervention LVOTG***. The choice of treatment strategy should be made after a thorough discussion of the procedures with the individual patient.
(J Am Coll Cardiol 2010;55:823–34)

DUAL-CHAMBER PACING

- Proposed benefit:
  - Pacing the RV apex will decrease the outflow tract gradient by decreasing projection of basal septum into LVOT
- Several RCTs have found that the improvement in subjective measures provided by dual-chamber pacing is likely a placebo effect
- Other treatment
  - EP
  - CRT
  - CARDIAC TRANSPLANTATION/VAD

EFFICACY OF THERAPEUTIC STRATEGIES

Table 3. Comparative Features of Septal-Reduction Therapies.

Therapy	Mortality	Residual Gradient	Effectiveness	Follow-up	Complications		Time to Resolution of Gradient
	%	*mm Hg*	*% of Patients*	*Yr*	*Type*	*% of Patients*	
Dual-chamber pacing	<1	<40	10–40	10	Infection or perforation	<2	4 wk
Septal myectomy*	<2–3	<10	>90	>30	Complete heart block Ventricular septal defect Aortic regurgitation	<3 <1 <1	Immediate
Septal ablation†	<2–3	<20	70–80	<5	Complete heart block Ventricular septal defect Large myocardial infarction	10–40 Unknown Unknown	8–12 wk

* Surgical septal myectomy is the only intervention that can treat concomitant problems, such as multivessel coronary disease, intrinsic mitral-valve disease, midventricular obstruction, and fixed subaortic obstruction.

† The true rates of death and complications may be underestimated, since complications may occur at a higher frequency in the inexperienced centers and may be underreported.

Nishimura et al. NEJM. 2004. 350(13):1323.

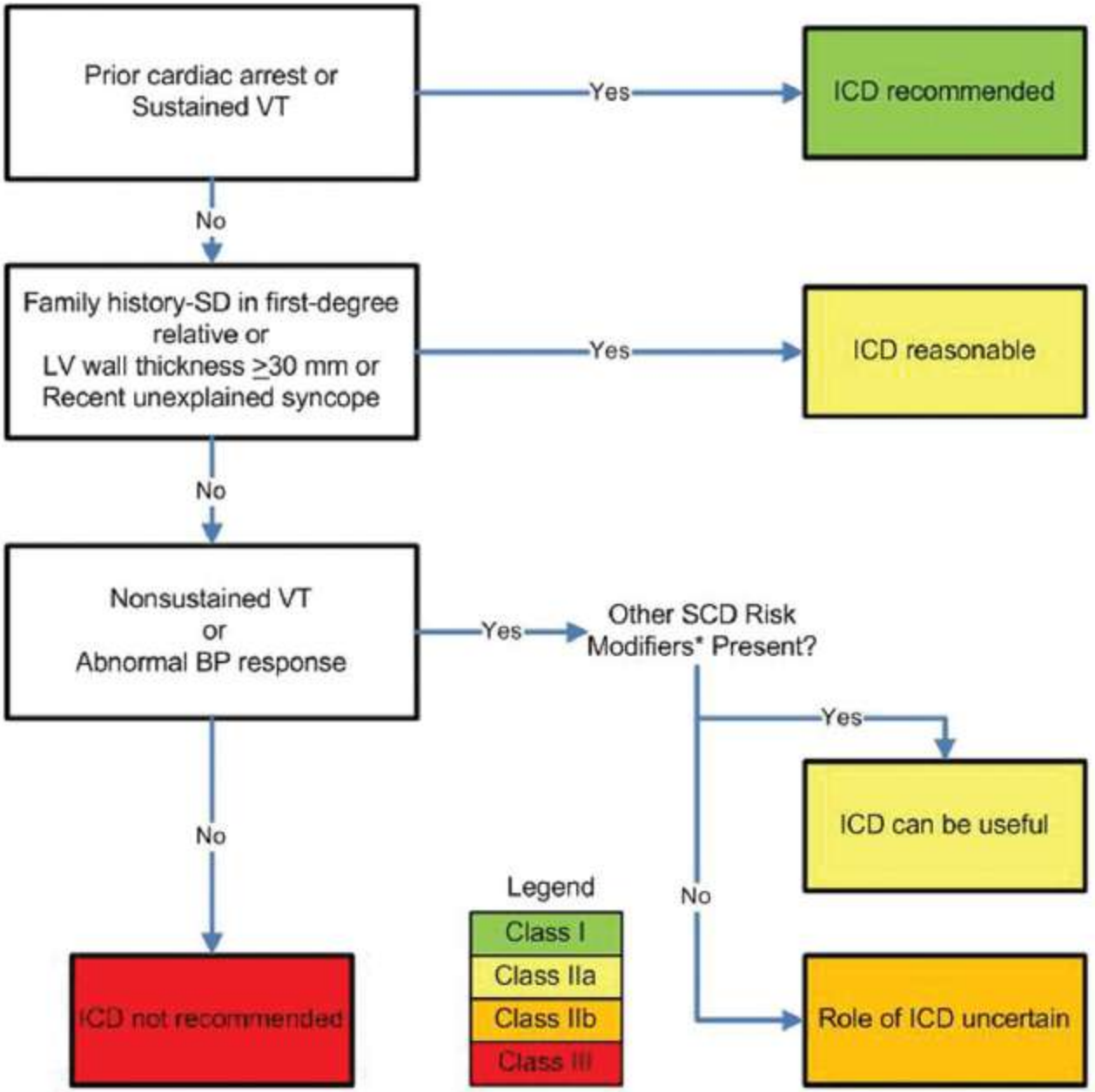
PRIMARY PREVENTION SECONDARY PREVENTION

HCM Risk-SCD Calculator

Age	**Years**	*Age at evaluation*
Maximum LV wall thickness	**mm**	*Transthoracic Echocardiographic measurement*
Left atrial size	**mm**	*Left atrial diameter determined by M-Mode or 2D echocardiography in the parasternal long axis plane at time of evaluation*
Max LVOT gradient	**mmHg**	*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 Bernouilli equation: Gradient= $4V^2$, where V is the peak aortic outflow velocity*
Family History of SCD	○ No ○ Yes	*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).*
Non-sustained VT	○ No ○ Yes	*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.*
Unexplained syncope	○ No ○ Yes	*History of unexplained syncope at or prior to evaluation.*

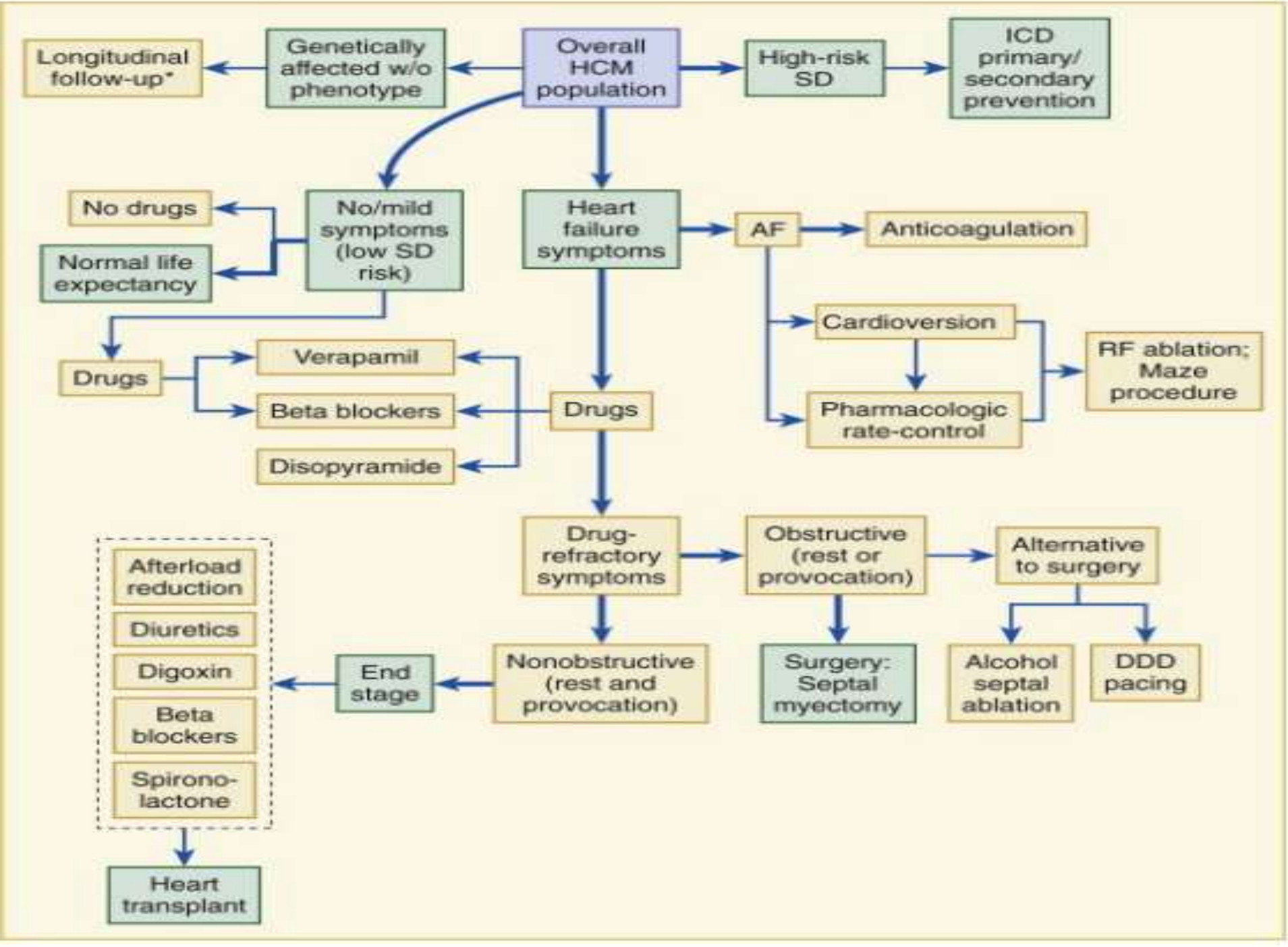
Risk of SCD at 5 years (%):	
ESC recommendation:	

Reset


Prior cardiac arrest or Sustained VT
Yes
ICD recommended
No
Family history-SD in first-degree relative or
LV wall thickness ≥30 mm or
Recent unexplained syncope
Yes
ICD reasonable
No
Nonsustained VT
or
Abnormal BP response
Yes
Other SCD Risk Modifiers* Present?
Yes
ICD can be useful
No
Role of ICD uncertain
No
ICD not recommended
Legend
Class I
Class IIa
Class IIb
Class III

Algorithm to identify at-risk HCM patients for implantation with an ICD

First degree risk factors	Definition
Positive family history of sudden cardiac death	Cases with SCD <45 years
Recurrent syncope	≥ 2 incidents
LVH	≥ 30 mm at any site in the LV
Abnormal blood pressure response during exercise	Increase <20 mm Hg or fall >20 mm Hg after transient increase
Non-sustained VT in Holter ECG	≥ 3 consecutive QRS complexes with a heart rate of ≥ 120 bpm.
Second degree risk factors	
Atrial fibrillations/atrial flutter	Any form, provided cannot be eliminated
LA dilatation	>45 mm (in m-mode ECG)
High LVOT gradient at rest	>80 mm Hg (CW Doppler)
Evidence of myocardial ischemia during exercise	
Early manifestation of HCM	<30 years of age
Myocardial bridging near the LAD	in younger patients (<45 years)
Marked fibrosis in cardiac MRI	fibrosis of ≥ 2 segments in a 17-segment model of the LV


Longitudinal follow-up*
Genetically affected w/o phenotype
Overall HCM population
High-risk SD
ICD primary/ secondary prevention
No drugs
Normal life expectancy
No/mild symptoms (low SD risk)
Heart failure symptoms
AF
Anticoagulation
Cardioversion
RF ablation; Maze procedure
Pharmacologic rate-control
Drugs
Verapamil
Beta blockers
Disopyramide
Drugs
Drug-refractory symptoms
Obstructive (rest or provocation)
Alternative to surgery
Afterload reduction
Diuretics
Digoxin
Beta blockers
Spirono-lactone
End stage
Nonobstructive (rest and provocation)
Surgery: Septal myectomy
Alcohol septal ablation
DDD pacing
Heart transplant

PREGNANCY/DELIVERY

- High risk for- LVOT gradient > 50 mmhg
- Class III for those with class III/IV systolic dysfunction
- No added risk for patients with controlled symptoms (II A)
- Continue drugs in prgnancy (class I)-watch or fetal bradycardia and growth abnormalities in fetus
- Guard against post delivery volume loss

PHYSICAL ACTIVITIES

- Low intensity aerobic exercises
- Avoid dehydration
- Avoid heavy meals
- Risk of syncope in high intensity sports
- Unpredictability of SCD→an other reason to avoid high intensity sports

What to expect in future!!!

- ASA OVERTAKING SURGERY
- ALTERNATIVES TO ALCOHOL
- INCLUSION OF CARDIAC MRI IN DIAGNOSIS AND RISK STRATIFICATION
- PRECLINICAL DIAGNOSIS WITH ECHO

FEW VIDEOS

HCM

MYOMECTOMY

ASA

Thank You