# Echocardiography in Hypertrophic Cardiomyopathy

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Morristown Medical Center/Atlantic Health System

Chanin T Mast Center for Hypertrophic Cardiomyopathy

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# No disclosures

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# Goals

- Understand the echocardiographic features of HCM
- Understand the importance of the mitral valve in HCM
- Learn imaging tips including the use of contrast
- Recognize SAM

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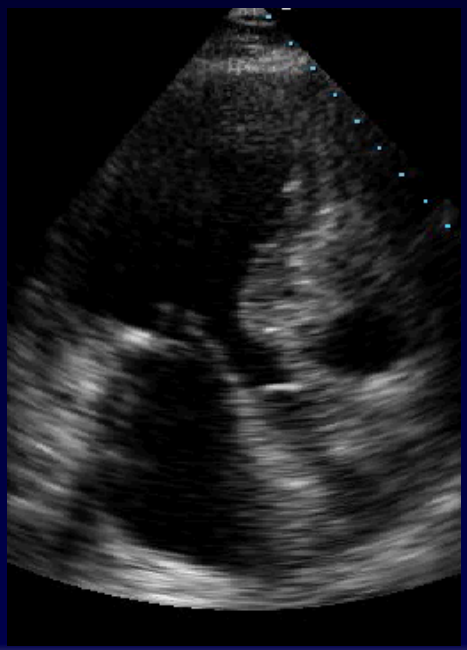

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# In Hypertrophic Cardiomyopathy, Echocardiography can ….

- Establish the diagnosis/characterize the disease
  - Define patterns of hypertrophy (LV and RV)
  - Assess systolic function
  - Assess diastolic function
  - Quantitate obstruction (at rest and with maneuvers)
  - Assess concomitant mitral valve abnormalities and regurgitation
  - Risk stratify
  - *Differentiate from athlete's heart*
  - *Differentiate from restrictive CM*
- Help guide myectomy and alcohol ablation
- Stress testing in HCM

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American Society of Echocardiography Clinical Recommendations for Multimodality Cardiovascular Imaging of Patients with Hypertrophic Cardiomyopathy

(J Am Soc Echocardiogr 2011;24:473-98.)

**Table 1** Echocardiographic evaluation of patients with HCM

| |
|---|
| 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 |

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**ACCF/AHA Guideline**

**2011 ACCF/AHA Guideline for the Diagnosis and Treatment of Hypertrophic Cardiomyopathy: Executive Summary**

**A Report of the American College of Cardiology Foundation/American Heart Association Task Force on Practice Guidelines**

*Developed in Collaboration With the American Association for Thoracic Surgery, American Society of Echocardiography, American Society of Nuclear Cardiology, Heart Failure Society of America, Heart Rhythm Society, Society for Cardiovascular Angiography and Interventions, and Society of Thoracic Surgeons*

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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 (France), Giuseppe Limongelli (Italy), Heiko Mahrholdt (Germany), William J. McKenna (UK), Jens Mogensen (Denmark), Petros Nihoyannopoulos (UK), Stefano Nistri (Italy), Petronella G. Pieper (Netherlands), Burkert Pieske (Austria), Claudio Rapezzi (Italy), Frans H. Rutten (Netherlands), Christoph Tillmanns (Germany), Hugh Watkins (UK).**

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# Defining "Hypertrophy"

- Wall thickness (any segment) >15 mm (?12 mm women)
- Septal to posterior wall thickness ratio >1.3 in normotensive pts. or 1.5 in hypertensive pts.

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## Define patterns of hypertrophy

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## Presence of hypertrophy and its distribution

- Classically asymmetric but can be in any pattern and at any location, including the right ventricle
- Although septal predominance is more common, hypertrophy can be isolated to the LV free wall or apex
- RV hypertrophy (may be in any location) is rare
  - >5 mm subcostal window

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# Asymmetric Septal Hypertrophy

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# Asymmetric septal hypertrophy (with SAM)


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# Asymmetric septal hypertrophy


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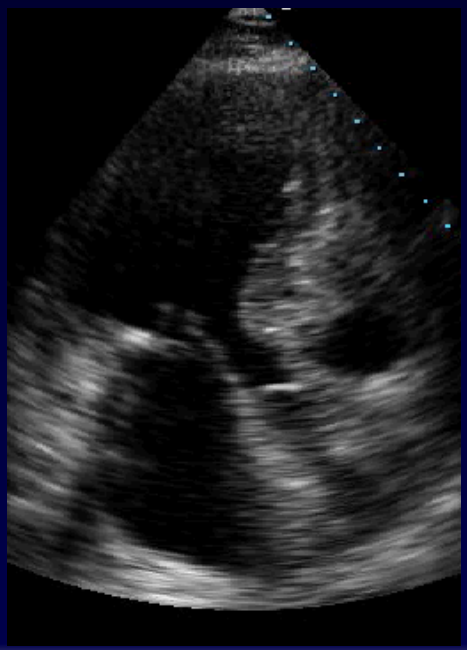

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# Multiple wall hypertrophy


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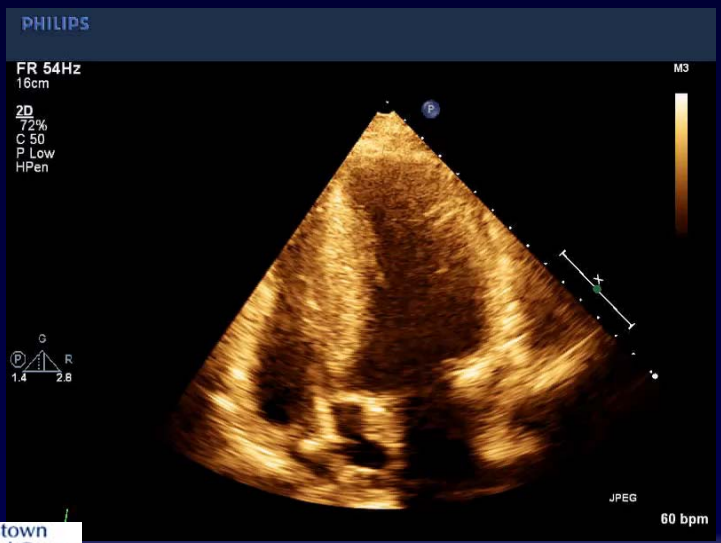
# Apical lateral wall hypertrophy

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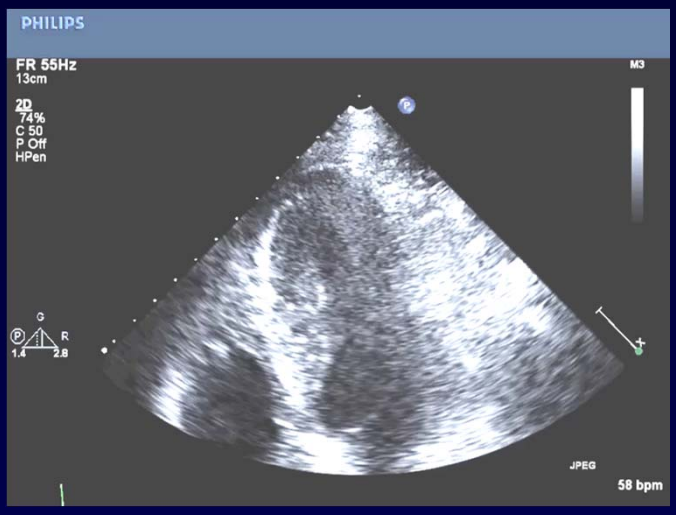
# Apical hypertrophy

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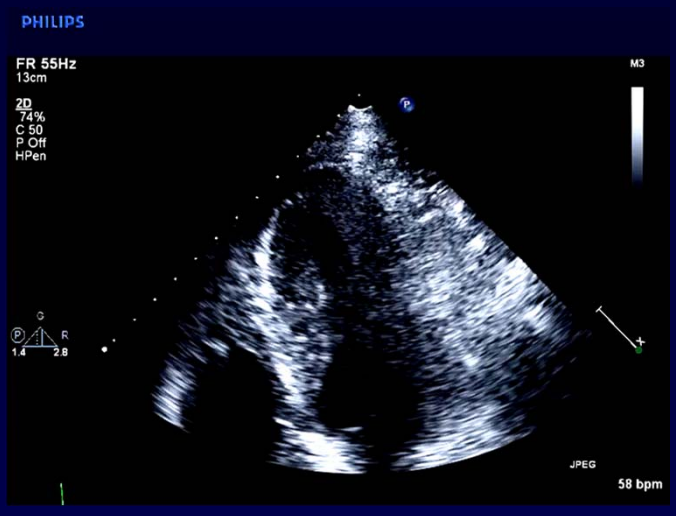
# Apical hypertrophy

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# Imaging

Use all windows, increase brightness, increase grey scale and use contrast

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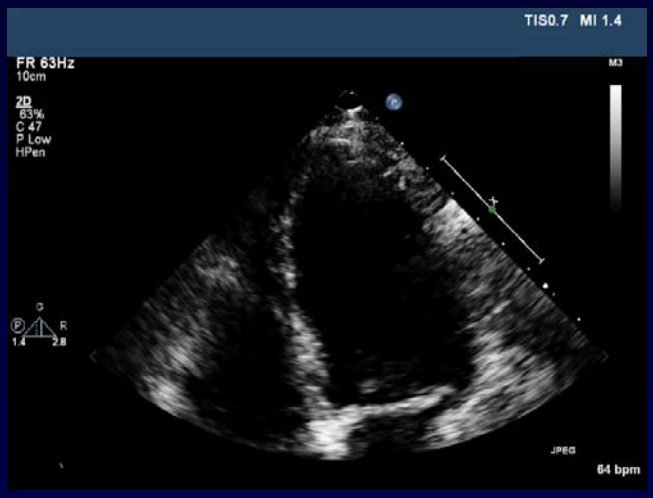


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## Reminder

- Not all genotypically positive pts have increased wall thickness
  - Troponin T mutations (mild)
  - Myosin binding protein C (late)
- Wall thickness > 30 mm predictive of SCD

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# Assessment of LV systolic function

- LV EF is usually normal or increased BUT DTI S wave and strain may be impaired
  - DTI S wave < 4 cm/sec  predictive of HF and death
- Overt LV systolic dysfunction, ‘‘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 markedly worse in the presence of LV systolic dysfunction

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# Strain

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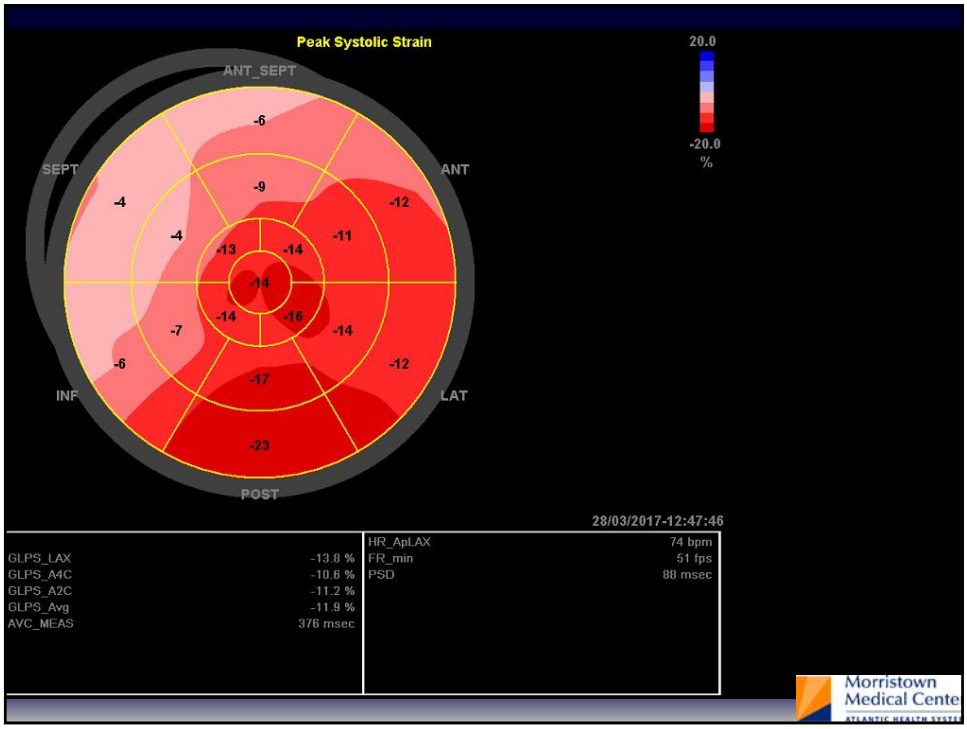


Dyssynchrony common (inter- and intra-ventricular)

## LV diastolic function

- Diastolic dysfunction has been reported in patients with HCM irrespective of the presence and extent of LV hypertrophy
- Correlations between the mitral inflow and pulmonary venous flow velocities and invasive parameters of LV diastolic function is relatively weak
- E/e' correlation with LV filling pressures is also modest

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## LA size

- LA size provides important prognostic information in HCM
- LA enlargement in HCM is multifactorial in origin,
  - mitral regurgitation
  - the presence of diastolic dysfunction
  - possibly atrial myopathy
- LA volume indexed to body surface area should be assessed in accordance with ASE guidelines

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# The Nature of the Obstruction

**Ultrasound Localization of Left Ventricular Outflow Obstruction in Hypertrophic Obstructive Cardiomyopathy**

By Pravin M. Shah, M.D., Raymond Gramiak, M.D., and David H. Kramer, M.D.

SUMMARY

Simultaneous recordings of reflected ultrasound from the anterior mitral leaflet and left ventricular outflow, the ECG, the phonocardiogram, and a recording of the carotid artery pulse were obtained in six patients with hypertrophic obstructive cardiomyopathy. Abnormal sharp systolic anterior movement (SAM) of the mitral leaflet was observed. This movement began with the onset of ventricular ejection and reached a peak with the initial peak in the arterial pulse. The leaflet was opposed to the interventricular septum up to 60% of the ejection period. In the latter part of systole as the mitral leaflet moved away from the interventricular septum, the arterial pulse showed a second systolic wave. Onset of SAM coincided with onset of the systolic murmur. Spontaneous variations in amplitude of SAM coincided with alterations in contour of the arterial pulse and in the intensity of the murmur. Administration of methoxamine to four patients resulted in disappearance of SAM. In one patient following surgery, the SAM of the mitral leaflet was noted only in the post-ectopic beats. This specific abnormality of mitral leaflet movement represents the localization of dynamic outflow obstruction in hypertrophic obstructive cardiomyopathy.

Additional Indexing Words:
Idiopathic hypertrophic subaortic stenosis (IHSS) Anterior mitral leaflet
Systolic anterior movement

THE physiologic behavior of left ventricular (LV) outflow in hypertrophic obstructive cardiomyopathy (HOCM) has been studied extensively over the last 10 years.[1-4] However, the anatomic site of obstruction has been elusive. It is generally difficult to appreciate the obstructive lesion in the exposed nonbeating heart.[2] Earlier angiographic demonstration of a probable site of obstruction was questioned by Criley and associates.[4] They introduced the concept of cavity obliteration as the cause of artifactual pressure gradients.

Based on observations at surgery, Björk[3] proposed that asymmetrical hypertrophy of the septum brings about an abnormal rotation of the anterior mitral leaflet. This leaflet balloons into the outflow tract and results in obstruction. This view of the mechanism of LV outflow obstruction did not find general support. More recently Dinsmore and co-workers[5] have demonstrated angiographically an abnormal anterior movement of the anterior mitral leaflet during systole. These observations were extended by Simon and associates[6] who em-

From the Cardiology Unit, Department of Medicine and the Department of Radiology, University of Rochester School of Medicine, and the Medical Clinic of Strong Memorial Hospital, Rochester, New York.

Work was supported in part by Grants HE-5500, HE-5506, and contract PH-43-68-1151 from the National Heart Institute, U.S. Public Health Service, the Genesee Valley Heart Association, the Rochester Fund, and the Ernest L. Woodward Fund. Work was accomplished during Dr. Kramer's and Shah's tenure of Special Research Fellowships from the National Heart Institute of the U.S. Public Health Service.

Circulation, Volume XL, July 1969 3

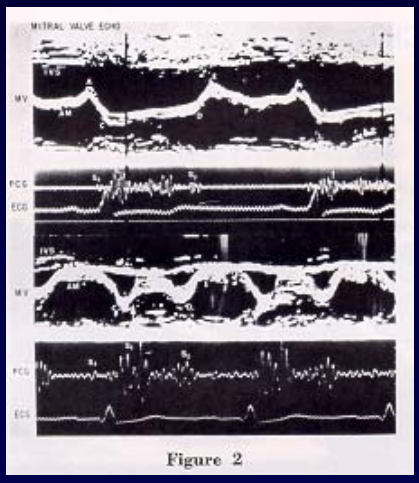


Figure 2

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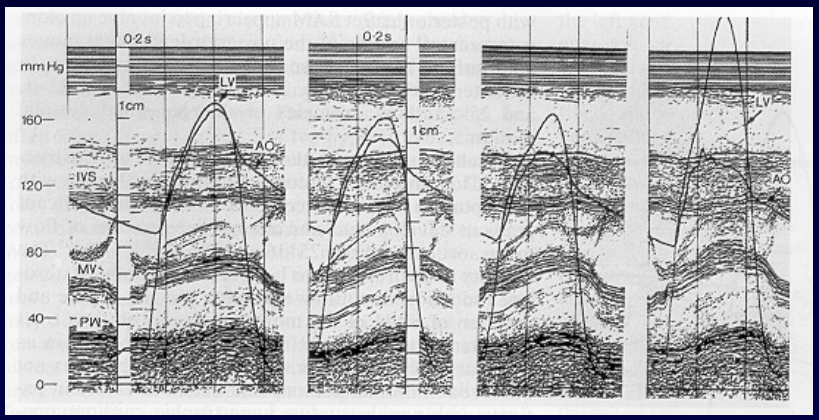


SAM-septal contact < 10% = mild, >30% = severe

Pollick C, Rakowski H, Wigle ED. Circulation 69:43, 1984

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# SAM-septal contact

- Associated with
  - notching of the aortic valve (M-mode)
  - Mitral regurgitation

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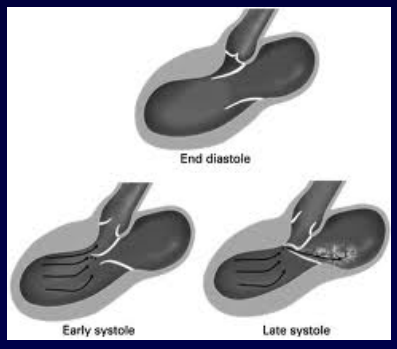
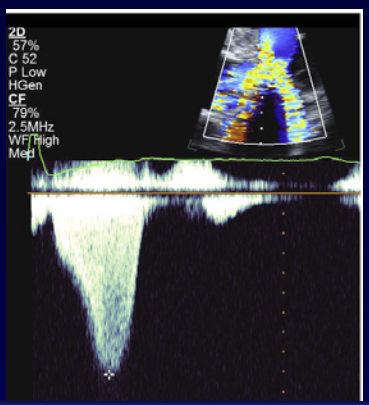
# Dynamic gradient

- Dynamic – increases gradually throughout systole to reach its peak in mid to late systole

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## Dynamic gradient

- Place of obstruction identified with PW/color
- Peak (measured with CW)influences treatment
  - >30 mmHg at rest a risk for SCD and HF

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## Provocative Maneuvers

- Valsalva
- Positional change
- Amyl nitrate
- Exercise

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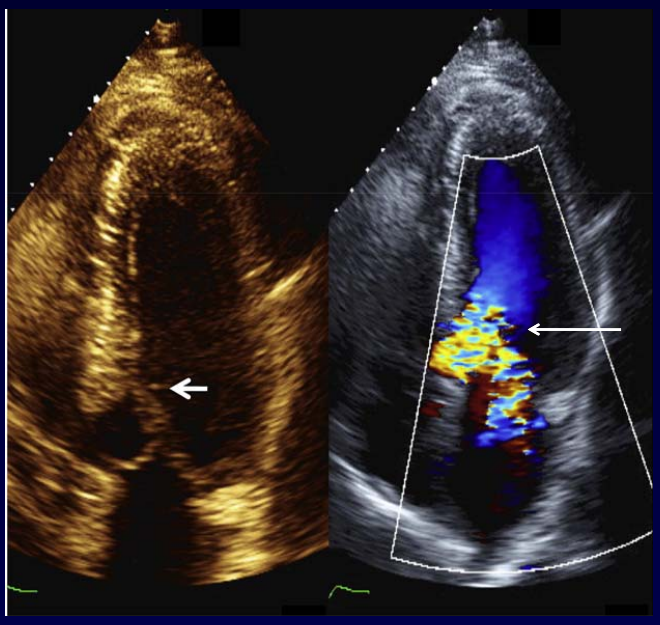


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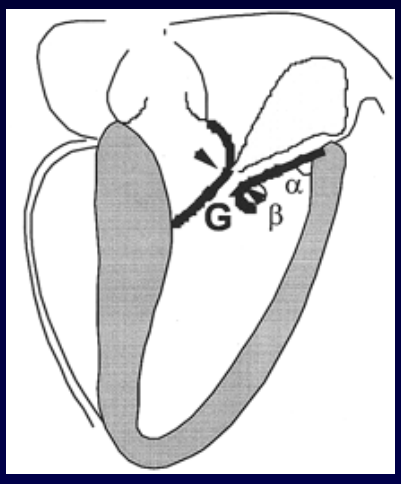
# SAM= Malcoaptation


Grigg LE, Wigle ED, Rakowski H. J Am Coll Cardiol 20:42, 1992

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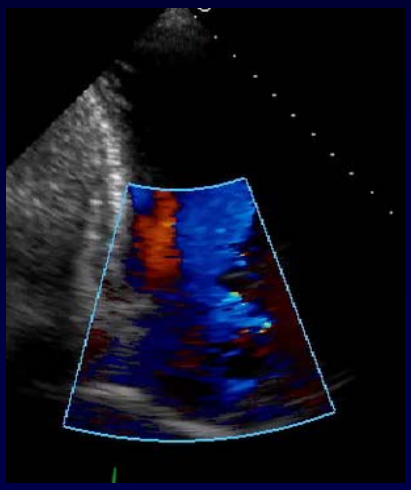
# HCM and MR



Why?

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Progress in Cardiovascular Diseases 54 (2012) 517–522
www.onlinepcd.com

ELSEVIER

## Diversity of Mitral Valve Abnormalities in Obstructive Hypertrophic Cardiomyopathy

João L. Cavalcante[a], Joaquim S. Barboza[b], Harry M. Lever[a,*]
[a]Cleveland Clinic Foundation, Department of Cardiovascular Medicine, Section of Cardiac Imaging, Cleveland, OH
[b]Henry Ford Hospital, Internal Medicine Department, Detroit, MI

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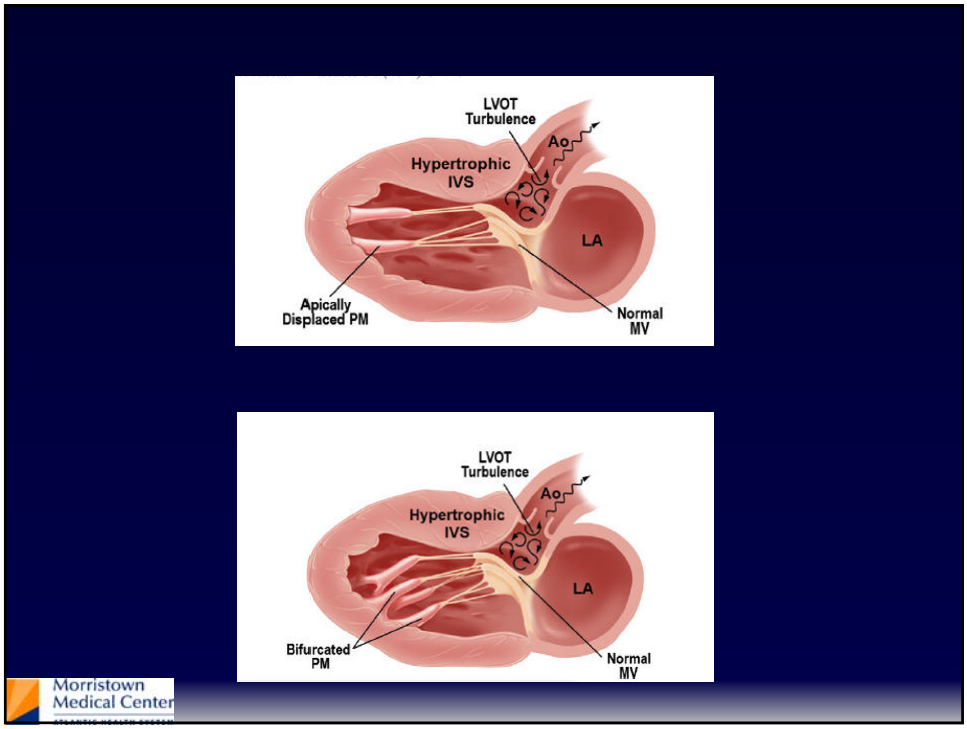
LVOT
Turbulence
Ao
Hypertrophic
IVS
LA
Apically
Displaced PM
Normal
MV
LVOT
Turbulence
Ao
Hypertrophic
IVS
LA
Bifurcated
PM
Normal
MV
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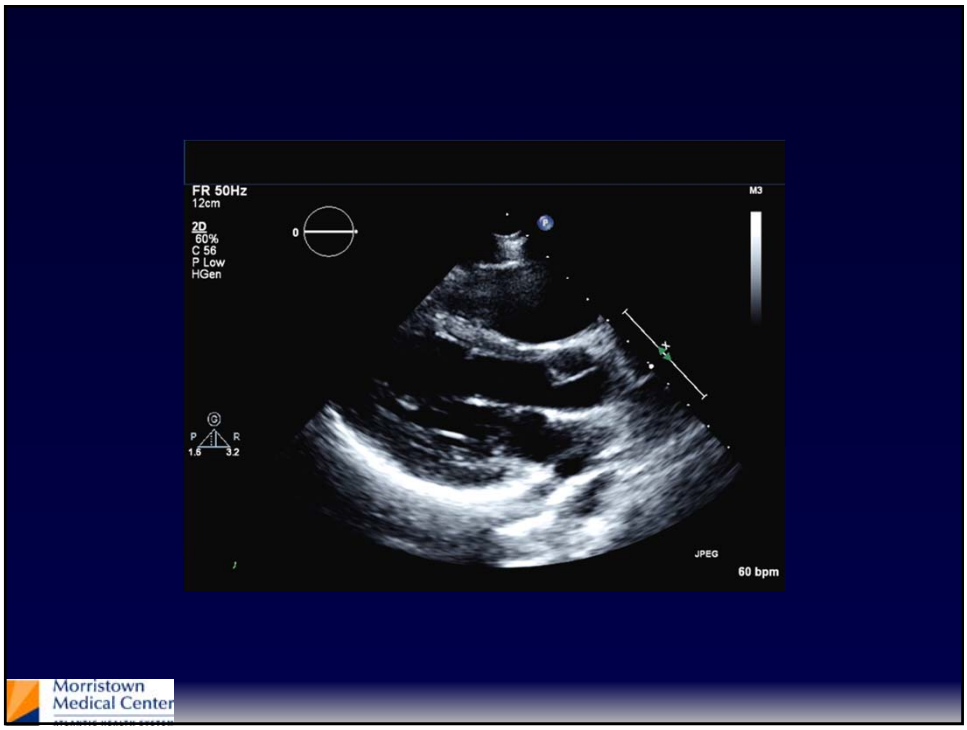
FR 50Hz
12cm
2D
60%
C 56
P Low
HGen
M3
JPEG
60 bpm
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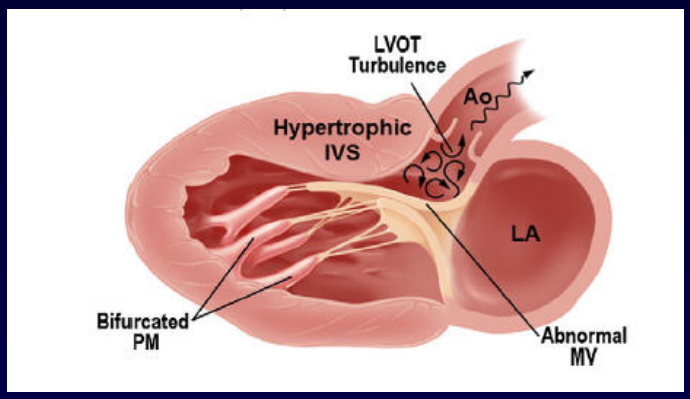


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# Mitral regurgitation

- Not all mitral regurgitation associated with HCM is related to SAM
- Patients with HCM can have intrinsic valvular abnormalities, such as
  - "degenerative" changes, mitral valve prolapse, leaflet thickening secondary to injury from repetitive septal contact or turbulent regurgitation jet, etc
- The presence of a central or an anteriorly directed jet should prompt careful evaluation of the mitral valve to identify intrinsic valvular abnormalities

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S5-1
72Hz
15cm
2D
71%
C 50
P Low
HGen
1.6 3.2
M3
49 bpm
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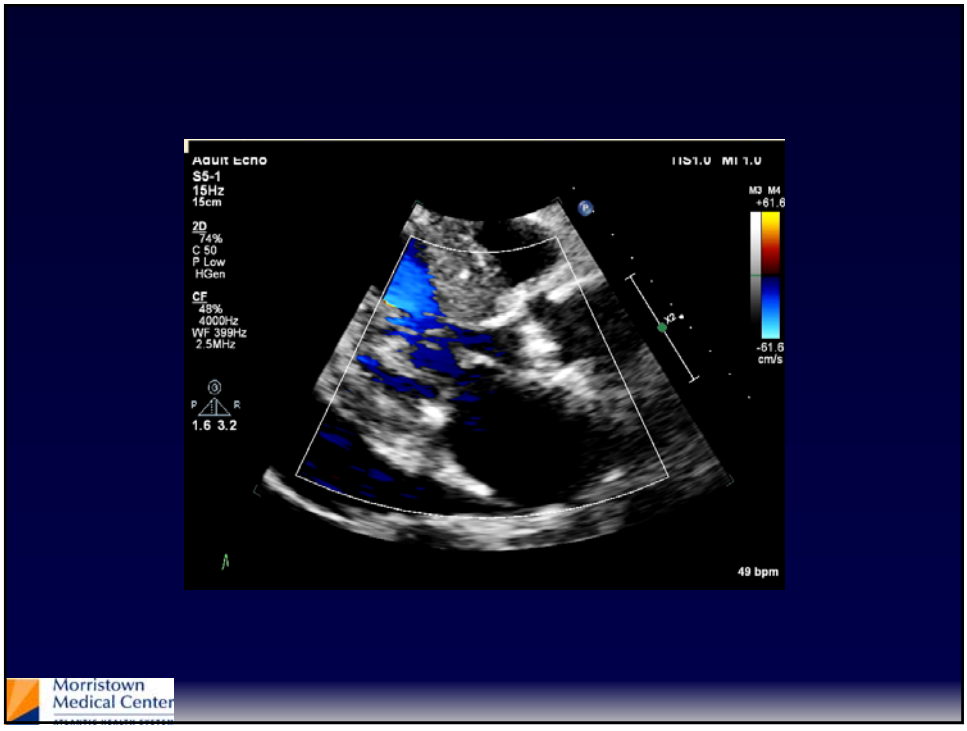
S5-1
15Hz
15cm
2D
74%
C 50
P Low
HGen
CF
48%
4000Hz
WF 399Hz
2.5MHz
1.6 3.2
M3 M4
+61.6
-61.6
cm/s
49 bpm
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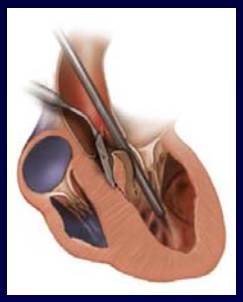
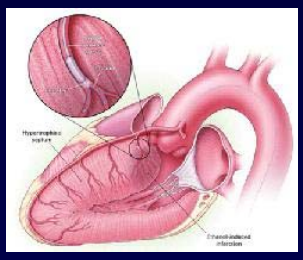
# Surgical Treatment

- Should take into consideration mitral valve anatomy and function





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# 174/1152 = 15%

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ORIGINAL INVESTIGATIONS

## Mitral Regurgitation in Patients With Hypertrophic Obstructive Cardiomyopathy

### Implications for Concomitant Valve Procedures

Joon Hwa Hong, MD, PhD,[a] Hartzell V. Schaff, MD,[a] Rick A. Nishimura, MD,[b] Martin D. Abel, MD,[c] Joseph A. Dearani, MD,[a] Zhuo Li, MS,[d] Steve R. Ommen, MD[b]

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# Take home points

- Mitral valve anatomic abnormalities are an intrinsic but often underappreciated component of HCM
- Mitral regurgitation and LVOT obstruction go hand in hand
- Mitral regurgitation may confound the noninvasive assessment of LVOT gradients
- MR due to HCM has a typical appearance
- The mitral valve should be carefully evaluated in surgical decision making for HCM

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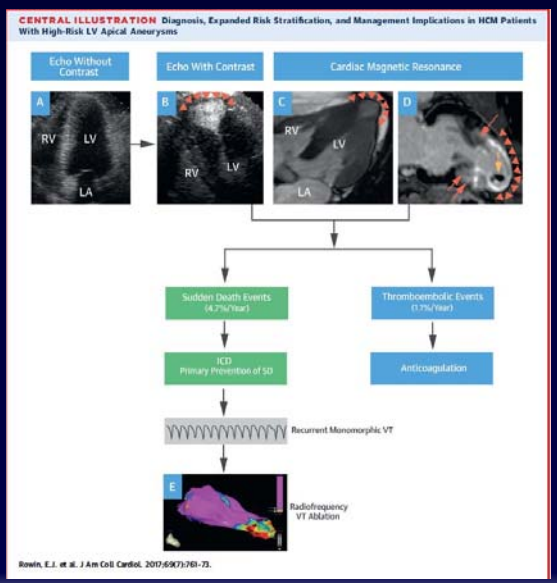
# Apical aneurysms

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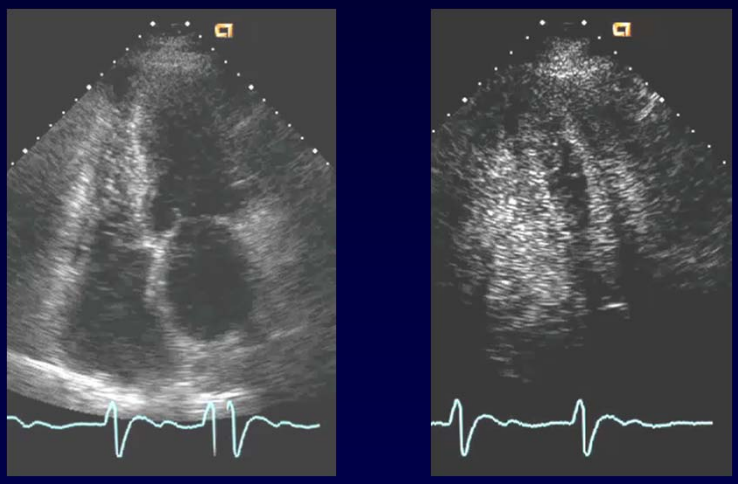
# Apical aneurysm??



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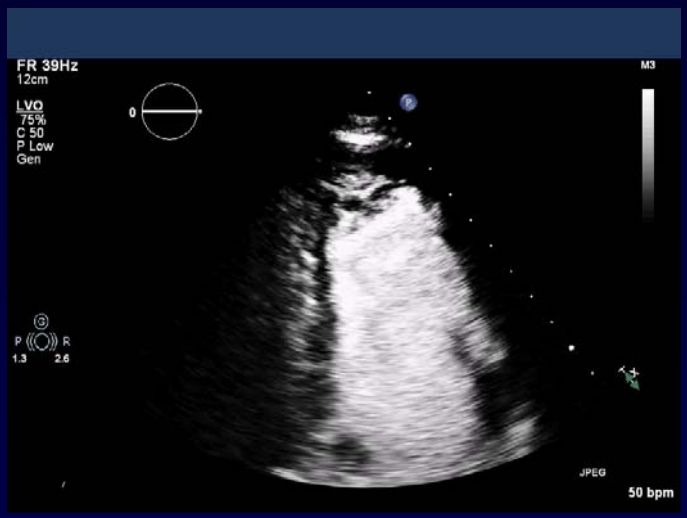
# Crypts

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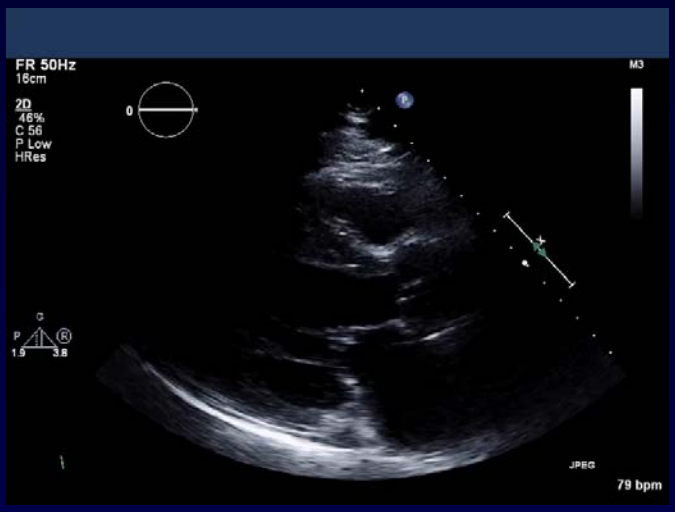
# Coronary Cameral Fistula

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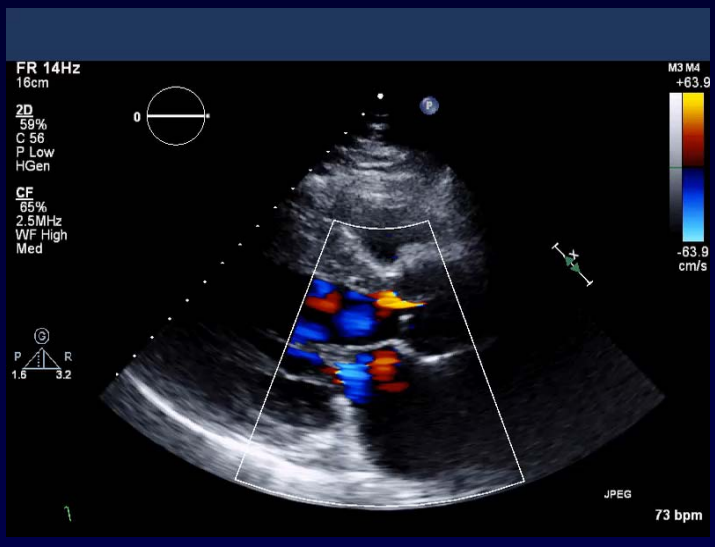


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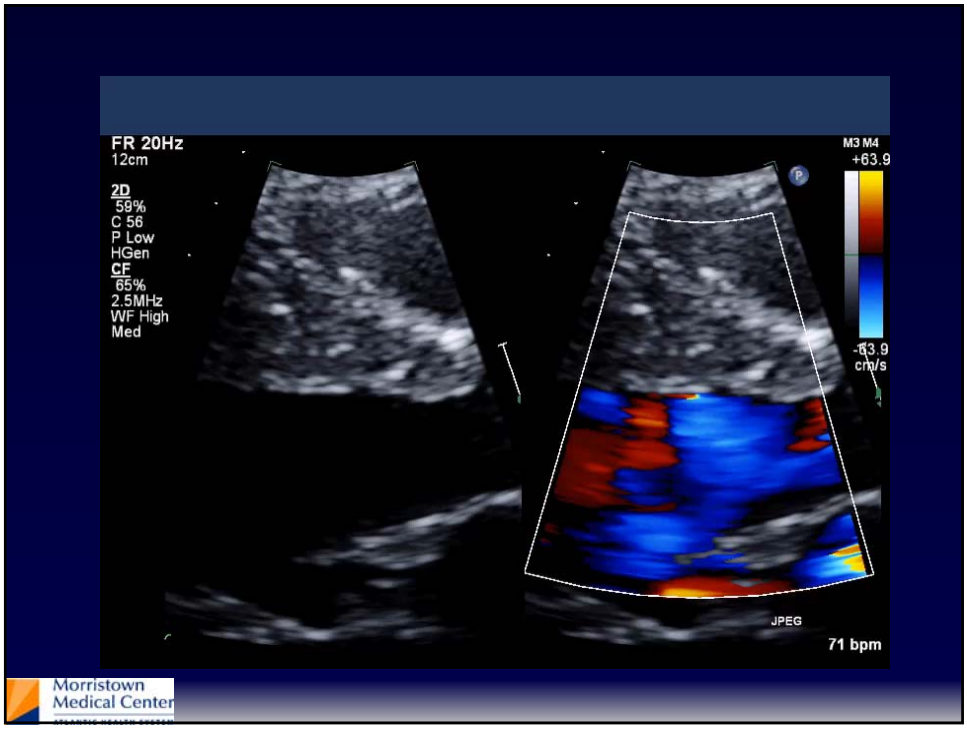
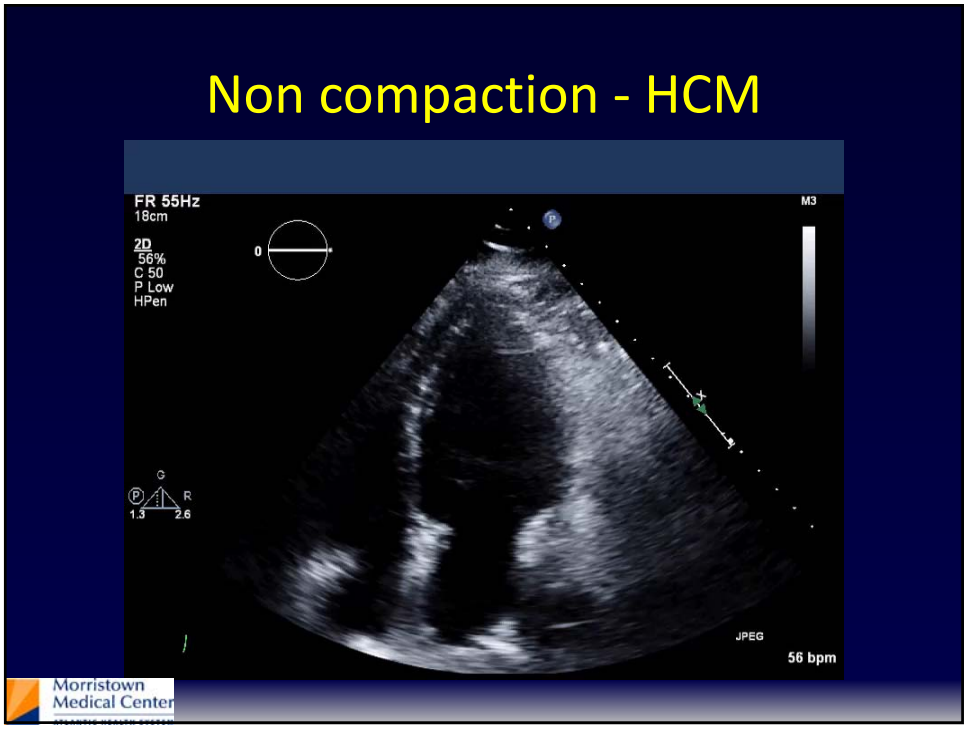
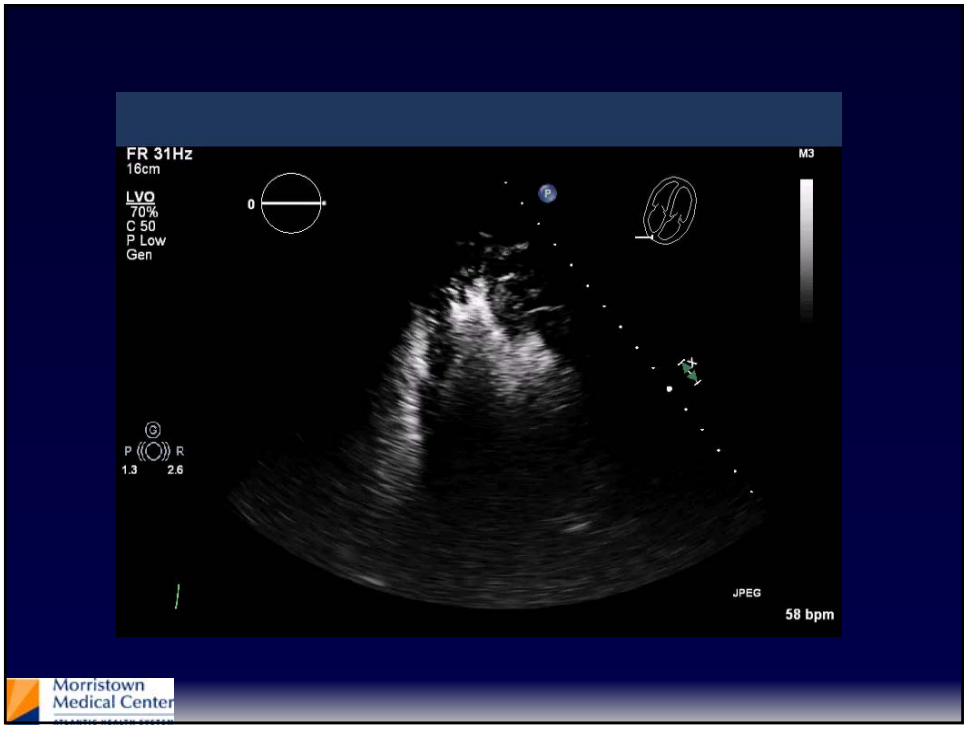



# Non compaction - HCM




FR 31Hz
16cm
LVO
70%
C 50
P Low
Gen
M3
P
R
1.3
2.6
JPEG
58 bpm
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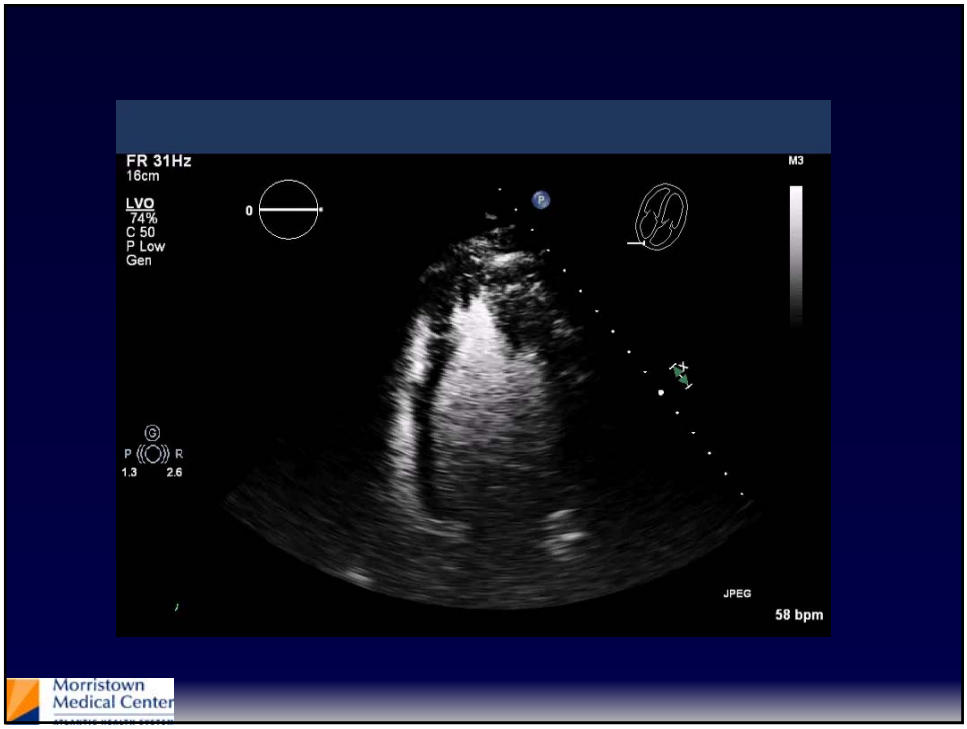
FR 31Hz
16cm
LVO
74%
C 50
P Low
Gen
M3
P
R
1.3
2.6
JPEG
58 bpm
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# Imaging Tips

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# Common Errors

- Confusing LVOT gradient with MR
- Failure to “see the SAM”
- Failure to see the true apex
- Failure to address the mitral valve apparatus

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# Common Errors

- Confusing LVOT gradient with MR
- Failure to "see the SAM"
- Failure to see the true apex
- Failure to address the mitral valve apparatus
- *Failure to see crypts*

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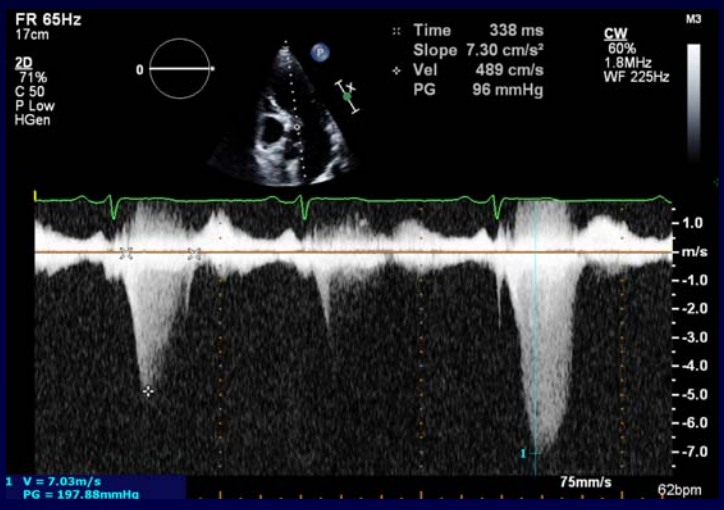
# Scan from LVOT to MR

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# Math check

- Use peak mitral velocity to estimate LVSP
- Estimated LVSP – SBP ≈ peak LVOT gradient

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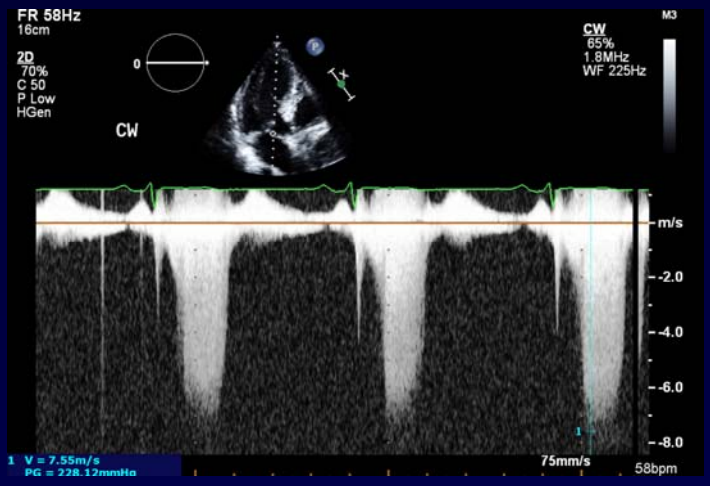
# Estimated LVSP ≈228 + 10 = 238

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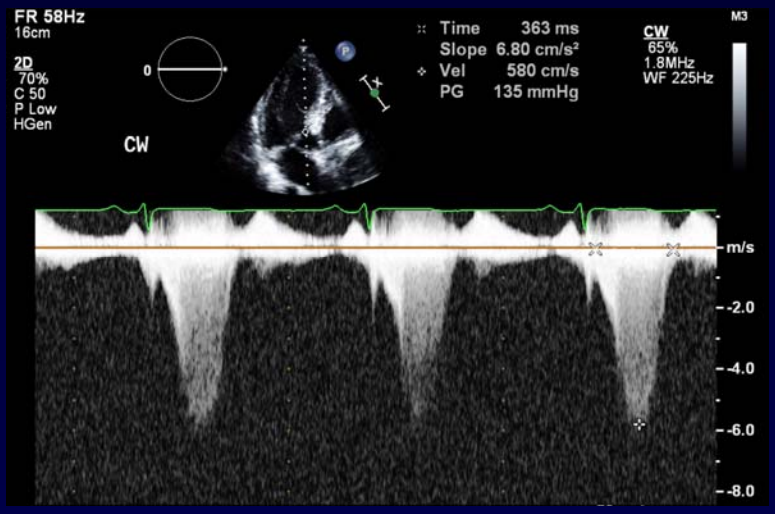
## LVSP-gradient = SBP
## 238-135 = 103!

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## Common Errors

- Confusing LVOT gradient with MR
- Failure to "see the SAM"
- Failure to see the true apex
- Failure to address the mitral valve apparatus

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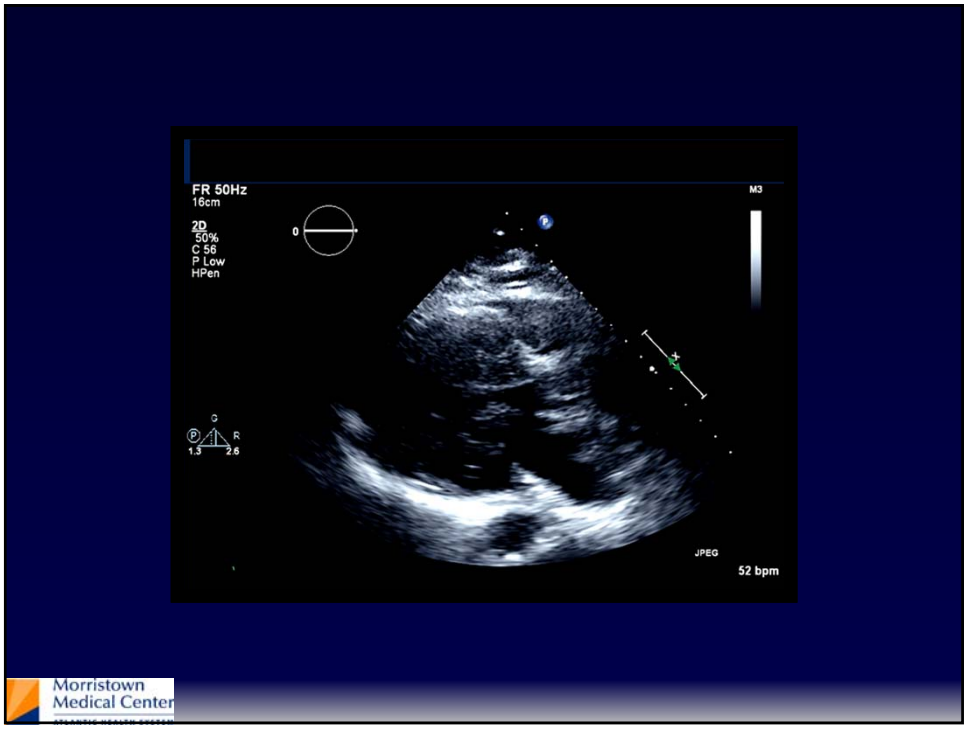
FR 50Hz
16cm
2D
50%
C 56
P Low
HPen
M3
JPEG
52 bpm
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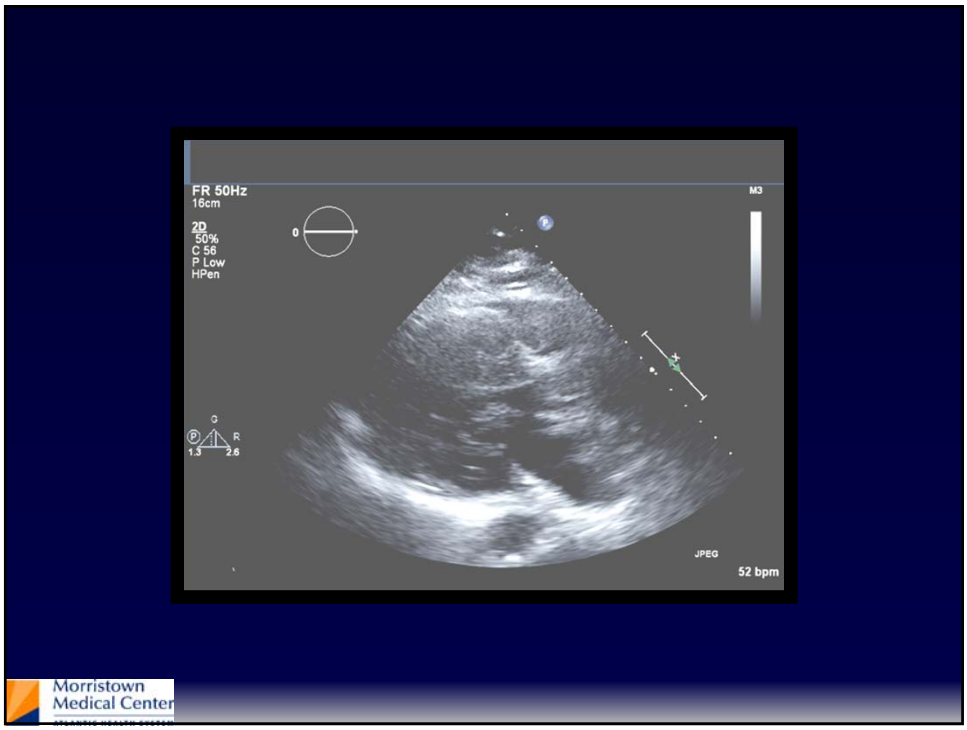
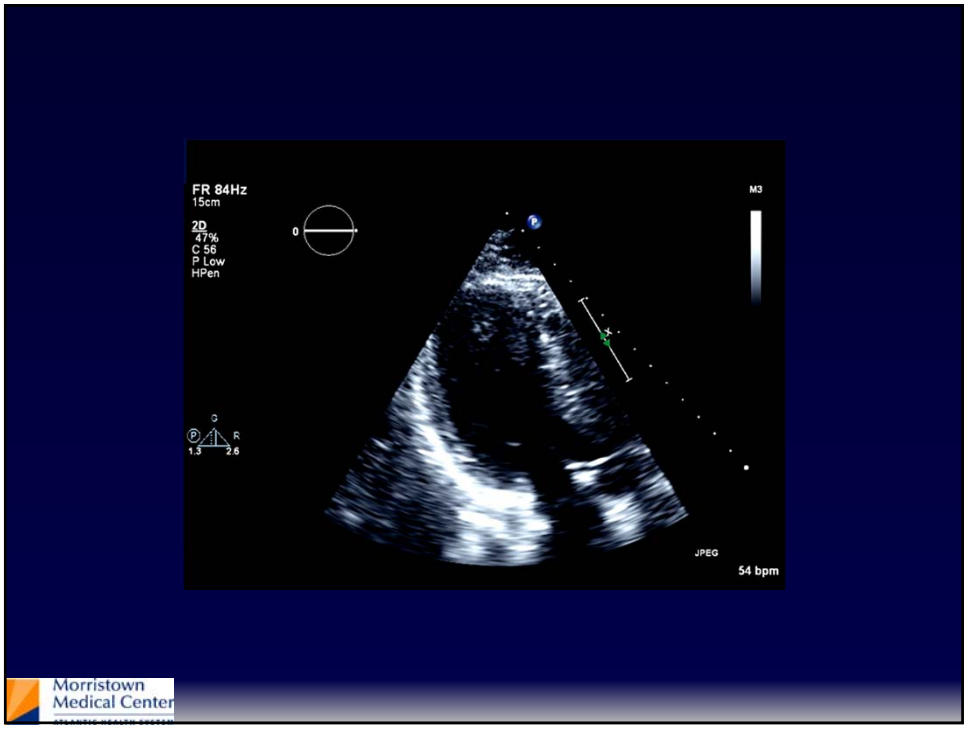
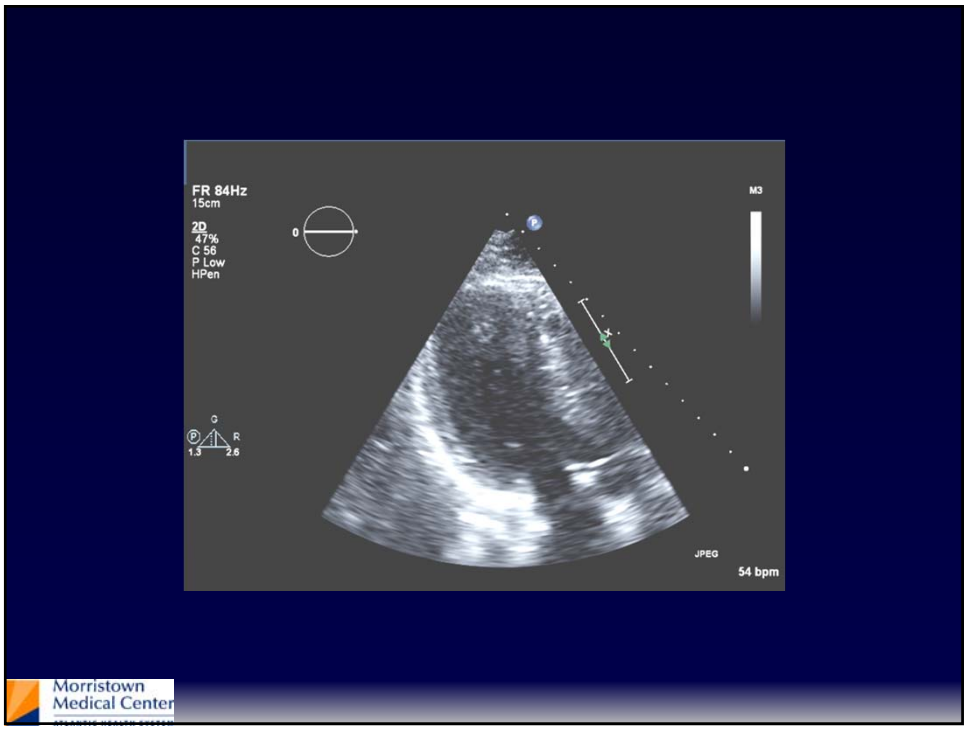
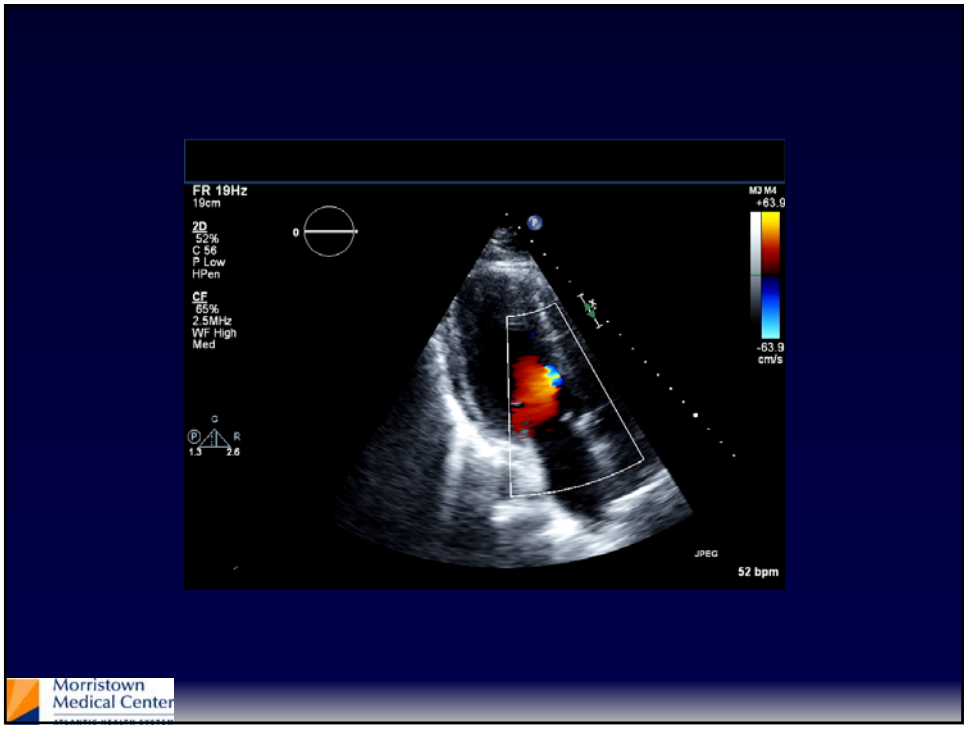

FR 50Hz
16cm
2D
50%
C 56
P Low
HPen
M3
JPEG
52 bpm
Morristown
Medical Center


FR 84Hz
15cm
2D
47%
C 56
P Low
HPen
M3
JPEG
54 bpm
Morristown
Medical Center


FR 84Hz
15cm
2D
47%
C 56
P Low
HPen
M3
JPEG
54 bpm
Morristown
Medical Center




# Common Errors

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# Easy to Miss

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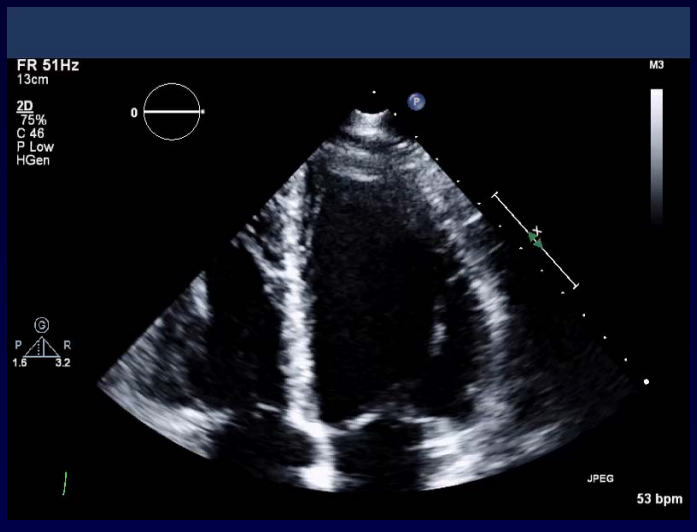


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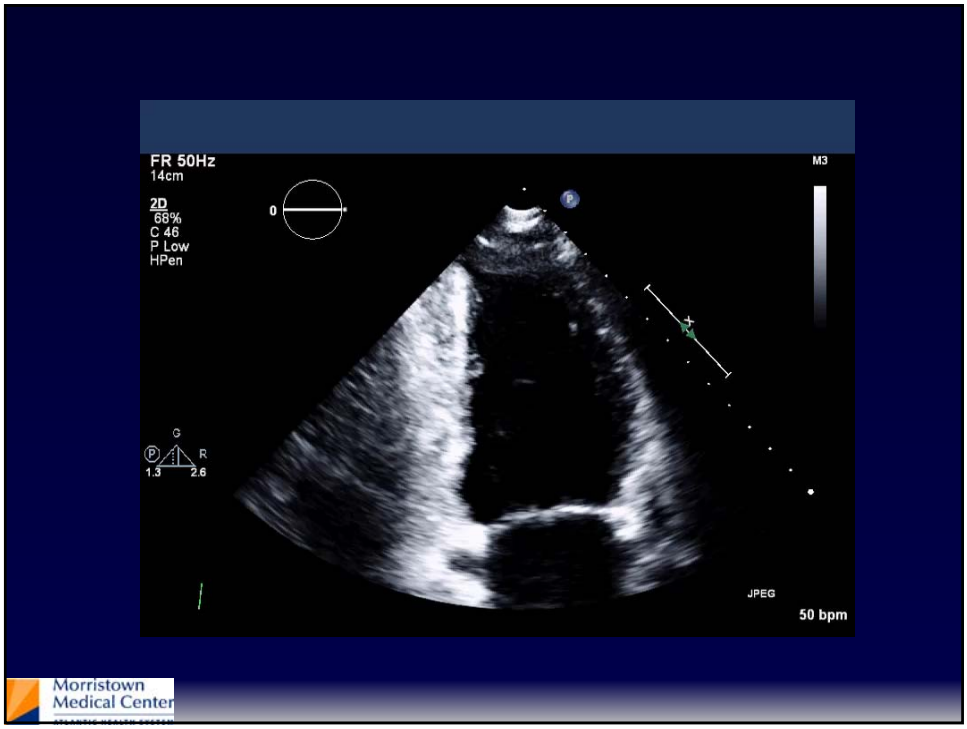
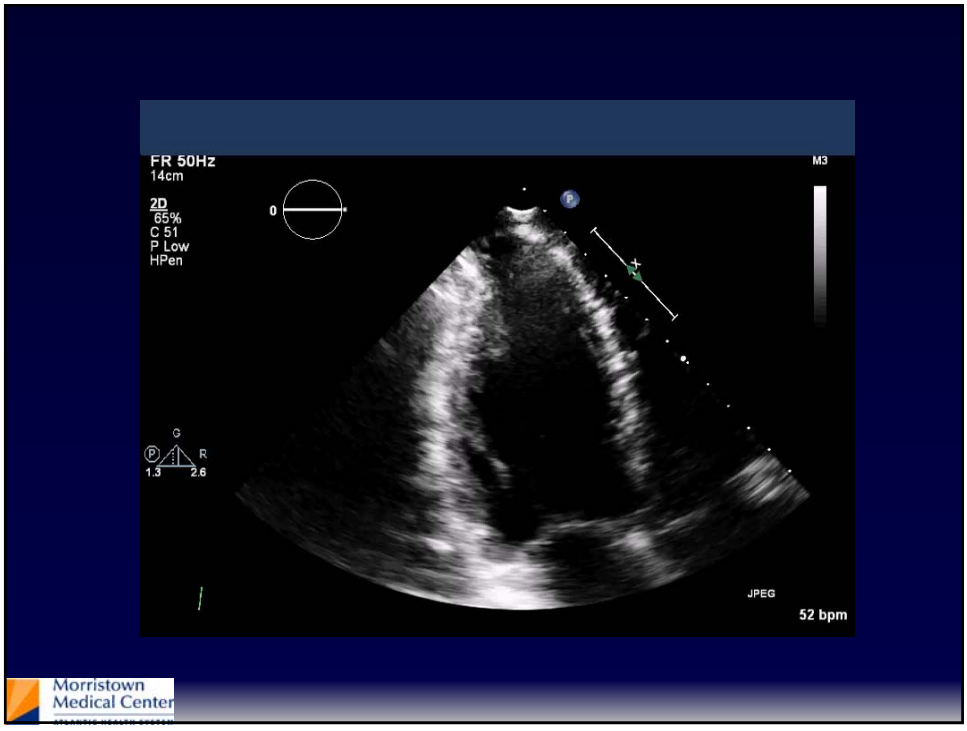

FR 50Hz
14cm
2D
68%
C 46
P Low
HPen
M3
JPEG
50 bpm
Morristown
Medical Center


FR 50Hz
14cm
2D
65%
C 51
P Low
HPen
M3
JPEG
52 bpm
Morristown
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## Contrast can help

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## Common Errors

- Confusing LVOT gradient with MR
- Failure to “see the SAM”
- Failure to see the true apex
- Failure to address the mitral valve apparatus

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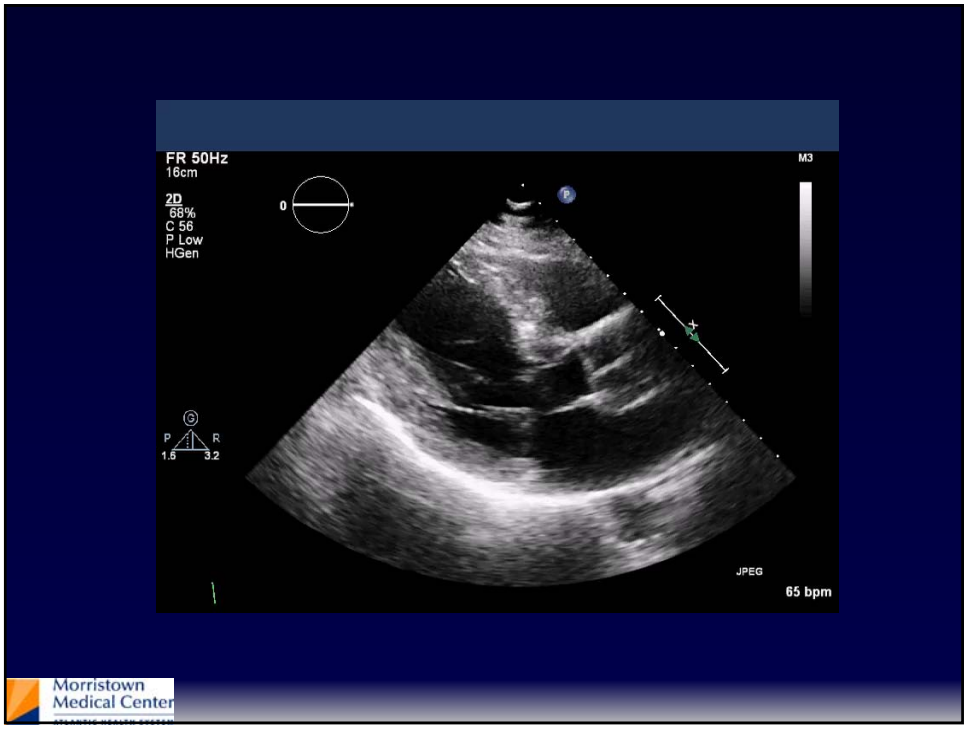
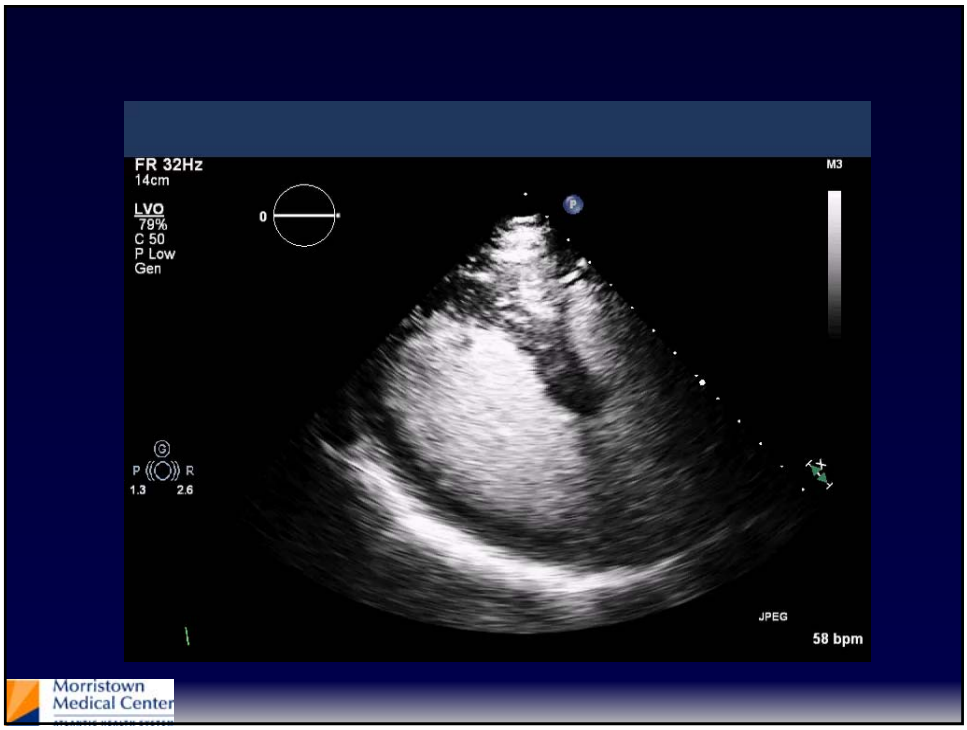
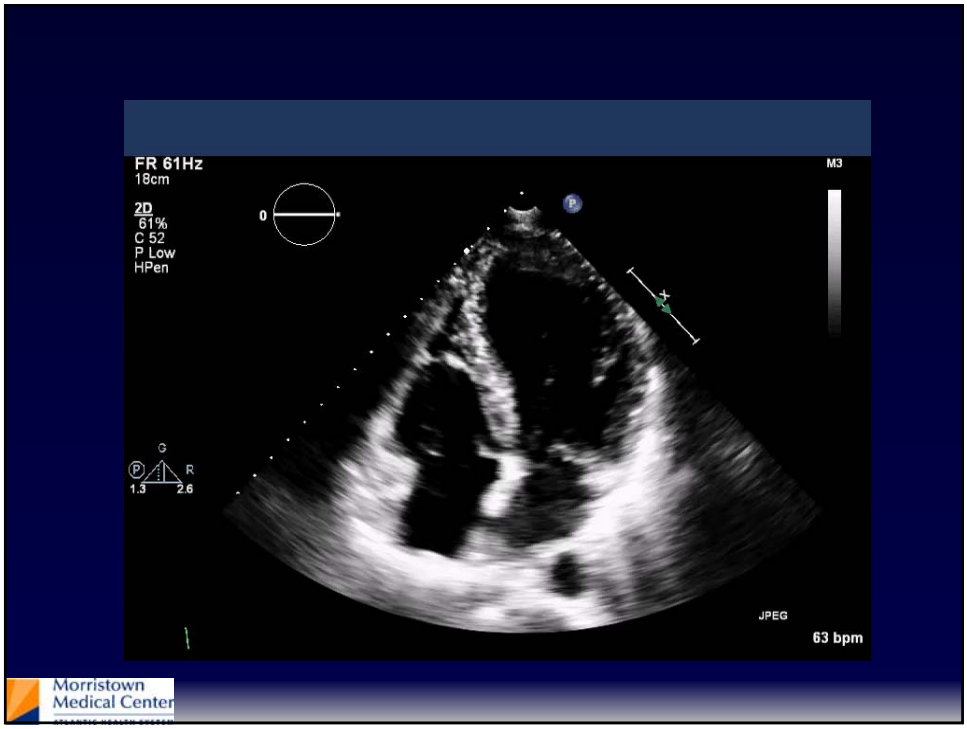
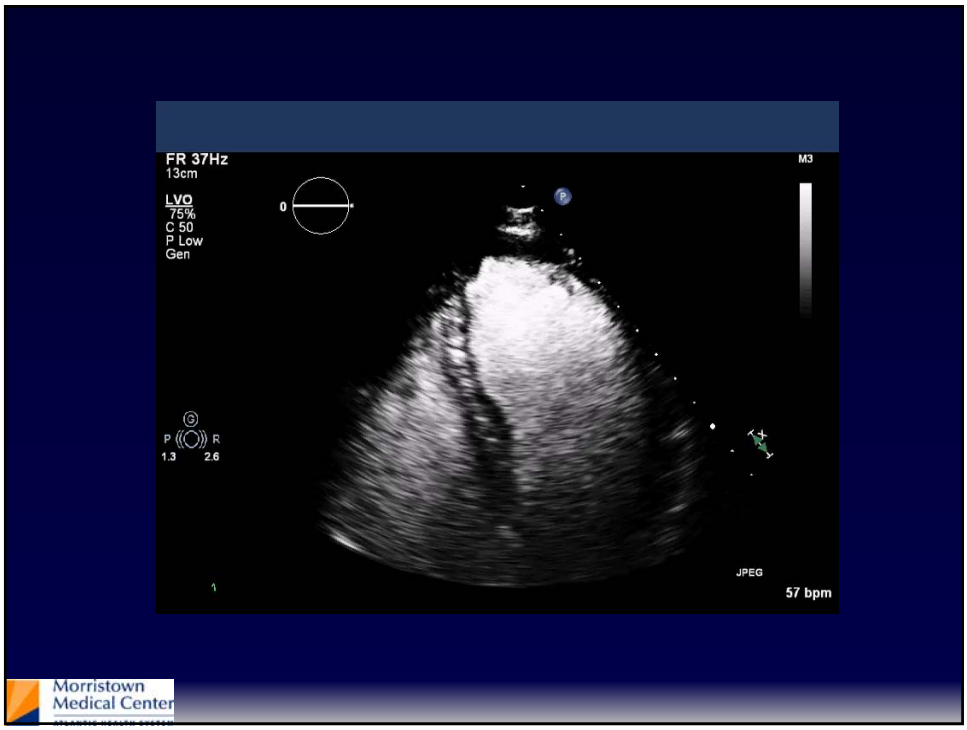
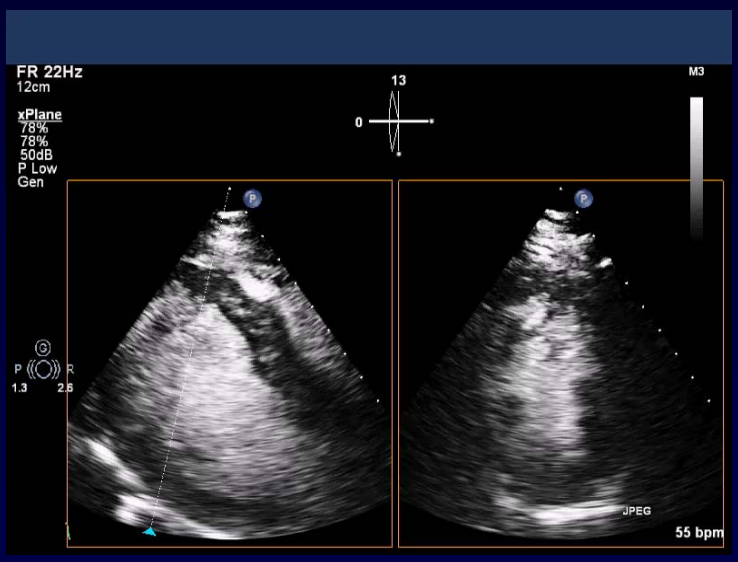

FR 50Hz
16cm
2D
68%
C 56
P Low
HGen
M3
P
R
1.6
3.2
JPEG
65 bpm
Morristown
Medical Center


FR 32Hz
14cm
LVO
79%
C 50
P Low
Gen
M3
P
R
1.3
2.6
JPEG
58 bpm
Morristown
Medical Center


FR 61Hz
18cm
2D
61%
C 52
P Low
HPen
M3
JPEG
63 bpm
Morristown
Medical Center


FR 37Hz
13cm
LVO
75%
C 50
P Low
Gen
M3
JPEG
57 bpm
Morristown
Medical Center

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## Contrast can help

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## Echocardiography is essential in HCM

- Defining structure and function
- Many features under appreciated and may be under-recognized
- Be on the look out for SAM and optimize images

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## Thank you!


Morristown Medical Center: Led by Drs. Martin and Barry Maron, Ethan Rowin

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