Structural Heart Disease

AAIM Triennial

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Peripartum Cardiomyopathy

Hypertensive Disorders of Pregnancy

Case Study #1

- 37 year old woman, nonsmoker
- 5’5” (165 cm), 216 lbs (98.1 kg), BMI 35.9
- Complicated obstetrical history
  - Miscarriage 2014
  - Uncomplicated pregnancy 2016
  - Pre-eclampsia leading to induction at 38 weeks in 2018
  - Episode of heart failure at delivery in 2020, full recovery
  - Currently pregnant, 14 weeks at time of application

What else would you want to know?

- Details about heart failure episode
- Chronic HTN?
- Most recent echo
- Family history
- Regular follow up?

Case Study #1: Additional Information

- Considered high risk due to complicated OB history and advanced maternal age
- Regular OB follow up
  - Single fetus
  - No cardiac symptoms noted
  - BPs normal
- Had pulmonary edema following last delivery in 2020 – thought to be due to volume overload
  - Initial echo: LV mildly dilated at 5.8, EF 45%
  - Echo at 3 month follow up: LV 5.5, EF 52%
  - No further cardiac follow up

Case Study #1: Discussion Questions

1. What are possible alternative reasons for the heart failure episode with last pregnancy?
2. What are hypertensive disorders of pregnancy? Are there long-term risks?
3. Are there long-term maternal risks with adverse pregnancy outcomes?
4. Is there a connection between hypertensive disorders of pregnancy, adverse pregnancy outcomes and peripartum cardiomyopathy?
5. Is there a risk of recurrence with peripartum cardiomyopathy?
6. What are the long-term risks associated with peripartum cardiomyopathy?

Hypertensive Disorders of Pregnancy (HDP)

- Include
  - Gestational hypertension
  - Pre-eclampsia
  - Eclampsia
  - HELLP syndrome (hemolysis, elevated liver enzymes, low platelet count)
- Incidence is rising – up to 10% American women experience at least 1 hypertensive pregnancy
- Most studies provide relatively short term follow up
- Risks of other CVD being identified with larger, longer term studies
  - CAD
  - Heart failure
  - Aortic stenosis
  - Mitral regurgitation

Long-Term Cardiovascular Risk After Hypertensive Pregnancy

- Honigberg study – prospective, observational study based on UK Biobank
- 220,024 women, 2808 had hypertensive disorder of pregnancy
- Mean age at baseline: 57
- Followed for 7 years: 7.0 (HPD) vs 5.7 (no HPD) incident CVD diagnoses per 1000 women-yrs

Condition	Hazard Ratio	CI	P-Value
Mitral regurg	5.0	1.5-17.1	0.01
Aortic stenosis	2.9	1.5-5.4	< 0.001
CAD	1.8	1.2-2.6	< 0.001
Heart failure	1.7	1.04-2.6	0.03

- Some but not all increased risk associated with prevalent HTN
- SFlt-1 (splice variant of VEGF, soluble fms-like tyrosine kinase-1) – antiangiogenic protein associated with pre-eclampsia and calcific AS

Adverse Pregnancy Outcomes and Long-Term CVD Risk

- Include
  - Hypertensive disease of pregnancy
  - Spontaneous pre-term birth
  - Intrauterine growth restriction
- Common: 10-20% of pregnancies
- Increasing in frequency. Significant racial disparities
- Likely shared pathogenic factors with hypertensive disorders of pregnancy
  - Defective placentation
  - Pro-inflammatory state
  - Production of anti-angiogenic proteins (sFlt-1)
  - Vascular dysfunction such as increased arterial stiffness, endothelial and myocardial dysfunction

Maternal Complications of Adverse Pregnancy Outcomes

- Increased risk of future CVD such as HTN, CAD, heart failure and possibly stroke
  - Endothelial dysfunction similar to what occurs in HFpEF
  - Higher central arterial stiffness
  - Higher Carotid Intimal Medial Thickness (CIMT)
  - Aberrant sympathetic neural activity and cardiac response (inability to increase stroke volume)
- Increased risk of renal disease
- Increased risk of hypertensive disorders with subsequent pregnancies
- Increased risk of peripartum cardiomyopathy with subsequent pregnancies

Peripartum Cardiomyopathy (PPCM)

Definition: *Idiopathic LV systolic dysfunction that develops during the last month of pregnancy or within 5 months of delivery*

- Incidence
  - US: 1:1000 to 1:4000
  - Haiti: 1:300
  - Japan: 1:20,000
- Risk factors
  - Maternal age > 30
  - African descent
  - Pregnancy-related HTN
  - Multiple fetuses
  - Multiparity

- Potential Causes
  - Pregnancy-related hemodynamic changes
  - Immunologic factors
  - Myocarditis
  - Altered prolactin processing
  - Angiogenic/vasculohormonal imbalance
  - Genetic predisposition

PPCM - Prognosis

Caveat: Studies are small

Acute Complications

- Major adverse event precedes diagnosis in about half of cases
- Short term complications
  - Heart failure
  - Arrhythmias
  - Thromboembolic events
  - Stroke
  - Death
- Mortality 4-11% at 1 year
- "Majority" show partial or complete recovery within 2-6 months

Chronic/Long Term Concerns

- Prognostic markers
  - EF at time of diagnosis (< 30% is unfavorable)
  - LV dilatation
  - LV thrombus
  - RV systolic dysfunction
  - Recovery EF < 50%
- Subsequent pregnancy
  - Worsening of LV function in about 1/3
    - Fully recovered: 20% risk of recurrence
    - EF < 50%: 50% have further deterioration
  - Mortality 2% - 19%
  - Increased risk of prematurity and miscarriage
- Long-term mortality 7-20% (over 8 years)

Case Study #1: Wrap Up

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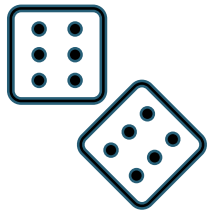

What would your recommendation be at this time?

What if she wasn’t currently pregnant?

If she was now age 55 and asymptomatic. Concerns?

Case Study #1: Val's Thoughts

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- Complicated obstetrical history
  - Miscarriage 2014
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What would your recommendation be at this time?

- Significant risk of recurrent PPCM
- Chance of worse outcome with recurrence
- High risk

What if she wasn't currently pregnant?

- At risk for recurrent PPCM with subsequent pregnancies
- Chance of worse outcome with recurrence
- High risk

If she was now age 55 and asymptomatic. Concerns?

- At increased risk for multiple CV impairments
- Would be concerned about current cardiac status and assess carefully

Case Study #1: Val's Thoughts on Discussion Questions

1. What are possible alternative reasons for the heart failure episode with last pregnancy?
   - Underlying cardiomyopathy or other heart disease
   - Peripartum cardiomyopathy
   - Myocarditis
   - Doubt infectious cause for pulmonary symptoms
2. What are hypertensive disorders of pregnancy? Are there long-term risks?
   - Gestational HTN, pre-eclampsia, eclampsia, HELLP syndrome
   - Long-term risks include chronic HTN, increased risk of recurrence of HDP, increased risk of PPCM, increased risk of CAD, heart failure, aortic stenosis and mitral regurgitation
3. Are there long-term maternal risks with adverse pregnancy outcomes?
   - Yes. Increased risk of aberrant vascular and sympathetic function.
4. Is there a connection between hypertensive disorders of pregnancy, adverse pregnancy outcomes and peripartum cardiomyopathy?
   - These conditions all appear to be related to production of antiangiogenic substances. Genetic predisposition is also likely
5. Is there a risk of recurrence with peripartum cardiomyopathy?
   - Overall, about 33% chance of recurrence with subsequent pregnancy. Those with fully recovered LV function (LVEF ≥ 50%) have about a 20% chance and those with LVEF < 50% have about a 50% chance of worsening LV function with subsequent pregnancy
6. What are the long-term risks associated with peripartum cardiomyopathy?
   - Risk of recurrence and increased risk of adverse pregnancy outcomes
   - Chronic heart failure
   - Arrhythmias and mortality in the range of 7-20% over about 8 year follow up

Useful References: HDP and APO

- Garovic VD et al. Incidence and long-term outcomes of hypertensive disorders of pregnancy. J Am Coll Cardiol 2020;75:2323.
- Wang Y et al. Hypertensive disorders of pregnancy and subsequent risk of premature mortality. J Am Coll Cardiol 2021;77:1302.
- Levine LD et al. Prospective evaluation of cardiovascular risk 10 years after a hypertensive disorder of pregnancy. J Am Coll Cardiol 2022;79:2401.
- Stuart JJ et al. Cardiovascular risk factors mediate the long-term maternal risk associated with hypertensive disorders of pregnancy. J Am Coll Cardiol 2022;79:1901.
- Honigberg MC et al. Long-term cardiovascular risk in women with hypertension during pregnancy. J Am Coll Cardiol 2019;74:2743.
- Lane-Cordova AD et al. Long-term cardiovascular risks associated with adverse pregnancy outcomes. J Am Coll Cardiol 2019;73:2106

Useful References: PPCM

- Davis MB et al. Peripartum cardiomyopathy – JACC state-of-the-art review. J Amer Coll Cardiol 2020;75:207
- Sliwa K et al. Peripartum cardiomyopathy: from genetics to management. Eur Heart J 2021;42:3094.
- Elkayam U. Risk of subsequent pregnancy in women with a history of peripartum cardiomyopathy. J Am Coll Cardiol 2014;64:1629.
- Goli R et al. Genetic and phenotypic landscape of peripartum cardiomyopathy. Circulation 2021;143(19):1852.
- Bauersachs J et al. Pathophysiology, diagnosis and management of peripartum cardiomyopathy: a position statement from the Heart Failure Association of the European Society of Cardiology Study Group on peripartum cardiomyopathy. Eur J Heart Fail 2019;21:827.
- Sliwa K et al. Incidence and prevalence of pregnancy-related heart disease. Cardiovasc Res 2014;101(4):554

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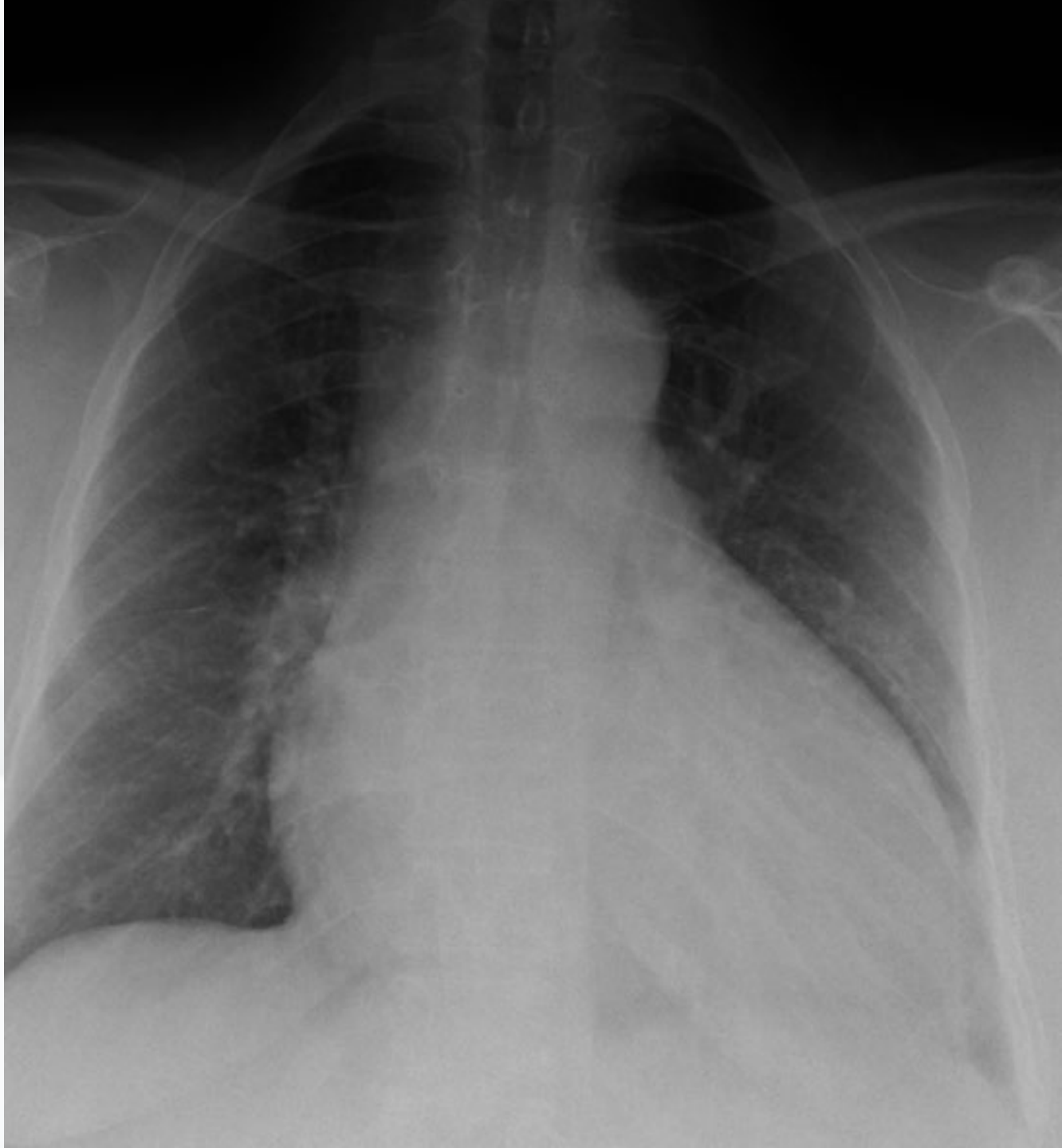
Asymptomatic Systolic Dysfunction

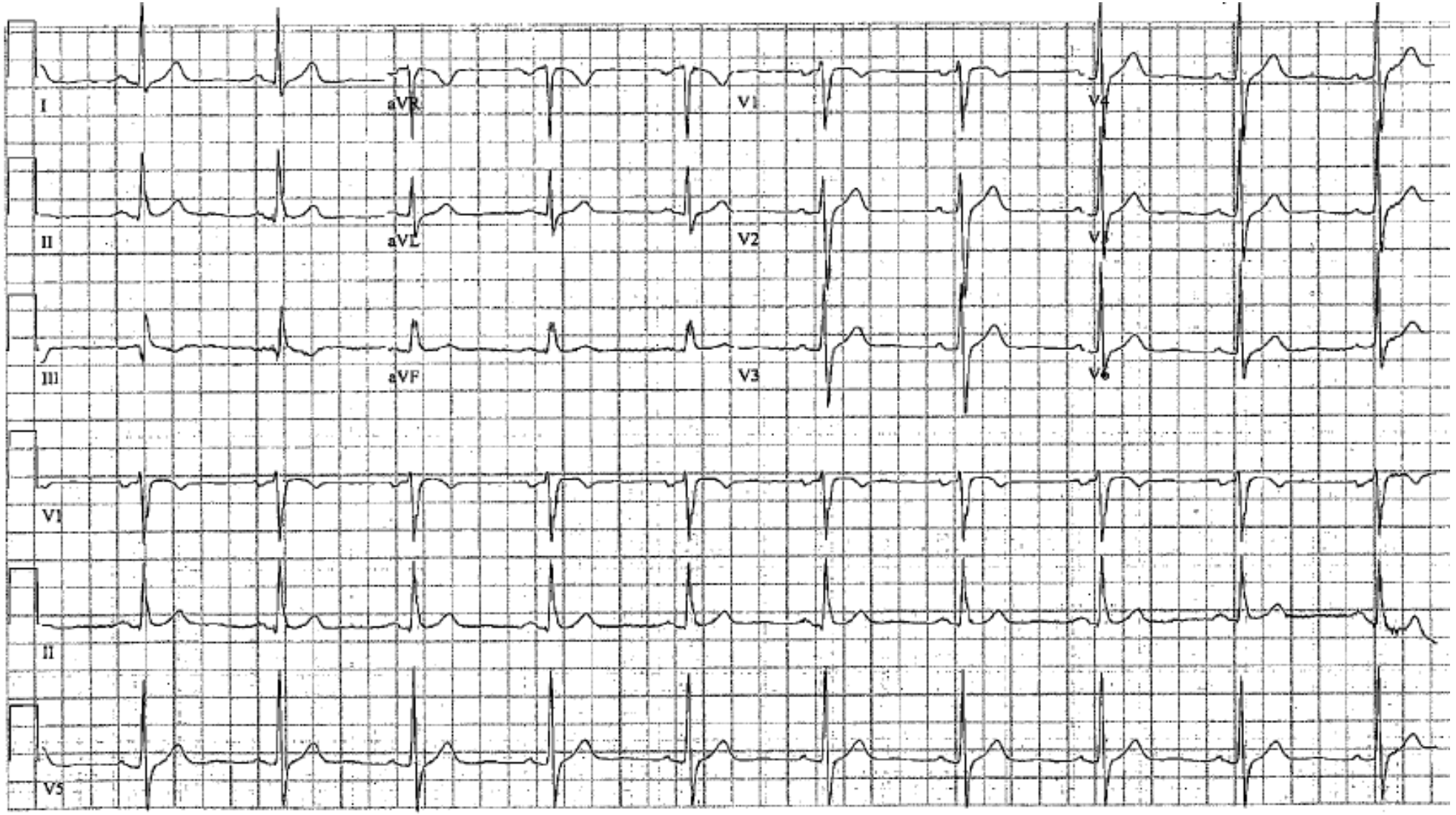
Dilated Cardiomyopathy



Case Study #2

- 52 year old man, nonsmoker
- Applying for $1,100,000
- 5’10” (1.78 m), 186 lbs (84.5 kg)
- BP 116/76
- No adverse family history
- Sleep apnea, uses CPAP nightly
- Had angiogram 10/19

Case Study #2: ECG



Case Study #2: APS

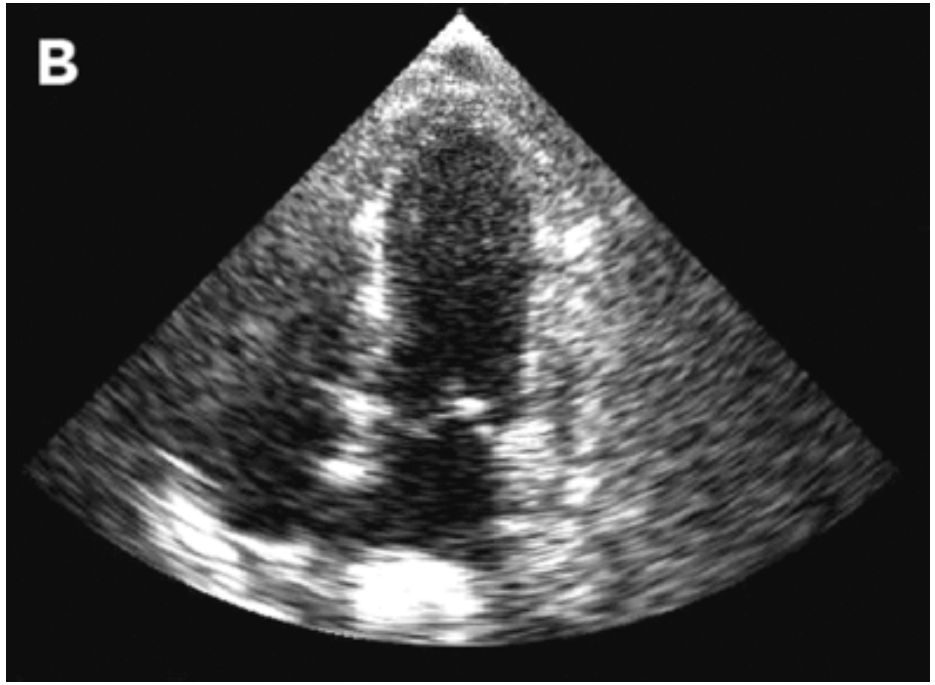
- 8/19: Chest pain and diaphoresis - went to Emergency Room
  - ECG normal
  - Portable (AP) CXR showed mildly enlarged heart, otherwise normal
  - Troponins were normal
- 9/19 Stress test
  - Exercised 10 minutes, 15 seconds to heart rate of 164 and peak BP 180/80
  - 1 mm downsloping ST depression in inferior leads, resolved by 2 minutes of recovery
  - Occasional PACs
  - SPECT cardiolyte
    - Left and right ventricles dilated
    - Normal perfusion
    - Left ventricular ejection fraction 42%
- 10/19 Cardiac cath
  - Normal coronary arteries
  - Mild lateral hypokinesis with LVEF 45%

What else would you want to know?

- How is he doing now?
- Meds?
- Any other medical conditions?
- Most recent echo
- Family history
- Regular follow up?
- NTproBNP

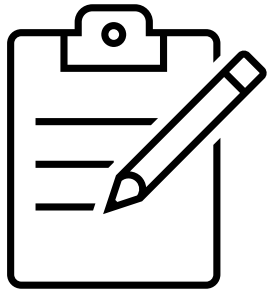
Case Study #2: Additional Information

- 1/20 Echo
  - LVIDd 6.1
  - LVIDs 4.0
  - LA 4.1
  - Normal RV size and function
  - Normal mitral valve with mild MR
  - LVEF 43%
- 1/22 - last cardiology follow up
  - No symptoms
  - On empagliflozin
  - Office echo – “stable”
- NTproBNP on insurance labs was 56



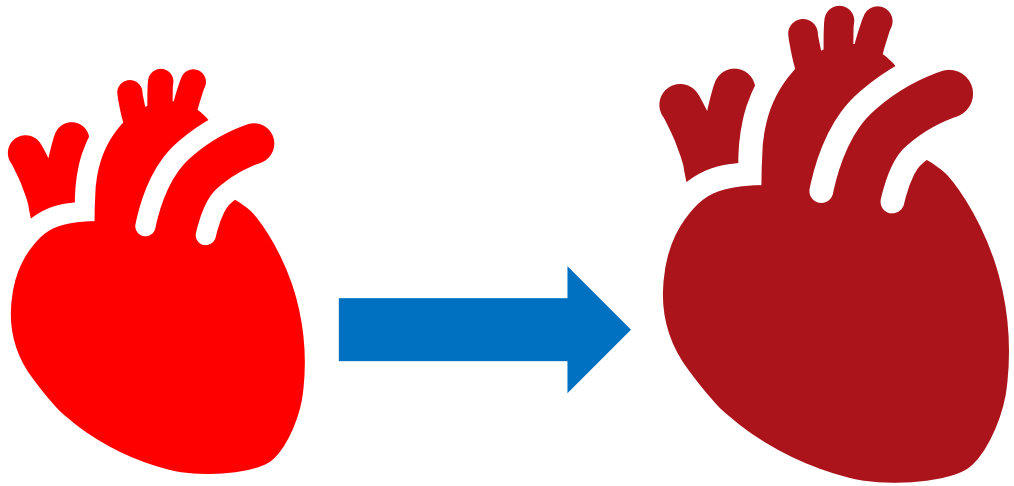
Case Study #2: Discussion Questions

1. What could be causing the low ejection fraction?
2. What other information would be helpful in assessing this risk?
3. An APS note states Stage B heart failure. Is this a typo?
4. What is appropriate treatment and follow up?
5. What is the risk of progression of asymptomatic systolic dysfunction to heart failure?
6. What factors are important in assessing this risk?
7. What is GLS and would it be helpful in this case?



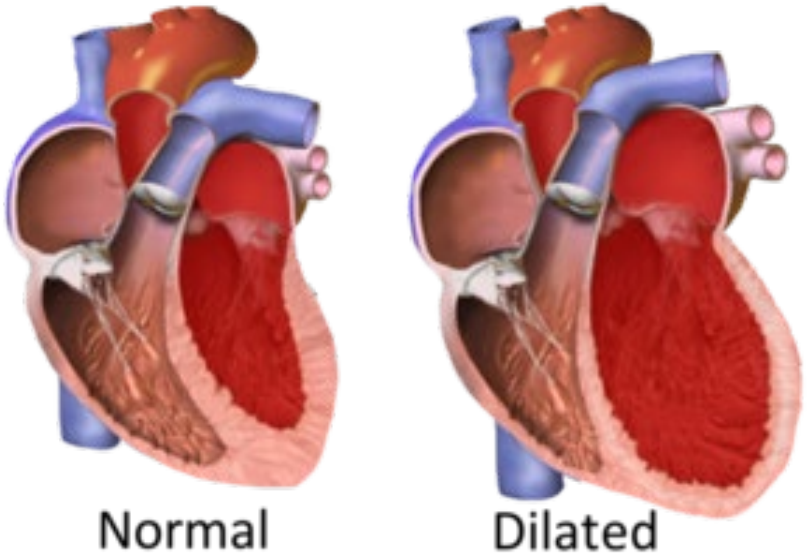
Dilated Cardiomyopathy (DCM)

- May also be known as nonischemic or idiopathic DCM
- Prevalence: 36/100,000 (1:2500) in US (estimated)
- Can occur at any age; most often presents between ages 20-40
- Often asymptomatic in early stages
- Accounts for 30-40% of cases in heart failure studies and registries
- About 1/3 of DCM cases are familial
- Mortality due to heart failure or arrhythmias



DCM: Diagnosis and Treatment

- Diagnosis
  - Echo or MRI
  - Dilatation of the ventricles or all 4 chambers
  - Systolic dysfunction
  - No other disease capable of causing the changes
  - (for example, no CAD, no valvular disease)
- Treatment (depends on stage)
  - Control of cardiovascular risk factors and comorbidities
  - ACE inhibitors or ARBs
  - Beta blockers
  - Diuretics, Na restriction
  - SGLT2i (sodium-glucose cotransporter-2 inhibitor)



Stages of Heart Failure

ACC/AHA Stage	Description	NYHA Class
A	At high risk for heart failure but without symptoms, structural heart disease or blood tests indicating myocardial injury (Ex: HTN)	None
B	Pre-heart failure. No signs or symptoms of heart failure, but evidence of one of the following: • Structural heart disease (Ex: reduced EF, LV enlargement, LVH, valvular heart disease) • Increased filling pressures as measured by cath or echo • Risk factors from Stage A plus elevated BNP/NTproBNP or persistently elevated troponin	None
C	Symptomatic heart failure. Structural heart disease with previous or current symptoms of heart failure	Could be Class I, II or III
D	Advanced heart failure with symptoms that interfered with daily life, are difficult to control and result in recurrent hospitalizations despite guideline-directed medical therapy	Class IV

Heidenreich PA et al. 2022 AHA/ACC/HFSA guideline for the management of heart failure. J Am Coll Cardiol 2022;79:e263

Risk of Progression to Symptomatic Heart Failure

2016 Metanalysis by Echouffo-Tcheugui

- Looked at risk of progression from asymptomatic LV systolic dysfunction to symptomatic heart failure
- 25,369 participants, followed for an average of 7.9 years
- Absolute risk of progression to heart failure
  - 8.4 per 100 person-years in those with asymptomatic LV systolic dysfunction
  - 1.04 per 100 person-years in those without systolic dysfunction
- Relative risk (adjusted): 4.6

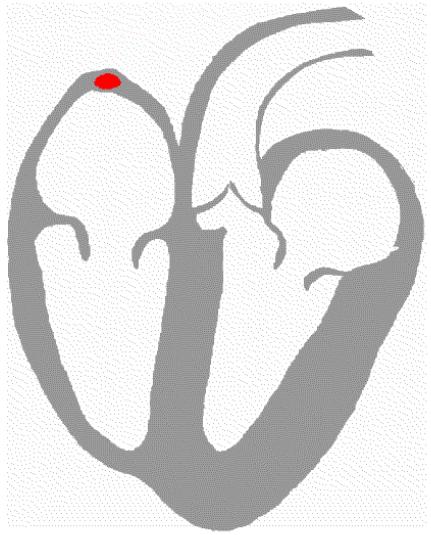
Echouffo-Tcheugui JB et al. Assessing the risk of progression from asymptomatic left ventricular dysfunction to overt heart failure. JACC HF 2016;4:237

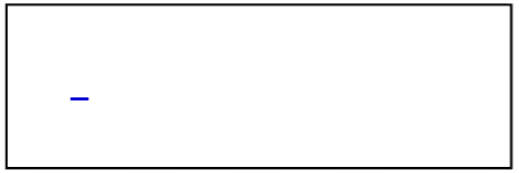
DCM: Unfavorable Prognostic Factors

- Male
- Black
- Increasing age
- NYHA Class III or IV at presentation
- Initial symptoms lasting > 3 months
- Greater LV enlargement
- Lower LV EF
- RV enlargement
- Low RV EF
- LBBB
- Markers of electrical instability
- Elevated biomarkers
- Lamin A/C mutation
- Myocardial fibrosis by MRI

Systolic Function: Global Longitudinal Strain (GLS)

- Measure of longitudinal shortening of LV during systole compared to baseline
- Measured by echocardiography: speckle strain imaging
- Detects systolic dysfunction earlier than EF
- May be expressed as a negative number – use the absolute value
- Range for GLS (may vary with software)
  - Normal: > 18%
  - Borderline: 16-18%
  - Significant systolic dysfunction: < 16%





Case Study #2: Wrap Up

- 1/20 Echo
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  - LVIDs 4.0
  - LA 4.1
  - Normal RV size and function
  - Normal mitral valve with mild MR
  - LVEF 43%
- 1/22 - last cardiology follow up
  - No symptoms
  - On empagliflozin
  - Office echo – “stable”
- NTproBNP on insurance labs was 56

What would your recommendation be at this time?

What would your recommendation be if NTproBNP was 456?

What would your recommendation be if NTproBNP was 1456?

Case Study #2: Val's Thoughts

- 1/20 Echo
  - LVIDd 6.1
  - LVIDs 4.0
  - LA 4.1
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  - Normal mitral valve with mild MR
  - LVEF 43%
- 1/22 - last cardiology follow up
  - No symptoms
  - On empagliflozin
  - Office echo – "stable, EF 45%"
- NTproBNP on insurance labs was 56

What would your recommendation be at this time?

- Stable idiopathic DCM
- Asymptomatic (Stage B heart failure)
- Well-followed
- NTproBNP favorable
- Increased risk, moderate to high substandard

What would your recommendation be if NTproBNP was 456?

- Significantly higher risk

What would your recommendation be if NTproBNP was 1456?

- Decompensation, likely overt heart failure, very high risk

Case Study #2: Discussion Questions

1. What could be causing the low ejection fraction?
   - Myocarditis, dilated cardiomyopathy (tachycardia-mediated, alcohol)
   - No indication of valvular disease (mitral regurg or aortic insufficiency would be most likely) on echo reports
2. What other information would be helpful in assessing this risk?
   - More recent echo, cardiac MRI
   - More details about sleep apnea and compliance/efficacy of CPAP
3. An APS note states Stage B heart failure. Is this a typo?
   - Stage B heart failure is called “pre-heart failure” and includes those with structural heart disease but no symptoms of heart failure (never had any symptoms of heart failure)
4. What is appropriate treatment and follow up?
   - Treat underlying cause if known
   - Manage cardiovascular risk factors and comorbidities
   - Beta-blockers, ACE/ARB inhibitors, diuretics, SGLT2i
5. What is the risk of progression of asymptomatic systolic dysfunction to heart failure?
   - 8.4 per 100 person-years
6. What factors are important in assessing this risk?
   - LV size, LVEF, stability, ECG findings, exercise capacity/symptoms, family history
7. What is GLS and would it be helpful in this case?
   - Global longitudinal strain is a measure of systolic function obtained by speckle strain imaging (a type of Doppler echocardiography)
   - GLS would not be useful in this case as it can help identify early systolic dysfunction before the EF decreases

Useful References: Asymptomatic Systolic Dysfunction and DCM

- Bozkurt B et al. Current diagnostic and treatment strategies for specific dilated cardiomyopathies. Circulation 2016;134:e579.
- Martin LD et al. Prevalence of asymptomatic left ventricular systolic dysfunction in at-risk medical inpatients. Am J Cardiol 2013;126:68.
- Echouffo-Tcheugui JB et al. Assessing the risk of progression from asymptomatic left ventricular dysfunction to overt heart failure. JACC HF 2016;4:237.
- Marwick TH. Ejection fraction pros and cons – JACC-state-of-the-art review. J Am Coll Cardiol 2018;72:2360.
- Luis SA et al. Echocardiographic assessment of left ventricular systolic function: an overview of contemporary techniques, including speckle-tracking echocardiography. Mayo Clin Proc 2019;94:125.
- Halliday BP et al. Personalizing risk stratification for sudden death in dilated cardiomyopathy – the past, present and future. Circulation 2017;136:215.

3

Ventricular Ectopy

RVOT vs ARVC

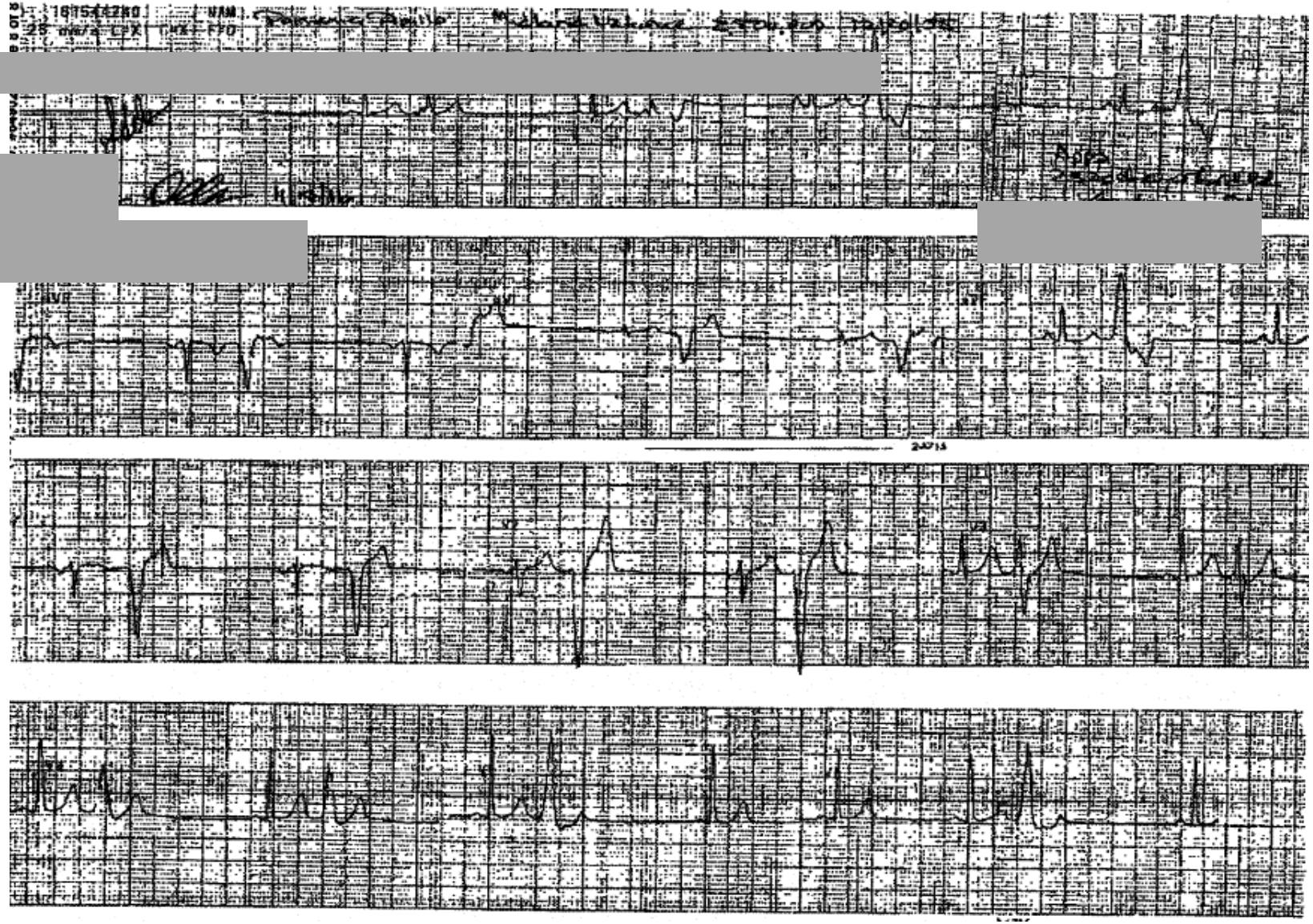
Case Study #3

- 30 year old woman
- 5’5” (1.65 m), 134 lbs (61 kg)
- No adverse FH noted in parents and siblings
- Had echo and Holter 6 months ago due to palpitations
- Insurance labs normal

Case Study #3: APS

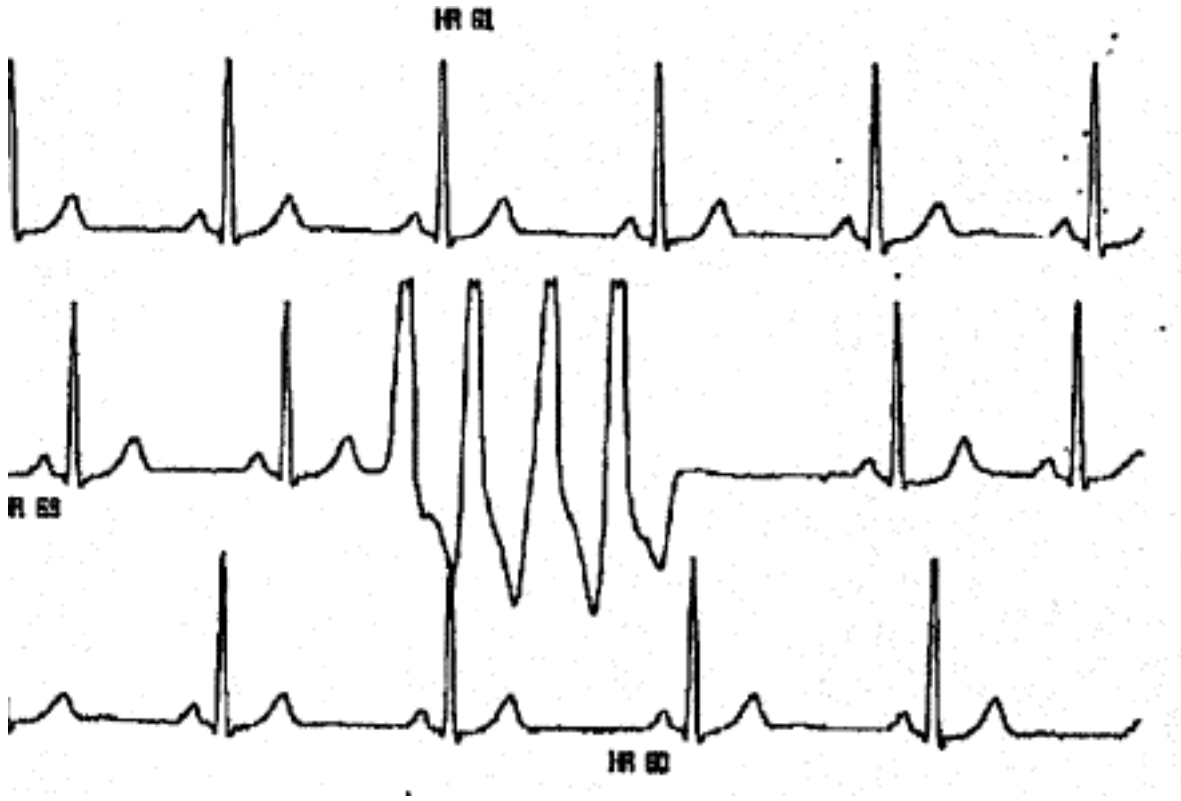
- Referred to cardiology due to palpitations, worse at night and with fatigue and stress, 6 months ago
- Additional family history: maternal grandfather died “young”
- EKG showed frequent PVCs
- Echo
  - Normal LV size and function
  - Valves normal with trace MR and TR
  - RV mildly enlarged with normal function
- Stress test
  - Exercised 12 minutes, Bruce protocol to peak heart rate of 165
  - BP increased from 112/78 to 148/68
  - No ST segment changes.
  - Occasional PVCs during exercise and recovery. Two couplets at peak exercise

Case Study #3: Resting ECG



Case Study #3: Holter Monitor

- Monitored for 23 hours, 12 minutes
- 83,520 total beats
- 3925 single PVCs
- 60 couplets
- Five 4-beat runs of VT at rates of 120 – 150
- 4.8% of beats were ventricular

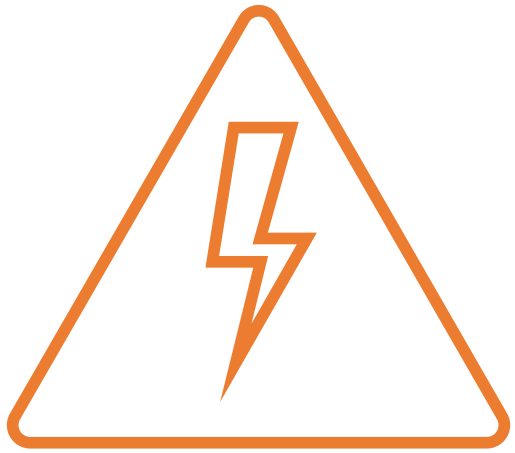


Case Study #3: Cardiologist Conclusion

- Cardiologist concluded benign right ventricular outflow tract (RVOT) tachycardia
- Begun on verapamil
- Discussed ablation if medication not effective
- Follow up planned in one year

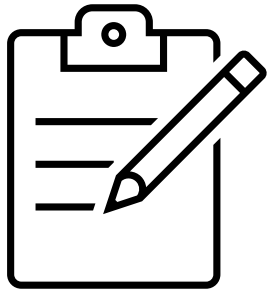
What else would you want to know?

- How is she doing now?
- Any follow up?



Case Study #3: Discussion Questions

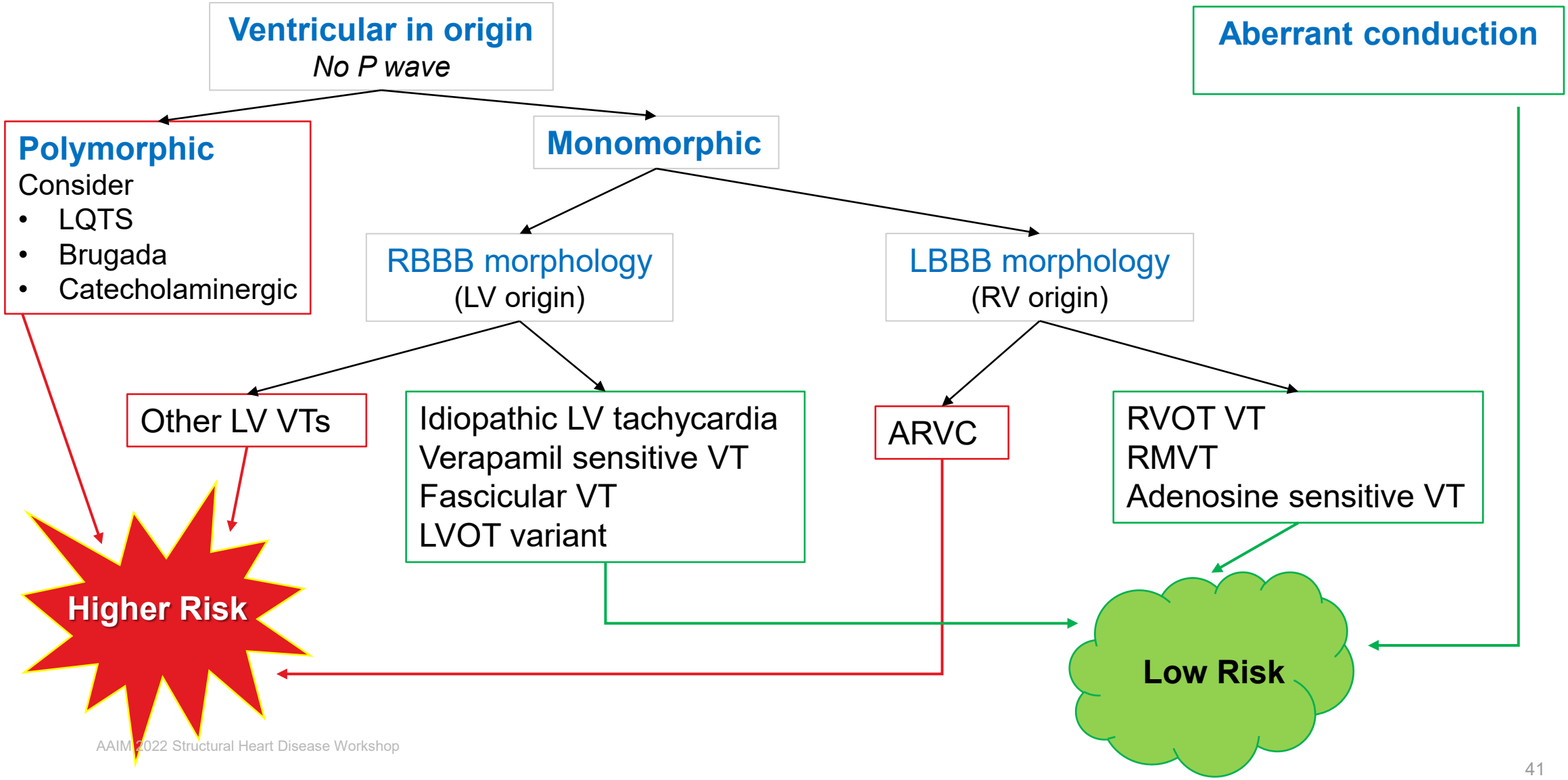
1. Can VT ever be “benign”? And how “benign” is benign?
2. How can higher risk cases be distinguished from lower risk cases?
3. Differential diagnosis for monomorphic VT arising from the right ventricle?
4. Is the number of PVCs worrisome by itself?
5. Was the stress test helpful? If so, how?



Assessment of Ventricular Ectopy

- Most important risk factor - presence or absence of structural heart disease
- PVCs are present in up to 75% of normal individuals on Holter monitor
- Nonsustained VT found in up to 4% of normal individuals
- Ventricular ectopy occurring in normal hearts – referred to as idiopathic
  - 10% of VT is idiopathic
  - Most idiopathic VT is “benign”
  - Must be distinguished from a few higher risk conditions
  - If frequent ectopy - at risk for tachycardia-mediated cardiomyopathy. Risk increases as PVC burden increases. “Frequent” defined in different ways:
    - > 20,000 ectopics/day
    - > 24% of total beats
    - > 30 PVCs per hour
- 2 types of idiopathic VT
  - Outflow tract VT, both RVOT and LVOT
  - Fascicular VT, also known as verapamil sensitive VT

Wide Complex Tachycardia, “Normal” Heart



Right Ventricular Outflow Tract (RVOT) Tachycardia

- RVOT tachycardia – also known as
  - RV tachycardia
  - RMVT – repetitive monomorphic VT
  - Catecholamine or adenosine sensitive VT
- 10-15% may arise from the left ventricular outflow tract
- Occurs in young/middle-aged individuals without structural heart disease
- Symptoms include palpitations and lightheadedness but frank syncope is uncommon
- Long term outcome - generally favorable
- Red flags

◀History of syncope

◀Very fast VT – faster than 230 beats/min

◀R on T PVCs (PVCs with short coupling interval)

Favorable Factors in Assessment of Ventricular Ectopy

Favorable Clinical Characteristics

- No underlying structural heart disease
- No history of syncope
- No FH of arrhythmia syndrome or other inherited cardiac conditions

Favorable Arrhythmia Characteristics

- Monomorphic (unifocal)
- RV origin (LBBB morphology)
- < 230 beats/minute
- Verapamil or adenosine sensitive
- Low frequency/burden of ectopic beats

Is There ANY Increased Risk with Idiopathic PVCs?

- Undiscovered or undeclared heart disease – MRI studies
- Tachycardia/PVC-mediated cardiomyopathy
- Atherosclerosis Risk in Communities (ARIC) study
  - PVCs present in 6.2% at baseline
  - Increased stroke risk (7.3% vs 4.8%)
  - Thrombotic stroke equal in those with and without PVCs; embolic stroke risk higher in those with PVCs
  - Increased risk of clinical CAD
  - HR 2.1 for sudden death with PVCs vs no PVCs

Arrhythmogenic Right Ventricular Cardiomyopathy

- Progressive disease characterized by
  - fibrofatty replacement of myocardium
  - wall motion abnormalities
  - RV dilatation
  - RV aneurysms
- LV often involved – Arrhythmogenic Cardiomyopathy?
- Clinical manifestations
  - Often asymptomatic
  - Palpitations, syncope, sudden cardiac death, ventricular arrhythmias
  - Chest pain
  - Dyspnea
  - Heart failure

ARVC

- Initial manifestations appear around age 12
- Mean age at presentation is 30
- Important cause of sudden cardiac death in the young
- Autosomal dominant with variable expressivity and incomplete penetrance
- Prevalence: 1/2000 – 1/5000
- Mortality: 2-4%/year
- Diagnosis: very complicated

Diagnosis of ARVC

2010 Revised Task Force Criteria

- 6 categories, with major and minor criteria in each
- Definite, Borderline and Possible diagnostic categories
- Categories
  - Global or regional dysfunction and structural alterations
  - Tissue characterization of ventricular wall
  - Repolarization abnormalities
  - Depolarization/conduction abnormalities
  - Arrhythmias
  - Family history

Marcus FI et al. Diagnosis of ARVC/D: proposed modification of Task Force criteria. Circulation 2010;121:533.

RVOT vs ARVC

	RVOT Tachycardia	ARVC
FH of arrhythmia or SCD	No	Frequently yes
Arrhythmias	VT at rest or with exercise	VT at rest or with exercise
Sudden cardiac death	Rare	1% per year
Frontal plane QRS	Positive in leads 3 and avF, negative in lead avL	Inferior or superior axis
T wave morphology	T wave upright V2-5	T wave inverted beyond V1
Epsilon wave V1-3	Absent	Present 30%
SAECG	Normal	Usually abnormal
Echocardiogram	Normal (?)	Increased RV size and/or wall motion abnormalities
RV ventriculogram	Usually normal	Usually abnormal
MRI	Usually normal, but data conflicting	Increased signal intensity of RV free wall; wall motion abnormalities
Response to therapy	Acute: vagal maneuvers, adenosine, verapamil, βblocker Chronic: βblocker , verapamil, class 1 antiarrhythmics	Sotalol, amiodarone, βblocker
RF ablation	Usually curative	Seldom curative

Johns Hopkins ARVD Center of Excellence, information for physicians, accessed 9/6/18

Another Approach: Scoring System

Based on a small study

- Baseline ECG: T wave inversions V1-3 → 3 points
- Ectopic beat: Lead 1 QRS duration > 120ms → 2 points
- Ectopic beat: QRS notching in multiple leads → 2 points
- Ectopic beat: R/S transition in V5 or later → 1 point

Score of 5 or higher predicts ARVC with sensitivity of 84%, specificity of 100%, PPV of 100% and NPV of 91%

Hoffmayer KS et al. An electrocardiographic scoring system for distinguishing RVOT arrhythmias in patients with ARVC from idiopathic tachycardia. Heart Rhythm 2013 April;10(4):477-82.

Case Study #3: Review Resting ECG

In normal beat, TWI V1-3?

NO

In ectopic, Lead 1 QRS duration > 120?

YES – 2 points

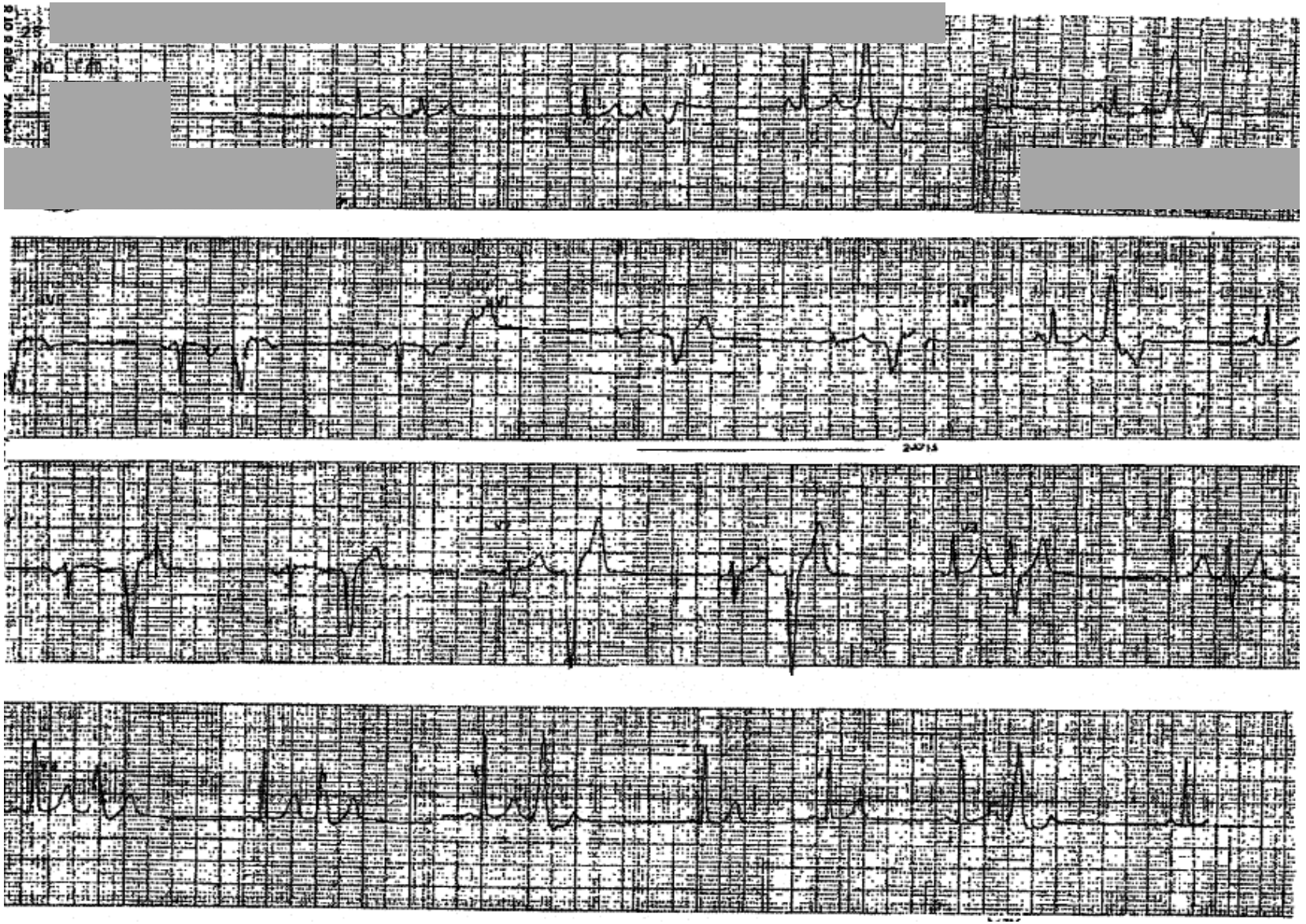
In ectopic, QRS notching in multiple leads?

NO

In ectopic, R/S transition in V5 or later?

NO

Unlikely to be ARVC



Case Study #3: Wrap Up

- 30 year old woman
- Palpitations lead to cardiac evaluation
- No adverse FH noted in parents and siblings; maternal grandfather “died young”
- EKG showed frequent PVCs, monomorphic, LBBB pattern
- Echo: normal except for mild RV enlargement with normal function
- Stress test: Occasional PVCs during exercise and recovery. Two couplets at peak

ARVC or RVOT?

What would your recommendation be at this time?

Case Study #3: Val's Thoughts

- 30 year old woman
- Palpitations lead to cardiac evaluation
- No adverse FH noted in parents and siblings; maternal grandfather "died young"
- EKG showed frequent PVCs, monomorphic, LBBB pattern
- Echo: normal except for mild RV enlargement with normal function
- Stress test: Occasional PVCs during exercise and recovery. Two couplets at peak

ARVC or RVOT?

- Pro ARVC (more concerning)
  - RV is enlarged
  - Grandfather "died young"
- Pro RVOT (more benign)
  - No syncope
  - Monomorphic, LBBB pattern
  - FH vague at best
  - No PVCs in recovery
  - Response to verapamil

Most likely RVOT/benign

What would your recommendation be at this time?

- Some increased risk, in the low range

Case Study #3: Discussion Questions

1. Can VT ever be “benign”? And how “benign” is benign?
   - Some types of VT are thought to be more benign than others. About 75% of “normal” individuals have PVCs on Holter, and up to 4% have nonsustained VT. This “idiopathic” ventricular ectopy is thought to be more benign than ventricular ectopy associated with heart disease or arrhythmic syndromes.
   - However, studies suggest some increased mortality for VT, even when thought to be idiopathic

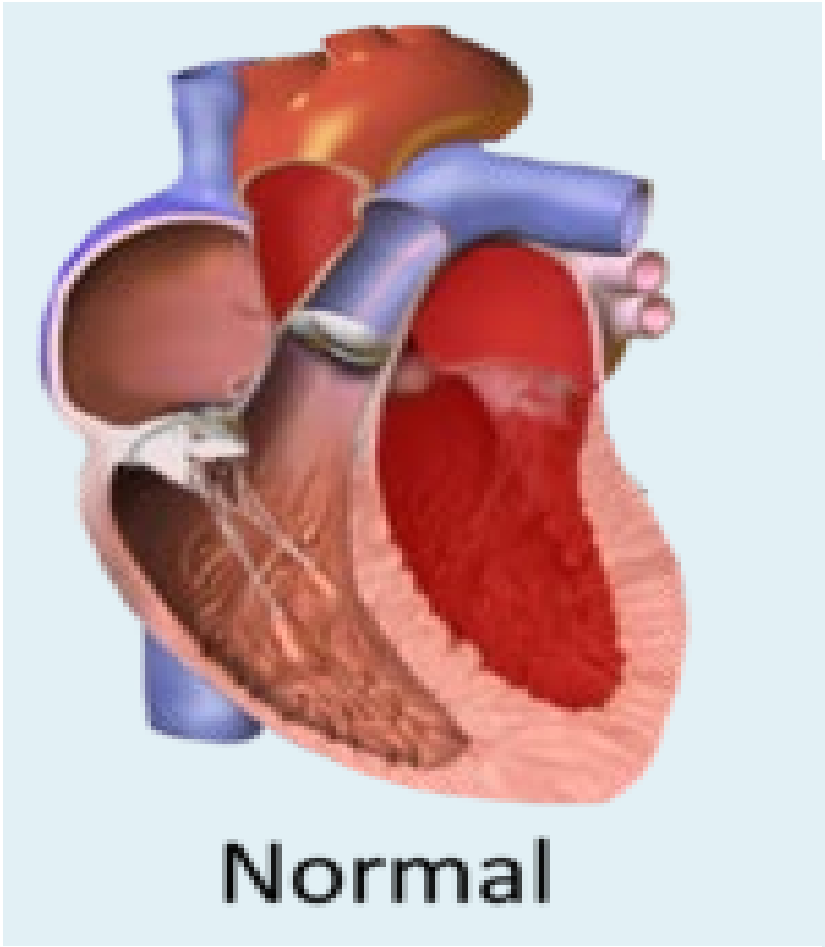
2. How can higher risk cases be distinguished from lower risk cases?
   - Most important distinction: is there underlying heart disease present or not?
   - With no underlying heart disease, the following are helpful for risk stratification:
     - RV origin
     - Monomorphic
     - Not frequent
     - No symptoms (mainly syncope)
     - If stress test, PVCs during exercise rather than during recovery only.
3. Differential diagnosis for monomorphic VT arising from the right ventricle?
   - Main considerations include RVOT and ARVC
4. Is the number of PVCs worrisome by itself?
   - Mortality increases with increasing frequency of PVCs
   - Risk of tachycardia or PVC-mediated cardiomyopathy increases with very frequent ectopy (> 30 PVCs/hour, > 20,000 PVCs/24 hours, >24% total beats)
5. Was the stress test helpful? If so, how?
   - Modestly helpful. Ventricular ectopy occurring only in recovery may portend higher risk of adverse cardiac events.

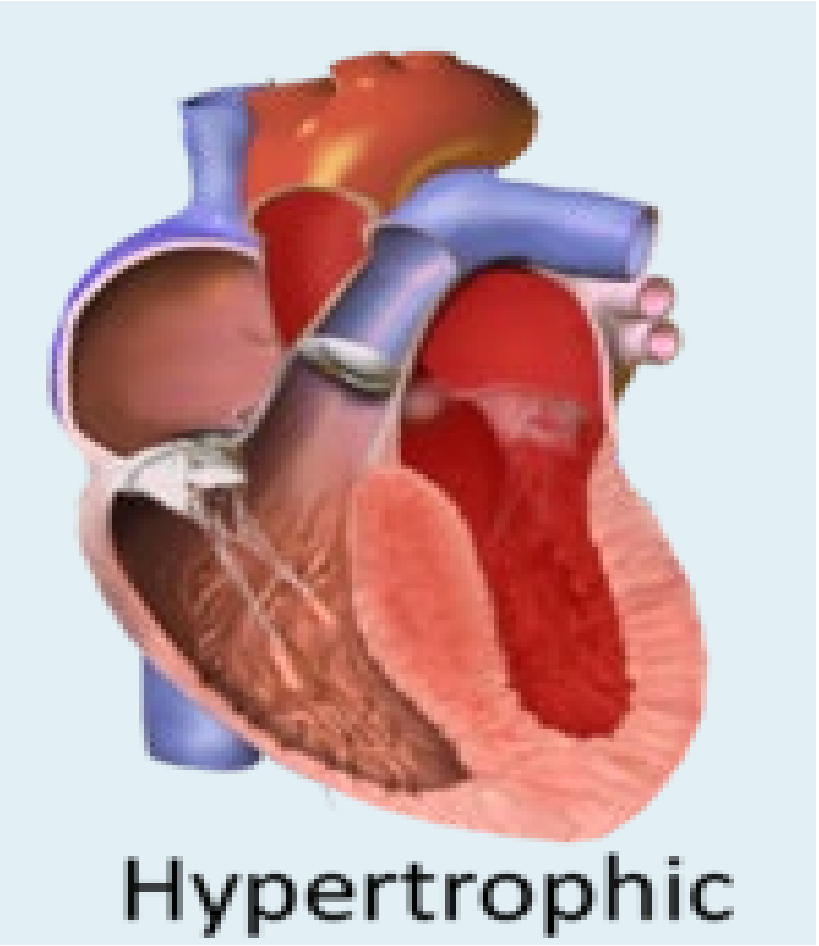
Helpful References

- Refaat MM et al. Exercise-induced ventricular ectopy and cardiovascular mortality in asymptomatic individuals. J Am Coll Cardiol 2021;78:2267.
- Saurav A et al. Premature ventricular contraction-induced cardiomyopathy. Clin Cardiol 2015;38(4):251.
- Al-Khatinib SM et al. 2017 AHA/ACC/HRS guideline for management of patients with ventricular arrhythmias and the prevention of sudden cardiac death. J Am Coll Cardiol 2017;72:e91.
- Marcus FI et al. Diagnosis of ARVC/D: proposed modification of Task Force criteria. Circulation 2010;121:533.
- Gandjbakhch E et al. Clinical diagnosis, imaging and genetics of ARVC/D. JACC 2018;72:784.
- Hoffmayer KS et al. An electrocardiographic scoring system for distinguishing RVOT arrhythmias in patients with ARVC from idiopathic tachycardia. Heart Rhythm 2013 April;10(4):477-82.

4

Hypertrophic Cardiomyopathy





Case Study #4

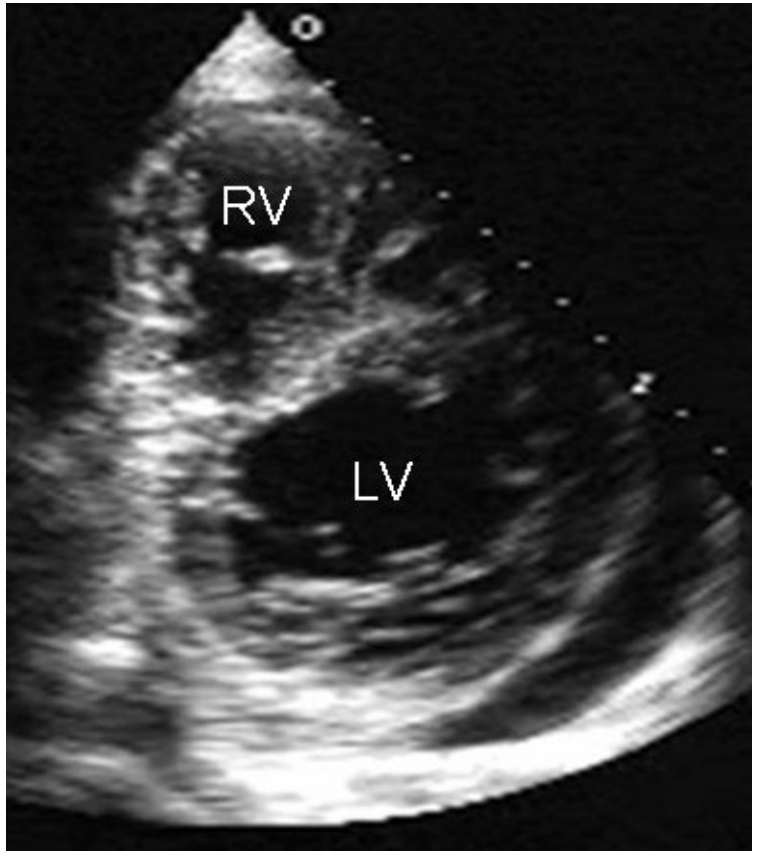
- 50 year old man, applying for $3 million
  - 5’11”, 193 lbs., BP 120/72
  - No medical history per part two
  - Last seen eight months ago for annual physical
  - FH: father died at age 70 due to lung cancer

    mother had breast cancer diagnosed at age 55
- Insurance labs all normal. NTproBNP 53
- APS: mother diagnosed age 68 with HCM. PI being monitored for this

Case Study #4: Additional APS Information

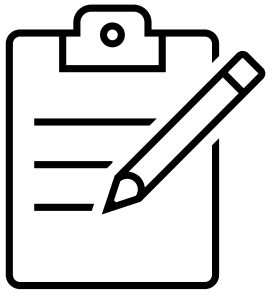
- No symptoms (palpitations, dyspnea, syncope)
- ECG normal
- Echos 2012 and 2014 normal

	Septum (mm)	**P. Wall** (mm)	**Other**
2012	11	10	Normal
2014	11	10	Normal
2016	14.1	11.5	**Possible HCM**
2/21	13	13	SAM, moderate MR
4/21 Mayo	15	11	No SAM, mild MR. **Diagnosed with HCM**
6/21 (CMR)	14	12	14 mm in multiple locations Mild MR, no convincing evidence of SAM



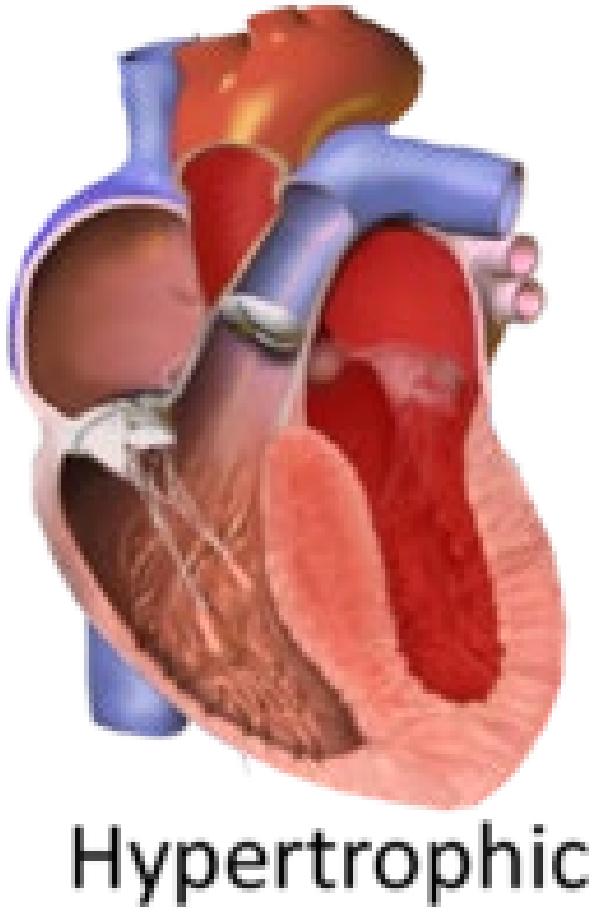
Case Study #4: Discussion Questions

1. Does this make sense? Can HCM “show up” at age 50?
2. Why are the echo values fluctuating?
3. How can the ECG be normal?
4. What, if any, additional testing could help?
5. What is appropriate follow up? Treatment?
6. What if the PI was a competitive distance runner (10K, ½ marathon)
7. Should the PI’s children be screened?
8. With increased genetic testing, what about those that are genotype positive (for HCM) but phenotype negative?



HCM Definition

"a 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"



Gersh BJ, Maron BJ et al. 2011 ACCF/AHA guideline for the diagnosis and treatment of hypertrophic cardiomyopathy. JACC 2011;58:2703-38.

About HCM

- Cause: Inherited
  - > 1500 mutations identified on 9 genes that encode for myocardial contractile proteins
  - Autosomal dominant, high penetrance, variable expression
- Prevalence: common, 1/200 - 1/500 in US population
- Signs and symptoms
  - Often asymptomatic (about half asymptomatic at presentation)
  - Classic presentation – exertional syncope
  - Dyspnea
  - Ventricular arrhythmias and palpitations
  - Abnormal screening ECG
- Complications
  - Sudden cardiac death
  - Heart failure

HCM Diagnosis

- Diagnosed by echo or cardiac MRI
  - Hypertrophy highly variable in location and pattern
    - Can be focal or more generalized
    - Often involves the anterior free wall of LV, difficult to “see” by echo
    - Can involve the RV in addition to the LV
    - Maximum wall thickness usually (but not always) > 15 mm
  - Normal or small LV diameter (LVIDd)
  - Normal or “supernormal” LVEF (cavity obliteration)
  - May have SAM (systolic anterior motion of the mitral valve)
  - May have LV outflow tract obstruction (gradient)
  - May have fibrosis on MRI
- ECG usually (90%) abnormal – increased voltage with ST–T changes

“Phenocopies”

- Cardiac amyloidosis
- Fabry disease
- Mucopolysaccharidoses
- Hypertensive heart disease

HCM Treatment

- Treatment aimed at reducing symptoms and preventing complications; has minimal impact on mortality
- All should avoid strenuous physical activities due to ↑ arrhythmia risk
- If asymptomatic, no specific treatment*
- With symptoms of LV outflow obstruction
  - Beta blockers
  - Calcium channel blockers
  - Myectomy
  - Alcohol septal ablation
  - Mavacamten*
- ICD to interrupt arrhythmias
- Heart transplant

HCM Mortality: Markers of Increased Risk

- Major Risk Markers
  - Prior cardiac arrest or sustained ventricular tachycardia
  - Unexplained syncope
  - Multiple runs of nonsustained VT (3 or more beats) on monitor
  - Family history of premature HCM-related sudden cardiac death
  - Extreme LVH with maximum wall thickness ≥ 3.0 cm
  - LGE (late gadolinium enhancements) ≥ 15% of LV mass on cardiac MRI
  - LV apical aneurysm
  - End stage (EF < 50%)

- Other Potential Risk Markers
  - Abnormally low or decrease in BP with exercise (especially in those < age 50) (high risk)
  - LV outflow tract gradient ≥ 30 mm Hg at rest (high risk)
  - Age ≥ 60 (lower risk, even with Major Risk Markers)
  - ↑ NTproBNP
  - ↑ troponin T

Risk Assessment in HCM

- Mortality about 1% per year or less in adults without high risk markers
- Morphologic changes usually begin to appear during adolescence, but not always
- Echo findings can be dynamic
- ECG changes can precede morphologic changes
- ECG may be normal
- Genetic changes have high penetrance but variable expression
- Genetic changes cannot be used to risk stratify
- First degree relatives should be screened regularly throughout life (50% risk)

HCM Testing and Follow Up

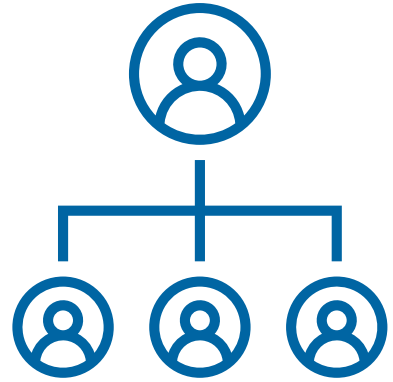
- Repeat echo is recommended
  - Every 1-2 years in those with stable symptoms
  - When there is a change in status or a new clinical event
- Heart rhythm assessment
  - ECG every 1-2 year is recommended
  - Holter every 1-2 years is recommended
- If palpitations or lightheadedness develop, extended monitoring or event recorder is recommended
- Cardiac MRI is recommended
  - For diagnosis if echo is inconclusive
  - If LVH is present with a concern for alternative diagnosis such as infiltrative/storage diseases or AHS
  - When risk of SCD is unclear, to assist in decision about ICD

Ommen SR et al. 2020 AHA/ACC guideline for the diagnosis and treatment of patients with hypertrophic cardiomyopathy. J Am Coll Cardiol 2020;76:e159.

Family Screening Guidelines

First Degree Relatives (when no genetic testing done)

- Under age 12 years – screening optional unless
  - "Malignant" clinical course in family
  - Competitive athlete in intense training
  - Symptoms
- Ages 12 to 18-21 years
  - ECG and echo every 12-18 months
- Age > 18-21 years
  - At onset of symptoms
  - If no symptoms, at least every 5 years
  - More frequent intervals in families with more "malignant" clinical course



Gersh BJ, Maron BJ et al. 2011 ACCF/AHA guideline for the diagnosis and treatment of hypertrophic cardiomyopathy. JACC 2011;58:2703-38.

Genetic Testing in HCM

- Most useful for screening relatives of those with the disease and an identified mutation
- Not recommended as primary diagnostic method in absence of known familial mutation
- Genotype positive, phenotype (hypertrophy) negative
  - Generally considered a “preclinical” phase
  - Serial ECGs and echos (every 1-2 years in children; every 3-5 years in adults)
  - Participation in athletics of any intensity is *reasonable*
- Ongoing clinical screening is not indicated in genotype-negative relatives in families with genotype-positive HCM

Risk Assessment of HCM

- Mortality
  - Generally about 1% / year or less in adults, if no high-risk markers
  - 2–6%/year in children

High Risk Markers

- Prior cardiac arrest or spontaneous sustained VT
- Multiple runs of non–sustained VT (three or more beats) on Holter
- Family history of HCM–related sudden cardiac death under age 50
- Unexplained syncope
- Decrease in BP with exercise
- Extreme LVH with maximum wall thickness 30 mm or more
- Late gadolinium enhancements (LGE) or fibrosis on MRI
- Regular participation in strenuous physical activity
- Left ventricular outflow gradient > 30 mm Hg

Case Study #4: Wrap Up

- 50–year–old man
- HCM diagnosed 2020
  - No symptoms
  - Normal ECG
  - Mother diagnosed at age 68
- With no high–risk markers

What would your recommendation be at this time?

Would any additional information allow for a more favorable offer?

High Risk Markers

- Prior cardiac arrest or spontaneous sustained VT
- Multiple runs of non–sustained VT (three or more beats) on Holter
- Family history of HCM–related sudden cardiac death under age 50
- Unexplained syncope
- Decrease in BP with exercise
- Extreme LVH with maximum wall thickness 30 mm or more
- Late gadolinium enhancements (LGE) or fibrosis on MRI
- Regular participation in strenuous physical activity
- Left ventricular outflow gradient > 30 mm Hg

Case Study #4: Val's Thoughts

- 50–year–old man
- HCM diagnosed 2020
  - No symptoms
  - Normal ECG
  - Mother diagnosed at age 68
- With no high–risk markers

What would your recommendation be at this time?
No high risk markers. Low to moderate increased risk

Would any additional information allow for a more favorable offer?
With confirmed diagnosis, no, additional information unlikely to help

High Risk Markers

- Prior cardiac arrest or spontaneous sustained VT
- Multiple runs of non–sustained VT (three or more beats) on Holter
- Family history of HCM–related sudden cardiac death under age 50
- Unexplained syncope
- Decrease in BP with exercise
- Extreme LVH with maximum wall thickness 30 mm or more
- Late gadolinium enhancements (LGE) or fibrosis on MRI
- Regular participation in strenuous physical activity
- Left ventricular outflow gradient > 30 mm Hg

Case Study #4: Discussion Questions

1. Does this make sense? Can HCM "show up" at age 50?
   - HCM typically manifests structural changes beginning in adolescence/young adulthood, but not always. It may appear at any time throughout life.
   - One of the most common sites of focal hypertrophy is the anterior free wall of the LV – which is difficult to image on echo.
2. Why are the echo values fluctuating?
   - HCM is dynamic. Findings can fluctuate. Also must consider variation from one echo report to next
3. How can the ECG be normal?
   - The ECG is usually abnormal in HCM, but may be normal in up to 10% of cases. Interestingly, the opposite may be true – ECG changes may precede the structural manifestations.
   - Must be aware of "phenocopies" that mimic some of the findings in HCM such as cardiac amyloidosis, Fabry disease, mucopolysaccharidoses and hypertensive heart disease.
4. What, if any, additional testing could help?
   - 24 hour Holter to check for arrhythmias
   - Possibly more detailed family history
5. What is appropriate follow up? Treatment?
   - If no new signs or symptoms, regular follow up every 1-2 years
   - If any new signs or symptoms, cardiac evaluation would be appropriate
   - Could consider beta blocker or calcium channel blocker therapy.
   - ? Mavacamten
   - Septal reduction therapy with alcohol septal ablation or myectomy if symptomatic obstruction

Case Study #4: Discussion Questions (cont)

1. What if the PI was a competitive distance runner (10K, ½ marathon)?
   - Strenuous exercise used to be contraindicated in HCM, but that is changing
   - Limited data suggests moderate exercise in asymptomatic HCM patients is safe and beneficial
   - No studies on those with HCM in strenuous/competitive sport – some data on ICDs suggests may be safe (with ICD in place)
2. Should the PI's children be screened?
   - Yes. Current recommendations are for lifelong surveillance of first degree relatives, unless genetic testing excludes the familial mutation
3. With increased genetic testing, what about those that are genotype positive (for HCM) but phenotype negative?
   - Generally considered to be presymptomatic. Not subject to activity restrictions
   - At risk for arrhythmias and long term complications
   - At this time, uncertain how to risk stratify
   - Advised regular cardiologic follow up and surveillance

Helpful References: HCM

- Maron BJ et al. Diagnosis and evaluation of hypertrophic cardiomyopathy – JACC state-of-the-art review. J Am Coll Cardiol 2022;79:372.
- Maron BJ et al. Management of hypertrophic cardiomyopathy – JACC state-of-the-art review. J Am Coll Cardiol 2022;79:390.
- Ommen SR et al. 2020 AHA/ACC guideline for the diagnosis and treatment of patients with hypertrophic cardiomyopathy. J Am Coll Cardiol 2020;76:e159.
- O'Mahony et al. A novel clinical risk prediction model for sudden cardiac death in HCM (HCM Risk-SCD). Eur Heart J 2014;35:2010.
- Maron BJ et al. Independent assessment of the European Society of Cardiology Sudden Death Risk Model for HCM. Am J Cardiol 2015;116:757.
- Semsarian C et al. Athletic activity for patients with hypertrophic cardiomyopathy and other inherited cardiovascular diseases. J Am Coll Cardiol 2022;80:1268.

Title of Presentation

Diastolic Dysfunction

Diastolic Function

- Includes
  - Ability of myocardium to relax between contractions (active, energy-dependent process)
  - Compliance (stretchiness) of myocardium (characteristic of the tissue)
  - Filling pressures
    - Left atrial pressure
    - Left ventricular end diastolic pressure
- Good diastolic function is necessary for optimal cardiac function
- If ventricle doesn’t fill properly, less blood gets pumped out

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- Good diastolic function is necessary for optimal cardiac function
- If ventricle doesn't fill properly, less blood gets pumped out

Ejection Fraction x End Diastolic Volume = Stroke Volume

↓ EF or ↓ EDV → ↓ SV

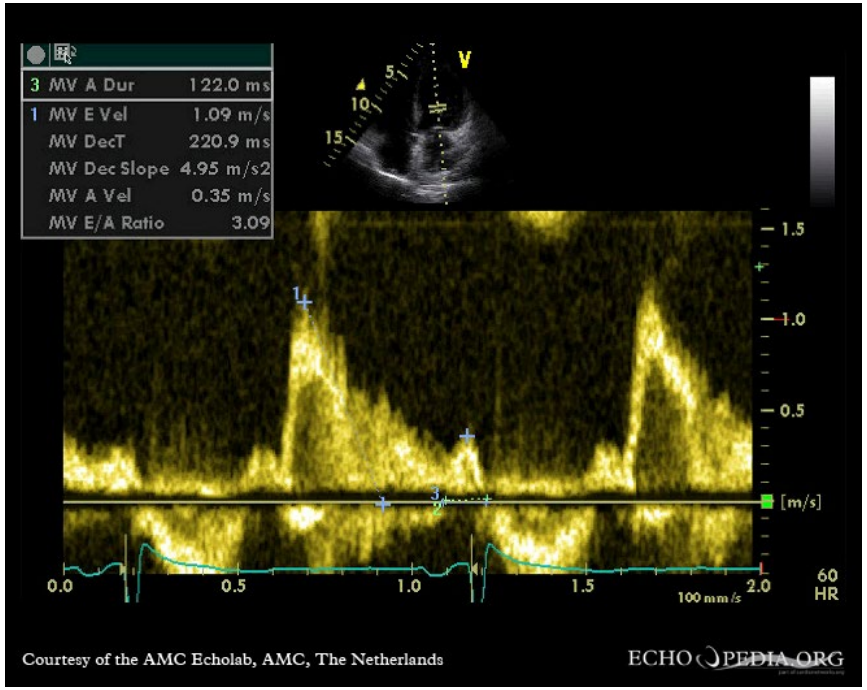
Stroke Volume x Heart Rate = Cardiac Output

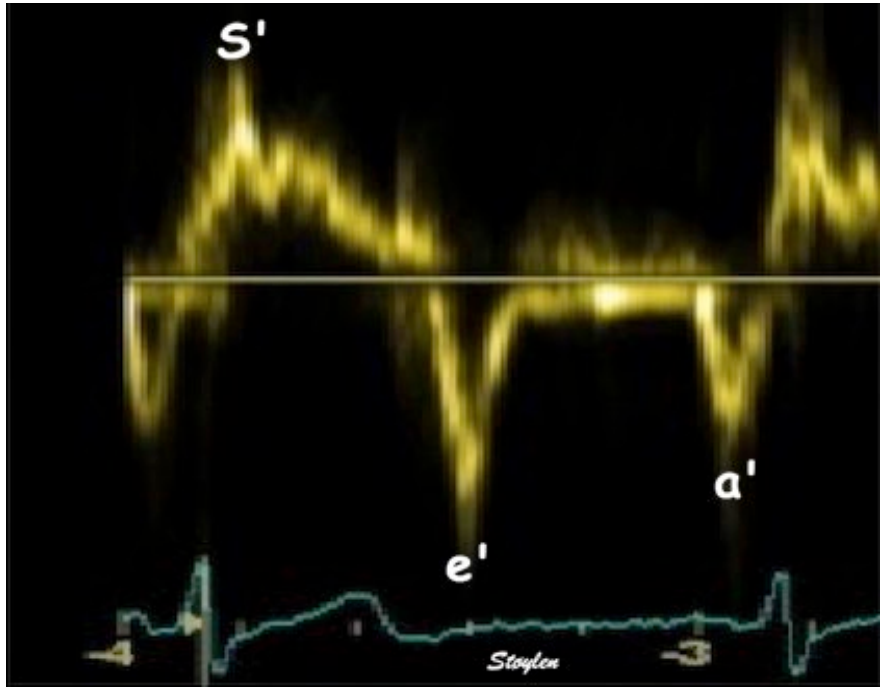
↓ SV or ↓ HR → ↓ CO

Assessment of Diastolic Function

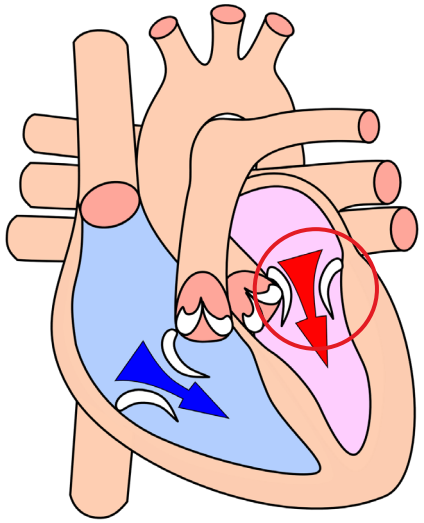
BAD NEWS

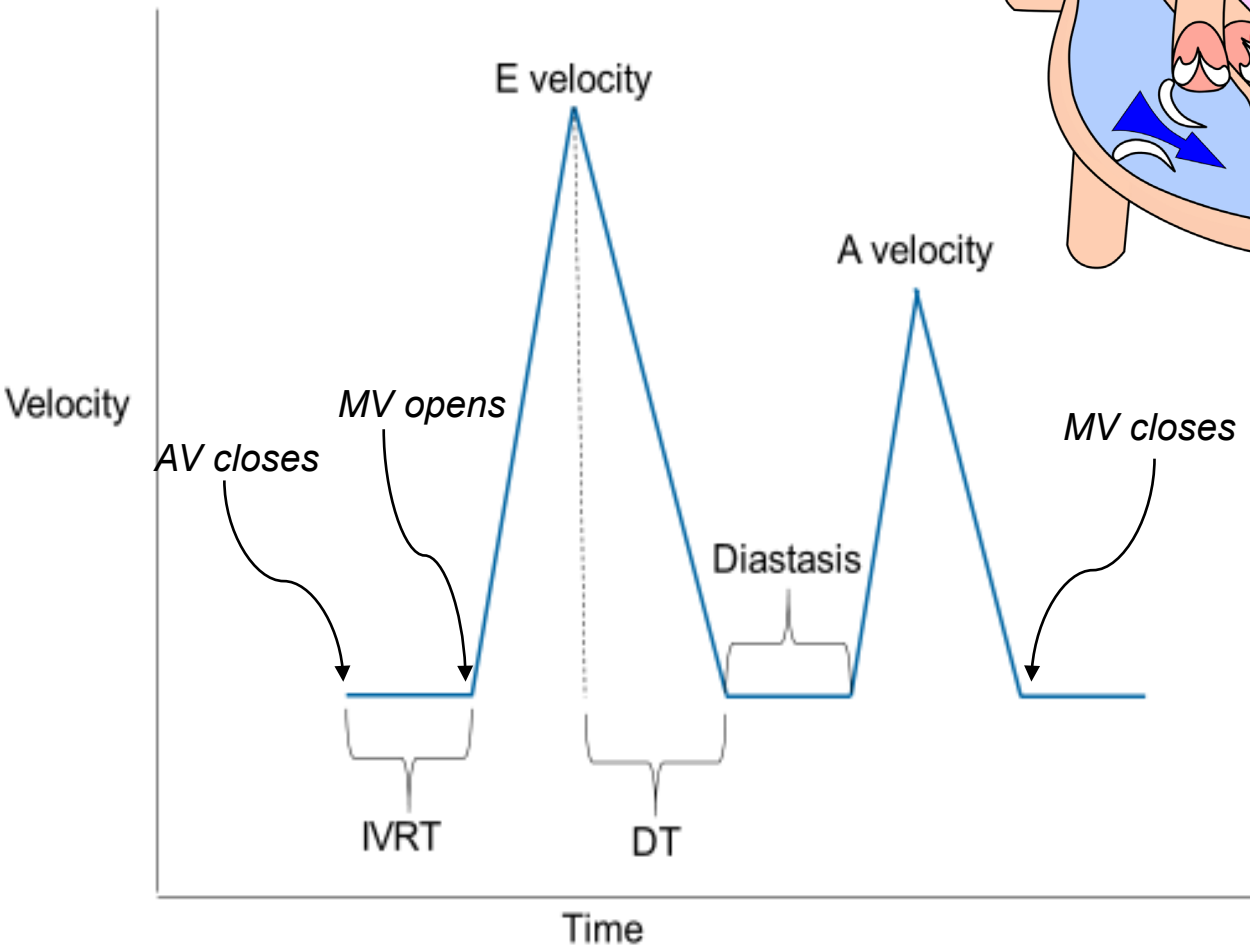
- Difficult, complicated. No single measurement like EF.
- Must consider multiple factors to determine if normal or abnormal
- If abnormal, other factors determine severity or grade
- Almost all factors dependent on Doppler echo





Doppler Transmitral Flow





Diastole - Time from aortic valve closure to mitral valve closure.
Four components:

1. IVRT
2. Early-rapid filling
3. Diastasis
4. Late filling

E velocity – Early-rapid filling *(decreases with age)*

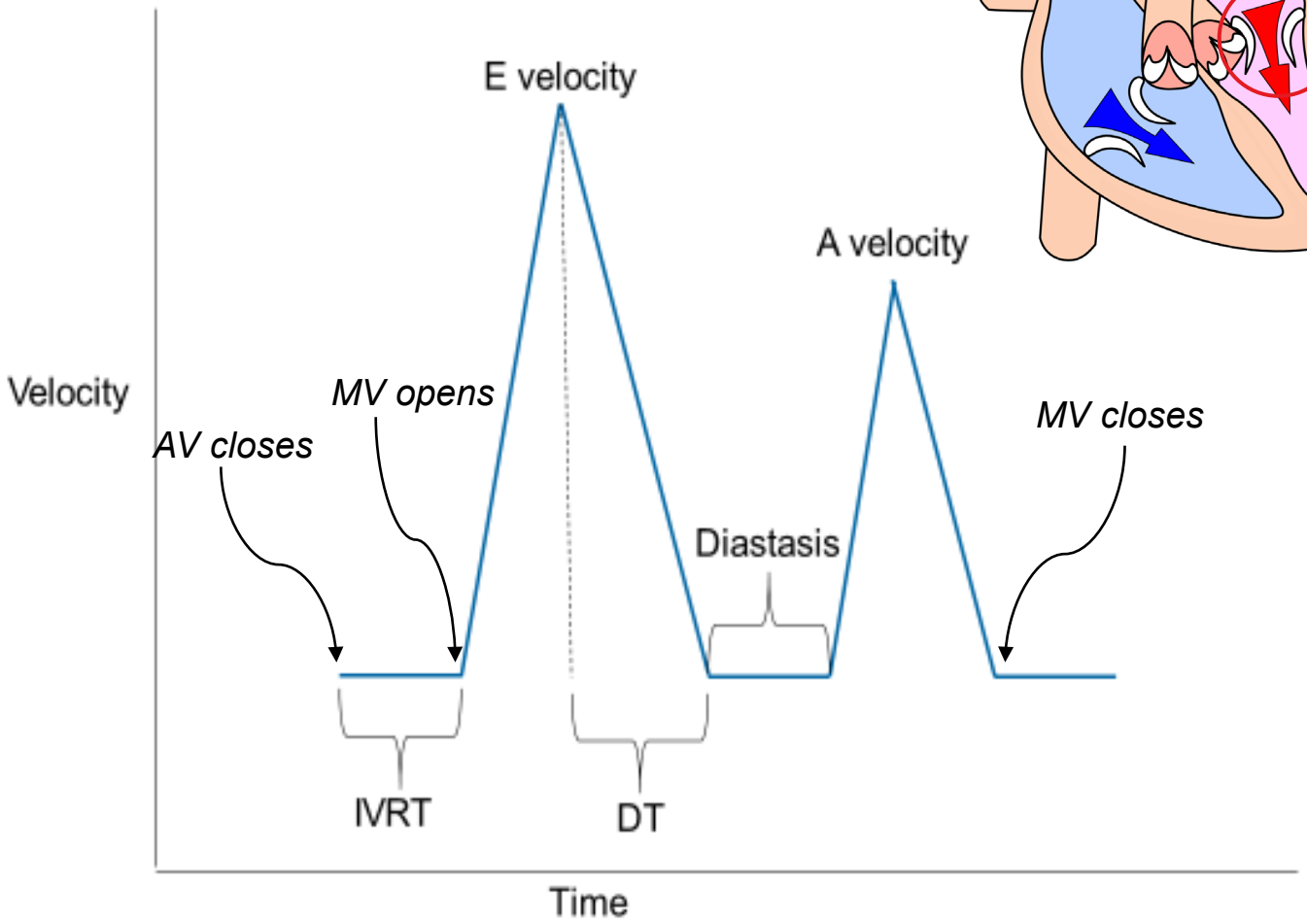
A velocity – Atrial contraction – late filling *(increases with age)*

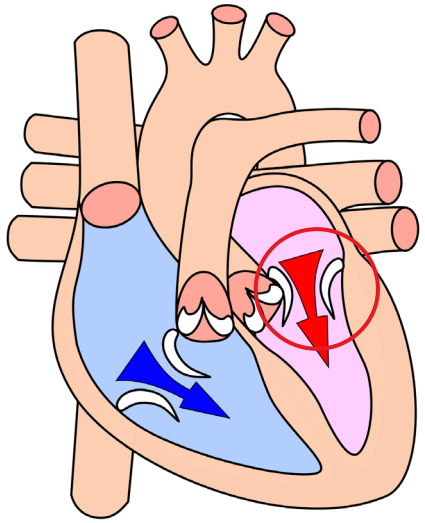
E/A ratio – Normal 0.8 – 2.0 *(decreases with age)*

IVRT – Isovolumic relaxation time

DT – Deceleration time

Doppler Transmitral Flow





***Cannot determine presence or absence of diastolic dysfunction by transmitral flow alone*

Diastole - Time from aortic valve closure to mitral valve closure.
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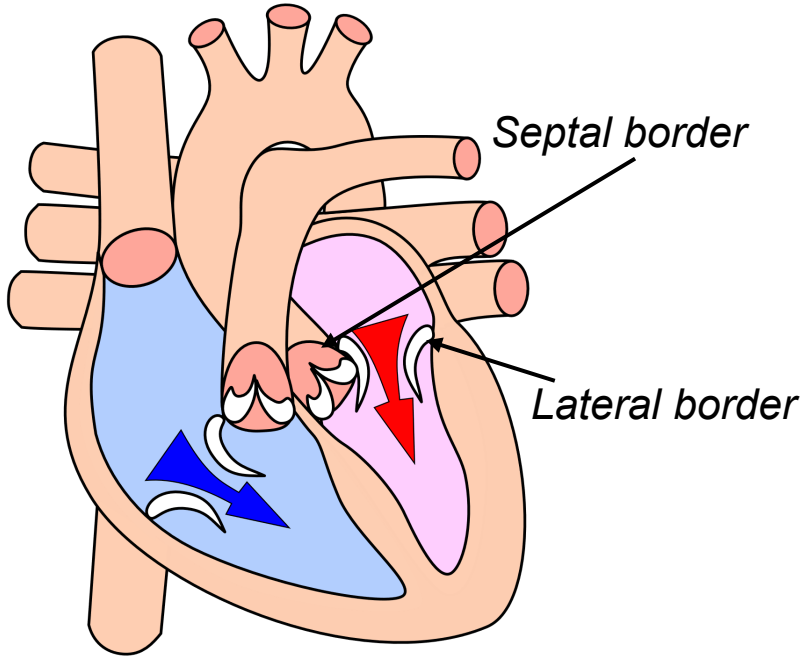
E/A ratio – Normal 0.8 – 2.0 *(decreases with age)*

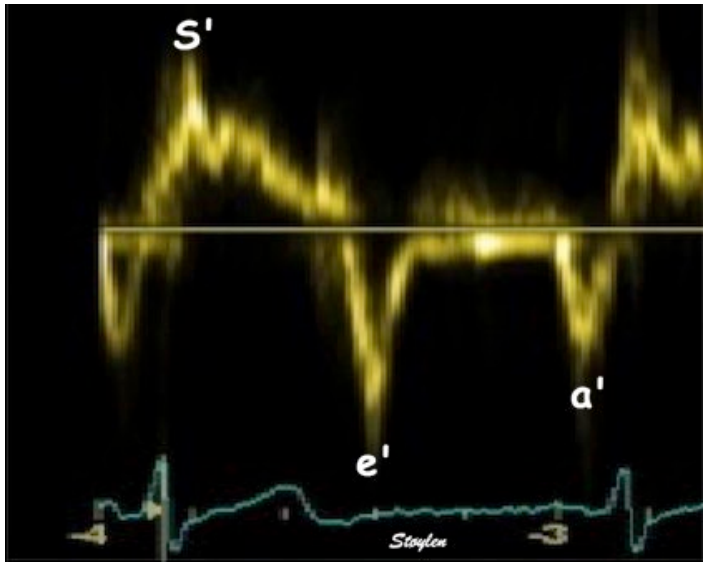
IVRT – Isovolumic relaxation time

DT – Deceleration time

Tissue Doppler Imaging (TDI)

- Doppler usually assesses flow of blood
- Tissue Doppler imaging (TDI) assesses motion of myocardium
- Assessment of diastolic function includes TDI of the septal and lateral borders of the mitral annulus
  - e' – early diastolic motion
  - a' – late diastolic motion
- e' – reflects relaxation of myocardium
- E/e' ratio – can be used to estimate left atrial pressure in those with diastolic dysfunction





Diagnosis of Diastolic Dysfunction with Normal EF

✓Average E/e' ≥ 14
✓Septal e' velocity < 7 cm/s or lateral e' velocity < 10 cm/s
✓Peak velocity of tricuspid regurgitation > 2.8 m/s
✓LA volume index > 34 ml/m^2

- If 0-1 of above criteria met: Normal diastolic function
- If 3-4 of above criteria met: Abnormal diastolic function
- If exactly 2 of above criteria met: Indeterminate diastolic function

Severity (Grade) of Diastolic Dysfunction

Diastolic Function	Normal	Grade 1 Impaired Relaxation	Grade 2 Pseudonormal	Grade 3 Restriction
LV relaxation	Normal	Impaired	Impaired	Impaired
LV compliance	Normal	Normal or ↓	Decreased	Decreased
LA pressure	Normal	Low or normal	Increased	Increased
Mitral inflow	E/A>0.8 but <2	E/A≤0.8 Peak E ≤50 cm/s	E/A>0.8 but <2	E/A≥2
IVRT	<70 msec	>90 msec	<90 msec	<70 msec
DT	>140 msec	>220 msec	<220 msec	<140 msec
Mitral inflow, Valsalva	ΔE/A <0.5	ΔE/A <0.5	ΔE/A ≥0.5	ΔE/A ≥0.5 (reversible) ΔE/A <0.5 (fixed)
Tissue Doppler	E/e'<10	E/e'<10	E/e' ≥10-14	E/e' ≥14
Pulmonary venous	S≥D	S>D	S<D	S<D
Peak TR velocity	<2.8	<2.8	>2.8	>2.8
LA volume index	Normal	Normal or ↑	Increased	Increased

Diastolic Dysfunction

- Prevalence
  - Mayo Clinic study, ages ≥ 45, no history of heart failure: up to 25%
- Signs
  - Echo Doppler abnormalities
  - Increased NTproBNP
- Symptoms
  - Often asymptomatic
  - Possibly reduced exercise capacity or DOE
  - In later stages, symptoms of heart failure
- Significance for underwriting: Risk of progression to heart failure

Redfield MM et al. Burden of systolic and diastolic ventricular dysfunction in the community – appreciating the scope of the heart failure epidemic. JAMA 2003;289:194-202.

Risk Assessment for Diastolic Dysfunction

- History of heart failure – high risk
- LVEF < 50% - rate for systolic dysfunction, no additional rating for DD
- Ratable cardiac impairment (valvular disease, cardiomyopathy, CAD, LVH) - rate for cardiac impairment, no additional rating for DD
- With LVEF ≥ 50%, no other cardiac impairment:

Grade of Diastolic Dysfunction	Rating
Grade 1: Impaired relaxation	Very low risk, preferred may be reasonable in some cases
Grade 2: Pseudonormal	Low risk
Grade 3: Restriction	Higher risk, refer to MD

Case Example

CONCLUSIONS

Normal left ventricular cavity size. Normal left ventricular wall thickness. Normal global left ventricular systolic function. EF estimated at 60-65%. Abnormal diastolic filling pattern for age. Normal pulmonary artery systolic pressure.
No significant valve abnormalities noted in current study.

1. Left Ventricle: Normal left ventricular cavity size. Normal left ventricular wall thickness. Normal global left ventricular systolic function. EF estimated at 60-65%. Abnormal diastolic filling pattern for age.
2. Right Ventricle: Normal right ventricular size. Normal right ventricular global systolic function.
3. Left Atrium: Normal left atrial size.
4. Right Atrium: Normal right atrial size. Right atrial pressure estimated at 3 mmHg.
5. Interatrial Septum: Intact interatrial septum.
6. Mitral Valve: Structurally/functionally normal mitral valve. No mitral regurgitation noted.
7. Aortic Valve: Structurally normal aortic valve. No aortic regurgitation noted.
8. Tricuspid Valve: Grossly normal appearing tricuspid valve. Trace tricuspid regurgitation noted. Normal pulmonary artery systolic pressure.
9. Pulmonic Valve: Pulmonic valve not well visualized. Trace pulmonary regurgitation.
10. Aorta: Aortic root is normal in size. Ascending aorta is normal in size.
11. Pericardium: No pericardial effusion.

MEASUREMENTS (Normal Values)

2D ECHO		
LV Diastolic Diameter PLAX	4.46 cm	4.2 - 5.9 / 3.9 - 5.3 cm
LV Systolic Diameter PLAX	2.43 cm	2.1-4.0 cm
LV Fractional Shortening PLAX	45.47 %	25-46 %
RV Internal Dim ED PLAX	2.11 cm	
IVS Diastolic Thickness	1.10 cm	
LVPW Diastolic Thickness	1.06 cm	
LA Systolic Diameter LX	3.19 cm	3.0-4.0 / 2.7-3.8 cm
LVOT Diameter	1.73 cm	
LVOT Area	2.34 cm2	
Aorta at Sinuses Diameter	2.98 cm	
LV Systolic Volume 2D Cubed	14.41 cm3	
Ascending Aorta Diameter	3.35 cm	
DOPPLER		
AV Velocity Time Integral	27.66 cm	
AV Peak Velocity	135.06 cm/s	
AV Peak Gradient	7.30 mmHg	
AV Mean Velocity	94.40 cm/s	
AV Mean Gradient	3.93 mmHg	
LVOT Velocity Time Integral	23.48 cm	
LVOT Peak Velocity	106.79 cm/s	
LVOT Peak Gradient	4.56 mmHg	
LVOT Mean Velocity	78.11 cm/s	
LVOT Mean Gradient	2.67 mmHg	
AV Area Cont Eq vti	1.99 cm2	
AV Area Cont Eq pk	1.85 cm2	
Mitral E Point Velocity	81.11 cm/s	
Mitral A Point Velocity	99.91 cm/s	
Mitral E to A Ratio	0.81	
MV Deceleration Time	220.03 ms	
TR Peak Velocity	208.27 cm/s	
TR Peak Gradient	17.35 mmHg	
PV Peak Velocity	88.88 cm/s	
PV Peak Gradient	3.16 mmHg	

Case Example

CONCLUSIONS

Normal left ventricular cavity size. Normal left ventricular wall thickness. Normal global left ventricular systolic function. EF estimated at 60-65%. Abnormal diastolic filling pattern for age. Normal pulmonary artery systolic pressure.

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9. Pulmoni
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11. Pericar

- *E/A 0.81*
- *Peak velocity of TR: 208 cm/s = 2.08 m/s*
- *LA size by measured diameter is normal*
- *No tissue Doppler (e' or a') values*

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Diagnosis of Diastolic Dysfunction with Normal EF

Case Details

- *E/A 0.81*
- *Peak velocity of TR: 208 cm/s = 2.08 m/s*
- *LA size by measured diameter is normal*
- *No tissue Doppler (e' or a') values*

Diagnostic Criteria for Diastolic Dysfunction

✓Average E/e' ≥ 14
✓Septal e' velocity < 7 cm/s or lateral e' velocity < 10 cm/s
✓Peak velocity of tricuspid regurgitation > 2.8 m/s
✓LA volume index > 34 ml/m^2

If 0-1 of above criteria met: Normal diastolic function
If 3-4 of above criteria met: Abnormal diastolic function
If exactly 2 of above criteria met: Indeterminate diastolic function

Summary
0 criteria met
2 unknown
At worst, indeterminate

Ratings for Diastolic Dysfunction

Grade of Diastolic Dysfunction	Rating
Grade 1: Impaired relaxation	Very low risk, preferred may be reasonable in some cases
Grade 2: Pseudonormal	Low risk
Grade 3: Restriction	Higher risk, Refer to MD

What should we do with this case?

Ratings for Diastolic Dysfunction

Grade of Diastolic Dysfunction	Rating
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Indeterminate Diastolic Function: *Very low risk or low risk most likely*