(PBL) Cardiomyopathies

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Learning Objectives

1. Practice applying new knowledge and skills gained from Cardiomyopathies sessions, through collaborative learning with peers and expert faculty.

2. Identify strategies that foster optimal management of cardiomyopathies, within the context of professional practice.

3. Formulate an action plan to implement practice changes, aimed at improving patient care.

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Associated Session

- Cardiomyopathies

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Case One:

A 24 year old Caucasian male medical student is presenting to your clinic for ‘cardiac eval.’

He volunteered to be the ‘patient’ in his small group as they learned about EKGs and electrophysiology.

The attending looked his EKG and sent him to you for evaluation and work-up.

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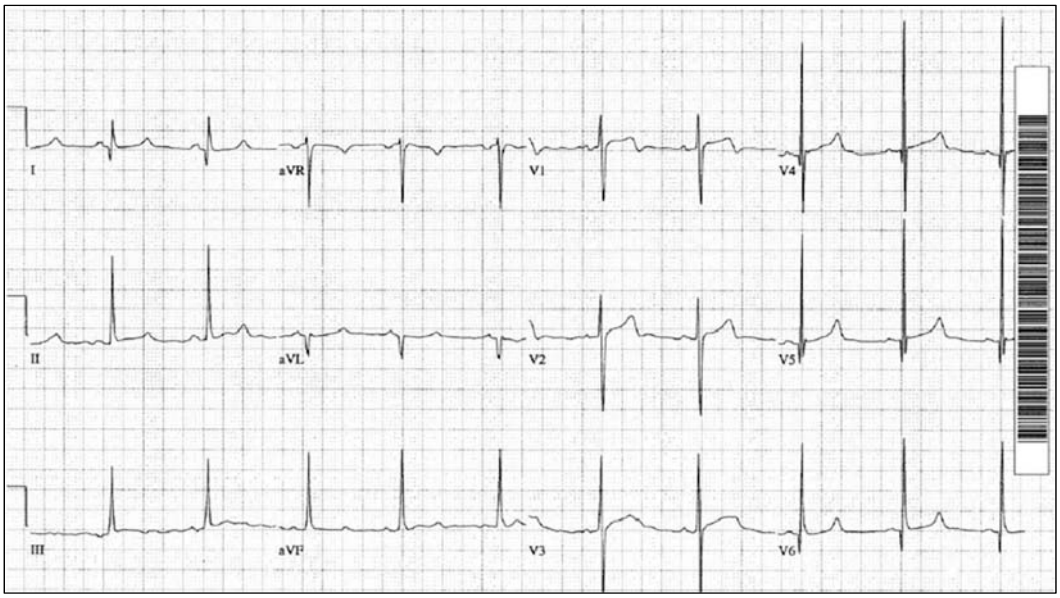
Here is his EKG*:*



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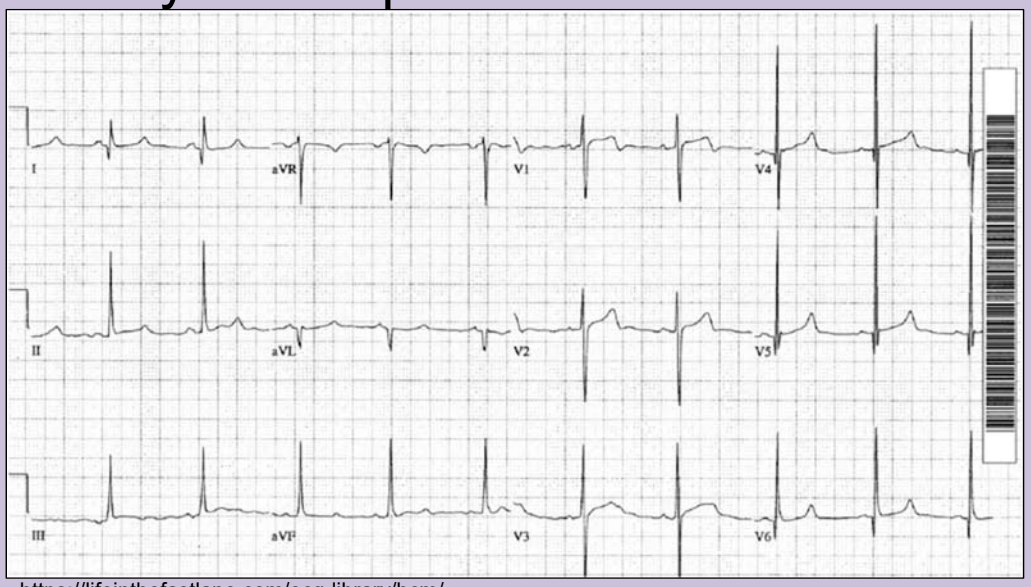
How would you interpret this EKG?



https://lifeinthefastlane.com/ecg-library/hcm/

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Case One:

You repeat an EKG with similar findings.

Past Medical History

- Mild asthma, well-controlled

Past Surgical History

- Wisdom Teeth removal, 2014
- Appendectomy, 2001

Medications

- Albuterol 2 puffs as needed
- Acetaminophen and ibuprofen prn

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Case One:

Social History:

- 2nd year medical student
- Rare alcohol on the weekends
- No tobacco use
- Lives with 4 other medical students in a house close to campus

What specific questions would you ask him?

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Case One:

He is an avid long-distance runner.

Denies syncope or chest pain.

Does note some recent lightheadedness and dyspnea with running, but attributes it to the hot summer weather and deconditioning from final exam season.

No family members with cardiac disease or sudden death.

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Case One:

Exam:

- HR 60, RR 14, BP 110/54, 100% on RA, 37° C
- Athletic, fit-appearing male in no acute distress
- Clear lungs
- Normal S1 and S2, no murmurs, rubs, or gallops
- Slightly displaced PMI

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Case One:

You have a high-performance athlete with a slightly displaced PMI and an EKG demonstrating LVH.

Physiologic left ventricular hypertrophy (athlete's heart) or pathologic left ventricular hypertrophy (HCM)?

- How can you differentiate between the two?
- What further testing would you like to order?

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Physiologic vs. Pathologic Left Ventricular Hypertrophy

History & Physical	EKG and GXT	Imaging
• Training regimen unlikely to cause adaptive changes • FH of sudden death or HCM • + Symptoms • Resting/provocative harsh systolic murmur	• Seattle Criteria findings on EKG • Symptoms reproduced on GXT • VO_2 max <50ml/kg/min	• LVH >16mm • **Disparity between LVH and LV cavity size (LVC <45mm)** • Evidence of diastolic dysfunction • Late gadolinium enhancement on MR

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Patient's Echocardiogram

- Asymmetric septal hypertrophy
- LVH 18mm
- Normal valves
- Diastolic dysfunction noted with E/e' >15
- No LVOT obstruction noted, even with provocative maneuvers

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Case One:

Patient returns to clinic after his echocardiogram to discuss the results.

Given his EKG findings, thickened septal wall, presence of diastolic dysfunction, and absence of secondary causes of LVH, you diagnosis him with hypertrophic cardiomyopathy.

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Case One:

The patient remembers that HCM can be genetic.

Should he undergo genetic screening?

What is the likelihood of identifying a pathogenic mutation?

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Case One:

Yes. Genetic testing is reasonable in the index patient to facilitate identification of family members at risk (LOE B/Class IIa).

Pathogenic mutations are identified in 60-70% of patients *with a positive family history*, but only in 10-30% of patients without a family history

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Case One:

His genetic panel returns with a mutation in the beta myosin heavy chain that is known to cause HCM.

He asks about screening his family members. His mom and dad are in their 50s and his sister is 18 years old.

- Who should be screened?
- How should they be screened?

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Case One:

- Screening (clinical, with or without genetic testing) is recommended in ALL first-degree relatives (Class I/LOE B)
- Since the patient has an indentified mutation, genetic screening of first degree relatives is reasonable AFTER discussion with a genetic counselor (Class I/LOE B)

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Case One:

His family members are all genotype-negative for the mutation, so ongoing screening is not indicated. (Class III/LOE B)

How would the screening have changed if the patient did NOT have an identified genetic mutation?

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Family Screening

- Genetic testing is NOT indicated in relatives when the index patient does not have a definitive pathogenic mutation (Class III/LOE B)
- Surveillance EKG and echocardiogram every 5 years starting with puberty and continuing through the 7th decade of life.
- EKG and echo every 12-18 months if participating in competitive sports

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Case One:

After extensive counseling, the patient has stopped running, and no longer participates in competitive sports. He has transitioned to playing golf after review of the 2011 AHA guidelines.

He is currently asymptomatic, but he is worried about sudden cardiac death (SCD) and wants to prevent it if at all possible.

- What further history and testing to stratify his SCD risk should be completed at this point?

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Case One: Prevention of SCD

- All patients should undergo a comprehensive SCD risk stratification at initial evaluation to determine presence of (Class I/LOE B):
  - Personal history of vfib, sustained VT, or SCD events
  - Family history of SCD
  - Unexplained syncope
  - Documented NSVT (3+ beats at rate >120bpm) on Holter
  - LV wall thickness ≥30mm
- It is reasonable to assess blood pressure response during exercise as part of SCD risk stratification in patients with HCM (Class IIa/LOE B)

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Case One: Prevention of SCD

- 24-hour ambulatory EKG monitor is recommended as part of the initial evaluation of patients with HCM to detect ventricular tachycardia (VT) and identify patients who may be candidates for ICD therapy. (Class I/LOE B)
- Consider repeating every 1-2 years (Class IIa/LOE C) or with development of symptoms (Class I/LOE B)

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Case One:

His 24-hour monitoring demonstrates normal sinus rhythm, sinus tachycardia, and isolated bursts of NSVT.

His blood pressure has a normal response to exercise.

- Should you refer him for AICD placement?
- Why or why not?

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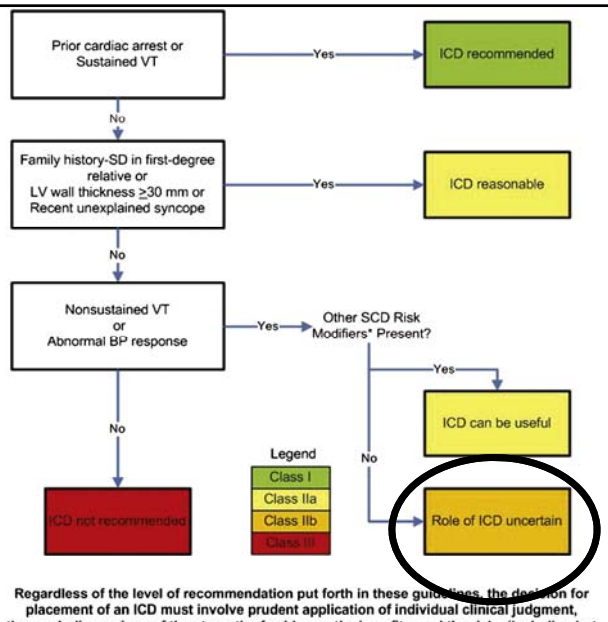
Case One: Selection of patients for ICDs



The Journal of Thoracic and Cardiovascular Surgery 2011 142, e153-e203DOI: (10.1016/j.jtcvs.2011.10.020)

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Case One:

You haven't seen the patient in 3-4 years when he returns to your clinic. He is now a 3rd year Family Medicine resident (inspired by your excellent care).

He has recently noticed new dyspnea and intermittent angina, prompting return for evaluation. He denies syncope or pre-syncope.

You repeat an echo that demonstrates interval worsening of septal hypertrophy and now has signs of LVOT obstruction with provocative maneuvers.

- What medications could you consider starting for symptomatic improvement?
- What medications should you avoid at all costs?

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Case One: Symptomatic patients

Goal of medication therapy is symptom reduction. There is currently NO role for medical therapy in asymptomatic patients.

- Beta-blockers are first line therapy for treatment of symptoms, titrated to symptom relief or a heart rate of less than 60-65 bpm. (Class I/LOE B)
- Verapamil is recommended for patients with an incomplete response to beta-blockers or who have side effects limiting beta blocker usage (Class I/LOE B)
- Consider combining beta-blocker or verapamil with disopyramide in patients who do not respond to single agent therapy.

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Case One: Symptomatic patients

Mediations listed at CLASS III: HARMFUL

- Nifedipine or other dihydropyridine calcium channel-blocking drugs (LOE C)
- Verapamil in a patient with obstructive physiology and hypotension or dyspnea at rest. (LOE C)
- Digitalis usage to treat dyspnea in the absence of atrial fibrillation (LOE B)
- Use of disopyramide alone without beta-blockers or verapamil may increase AV conduction and increase ventricular rate in patients with underlying atrial fibrillation (LOE B)

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Case One:

You start the patient on an oral beta-blocker with complete symptom relief.

- When would you refer for evaluation for surgical management (either septal myectomy or alcohol septal ablation)?

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Case One: Invasive Therapy

- Severe dyspnea or chest pain (NYHA Class III or IV) or occasionally other exertional symptoms (syncope or near-syncope) that interfere with everyday activity despite *optimal medical therapy.*

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Case Two:

A 26 year old Italian soccer player is rushed into your Emergency Room. He collapsed on the field and was found to be pulseless. After 3 rounds of CPR, a shock was administered with return of circulation.

The medical team noted ventricular tachycardia on the monitor prior to shock.

He is sitting up, conversational, complaining of chest pain, but otherwise asymptomatic.

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Case Two:

What are the common causes of sudden cardiac death in young athletes?

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Case Two: Causes of SCD in Athletes

Structural	Electrical	Acquired
• HCM • Arrhythmogenic right ventricular cardiomyopathy • Marfan's • Congenital coronary abnormalities • Valvular disease	• WPW • Congenital long QT syndrome • Brugada sydrome • Catecholaminergic polymorphic ventricular tachycardia	• Infection (myocarditis) • Trauma • Toxicity (illicit/performance enhancing drugs) • Environment (hyper/hypothermia)

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Case Two:

Is pre-participation screening of athletes recommended?

And, if so, what is the protocol?

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Case Two:

YES!

Pre-participation cardiovascular screening is recommended by the American Heart Association AND the European Society of Cardiology.

In the United States, the recommendation is for health care professionals to use the AHA 14-element screening checklist for athletes ages 12-25 years old.

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Case Two: 14 Element Screening

Personal History:

- Chest pain with exertion, unexplained pre-syncope or syncope, excessive exertional dyspnea or palpitations, history of heart murmur, elevated SBP, prior restriction from sports by a physician, prior testing of the heart

Family History:

-Premature death before the age of 50 due to heart disease, close relative with disability due to heart disease, ANY known cardiac conditions in family members (ie HCM, Marfans, Long-QT)

Physical Examination:

- Concerning heart murmur, femoral pulses, stigmata of Marfan's, blood pressure abnormalities

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Case Two:

An EKG is not part of the 14 Element Screening described on the prior slide…

- Should you get an EKG on young athletes as part of the pre-participation screening?
- What would you be looking for?

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To get or not to get an EKG…

- Evaluate and diagnosis electrical abnormalities like Wolff-Parkinson-White and congenital long-QT.
- Will be abnormal in >90% of patients with underlying hypertrophic cardiomyopathy
- Will be abnormal in >75% of patients with arrhythmogenic right ventricular cardiomyopathy

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To get or not to get an EKG…

Incorporating an EKG into screening protocol improves efficacy in identifying conditions that may cause SCD.

The American Heart Association currently does NOT support the routine use of EKG as part of the pre-participation screening.

- High false-positive rates
- Cost-effectiveness
- Psychological implications for athletes and families

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If you do get an EKG…

Normal EKG findings in athletes
Sinus bradycardia >30 bpm
Sinus arrhythmia
Ectopic atrial rhythm
Junctional escape rhythm
1st degree AV block
Mobitz I 2nd degree AV block
Incomplete RBBB
Isolated voltage criteria for LVH
Early repolarization

Abnormal EKG findings in athletes
T-wave inversion
ST depression (≥0.5mm in 2+ leads)
Sinus tachycardia
Pathologic Q-waves
LBBB
Left atrial enlargement or RVH
Ventricular pre-excitation
Sinus bradycardia <30 bpm
PVCs
Brugada-like EKG pattern

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Case Three:

You are called to evaluate 22 year old female in the ER. She is a college student home for the summer.

She is complaining of 2 weeks of fatigue, worsening dyspnea, decreased exercise tolerance, and intermittent chest tightness.

She is hypoxic on room air and has not responded to continuous nebulizer treatments. Her CXR is pending.

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Case Three :

History of Present Illness:

- Family just moved to North Carolina from Virginia about 4 weeks ago. No one else is sick
- Has been to urgent care and ER twice with similar sx and was started on allergy medications and albuterol with no improvement.
- + Slight cough (?), +chills, subjective fevers
- No rash, weight loss.

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Case Three :

Past Medical History

- None

Medications

- Albuterol 2 puffs as needed
- Loratadine 10 mg daily for allergies

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Case Three :

Social History:

- College student, varsity soccer player
- Denies tobacco, alcohol, or recreational drug use

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Case Three :

Exam:

- HR 110, RR 27, BP 100/62, 88% on 4L NC, 37^{o} C
- Fatigued, ill-appearing, mild distress
- Crackles in bilateral lung fields
- 2+ peripheral edema bilaterally to the knees

Any other exam maneuvers or bedside tests?

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Case Three :

Bedside Echo

Cardiac

- EF approximately 20-25%
- Severe dilation of left ventricle with global reduction in systolic function

Lung

- Lung slide present, diffuse B-lines bilaterally

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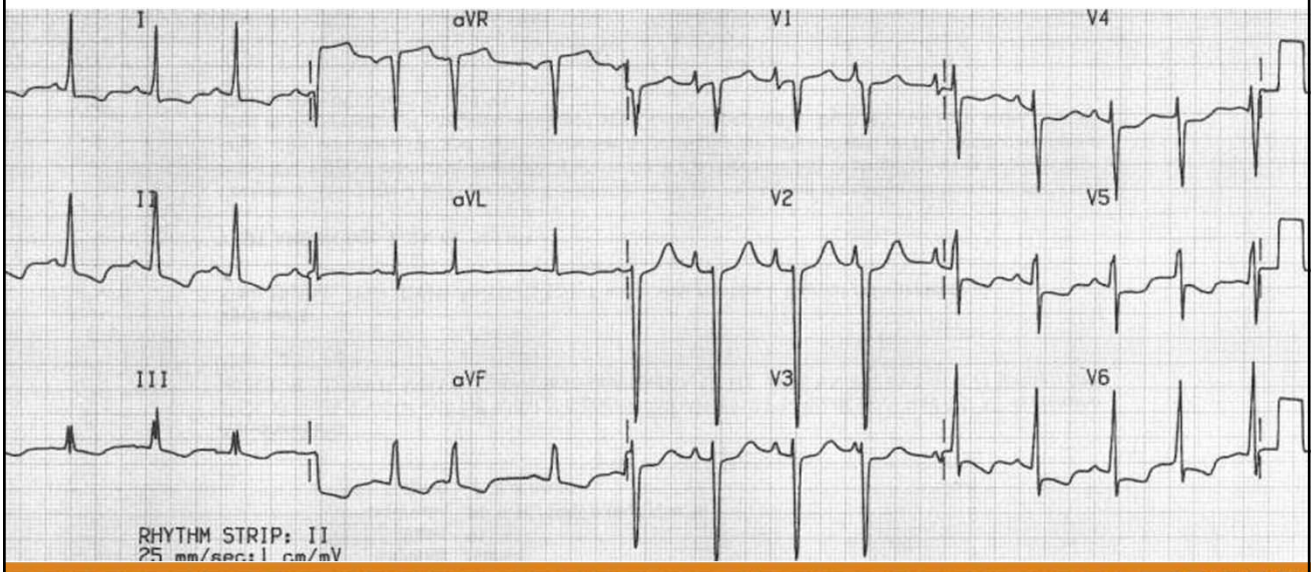
Rhythm Stripe



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Case Three :

Labs

- WBC WNL, but lymphocytic predominance
- BMP with elevated Cr of 1.8 (baseline 0.9)
- TnT 0.358
- BNP 800

Chest X-Ray

- Enlarged cardiac silhouette, evidence of cardiopulmonary volume overload

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Case Three :

- What is the differential diagnosis for this patient?

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Differential Diagnosis:

- Infectious myocarditis
- Ischemic heart disease
- Valvular heart disease
- Right ventricular cardiomyopathy
- Post-partum cardiomyopathy
- Drug-induced cardiomyopathy
- Thyroid disorder
- Giant cell myocarditis
- Pericarditis
- Endocarditis

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Myocarditis

→*Inflammatory disease of the myocardium*

Causes:

-Vary significantly by region, but most commonly due to infectious etiology

North America:

Parvo B19, HHV-6, Coxsackie B, Adeno, other enteroviruses, Lyme disease, Flu A/B, Histo, Coccidio, Syphilis

Worldwide:

HIV, Chagas disease, post-streptococcal rheumatic heart disease, diphtheria, typhoid, rubella, Leishmaniasis, Cholera, schistosomiasis

Bozkurt B, Colvin M, Cook J, et al. 2016 AHA scientific statement for the current and diagnostic treatment strategies for specific dilated cardiomyopathies Circ 2016; 134e579-e464

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Myocarditis

→*Inflammatory disease of the myocardium*

Clinical Features:

Chest pain, tachycardia, respiratory distress/tachypnea, new S3/S4, abnormal echo, abnormal EKG, new cardiomegaly, new partial or complete heart block, new heart failure, acute pericarditis, cardiogenic shock, cardiac arrest

*Technically…*requires histological or immunohistological confirmation by EMB, surgical heart specimens, or an autopsy

Bozkurt B, Colvin M, Cook J, et al. 2016 AHA scientific statement for the current and diagnostic treatment strategies for specific dilated cardiomyopathies Circ 2016; 134e579-e464

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Diagnostic Criteria for Clinically Suspected Myocarditis

Clinical Presentation	Diagnostic criteria
• Acute chest pain, pericarditic, or pseudoischemic • Worsening dyspnea, fatigue, or right- or left-sided HF • Palpitations and/or unexplained arrhythmia or syncope • Aborted sudden cardiac death • Unexplained cardiogenic shock	1. EKG/Holter/Stress test features 2. Myocardiocytolysis markers (elevated TnT) 3. Function and structural abnormalities (echo/angiography/CMR) 4. Tissue characterization by cardiac MR

Bozkurt B, Colvin M, Cook J, et al. 2016 AHA scientific statement for the current and diagnostic treatment strategies for specific dilated cardiomyopathies Circ 2016; 134e579-e464

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Case Three :

- Does the patient have myocarditis?
- What other studies would you need to support or confirm your diagnosis?

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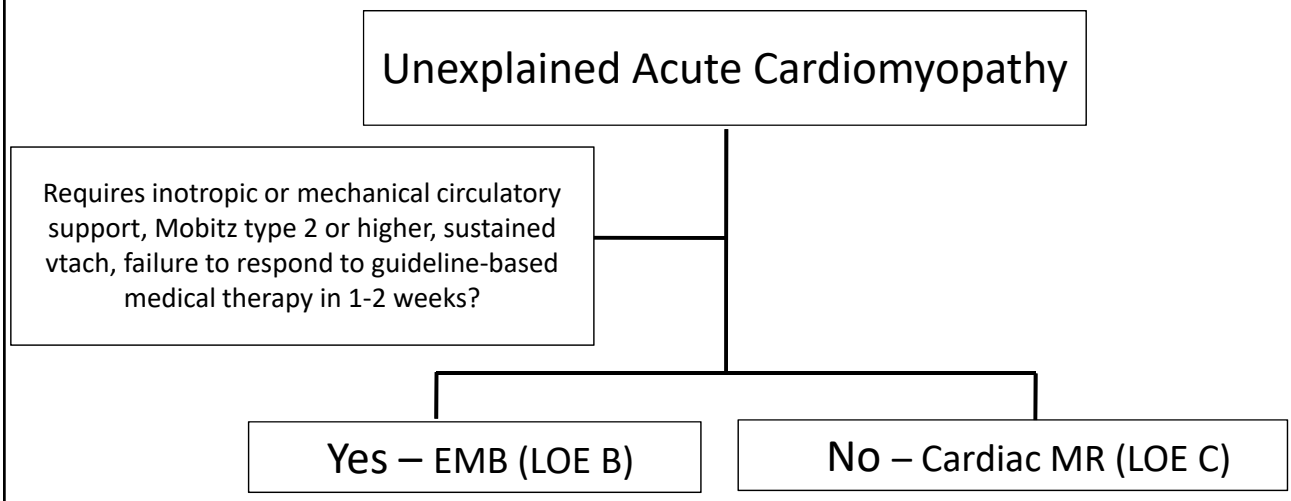
Who gets an endomyocardial biopsy (EMB)?



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Case Three :

- She is admitted to the hospital. What would your initial management include?

Treatment of Myocarditis

→*Myocarditis that presents at DCM should be treated per current guidelines for systolic heart failure*

- Immunosuppression is not indicated
- Role for IVIG (?)
- Supportive management of arrhythmias
- Competitive sports should be avoided for 3-6 months
- Reassessment with clinical evaluation AND functional testing is indicated before returning to competition
- NSAIDS should be avoided

Bozkurt B, Colvin M, Cook J, et al. 2016 AHA scientific statement for the current and diagnostic treatment strategies for specific dilated cardiomyopathies Circ 2016; 134e579-e464

At discharge:

- Formal echocardiogram confirmed severely depressed EF with dilated ventricles
- Patient significantly improved with diuresis
- Started on ACE-I and beta blocker

She is asking about her follow-up, prognosis, and when she can return to playing soccer.

What do you tell her?

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Prognosis and Follow-Up

- Most patients with acute myocarditis have partial or full recovery.
- Consider outpatient MRI
- Initially, echocardiograms every 1-3 months
- No competitive sports for 3-6 months, then gradual return after functional testing

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Questions

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Contact Information

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