Cardiomyopathies: PBL

Craig Barstow, MD, FAAFP

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Craig Barstow, MD, FAAFP

Program Director, Hospitalist Fellowship at Womack Army Medical Center, Fort Bragg, North Carolina; Assistant Professor of Family Medicine at Uniformed Services University of Health Sciences, Bethesda, Maryland; Director of Ultrasound Education, Family Medicine Residency Program, Womack Army Medical Center; Physician, Scotland Memorial Hospital Emergency Department, Laurinburg, North Carolina.

Dr. Barstow is a graduate of the Uniformed Services University of the Health Sciences – F. Edward Herbert School of Medicine in Bethesda, Maryland. He completed undergraduate studies at the U.S. Military Academy. Dr. Barstow joined the Womack Army Medical Center Family Medicine Residency Program in 2012, and created the fellowship program, accepting the first fellow in July 2015. His areas of interest include inpatient family medicine, newborn care, and point-of-care ultrasound teaching.

Learning Objectives

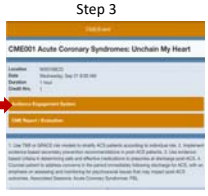
1. Practice applying new knowledge and competencies gained from cardiomyopathies sessions, and receive feedback from expert faculty.
2. Interact collaboratively with peers to solve complex and challenging case-study scenarios.
3. Develop problem-solving skills that promote effective reasoning to manage cardiomyopathy within the context of professional practice.

Audience Engagement System

Step 1

Step 2

Step 3



What is Cardiomyopathy?

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Cardiomyopathy

- Disease of the myocardium
- Mechanical or electrical dysfunction
- Usually with ventricular hypertrophy or dilation
- Frequently genetic
- Primarily cardiac or part of systemic disorder
- Often leads to cardiac death or heart failure

Maron BJ, Towbin JA, Thiene G, et al. Contemporary definitions and classification of the cardiomyopathies: An American Heart Association scientific statement from the council on clinical cardiology, heart failure and transplantation committee; quality of care and outcomes research and functional genomics and translational biology interdisciplinary working groups; and council on epidemiology and prevention. Circulation. 2006;113:1807-1816.

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Case

Nick is a 17-year-old male who is brought to your office by his parents. They are concerned because he passed out during soccer practice.

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What are you concerned about?

FMX

What type of questions do you want to ask?

FMX

PPE History

- Have you ever passed out or nearly passed out DURING or AFTER exercise?
- Have you ever had discomfort, pain, tightness, or pressure in your chest during exercise?
- Does your heart ever race or skip beats (irregular beats) during exercise?
- Has a doctor ever ordered a test for your heart? (For example, ECG, echocardiogram)
- Do you get lightheaded or feel short of breath more than expected during exercise?
- Have you ever had an unexplained seizure?
- Do you get tired or short of breath more quickly than your friends during exercise?

Preparticipation Physical Evaluation History Fort. AAFP, AAP, ACSM, AMSSM, AOSSM, AOASM. 2010

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PPE Family History

- Has any family member or relative died of heart problems or had an unexpected or unexplained sudden cardiac death before age 50 (including drowning, unexplained car accident, or sudden infant death syndrome)?
- Does anyone in your family have hypertrophic cardiomyopathy, Marfan syndrome, arrhythmogenic right ventricular cardiomyopathy, long QT syndrome, Brugada syndrome, or catecholaminergic polymorphic ventricular tachycardia?
- Does anyone in your family have a heart problem, pacemaker, or implanted defibrillator?
- Has anyone in your family had unexplained fainting, unexplained seizures, or near drowning?

Preparticipation Physical Evaluation History Fort. AAFP, AAP, ACSM, AMSSM, AOSSM, AOASM. 2010

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Case

He was running drills when he suddenly lost consciousness. On further questioning, he admits to having mild chest discomfort while exercising, but has attributed this to the difficult workouts during soccer practice. He has an uncle who died unexpectedly at a young age.

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What do you look for on the physical exam?

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Case

Physical examination reveals a soft systolic heart murmur. The remainder of the exam is unremarkable.

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What do you want to do next?

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What do you want to do next?

- Consult pediatric cardiology?
- Order an ECG?
- Order an echocardiogram?

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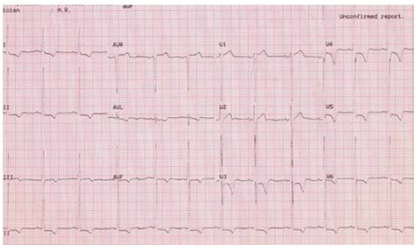

ECG courtesy of Jonathan Drezner MD (University of Washington)

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So is this ECG normal?

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Normal ECG Findings in Athletes

- *Common training-related ECG alterations*
- *Physiological adaptations to regular exercise*
- *Considered normal variants in athletes*
- *Do not require further evaluation*

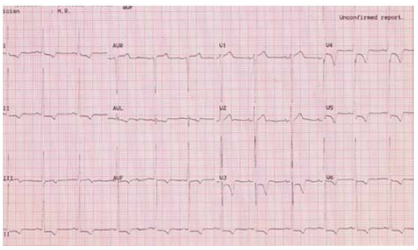
1) Sinus bradycardia (≥ 30 bpm)
2) Sinus arrhythmia
3) Ectopic atrial rhythm
4) Junctional escape rhythm
5) 1° AV block (PR interval > 200 ms)
6) Mobitz Type I (Wenckebach) 2° AV block
7) Incomplete RBBB
8) QRS voltage criteria for left ventricular hypertrophy (Except: QRS voltage criteria for LVH AND any non-voltage criteria for LVH such as left atrial enlargement, left axis deviation, ST segment depression, T-wave inversion, or pathologic Q waves)
9) Early repolarization (ST elevation, J-point elevation, J-waves, or terminal QRS slurring)
10) Convex ("domed") ST segment elevation combined with T-wave inversion in leads V1-V4 in black/African athletes

Slide provided by Jonathan Drezner MD (University of Washington). Used with permission

ECG Changes in Cardiomyopathy

T wave inversion	>1 mm 2 or more leads; V2-V6, II and aVF or I and AVL
ST segment depression	> 0.5 mm in two or more leads
Pathological Q waves	> 3 mm depth or > 40ms duration two or more leads
Complete left bundle branch block	QRS > 120 ms, negative QRS complex in V1 and upright monophasic R 1 and V6
Intraventricular conduction delay	QRS > 140 ms
Left axis deviation	-30° - -90°
Left atrial enlargement	Prolonged P wave duration of > 120 ms in leads I or II with negative portion of the P wave >= 1 mm in depth and >= 40 ms in duration in lead V1
Right ventricular hypertrophy pattern	R-V_1+S-V_5 > 10.5 mm AND right axis deviation > 120°
Premature ventricular contractions	> 2 PVCs per 10 sec tracing
Ventricular arrhythmias	Couplets, triplets and non-sustained VT

Drezner JA, Ashley E, Baggish AL. Abnormal electrocardiographic findings in athletes: recognizing changes suggestive of cardiomyopathy. Br J Sports Med. 2013;47:137-152.



ECG courtesy of Jonathan Drezner MD (University of Washington)

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What do you want to do next?

- Consult pediatric cardiology?
- Order an echocardiogram?
- Both?

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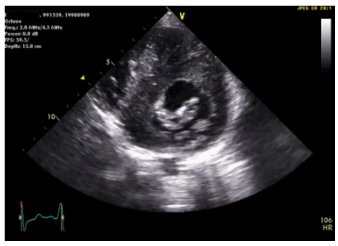

Echocardiogram courtesy of Ryan Flanagan, MD FAAP FACC (Womack Army Medical Center)

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What's your diagnosis?

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Hypertrophic Cardiomyopathy

- Incidence of 1 in 500 adults
- Hypertrophy of the ventricular septum
- Autosomal dominant inheritance (half are denovo mutations)
- Leading cause of sudden death among young athletes
- Mortality is less than 1% per year

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Hypertrophic Cardiomyopathy

- Systolic ejection murmur
- Becomes louder with reduced preload, softer with increased preload (opposite other murmurs)
- ECG may show signs of ventricular hypertrophy
- Diagnosis made with echocardiogram

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So what's next for Nick?

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Hypertrophic Cardiomyopathy

- Death is from ventricular arrhythmias
- Treatment is with beta blockers
- Other treatments
  - Myomectomy
  - Septal ablation
  - Implantable defibrillator

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What else is on your differential?

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Cardiomyopathies

Primary Cardiomyopathies			Secondary Cardiomyopathies		
Genetic	**Mixed**	**Acquired**	**Infiltrative/ Storage**	**Endocrine**	**Other**
Hypertrophic cardiomyopathy	Dilated cardiomyopathy	Myocarditis	Amyloidosis	Diabetes mellitus	Sarcoidosis
Arrhythmogenic right ventricular dysplasia	Restrictive	Takotsubo	Gaucher disease	Hyperthyroidism	Neuromuscular
LV Noncompaction		Peripartum	Hurler's disease	Hypothyroidism	Neurological
Glycogen storage diseases		Tachycardia induced	Hunter's disease	Hyperparathyroidism	Nutritional deficiencies
Conduction defects			Hemochromatosis	Pheochromocytoma	Dermatomyositis
Mitochondrial myopathies			Fabry's disease	Acromegaly	Scleroderma
Ion channel disorders			Glycogen storage disease		Electrolyte imbalance
			Niemann-Pick disease		Cancer therapy

Maron BJ, Towbin JA, Thiene G, et al. Contemporary definitions and classification of the cardiomyopathies: An American Heart Association scientific statement from the council on clinical cardiology, heart failure and transplantation committee; quality of care and outcomes research and functional genomics and translational biology interdisciplinary working groups; and council on epidemiology and prevention. Circulation. 2006;113:1807-1816.

Cardiomyopathies
Genetic

- Hypertrophic cardiomyopathy
- Arrhythmogenic right ventricular dysplasia
- LV Noncompaction
- Glycogen storage disease
- Mitochondrial myopathies
- Ion channel disorders

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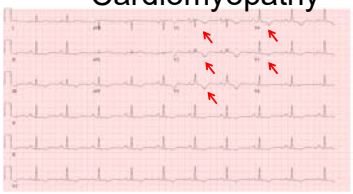
Arrhythmogenic Right Ventricular Cardiomyopathy



Inverted T-waves in leads V1-V5.

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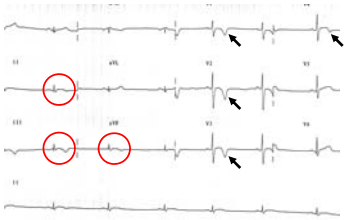
ARVC



Delayed S-wave upstroke in V1, low voltages in limb leads < 5 mm (circles), and inverted T-waves in anterior precordial leads (V1-V4; arrows) and inferior leads (III and aVF).

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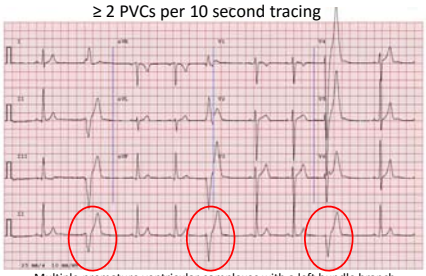
Premature Ventricular Contractions
≥ 2 PVCs per 10 second tracing



Multiple premature ventricular complexes with a left bundle branch pattern and superior axis (negative QRS vector in inferior leads).

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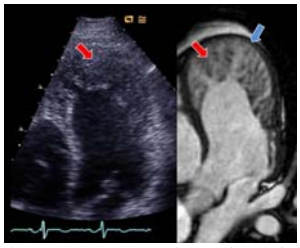
Left Ventricular Non-Compaction



Echocardiographic (left panel) and cardiac magnetic resonance (right panel) images of isolated left ventricular non-compaction. Note the prominent trabeculations in the left ventricular apex (red arrow), with a thin layer of compact myocardium (blue arrow) and prominent inter-trabecular recesses.

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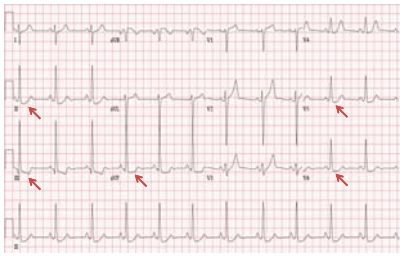

ST segment depression in the inferolateral leads (II, III, aVF, V5-V6).

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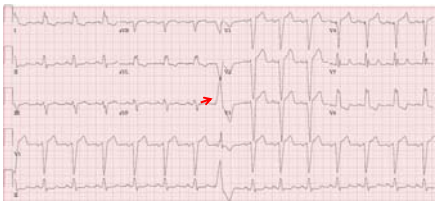
Left Bundle Branch Block – Abnormal



ECG from a patient with left ventricular non-compaction, demonstrating complete left bundle branch block (QRS ≥ 120 ms with predominantly negative QRS complex in lead V1). An isolated premature ventricular complex is also present (arrow). LBBB warrants further testing to evaluate for cardiomyopathy.

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Channelopathies

- Long QT syndrome
- Brugada syndrome
- Catecholaminergic polymorphic ventricular tachycardia
- Short QT syndrome
- Idiopathic ventricular fibrillation

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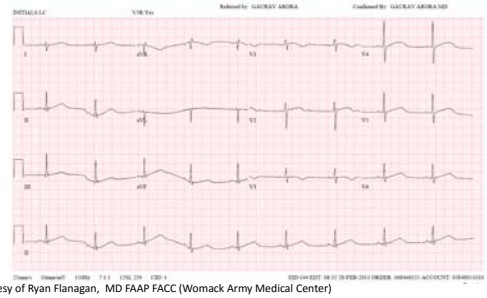
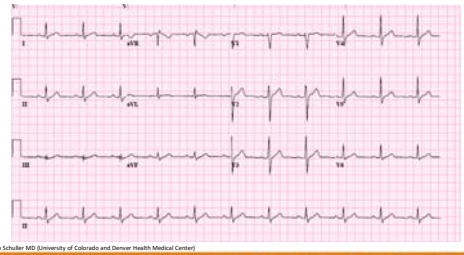
17-year-old who passed out at a haunted house



ECG courtesy of Ryan Flanagan, MD FAAP FACC (Womack Army Medical Center)

Male with syncope



ECG courtesy of Joseph Schuller MD (University of Colorado and Denver Health Medical Center)

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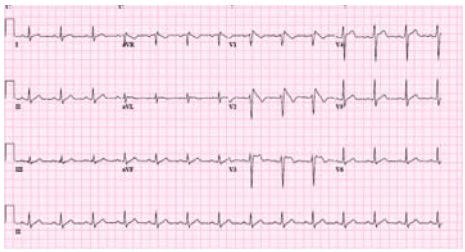
Same patient with a fever



ECG courtesy of Joseph Schuller MD (University of Colorado and Denver Health Medical Center)

Questions?

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ICD-10 Codes

I42.– Cardiomyopathy
- I42.0 Dilated cardiomyopathy
- I42.1 Obstructive hypertrophic cardiomyopathy
- I42.2 Other hypertrophic cardiomyopathy
- I42.6 Alcoholic cardiomyopathy
- I42.7 Cardiomyopathy due to drug agent and external agent
- I42.9 Cardiomyopathy, unspecified

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Contact Information

Craig Barstow, MD FAAFP
Email: craig.h.barstow.mil@mail.mil

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Associated Sessions

- Cardiomyopathies
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