

The Heart of the Matter

Is the Heart the Matter?

--Chest pain in the Pediatric Patient--

19th Interregional Symposium

November 2, 2018

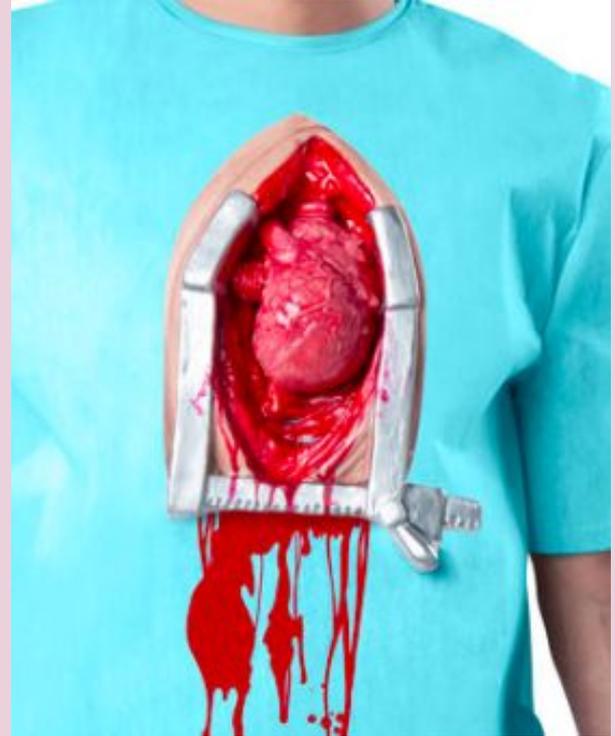
Session A, 9:35-10:50 am

John-Charles Loo, MD

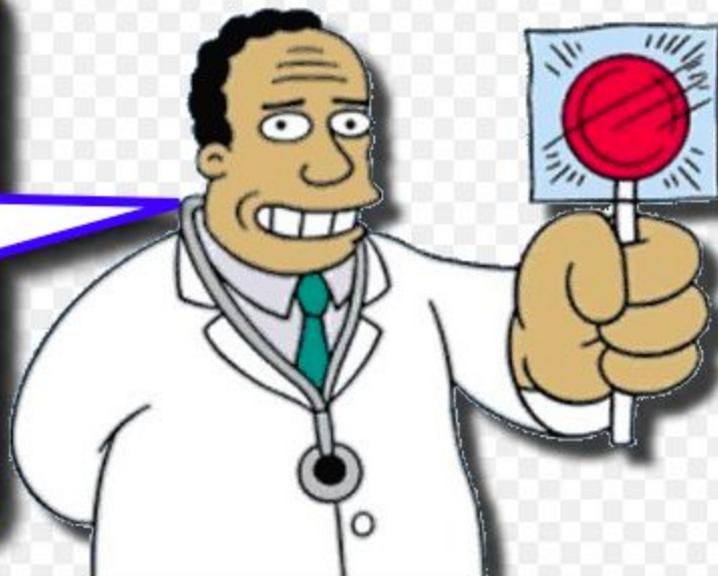
Pediatric Cardiology, KP--OC

It's about holding hands.
And people don't get lost if
there are enough people to
hold hands

--Jean Vanier, L'arche



Oh, that's cute!
A child with
Chest Pain! I
am sure it is
nothing. Take
one lollipop
and call
someone else
in the morning.



A 15 year old presents with chest pain

The next appropriate step is:

1. Order an ECG
2. Order an echocardiogram
3. Obtain labs, including troponin
4. Obtain more history



A 15 year old presents with chest pain

A 15 year old comes in with 3-4 year history of chest pain. Pain occurs every 1-2 weeks. Pain is at rest. The pain is sharp quality, located left upper parasternal area. The pain lasts for less than 1 minute, usually just a few seconds. The pain is worse when he takes a deep breath. It hurts more to twist his body. It occurred last night while he was on his side in bed on his phone. He changed position, and it improved. The pain has not occurred with exercise, except for one time a year ago when he was in the first week of football conditioning, after a long series of sprints and drills, he was out of breath and felt a tightness in his chest, but that was a different pain. He played the rest of the season without any problem and still works out daily without any chest pain. He has had wheezing in the past and has taken Albuterol for it, but this is with URI, not really with exercise. He has no heartburn symptoms.

What are the history details that seem most helpful?

A 15 year old comes in with 3-4 year history of chest pain. Pain occurs every 1-2 weeks. Pain is at rest. The pain is sharp quality, located left upper parasternal area. The pain lasts for less than 1 minute, usually just a few seconds. The pain is worse when he takes a deep breath. It hurts more to twist his body. It occurred last night while he was on his side in bed on his phone. He changed position, and it improved. The pain has not occurred with exercise, except for one time a year ago when he was in the first week of football conditioning, after a long series of sprints and drills, he was out of breath and felt a tightness in his chest, but that was a different pain. He played the rest of the season without any problem and still works out daily without any chest pain. He has had wheezing in the past and has taken Albuterol for it, but this is with URI, not really with exercise. He has no heartburn symptoms.

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What I'm thinking

(not necessarily what you should be thinking)

At first I just want to hear the story (but this takes time)

I'm not yet trying to fit it into a box or diagnosis (it does it by itself)

Then I ask specific questions to clarify which box it fits in

The first distinction is whether it seems cardiac or not (frequent with exertion)

The next distinction is determining the actual diagnosis

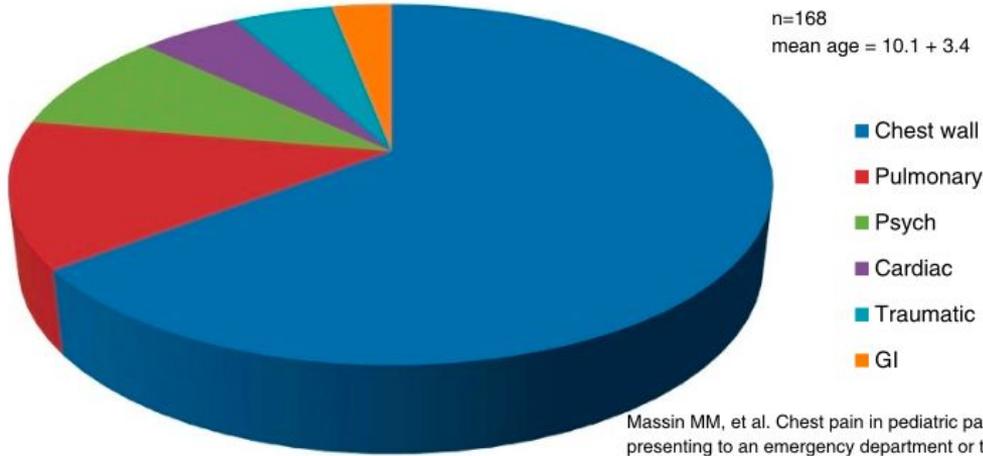
1 big idea:

Is it exertional?

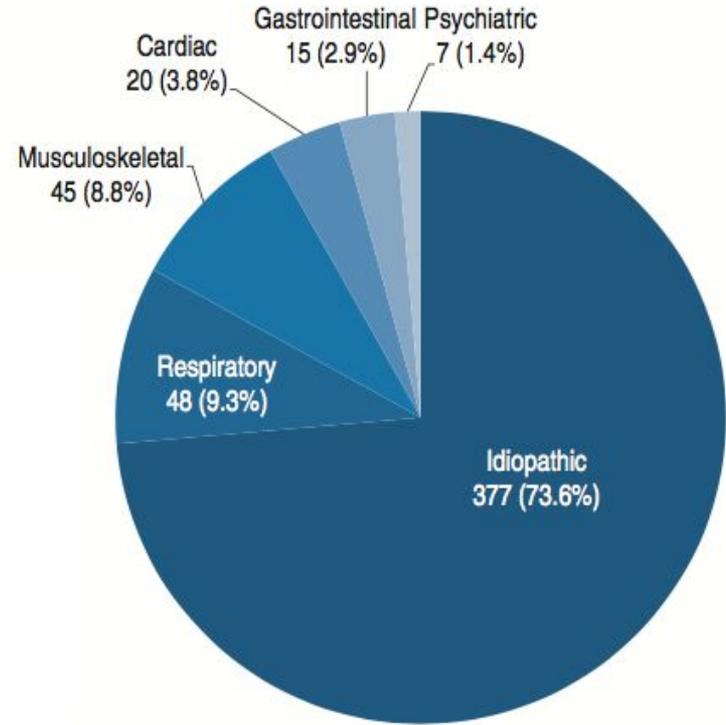
Let's take a look at the causes

Chest pain etiology in younger populations

n=168
mean age = 10.1 ± 3.4



Massin MM, et al. Chest pain in pediatric patients presenting to an emergency department or to a cardiac clinic. *Clinical Pediatrics*. 2004; 43:231-8.



Analysis of clinical characteristics and causes of chest pain in children and adolescents

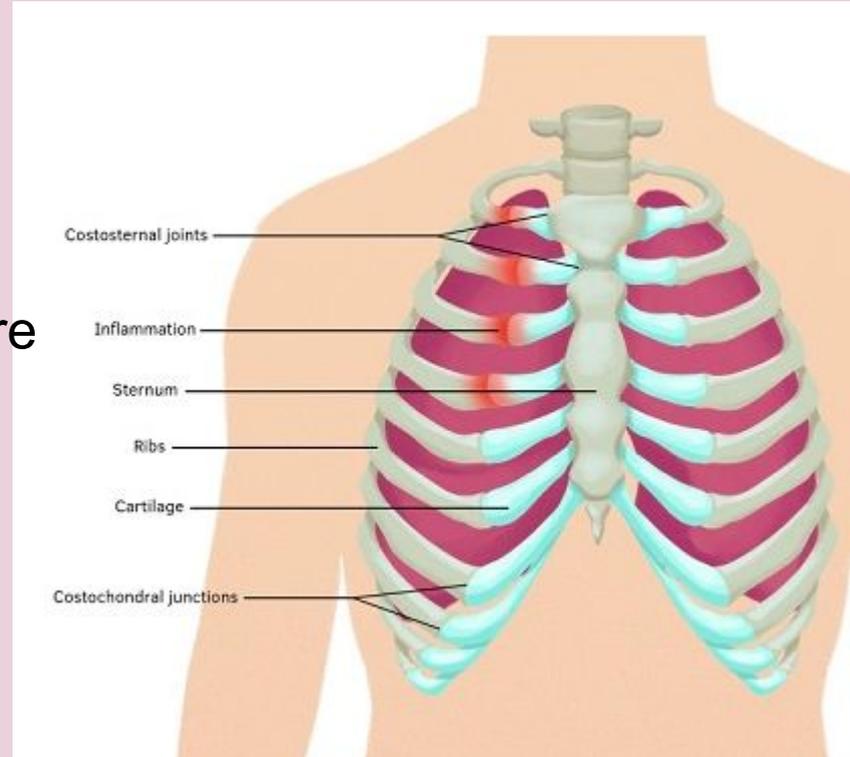
Korean J of Peds 2015

Differential diagnosis list for noncardiac causes of chest pain in athletes.

Musculoskeletal	GI	Respiratory	Miscellaneous
Rib Stress Fracture	GERD	Asthma (exercise induced, intermittent, or persistent)	Malignancy (primary or metastatic)
Costochondritis	Esophagitis	Pneumothorax (spontaneous or posttraumatic)	Drug induced
Intercostal Strain	Peptic ulcer	Pulmonary embolus	Zoster
Serratus Anterior Strain	Cholecystitis	Vocal cord dysfunction	Psychogenic (example: anxiety)
Trigger Points	Pancreatitis	Pleurisy	
Slipping Rib Syndrome	Esophageal stricture	Pneumonia (viral, bacterial, atypical)	Pneumomediastinum (spontaneous or posttraumatic)
Thoracic Radiculopathy	Hiatal hernia	Bronchitis (acute or chronic)	ACS (due to sickle cell disease)
Osteomyelitis (of Thoracic Spine, Sternum, or Rib)		Empyema	

Musculoskeletal (costochondritis, precordial catch)

- Days, months, or years of chest pain
- Seconds to several minutes
- Generally **sharp** pain
- Exacerbated by **movement** or pressure
- Exacerbated by **deep breath**
- “shortness of breath” is due to pain
- Palpitations follow pain



	Diagnoses	Distinguishing factors on history	Distinguishing factors on physical exam
Musculoskeletal	Muscle strain, trauma, xiphoid syndrome, costochondritis, Tietze's syndrome, slipping rib syndrome, precordial catch syndrome, myositis, scoliosis	Typically localized Exacerbated with palpation of area Noted at rest Improved by NSAID or acetaminophen use	Focal tenderness over an area (often costochondral junctions), reproducible tenderness, bruising, limited range of motion in upper extremities, visible deformities

**“WHEN I HAVE AN
ASTHMA ATTACK
I FEEL LIKE A FISH
WITH NO WATER.”**

-JESSE, AGE 5



Asthma

History of asthma

Chest tightness, pressure, squeezing

Exertional, seasonal, URI

Wheezing, Cough

Allergies

Proper inhaler use

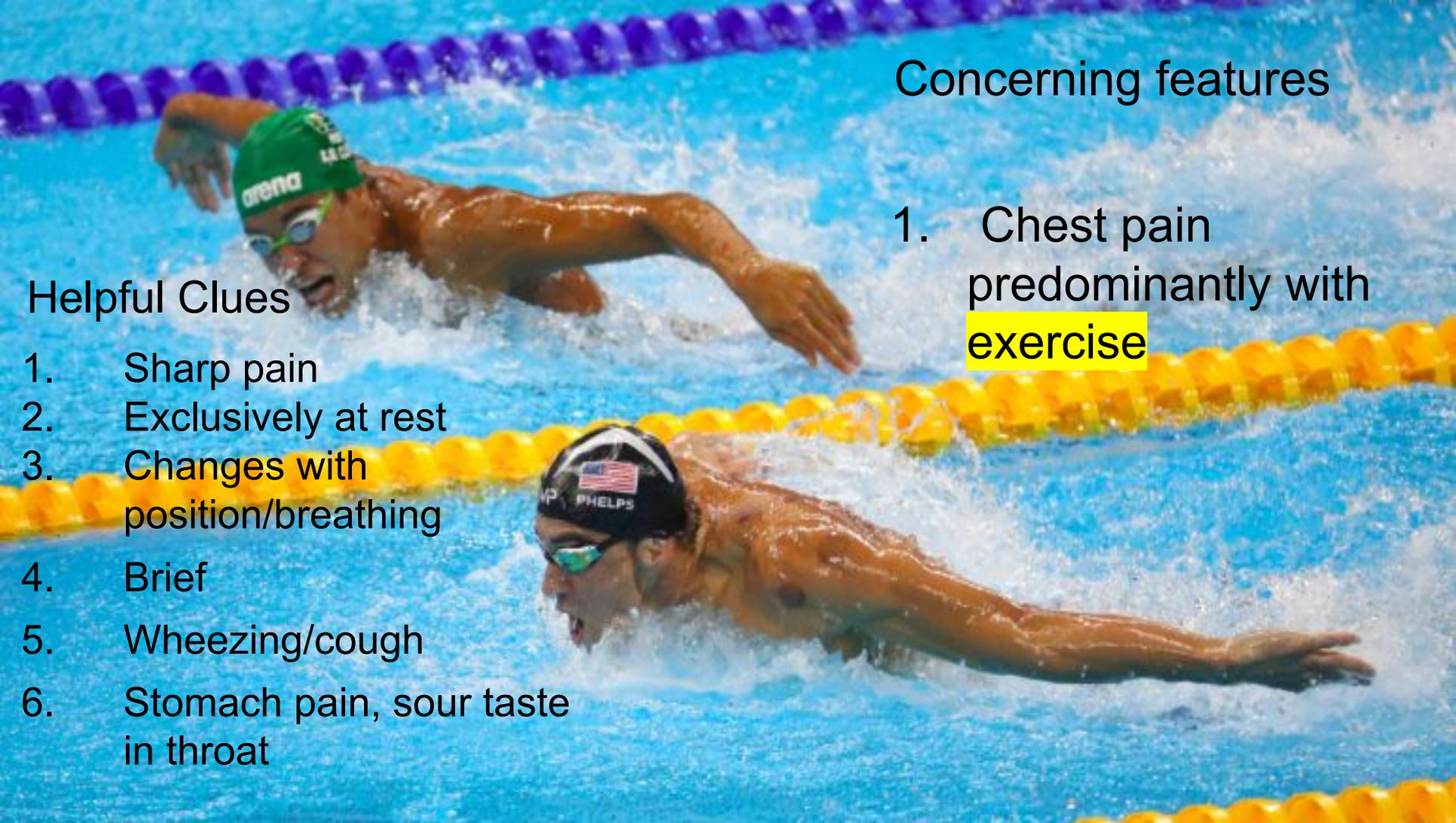
	Diagnoses	Distinguishing factors on history	Distinguishing factors on physical exam
Respiratory	Asthma, pneumonia, pleural effusion, pneumothorax, pulmonary embolism, empyema, foreign body in airway	Pleuritic pain Associated with coughing or respiratory distress Not reproducible to palpation	Tachypnea, hypoxia, retractions/nasal flaring/abdominal breathing, crepitus, diminished breath sounds, asymmetry on auscultation, adventitious lung sounds (wheeze, rales)

Gastroesophageal reflux

- Abdominal pain
- Heartburn symptoms
- Spicy, fried, fatty foods
- Sour taste in mouth
- Burning in chest



	Diagnoses	Distinguishing factors on history	Distinguishing factors on physical exam
Gastrointestinal	Esophagitis, gastritis, foreign body ingestion, caustic ingestion, gastroesophageal reflux, diaphragmatic irritation, hiatal hernia, esophageal spasm/dysmotility	Midsternal or epigastric Dyspepsia Burning quality Temporally related to food intake	Nonreproducible epigastric pain, hepatosplenomegaly

A photograph of two swimmers in a pool. The swimmer in the foreground is wearing a black swim cap with an American flag and the name 'PHELPS' on it, and is swimming with a determined expression. The swimmer in the background is wearing a green swim cap with 'arena' written on it. The pool has blue water and yellow lane dividers.

Concerning features

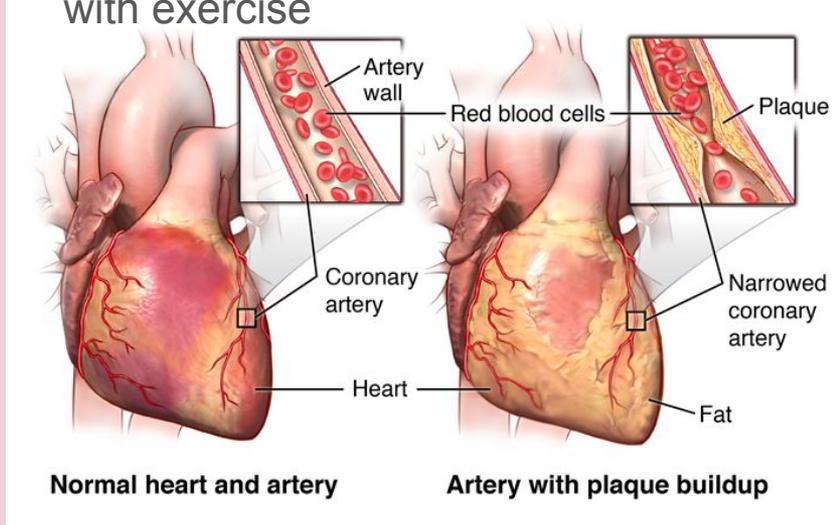
1. Chest pain predominantly with **exercise**

Helpful Clues

1. Sharp pain
2. Exclusively at rest
3. Changes with position/breathing
4. Brief
5. Wheezing/cough
6. Stomach pain, sour taste in throat

Why does exertional chest pain matter?

- Let's start with (but then move away from) a classic adult with atherosclerotic heart Dz
- Ischemia results from relatively decreased blood flow, typically not occurring at rest
- Ischemia increases in proportion/frequency with exercise



But why doesn't this make sense for a child?

- Children are too young to develop atherosclerosis.
- What else can kids “acquire”?
- What congenital heart disease?

Cardiac causes of pediatric chest pain

Hypertrophic Cardiomyopathy

Anomalous Coronary Arteries

Pericarditis

Aortic stenosis

Kawasaki's

Drugs

Cardiac causes of chest pain in athletes.

Acquired

Ischemic heart disease

Heart failure (diastolic or systolic)

Coronary Vasospasm

Arrhythmias

Supraventricular tachycardias (including atrial fibrillation/flutter)

Exercise-associated (catecholamine-induced) ventricular tachycardia

WPW syndrome

Prolonged QT syndrome

Brugada syndrome

Autoimmune

Kawasaki disease-related coronary artery aneurysm

Systemic lupus erythematosus related pleuritis

Congenital causes

Aortic stenosis

Left main coronary artery arising from the right sinus of valsalva

Right main coronary artery arising from the left sinus of valsalva

Anomalous origin of the left coronary artery from the main pulmonary artery

Coronary artery ostial stenosis

Developmental

Pregnancy

Drug-induced

Cocaine, amphetamines, anabolic steroids, marijuana, methylphenidate (Ritalin, Concerta), pseudo-ephedrine, ephedra (Ma Huang)

Genetic

Sickle cell crisis

HCM

Marfan's syndrome-related thoracic aortic dissection

Arrhythmogenic right ventricular cardiomyopathy/dysplasia

Infectious

Myocarditis

Pericarditis

Traumatic

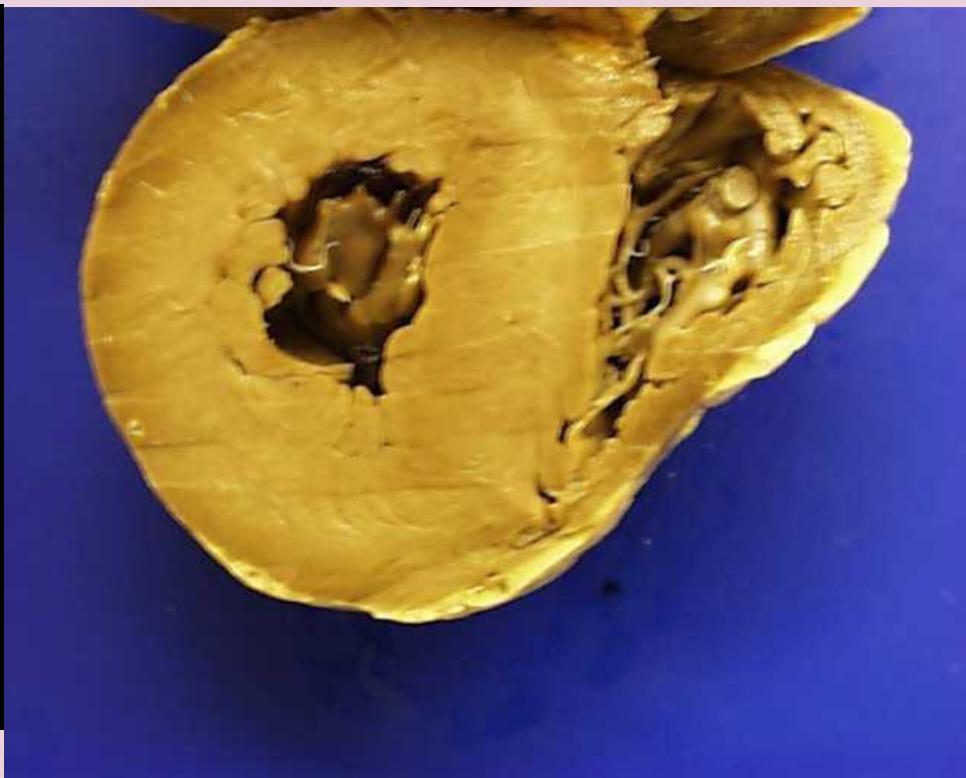
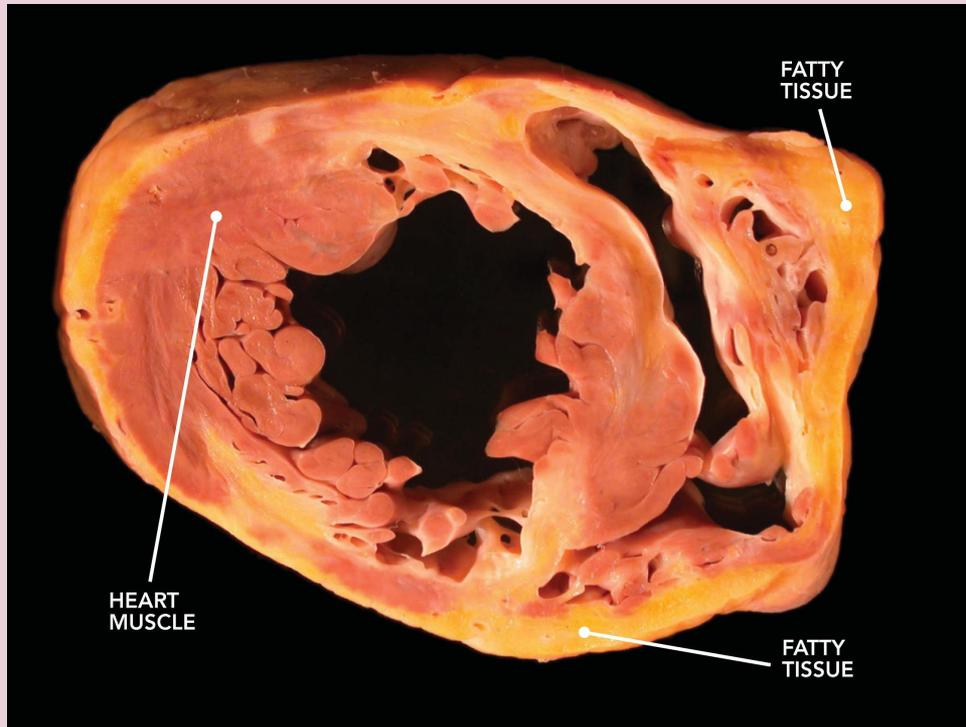
Cardiac contusion

Comotio cordis

Hemopericardium with tamponade

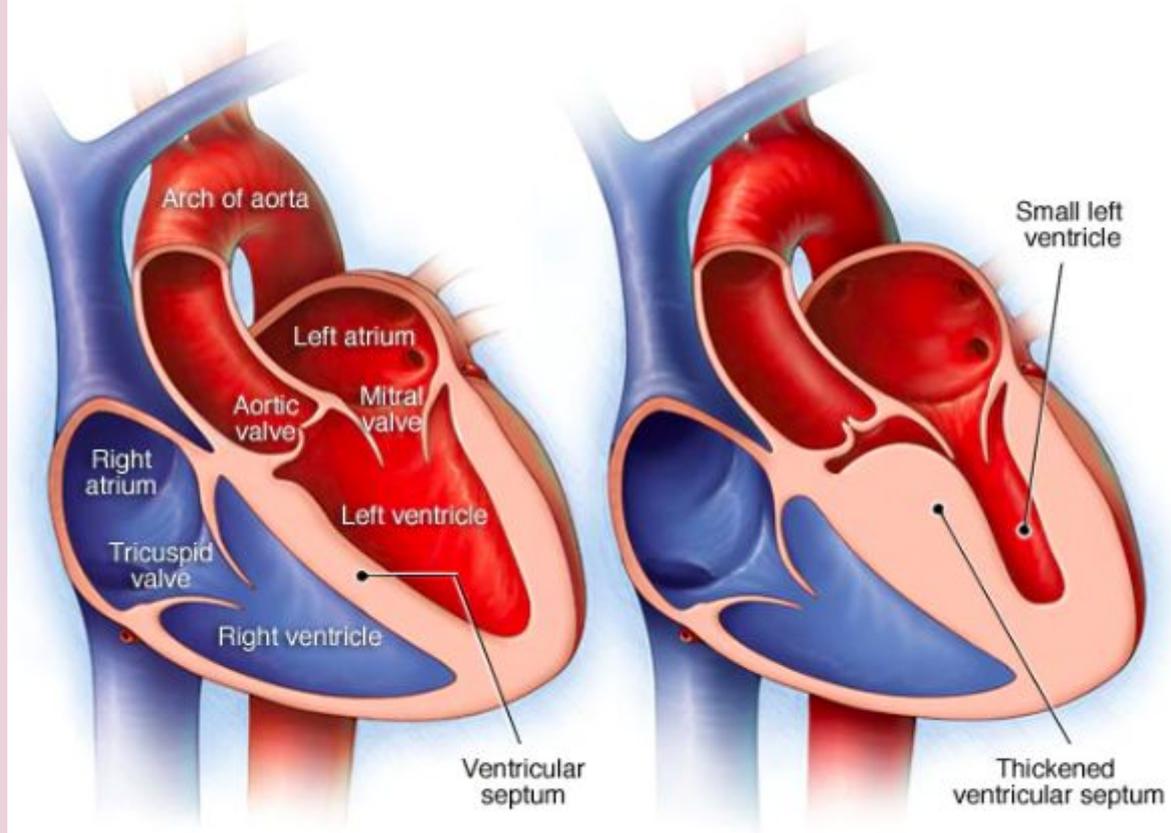
Valvular

MVP

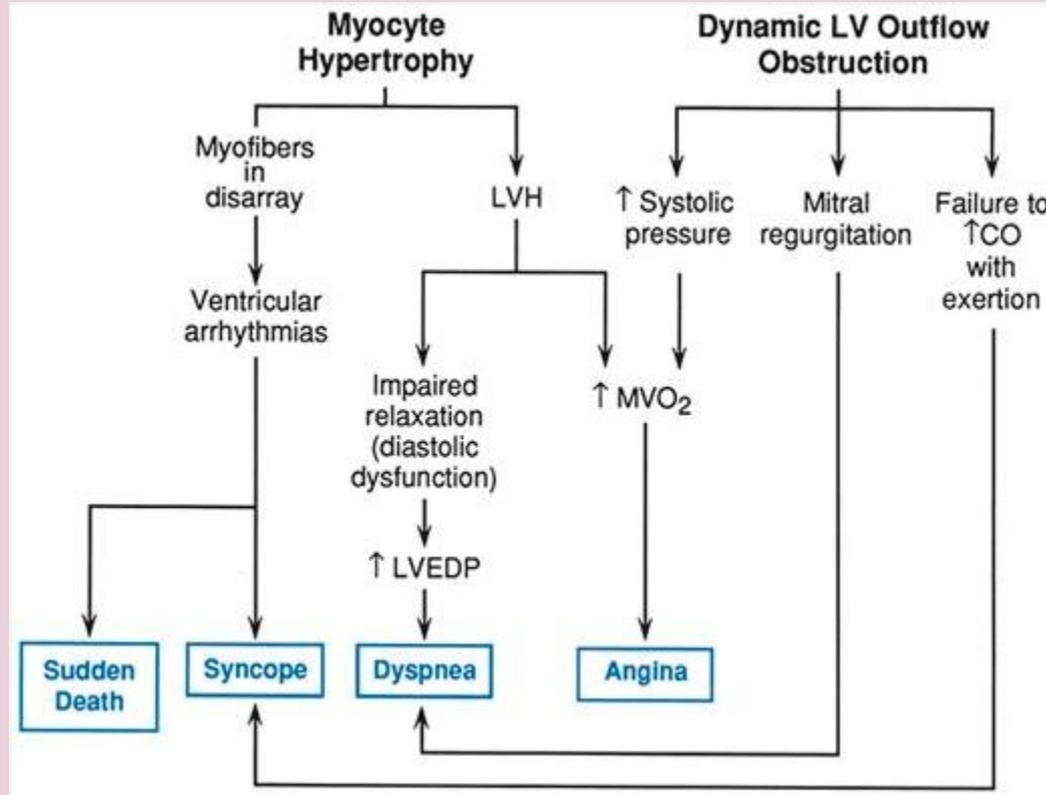


Normal

Hypertrophic cardiomyopathy



Pathophysiology of Hypertrophic Cardiomyopathy



Anomalous coronary artery (congenital)

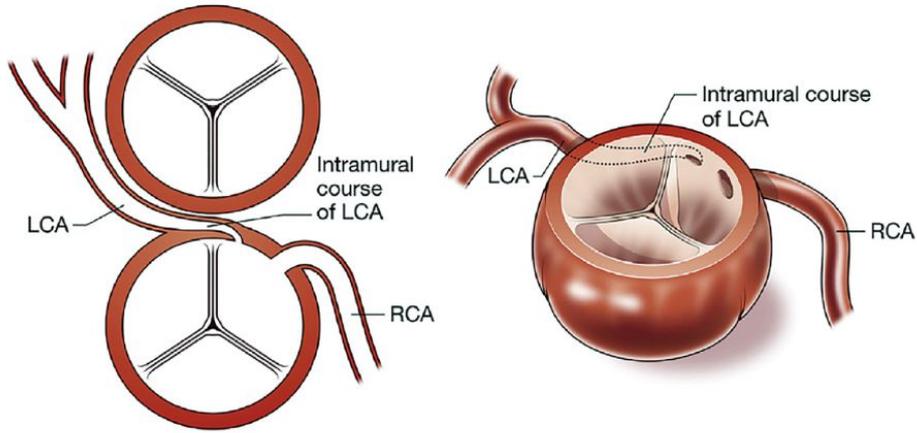
Adult vs pediatric coronary artery disease

Anomalous left or right coronary artery (from aorta or from pulmonary artery)

Obstruction -- decreased blood flow

Deoxygenated blood -- hypoxic blood flow

Surgically reparable

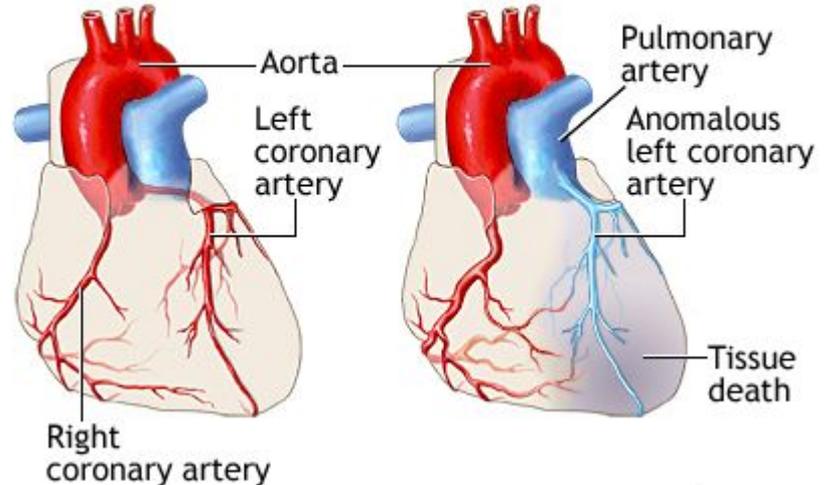


Anomalous left coronary from wrong sinus

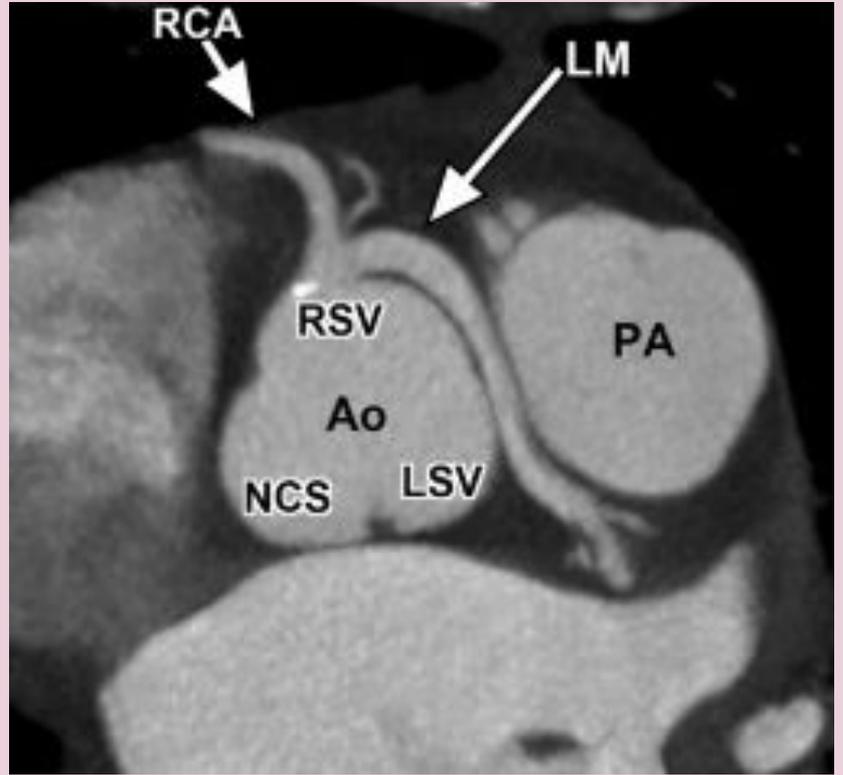
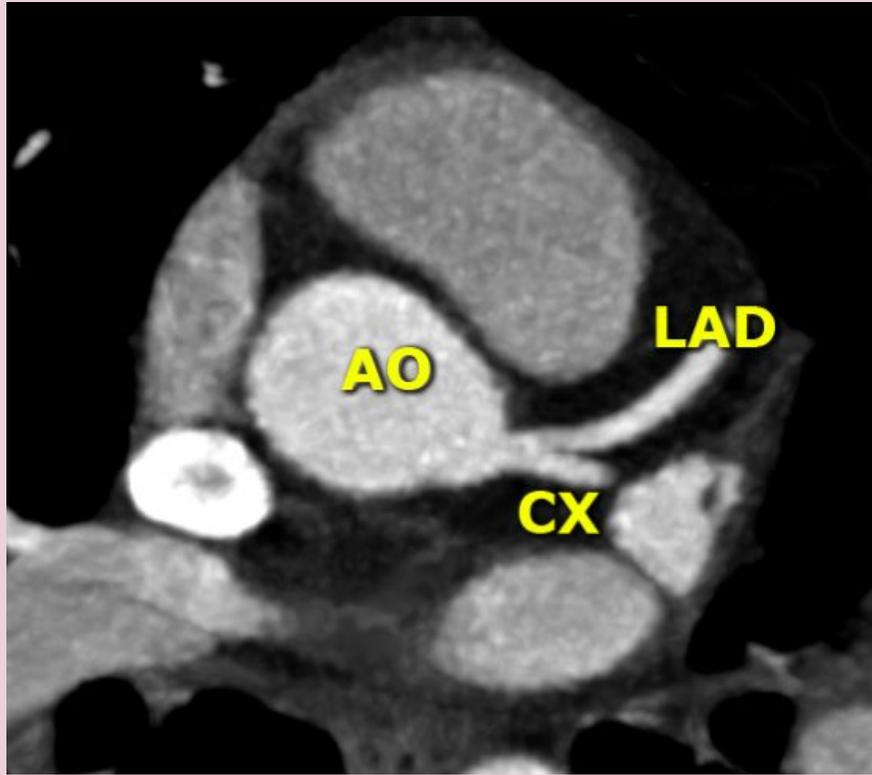
Anomalous left coronary from pulmonary artery

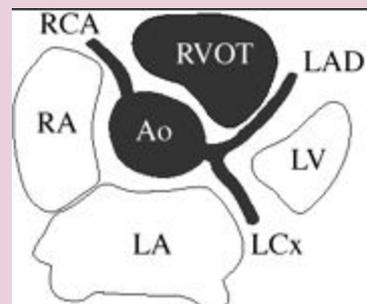
Normal heart

Anomalous left coronary artery

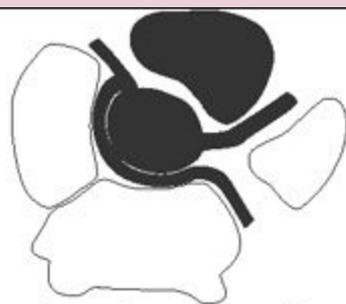




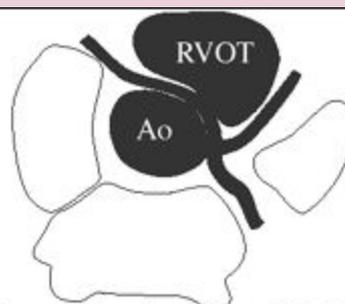




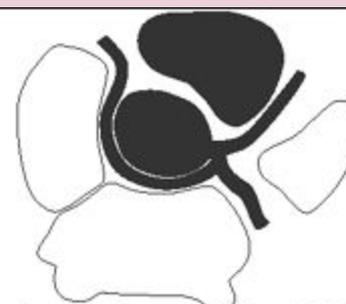
Normal Anatomy



Anomalous Circumflex



Anomalous interarterial RCA



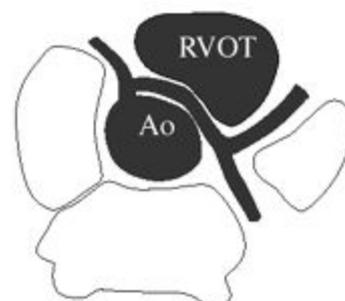
Anomalous posterior RCA



Anomalous anterior LCA



Anomalous posterior LCA



Anomalous interarterial LCA



Anomalous intramural LCA

Beyond symptoms

- Why does the family history matter? What Dx are we worried about?
 - Congenital Heart Disease?
 - Coronary atherosclerotic heart disease?
 - Arrhythmias?
 - **Cardiomyopathies?**
 - Asthma
- Family history
 - Sudden, aborted, or unexplained death or cardiomyopathy
 - Most will not know the diagnosis
 - Deeper questions--risk factors for MI, defibrillator, age, medications, surgery?
 - First degree relatives
 - Autosomal dominant vs autosomal recessive transmission
 - What about grandma? Cousin? Half-brother? **Mother?**

Physical exam

Could be

- Reproducible tenderness → Musculoskeletal
- Wheezing → Asthma
- Abdominal tenderness → GERD
- Pectus excavatum → Musculoskeletal
- Murmur → HCM

Who needs an EKG?



1. A 5 year old boy entering his first soccer season, asymptomatic, presents for pre-participation physical.
2. A 16 year old had 2 episodes of shortness of breath and chest tightness during conditioning week of football 1 year ago. None for the rest of the season nor the current basketball season.
3. A 12 year old has mid-sternal chest pain each time he runs the mile at school. He has asthma and has been using his inhaler prior to running.
4. 14 year old with sharp left sided chest pain, at rest, lasting a few seconds. His brother has hypertrophic cardiomyopathy diagnosed at 28 years of age.

Common athlete EKG findings

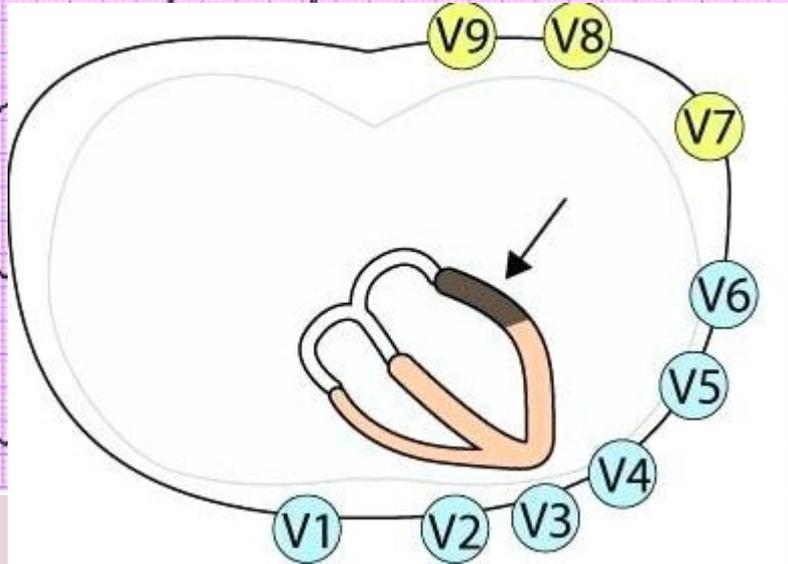
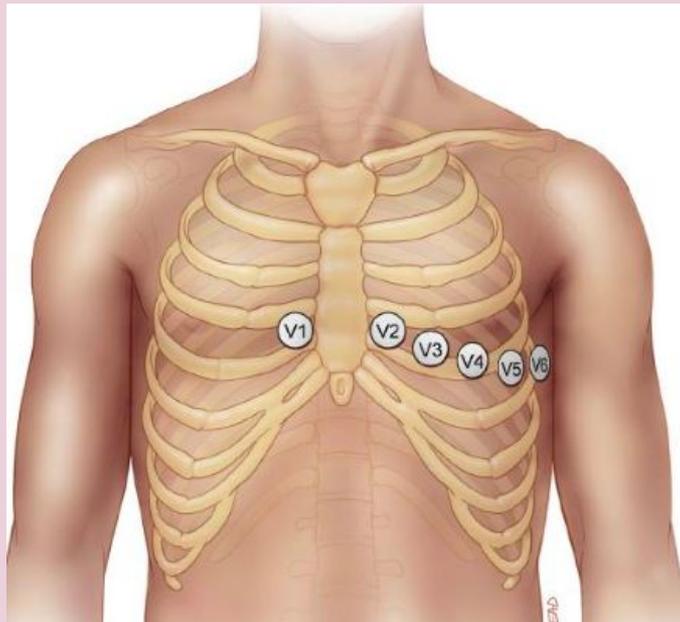
(should not preclude participation)

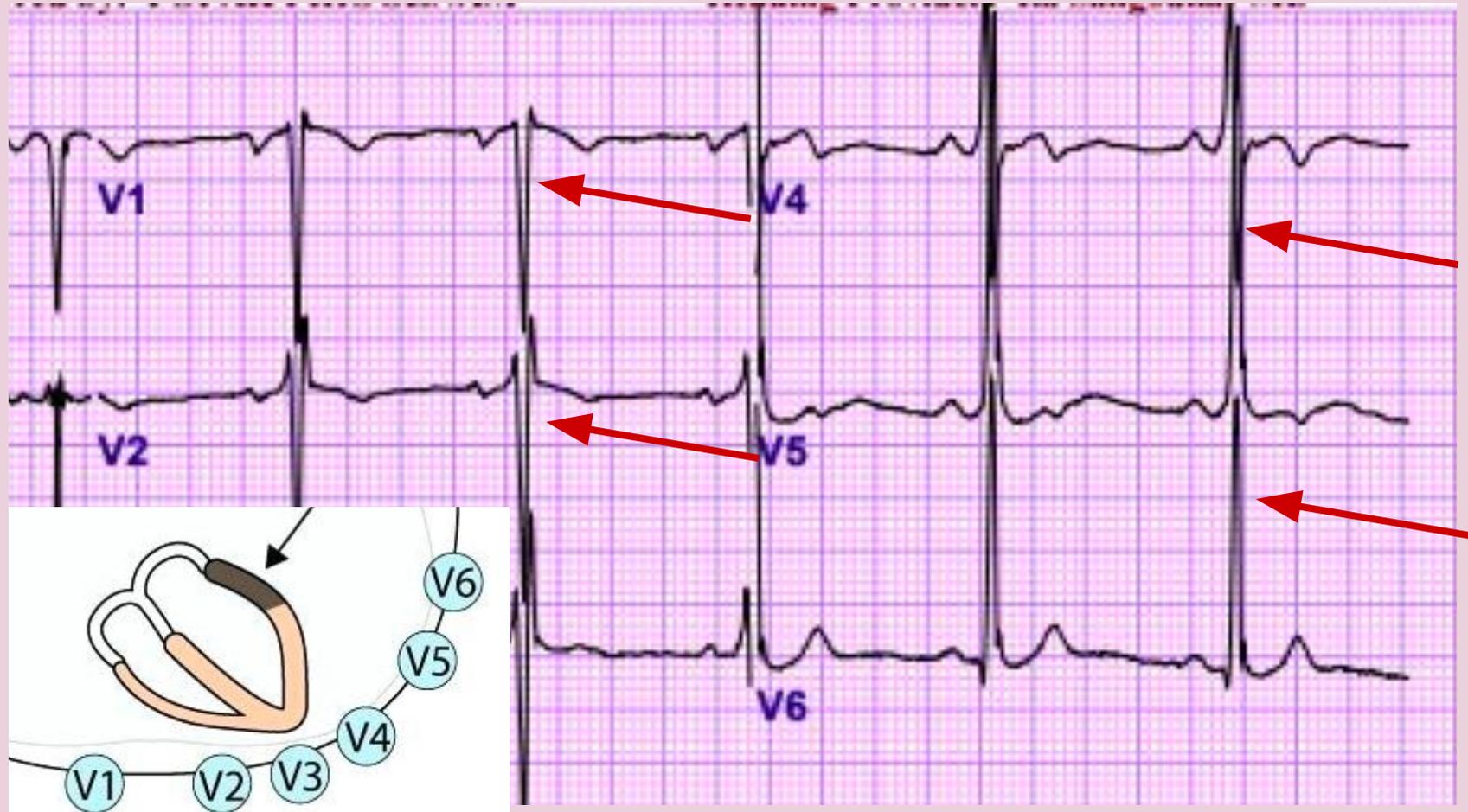
- Sinus bradycardia (rate 30-60 bpm)
 - 1st degree AV block (up to 35%)
 - 2nd degree, mobitz I (up to 10%)
 - Voltage LVH (24%)
 - IRBBB (35-50%)
 - Early repolarization (50-80%)
- Sinus arrhythmia is a normal finding

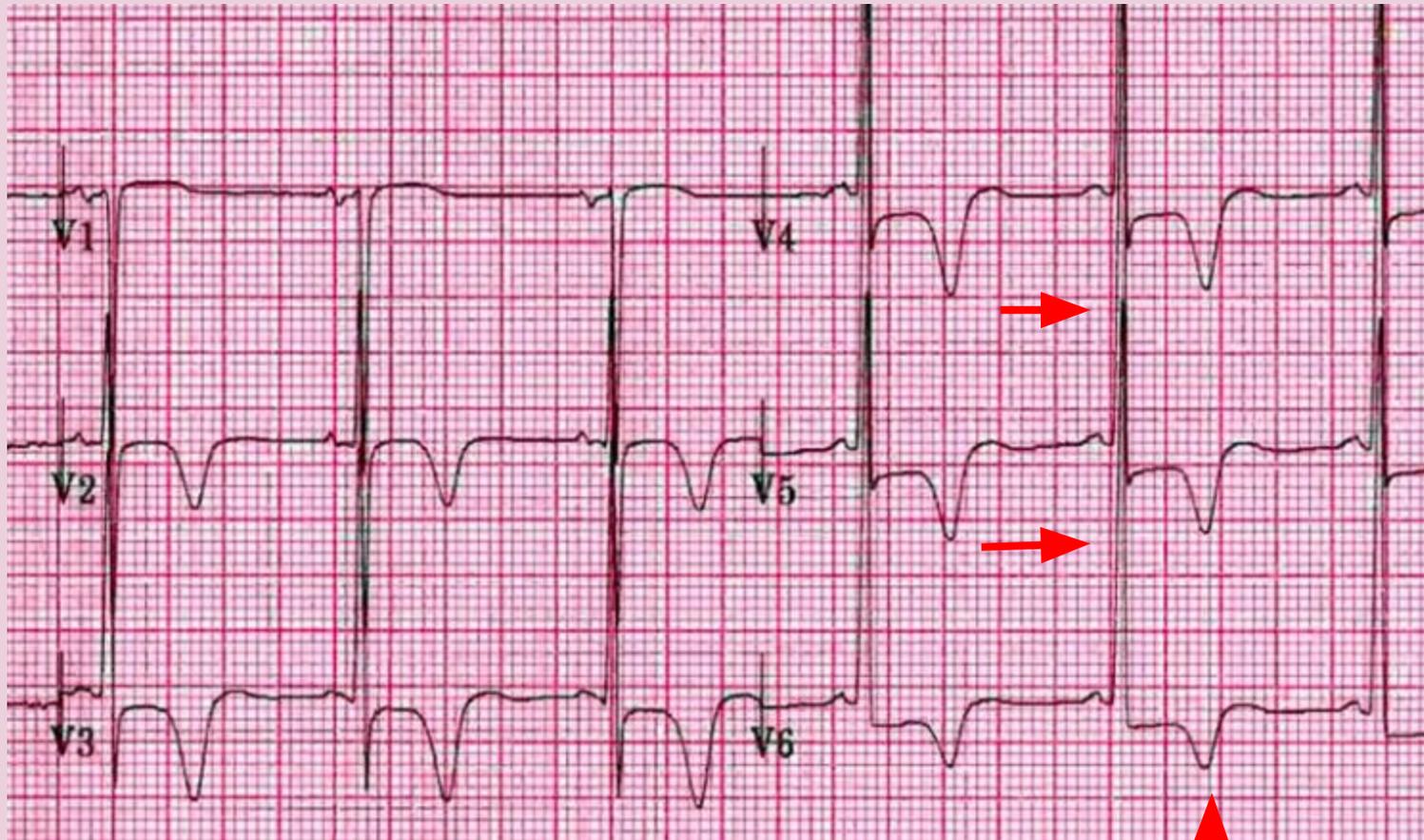
EKG for HCM

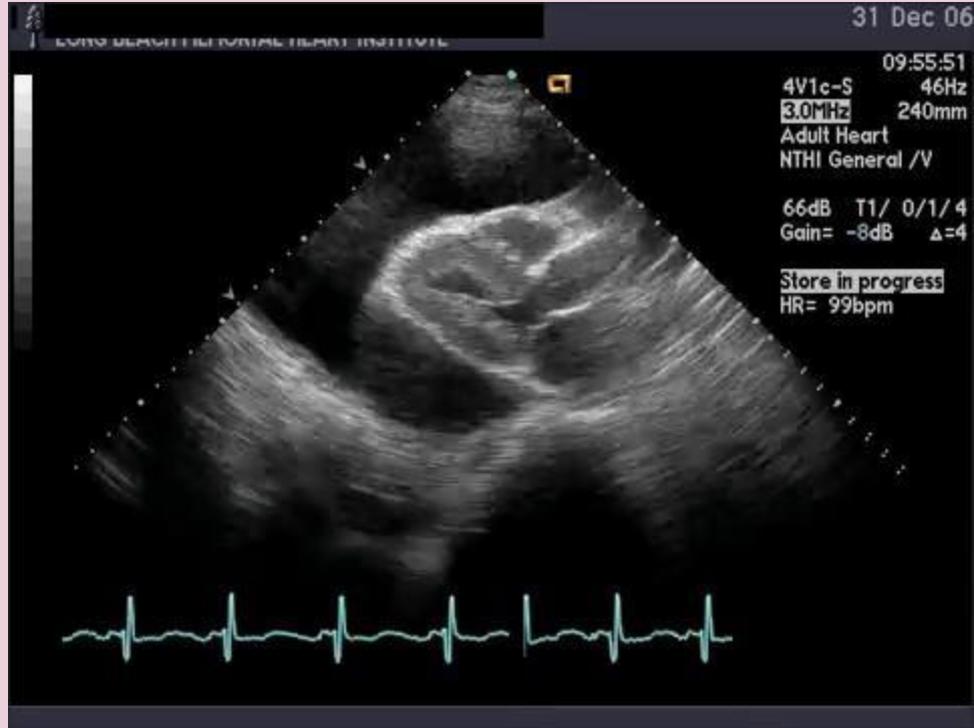
- EKG is abnormal in 95% of HCM
- The 5% with a normal EKG has limited risk of sudden death
- So if the chest pain is not predominantly exertional and the EKG is normal, reassure

- A computer “abnormal” is often incorrect
- Call, page, fax EKG









Who gets an echocardiogram?

Why are we talking about echos when I don't
order echos for kids?

Boston Children's 2006

Table 2. SCAMP initiative for chest pain

Significant history	Significant family history	Significant physical findings	Significant ECG findings
Association with exertion or exertional syncope Radiation to back, jaw, left arm, or left shoulder More pain with supine position Temporal association with fever History of systemic inflammatory disease, malignancy, hypercoagulable state myopathy or coagulable state, myopathy, or prolonged immobilization	Sudden or unexplained death Aborted sudden death Cardiomyopathy Severe familial hyperlipidemia Pulmonary hypertension	Pathologic murmur, gallop, rub, abnormal second sound, distant heart sounds Hepatomegaly Decreased peripheral pulses Peripheral edema Tachypnea Fever over 38.4°C	Ventricular hypertrophy, atrial enlargement ST enlargement, ST-T abnormalities, high-grade A-V block, ventricular or supraventricular ectopy, axis deviation, ventricular preexcitation. Most athletes (60%) in this large cohort had ECGs that were completely normal or showed only minor alterations. A variety of abnormal ECG patterns occurred in 40%; this was usually indicative of physiological cardiac remodeling. A small but important subgroup of athletes without cardiac morphological changes showed striking ECG abnormalities that suggested cardiovascular disease; however, these changes were likely an innocent consequence of long-term, intense athletic training and, therefore, another component of athlete heart syndrome. Such false-positive ECGs represent a potent

Appropriate Use Criteria for Pediatric Echocardiography

American Society of Echocardiography

TABLE 3 Chest Pain

Indication		Appropriate Use Rating
28.	Chest pain with no other symptoms or signs of cardiovascular disease, a benign family history, and a normal ECG	R (2)
29.	Chest pain with other symptoms or signs of cardiovascular disease, a benign family history, and a normal ECG	M (6)
30.	Exertional chest pain  	A (8)
31.	Non-exertional chest pain with no recent ECG	R (3)
32.	Non-exertional chest pain with normal ECG	R (1)
33.	Non-exertional chest pain with abnormal ECG  	A (7)
34.	Chest pain with family history of sudden unexplained death or cardiomyopathy  	A (8)
35.	Chest pain with family history of premature coronary artery disease	M (4)
36.	Chest pain with recent onset of fever	M (6)
37.	Reproducible chest pain with palpation or deep inspiration	R (1)
38.	Chest pain with recent illicit drug use	M (6)

The number in parentheses next to the rating reflects the median score for that indication.



Application of the Pediatric Appropriate Use Criteria for Chest Pain

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Objectives To characterize the subgroup of outpatient pediatric patients presenting with chest pain and to determine the effectiveness of published pediatric appropriate use criteria (PAUC) to detect pathology.

Study design The Pediatric Appropriate Use of Echocardiography study evaluated the use and yield of transthoracic echocardiography (TTE) before and after PAUC release. Data were reviewed on patients ≤ 18 years of age who underwent TTE for chest pain. Indications were classified as appropriate (A), may be appropriate (M), and rarely appropriate (R) based on PAUC ratings, and findings were normal, incidental, or abnormal.

Results Chest pain was the primary indication in 772 of 4562 outpatient TTE studies (17%) (median age 14 years, IQR 10-16) ordered during the study period: 458 of 772 before (59%) and 314 of 772 after (41 %) the release of PAUC with no change in appropriateness. In A indications ($n = 654$), 642 (98%) were normal, 5 (1%) had incidental findings, and 7 (1%) were abnormal. A and M detected 100% of all abnormal findings (A: $n = 7$; M: $n = 6$; R: $n = 0$), with an association between ratings and findings ($P < .001$). There was no association between R rating and any pathology.

Conclusions There was no change in ordering patterns with publication of the PAUC. Despite the high rate of TTEs ordered for indications rated A, most studies were normal. Studies that detected pathology were performed for indications rated A or M, but not R. This study supports PAUC as a useful tool in pediatric chest pain evaluation that may subsequently improve the use of TTE. (*J Pediatr* 2017;185:124-8).

Appropriate Use Criteria evaluated

1 year time period

68 pediatric cardiologists

772 echocardiograms for chest pain

Median 8 echo per cardiologist

13 abnormal echocardiograms (1.7%)

3 abnormal related to chest pain (0.39%)

2 abnormal, moderate/severe (0.26%)

98% of “A” indications were normal

All significant abnormal findings were “A” indication, no abnormal findings in “R” group

Table II. Pathologic findings on TTE

	Ages (y)		AUC indication and rating	Pathology	Severity	Related/unrelated to indication
1	16	30	A Exertional chest pain	Mitral valve prolapse with mild mitral regurgitation	Minor	Unrelated
2	17	30	A Exertional chest pain	Echo dense pericardium	Minor	Related
3	18	30	A Exertional chest pain	Aortic arch abnormality	Minor	Unrelated
4	15	30	A Exertional chest pain	Mild aortic insufficiency in an anatomically normal valve	Minor	Unrelated
5	7	30	A Exertional chest pain	Coronary abnormality	Minor	Unrelated
6	15	34	A Non-exertional chest pain with abnormal ECG	Moderate pericardial effusion	Moderate	Related
7	15	34	A Chest pain with family history of sudden unexplained death or cardiomyopathy	Cardiac tumor on mitral valve with mitral stenosis and mitral regurgitation	Severe	Related
8	14	29	M Chest pain with other symptoms or signs of CV disease	Mitral valve prolapse	Minor	Unrelated
9	14	29	M Chest pain with other symptoms or signs of CV disease	Mitral valve prolapse	Minor	Unrelated
10	17	29	M Chest pain with other symptoms or signs of CV disease	Mitral valve prolapse	Minor	Unrelated
11	17	29	M Chest pain with other symptoms or signs of CV disease	Mitral valve prolapse	Minor	Unrelated
12	16	29	M Chest pain with other symptoms or signs of CV disease	Mildly dilated aortic root	Minor	Unrelated
13	18	29	M Chest pain with other symptoms or signs of CV disease	Mitral valve prolapse	Minor	Unrelated

Evaluating Appropriate Use of Pediatric Echocardiograms for Chest Pain in Outpatient Clinics



Reid C. Chamberlain, MD, Jonathan H. Pelletier, MD, Sarah Blanchard, MD, Christoph P. Hornik, MD, MPH, Kevin D. Hill, MD, MS, and Michael J. Campbell, MD, *Durham, North Carolina*

Background: Echocardiography is often used in the evaluation of pediatric chest pain, but the incidence of cardiac pathology is low. In 2014, the American College of Cardiology published appropriate use criteria (AUC) for echocardiography including recommendations for pediatric chest pain. We evaluated the frequency and diagnostic yield of echocardiograms performed for each AUC indication and cost associated with echocardiograms performed for indications meeting the “rarely appropriate” criteria.

Methods: Retrospective, single-institution study of all patients 18 years or younger undergoing an initial evaluation for chest pain by a pediatric cardiologist (2014-15). We categorized the appropriateness of indications for echocardiograms on the basis of the AUC. We used multivariable logistic regression to determine factors associated with performance of an echocardiogram with the “rarely appropriate” indication. Excess costs associated with nondiagnostic echocardiograms meeting the “rarely appropriate” criteria were estimated using the Healthcare Bluebook to estimate a fair market price.

Results: The cohort included 539 patients, median age 13 years (range, 3-18) and 51.0% female. With retrospective application of the AUC, echocardiogram indications were classified as “appropriate” (304/539, 56.4%), “maybe appropriate” (68/539, 12.6%), and “rarely appropriate” (167/539, 31.0%). Echocardiograms were performed in 70.5% (380/539) of patients overall and in 35.9% (60/167) of patients with “rarely appropriate” indications. Of those undergoing echocardiography, abnormal findings were present in 5.0% (19/380) and incidental findings in 2.6% (10/380); however, only one echocardiogram (0.3%) led to a diagnosis considered to be contributory to the patient’s chest pain. There were no abnormal findings in the “rarely appropriate” subgroup. Provider use of echocardiography for “rarely appropriate” indications varied widely from 0 to 75% across 15 providers ($P = .004$). In multivariable analysis, provider clinical experience of ≥ 20 years was associated with a lower rate of echocardiograms for “rarely appropriate” indications (odds ratio, 0.21 [95% CI, 0.09-0.47] vs. providers with < 10 years’ experience, $P < .001$). There was no significant association between race, ethnicity, age, sex, payer status, or total number of patients seen and performance of an echocardiogram meeting the “rarely appropriate” indications. Echocardiograms with “rarely appropriate” indications resulted in \$47,578 in excess costs over the 1-year study.

Conclusions: Echocardiogram use in patients meeting the “rarely appropriate” indication criteria is of little diagnostic utility and contributes to additional cost to the patient and health care system. (*J Am Soc Echocardiogr* 2017;30:708-13.)

Findings consistent

539 CP patients, 380 echos

5% abnormal, minor

1 abnormal related to CP, 0.18%

No abnormal in the “R” group

No significant abnormal nor related in “M”

Excess cost of >\$47,000

Table 2 Summary of abnormal echocardiogram results

	Total, N = 380	Rarely appropriate, n = 60	Maybe appropriate, n = 52	Appropriate, n = 268
Abnormal, n (%)	19 (5.0)	0 (0.0)	2 (3.8)	17 (6.3)
Patent ductus arteriosus	1	—	—	1
Atrial septal defect	1	—	—	1
Ventricular septal defect	2	—	—	2
Bicuspid aortic valve*	5	—	—	5
Valvar regurgitation	3	—	1	2
Anomalous right coronary artery	1	—	—	1
Abnormal aortic origin of coronary†	2	—	—	2
Partial anomalous pulmonary venous return	1	—	—	1
Aortic dilation	2	—	—	2
Situs inversus	1	—	1	—

Bottom line--our view

- If it hurts to breathe, move, or press on it, it's non-cardiac
- If you think it's non-cardiac, I will agree
- Parents (and kids) need confident reassurance
- EKG not required but may be helpful
- It may or may not go away but not to worry unless it's predominantly with exercise
 - 60% improved by 2 years

Communicating the plan

- Convincingly reassuring the family
- Common and frequent: up to 50% of children with chest pain
- “I’ve seen this a lot”, “I get that too”, 80 year old vs 13 year old
- Explain physiology: sternal joint, costochondral junction, GERD, asthma
- Trial of ibuprofen (even tylenol) PRN
- Things to watch for: regularly exertional
- Let them play

What about pericarditis?

And myocarditis?

Aortic stenosis?

Marfan Syndrome?

Pericarditis

Myocarditis

Aortic Stenosis

Marfan Syndrome

While the primary question of exertional chest pain does not apply to these, the history or exam for these will point to the diagnosis.

Pericarditis

Myocarditis



Acute and constant chest pain, URI

Pain while supine, fatigue, fever

Aortic Stenosis

Progressive dyspnea, murmur

Marfan Syndrome

Typical features, dilation vs dissection

Shaun White

- Tetralogy of Fallot
- S/p 2 surgeries
- 3x olympics, gold medalist in snowboarding
- Professional skateboarder and snowboarder

