Sudden Cardiac Arrest in Young Athletes

Arash Sabati MD FACC

10/11/2023

Disclosures

- No financial disclosures

Context

- Sudden Cardiac Arrest (SCA)/Sudden Cardiac Death (SCD)
  - Also referred to as aborted SCD
  - Culmination of multiple different etiologies that result in VT/VF or myocardial infarction
- Rare but tragic events
- Often highly publicized and may generate attention and other emotional reactions from the community

Recent events/news stories

The Washington Post
Democracy Dies in Darkness

Bronny James in stable condition after cardiac arrest during practice at USC

By Cindy Boren and Ben Golliver
Updated July 25, 2023 at 11:56 a.m. EDT | Published July 25, 2023 at 11:08 a.m. EDT

Bronny James, shown during a high school basketball game this winter, was hospitalized Monday after going into cardiac arrest. (Gregory Payan/AP)

Washingtonpost.com

The Washington Post
Democracy Dies in Darkness

kgormley

Bronny James has treatable heart defect, eyes basketball return

By Ben Golliver
August 25, 2023 at 8:42 p.m. EDT

USC freshman Bronny James, son of NBA superstar LeBron James, could make a swift return to the court after being diagnosed with a congenital heart defect that doctors believe can be treated. (AP Photo/Gregory Payan, File)

Recent events/news stories

Sudden Cardiac Arrest: Survivors stress importance of CPR

12-year-old Phoenix soccer player returns home month after sudden cardiac arrest

A 12-year-old Phoenix soccer player who collapsed on the field due to a rare medical condition is now out of the hospital.

By Sarah Robinson

Published: Jun. 2, 2023 at 5:12 PM MST

Fox10phoenix.com
Azfamily.com

Incidence of SCA/SCD

True incidence is controversial because the denominator is not clear and the events are rare.

Estimates are 1 in 50-100k athlete-years

TABLE 1 Studies of SCA and SCD Incidence in Athletic Popul

First Author (Ref. #)	Year	Incidence (per Athlete-Person Yrs)	Total Number of Cases	De
Van Camp et al. (1)	1995	1/281,000	107	
Corrado et al. (2)	2006	1/115,000 over the final 11 yrs (1/53,000 over the total period)	55	
Maron et al. (8)	2009	1/103,000	22	
Maron et al. (8)	2009	1/164,000	1,049	
Holst et al. (3)	2010	1/83,000	15	
Solberg et al. (4)	2010	1/117,000	23	
Steinvil et al. (9)	2011	1/39,000	24	
Marijon et al. (10)	2011	1/102,000	50	
Roberts and Stovitz (5)	2013	1/417,000	4	
Drezner et al. (11)	2014	1/71,000	13	
Maron et al. (6)	2014	1/63,000	64	
Harmon et al. (7)	2015	1/54,000	79	
Harmon et al. (12)	2016	1/101,000 SCD 1/67,000 SCD + SCA	69 SCD 104 SCA + SCD	5

C/T — competition/training (defined as during or within 1 h of cessation of a
Catastrophic Sports Injury Research; NHLBI — National Heart, Lung, Blood In

JACC: HEART FAILURE VOL. 6, NO. 1, 2018Sudden Cardiac Death in Athletes JANUARY 2018:30–40

Risk Factors

- Sports activity and training – 2.4 to 4.5x increased risk of SCA/SCD compared to non-athletes or recreational athletes
- Male athletes 3-5x higher SCD
- Black athletes 3.2x higher
- Athletes playing basketball and football have highest incidence:
  - NCAA Division 1 Mens basketball
    - Incidence 1 in 5,200 Athlete years (all males)
    - Incidence 1 in 4,380 Athlete years (black males)

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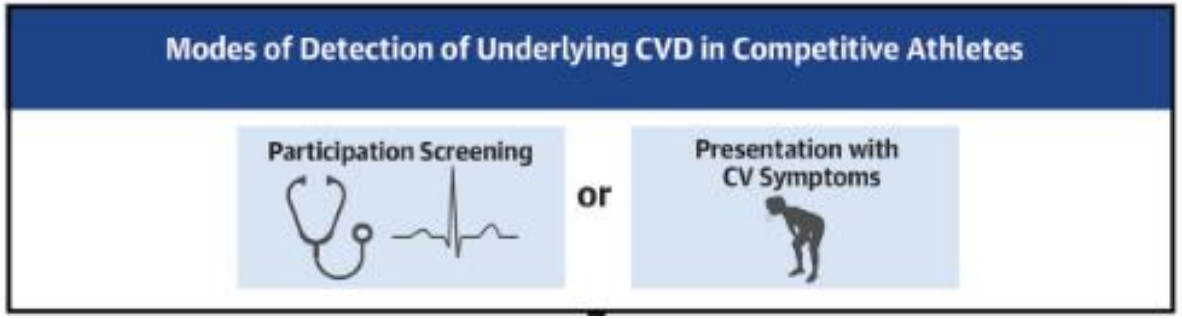
Screening and Prevention

- Pre-Participation Evaluation (PPE)
- Recommended by multiple societies
- 14-point AHA history and physical is a standard



	Sensitivity/Specificity
History	20%/94%
Physical examination	9%/97%

- Initial positive response rate is high 24-68% - not all require evaluation
- No evidence that ECG screening adds benefit to this screening process

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JACC VOL. 82, NO. 8, 2023 Martinez et al AUGUST 22, 2023:661–670

The 14 Element AHA Cardiovascular Screening Checklist for Congenital and Genetic Heart Disease (Recommended for Pre-Participation Screening of Competitive Athletes)

Personal History

Yes	No	
☐	☐	1. Chest pain/discomfort/tightness/pressure related to exertion
☐	☐	2. Unexplained syncope/near-syncope*
☐	☐	3. Excessive exertional and unexplained dyspnea/fatigue or palpitations, associated with exercise
☐	☐	4. Prior recognition of a heart murmur
☐	☐	5. Elevated systemic blood pressure
☐	☐	6. Prior restriction from participation in sports
☐	☐	7. Prior testing for the heart, ordered by a physician

Family History

Yes	No	
☐	☐	8. Premature death (sudden and unexpected, or otherwise) before age 50 attributable to heart disease in ≥1 relative
☐	☐	9. Disability from heart disease in close relative <50 y of age
☐	☐	10. Hypertrophic or dilated cardiomyopathy, long-QT syndrome, or other ion channelopathies, Marfan syndrome, or clinically significant arrhythmias; specific knowledge of certain cardiac conditions in family members

Physical Examination

Yes	No	
☐	☐	11. Heart murmur**
☐	☐	12. Femoral pulses to exclude aortic coarctation
☐	☐	13. Physical stigmata of Marfan syndrome
☐	☐	14. Brachial artery blood pressure (sitting position)***

* When determined to be not of neurcardiogenic (vasovagal) in origin. Of particular concern is syncope post-strenous activity.

** Auscultation should be performed in both sitting and standing positions (or with Valsalva maneuver). Objective is to identify murmurs of dynamic LV outflow tract obstruction.

*** Should be taken in both arms.

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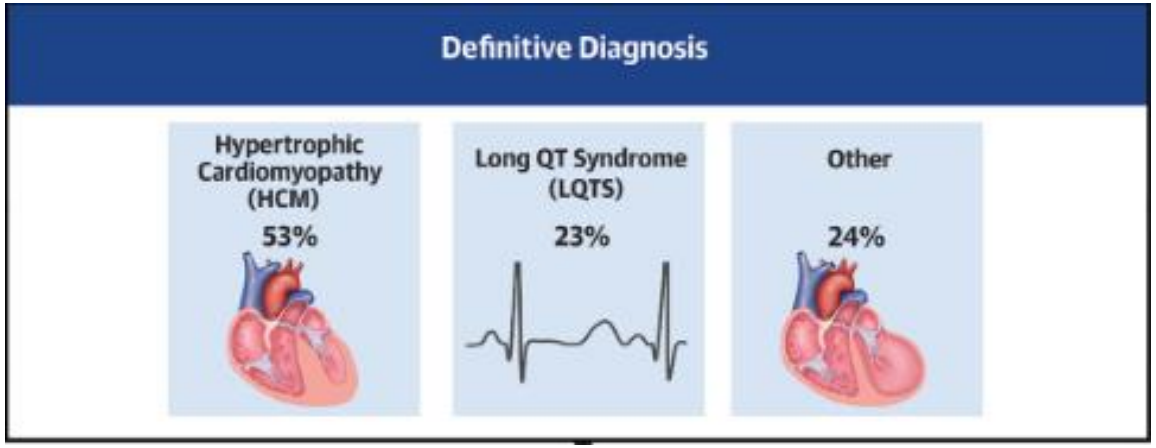
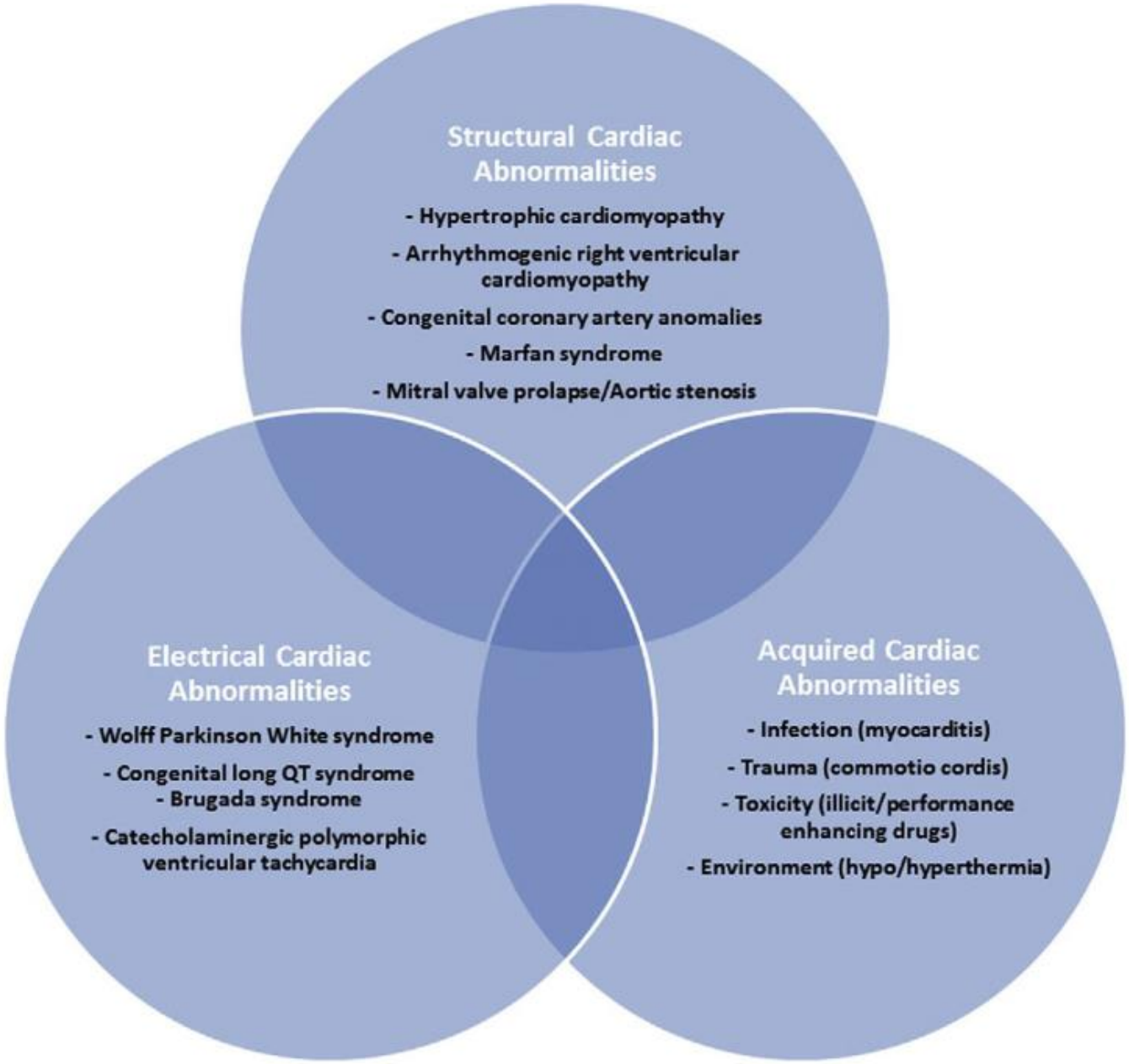
Maron, et al; JACC 2014;64(14) 1479-1514.

Causes of SCA/SCD

Incidence has been variable depending on the study or registry





JACC: HEART FAILURE VOL. 6, NO. 1, 2018Sudden Cardiac Death in Athletes JANUARY 2018:30–40

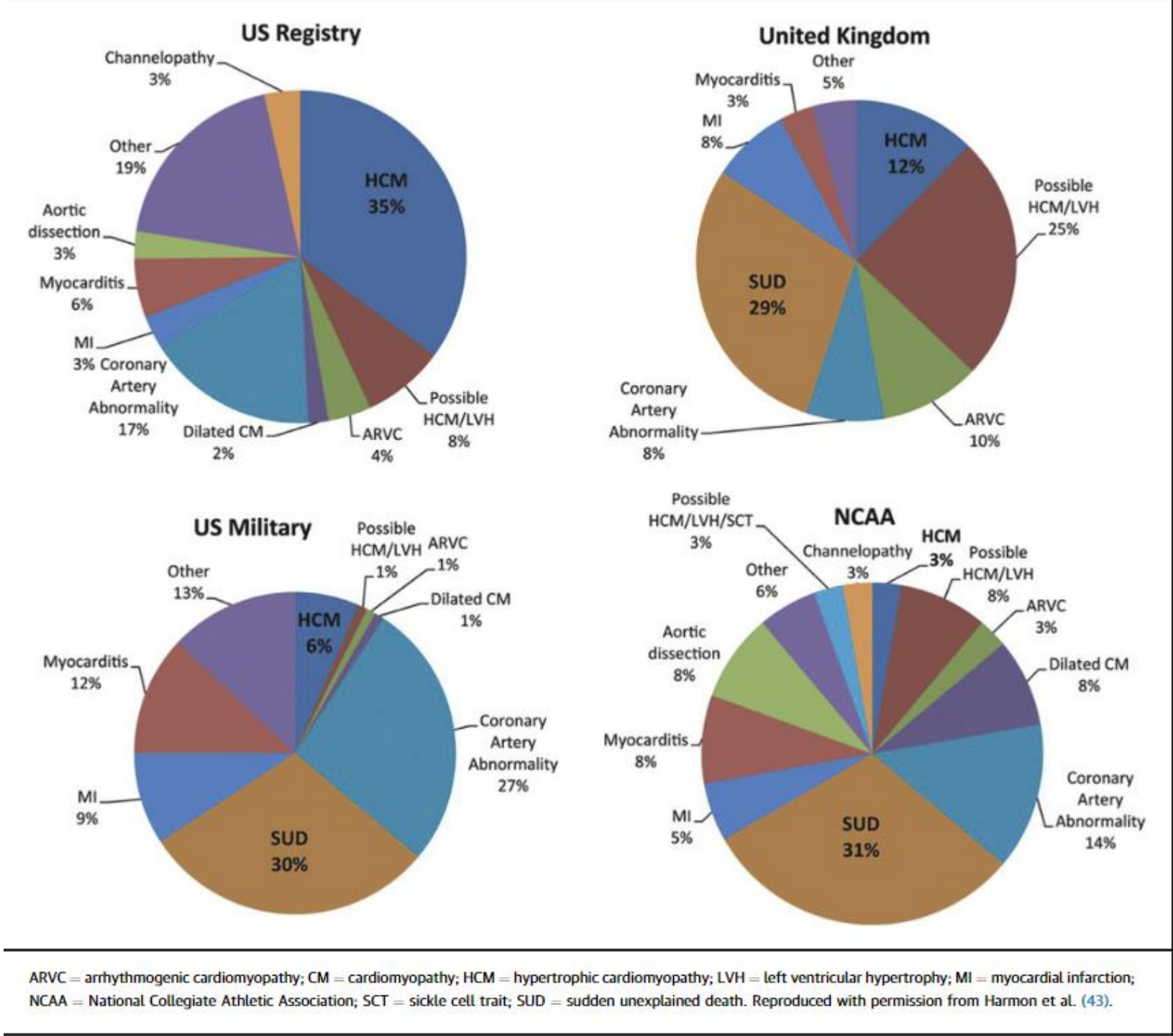
JACC VOL. 82, NO. 8, 2023 Martinez et al AUGUST 22, 2023:661–670

Hypertrophic cardiomyopathy is identified as the most common cause in several registries with rates anywhere from 8% - 50%

Coronary artery anomalies are the second most common in the structural group

Electrical abnormalities such as Long QT is also variable but newer data suggests rates are <5%

Myocarditis is reported at 3-12%

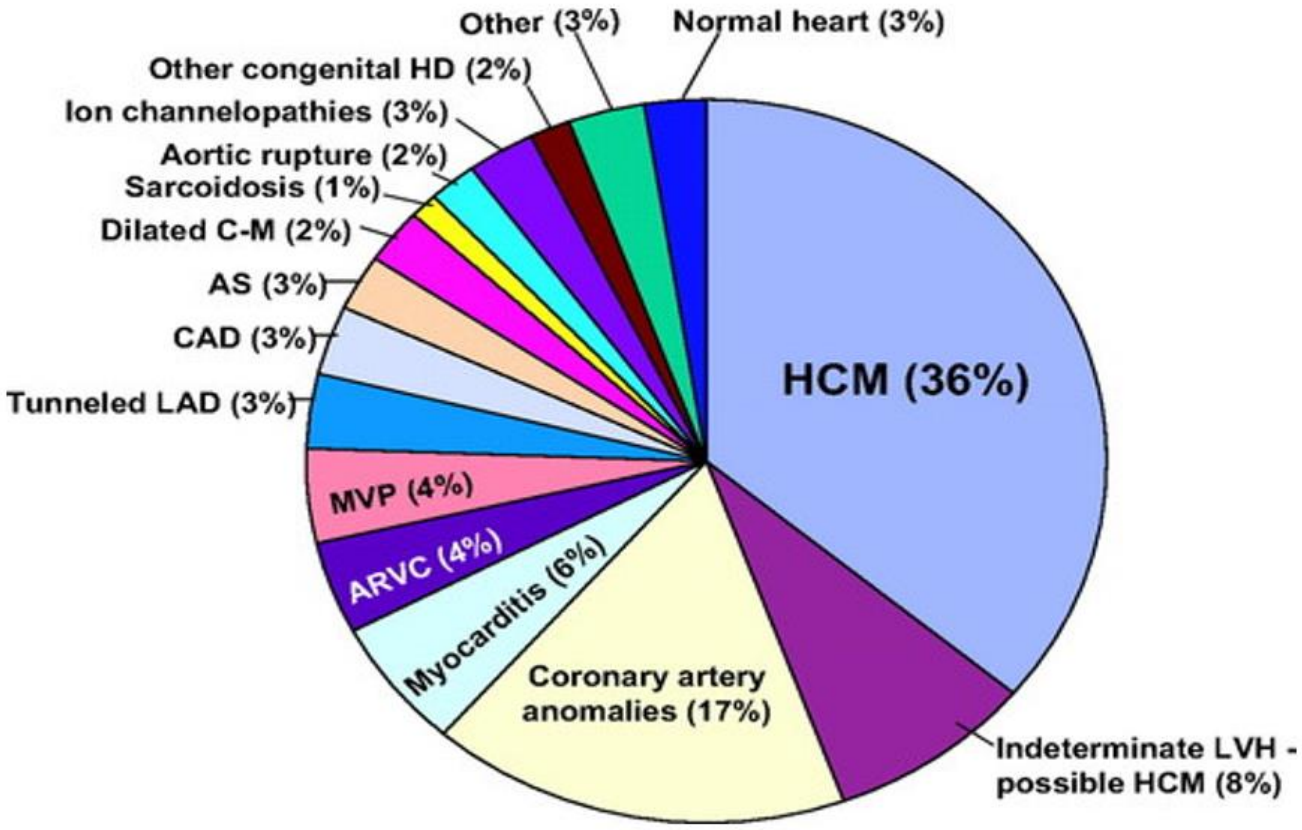


ARVC = arrhythmogenic cardiomyopathy; CM = cardiomyopathy; HCM = hypertrophic cardiomyopathy; LVH = left ventricular hypertrophy; MI = myocardial infarction; NCAA = National Collegiate Athletic Association; SCT = sickle cell trait; SUD = sudden unexplained death. Reproduced with permission from Harmon et al. (43).

JACC: HEART FAILURE VOL. 6, NO. 1, 2018Sudden Cardiac Death in Athletes JANUARY 2018:30–40

Causes of SCD

- In the US the most common causes (<35 years old) are:
  - Hypertrophic cardiomyopathy
  - Coronary artery anomalies
  - Myocarditis
- Ventricular arrhythmias seen in 78% of patients with myocarditis
- Can also present as acute myocardial infarction-like syndrome
- Some cases result in SCD without antecedent symptoms or macroscopic abnormalities



Barry J. Maron. Circulation. 2007 Update, Volume: 115, Issue: 12, Pages: 1643-1655,
Peretto G, Sala S, Rizzo S, et al. J Am Coll Cardiol. 2020;75:1046-1057

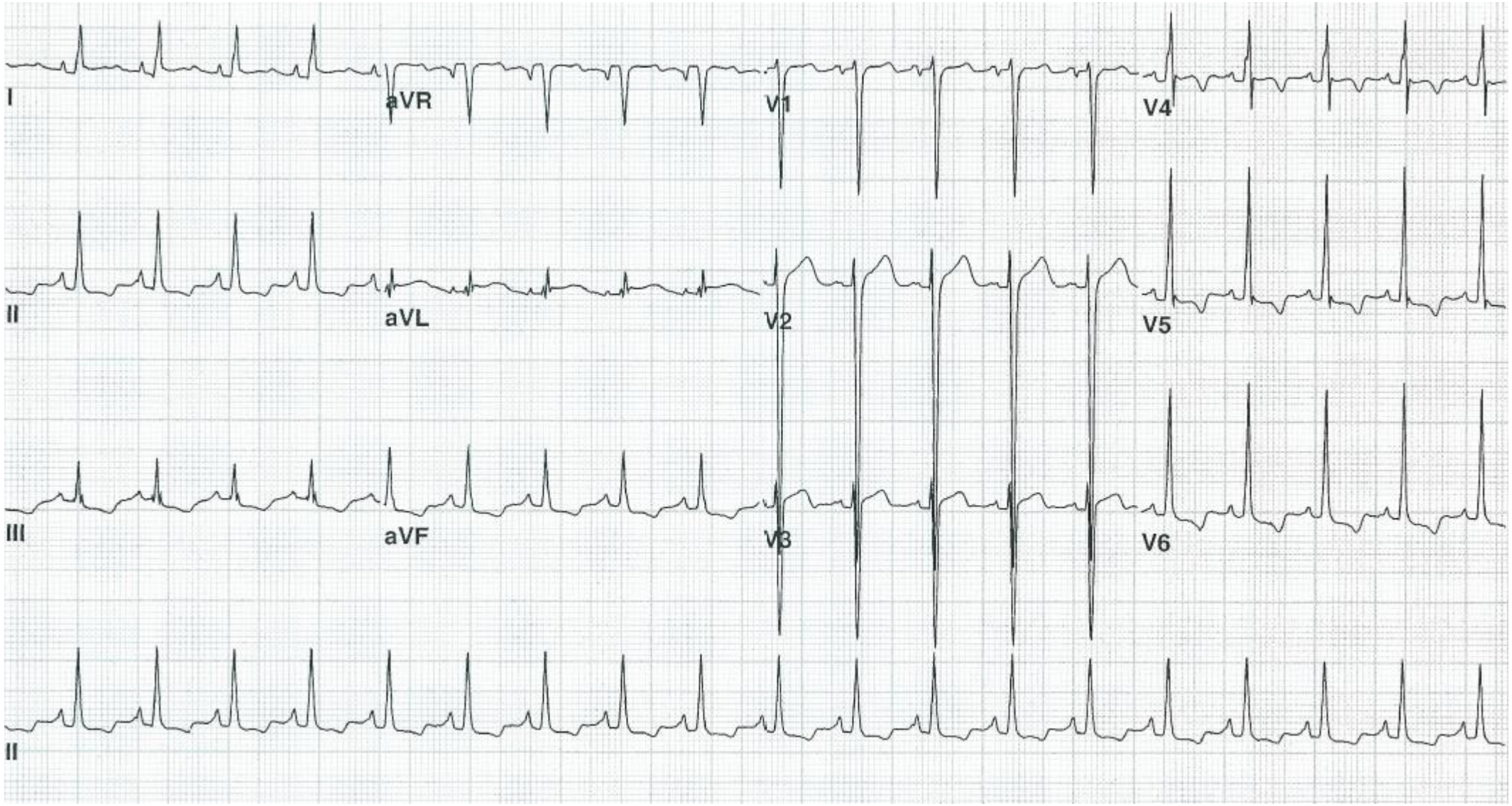
Structural heart disease

- Hypertrophic cardiomyopathy
- Coronary artery anomalies
- LVOT obstruction (Aortic stenosis and subaortic stenosis)
- Marfan Syndrome
- Dilated cardiomyopathy

Hypertrophic cardiomyopathy

- Most common causes for SCD in adolescents and young athletes
- Autosomal dominant inheritance in most cases (Family Hx)
- Newer studies show high prevalence in adults up to 0.5% (1/200)
- History:
  - Symptoms are typically during exercise from LVOT obstruction or arrhythmia
  - May have associated chest pain symptoms but many are asymptomatic
- Examination: Murmur – Harsh crescendo decrescendo systolic murmur at the LLSB- Apex. Increases in intensity with standing or Valsalva, decreases with squatting.
- ECG – Left axis deviation, LVH, left atrial enlargement, LV strain pattern

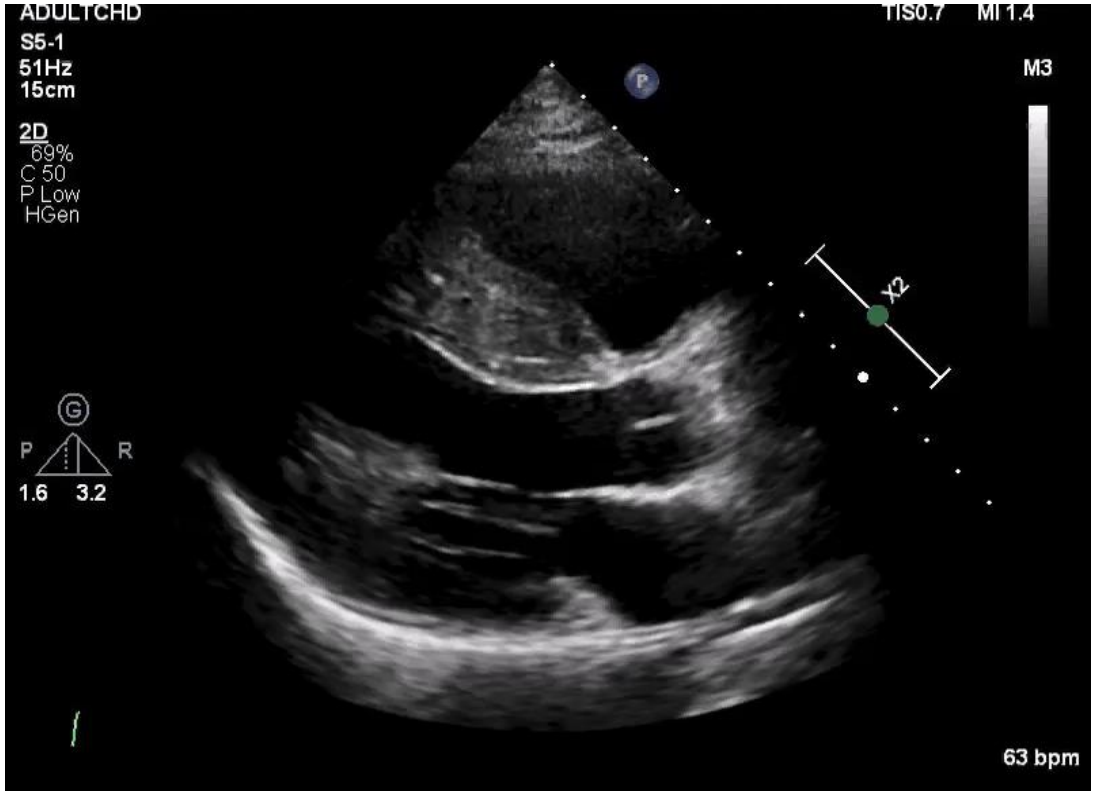
Semsarian C, JACC. 2015 Mar;65(12):1249-54. Maron BJ, JAMA 1996;276:199-204.

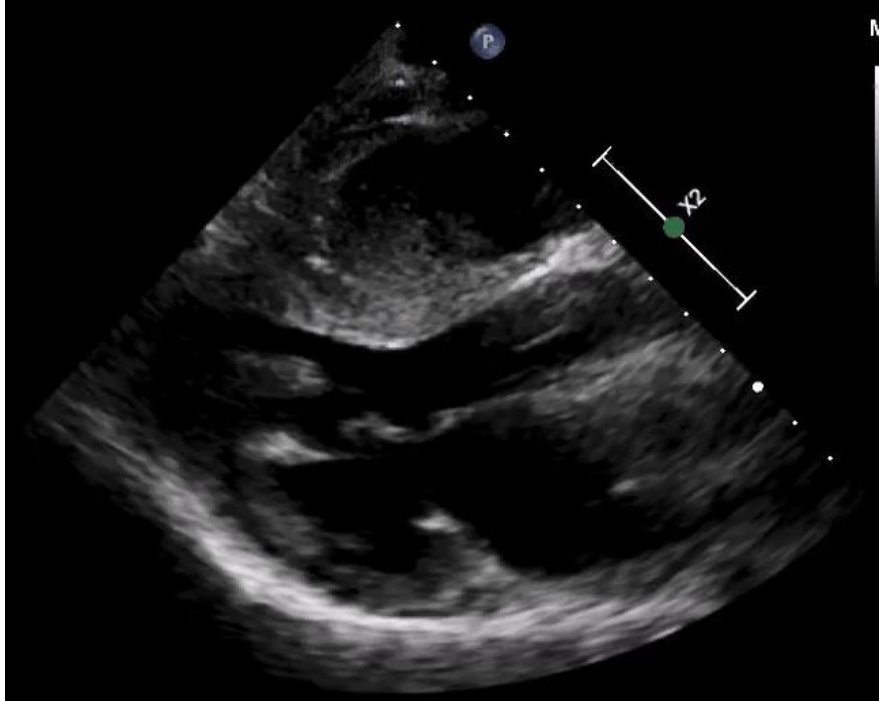

I
aVR
V1
V4
II
aVL
V2
V5
III
aVF
V3
V6
II

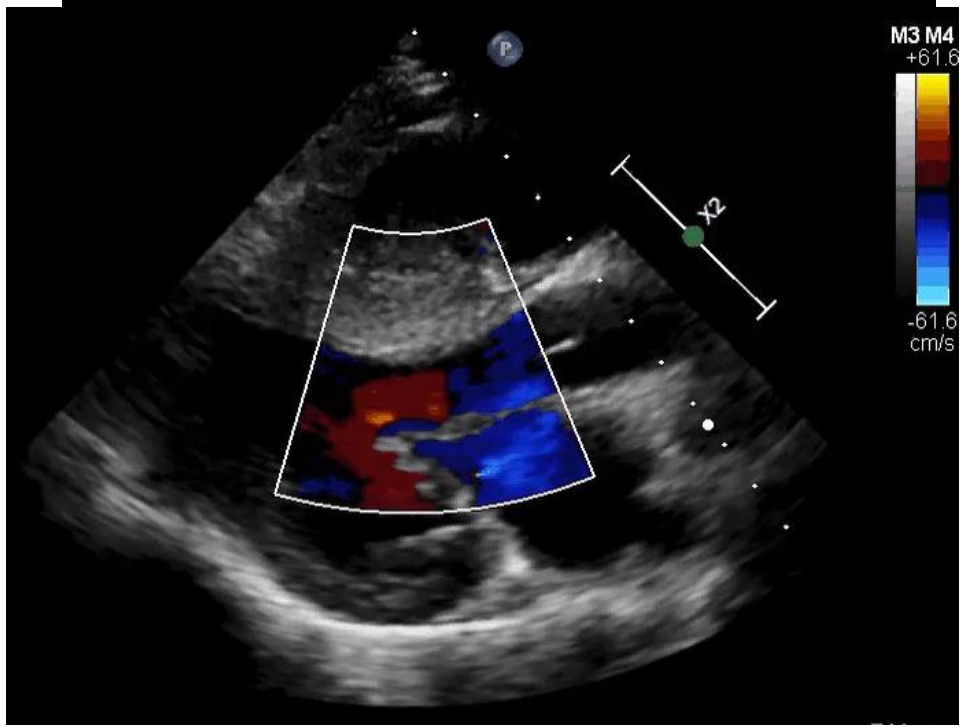
Hypertrophic cardiomyopathy

- Defined by the presence of a hypertrophied, nondilated ventricle in the absence of a hemodynamic cause
  - Histopathology – myocardial disarray
- Maximum diastolic septal or LV free wall thickness
  - Adults use 15 mm as threshold
  - Adjust for body size in children (z score)
- Ventricular systolic function generally normal or hyperdynamic
- LVOT obstruction (≥30 mmHg) is present at rest or with provocation in 75% of patients with HCM
- Systolic anterior motion of the MV and mitral regurgitation
- Diastolic dysfunction
- Myocardial ischemia
- Arrhythmias
- Autonomic dysfunction (abnormal BP response to exercise)

Semsarian C, JACC. 2015 Mar;65(12):1249-54. Maron BJ, JAMA 1996;276:199-204.


ADULTCHD
S5-1
51Hz
15cm
2D
69%
C 50
P Low
HGen
TIS0.7 MI1.4
M3
X2
P
R
1.6 3.2
63 bpm


X2
P


M3 M4
+61.6
-61.6
cm/s
X2
P

Hypertrophic cardiomyopathy

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TABLE 7 Established Clinical Risk Factors for HCM Sudden Death Risk Stratification

Family history of sudden death from HCM	Sudden death judged definitively or likely attributable to HCM in ≥1 first-degree or close relatives who are ≤50 years of age. Close relatives would generally be second-degree relatives; however, multiple SCDs in tertiary relatives should also be considered relevant.
Massive LVH	Wall thickness ≥30 mm in any segment within the chamber by echocardiography or CMR imaging; consideration for this morphologic marker is also given to borderline values of ≥28 mm in individual patients at the discretion of the treating cardiologist. For pediatric patients with HCM, an absolute or z-score threshold for wall thickness has not been established; however, a maximal wall that corresponds to a z-score ≥20 (and >10 in conjunction with other risk factors) appears reasonable.
Unexplained syncope	≥1 Unexplained episodes involving acute transient loss of consciousness, judged by history unlikely to be of neurocardiogenic (vasovagal) etiology, nor attributable to LVOTO, and especially when occurring within 6 months of evaluation (events beyond 5 years in the past do not appear to have relevance).
HCM with LV systolic dysfunction	Systolic dysfunction with EF <50% by echocardiography or CMR imaging.
LV apical aneurysm	Apical aneurysm defined as a discrete thin-walled dyskinetic or akinetic segment of the most distal portion of the LV chamber; independent of size.
Extensive LGE on CMR imaging	Diffuse and extensive LGE, representing fibrosis, either quantified or estimated by visual inspection, comprising ≥15% of LV mass (extent of LGE conferring risk has not been established in children).
NSVT on ambulatory monitor	It would seem most appropriate to place greater weight on NSVT as a risk marker when runs are frequent (≥3), longer (≥10 beats), and faster (≥200 bpm) occurring usually over 24 to 48 hours of monitoring. For pediatric patients, a VT rate that exceeds the baseline sinus rate by >20% is considered significant.

CMR indicates cardiovascular magnetic resonance; ICD, implantable cardioverter-defibrillator; LGE, late gadolinium enhancement; LV, left ventricular; LVH, left ventricular hypertrophy; LVOTO, left ventricular outflow tract obstruction; NSVT, nonsustained ventricular tachycardia; and SCD, sudden cardiac death.

2020 AHA/ACC Guideline for the Diagnosis and Treatment of Patients with Hypertrophic Cardiomyopathy. JACC. 76(25). 2020

Hypertrophic cardiomyopathy

Table 9. Survival Rate From Time of Diagnosis of HCM by Pathogenesis

Pathogenesis	Survival Since Diagnosis of HCM, %*			
	1 y	2 y	5 y	10 y
Inborn error of metabolism	53.6	44.9	41.7	…
Malformation syndrome	82.4	76.6	74.4	74.4
Neuromuscular disease	98.2	98.2	98.2	91.7
Idiopathic disease	94.4	92.8	89.8	85.3
Infantile idiopathic disease	85.8	84.3	82.2	82.2
Noninfantile idiopathic disease	99.2	97.6	93.9	85.9

HCM indicates hypertrophic cardiomyopathy.
*Maximum follow-up was only 9 years.
Reprinted from Colan et al.[9] Copyright © 2007, American Heart Association, Inc.

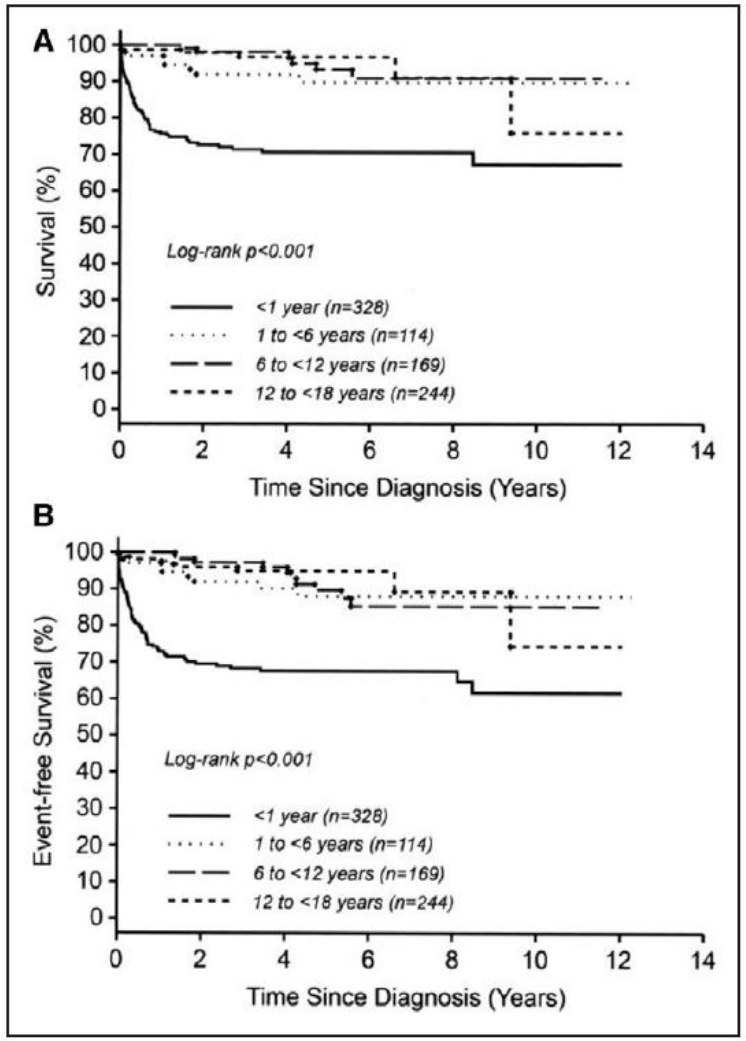
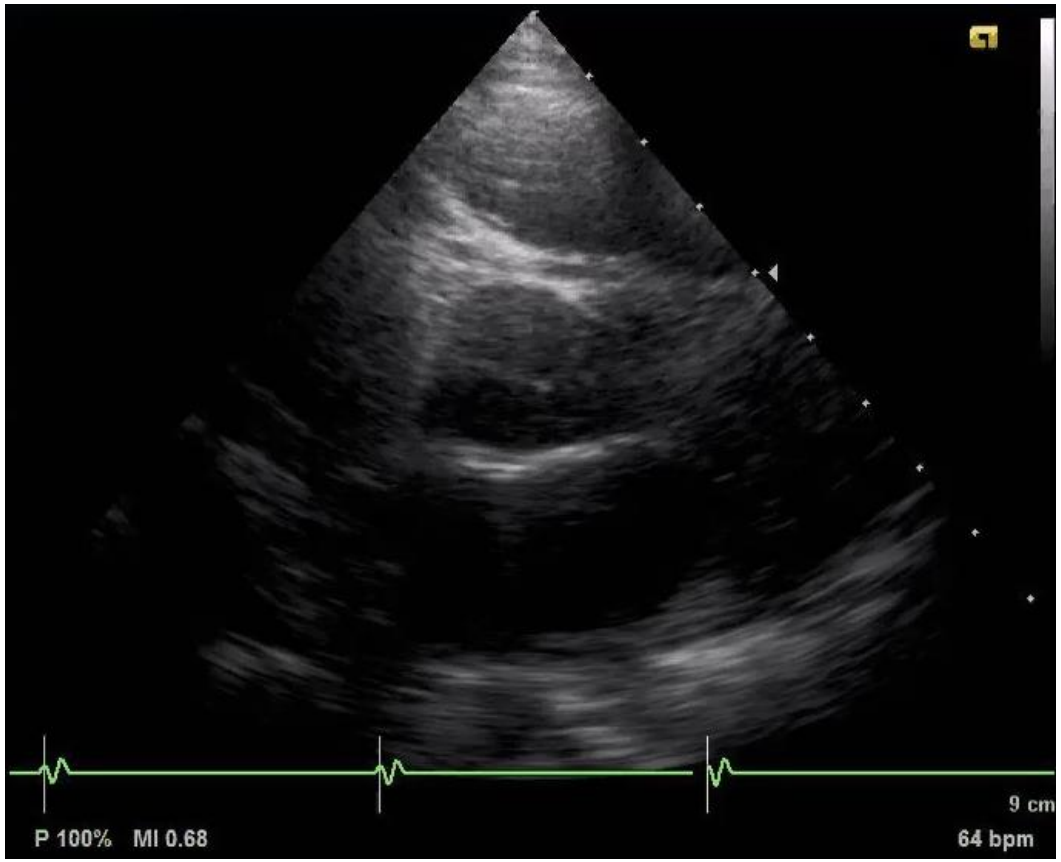


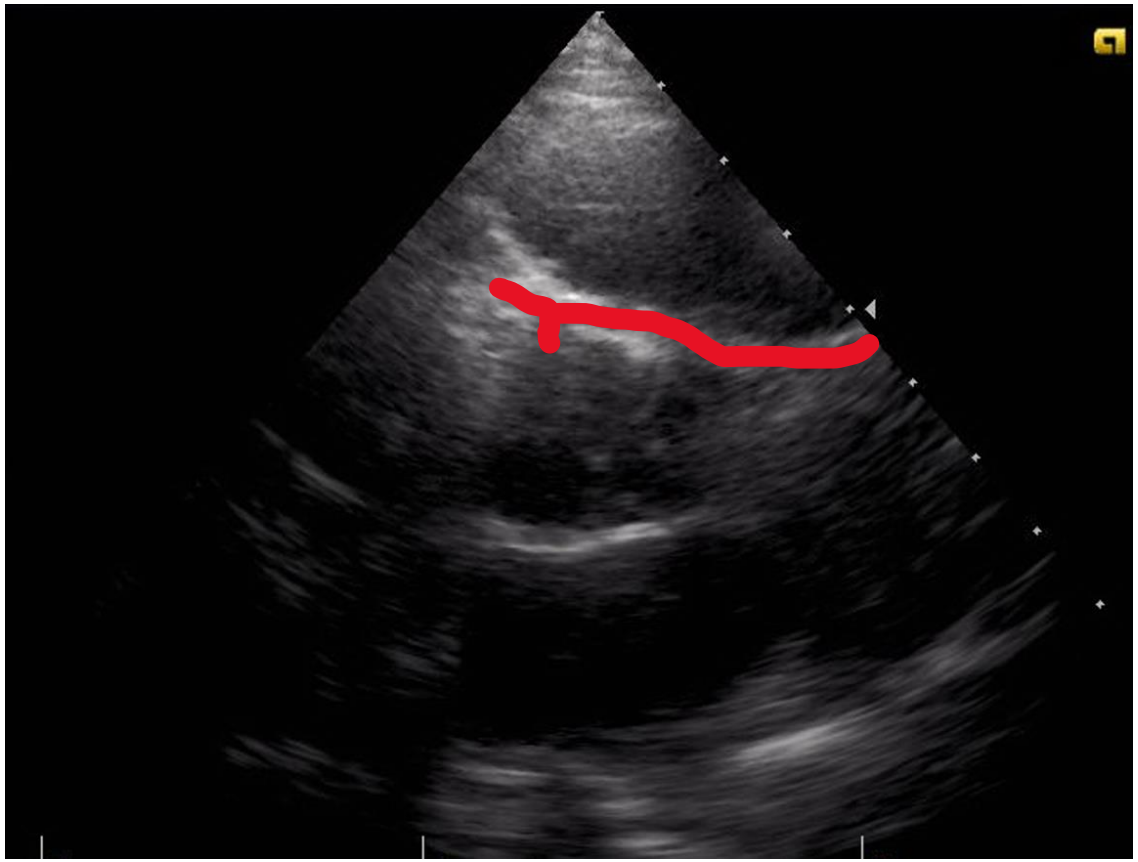
Figure 4. Survival rates from diagnosis to (A) death resulting from cardiomyopathy and to (B) death or transplantation by age at diagnosis. Reprinted from Colan et al.[9] Copyright © 2007, American Heart Association, Inc.

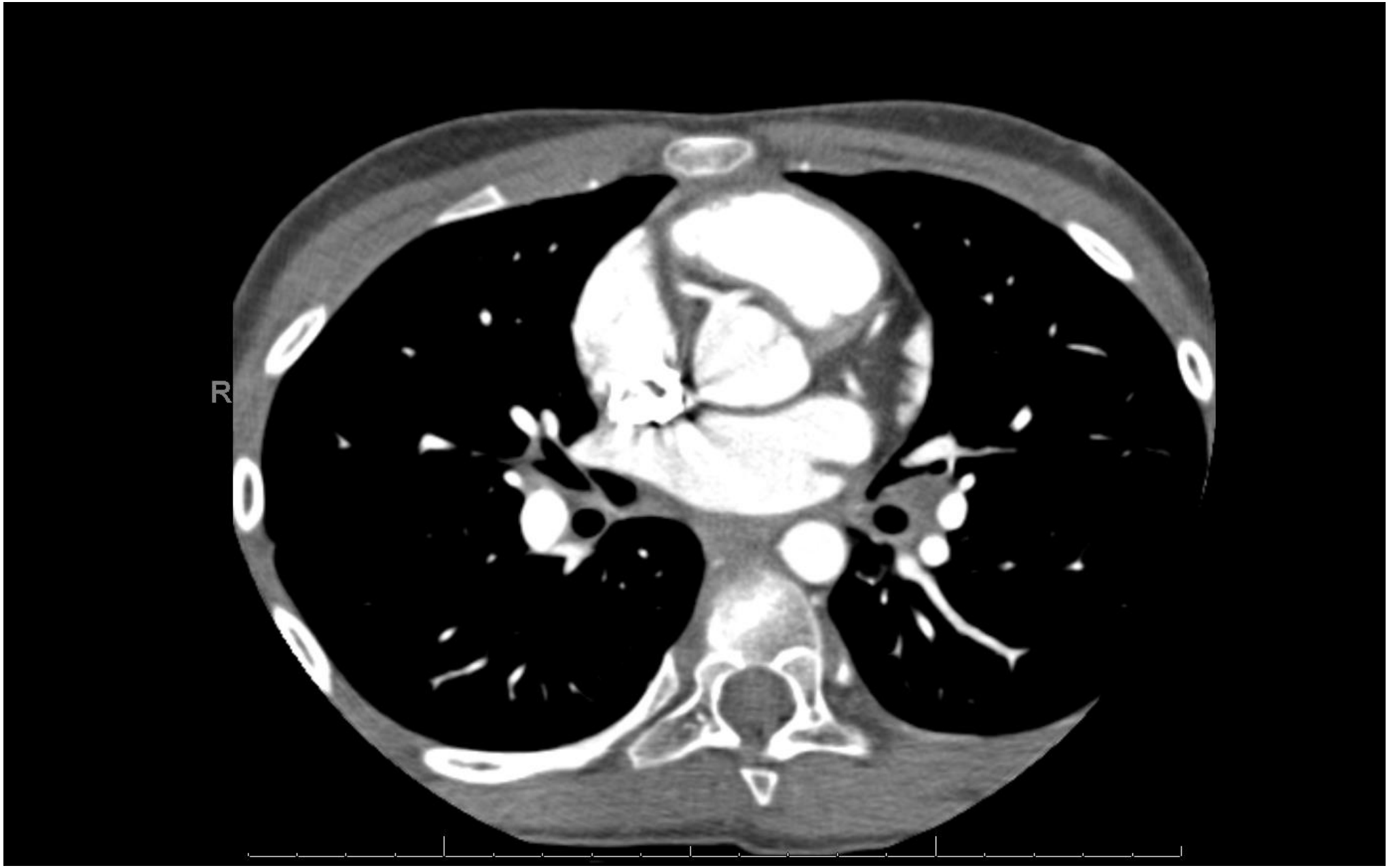
Coronary artery anomalies

- There are a variety of coronary anomalies that can cause exertional syncope or sudden cardiac death
- Multiple variants but generally anomalous Left coronary artery with inter-arterial course is most concerning.
  - Anomalous Right coronary artery – most often benign, incidence 0.23%
  - Anomalous Left coronary artery – often lethal but far more rate, incidence 0.03%
- Only one-third of patients with this anomaly who died suddenly reported prior symptoms of angina or exertional syncope
- History is typically positive
  - exertional chest pain (red flag) – Sudden onset, angina type pain.
  - exertional syncope (red flag) – Sudden syncope no prodrome, no post event symptoms
- Physical exam and ECG at rest are normal

Moss and Adams' Heart Disease in Infants, Children and Adolescents, 7th Edition, 2008.


9 cm
P 100% MI 0.68
64 bpm

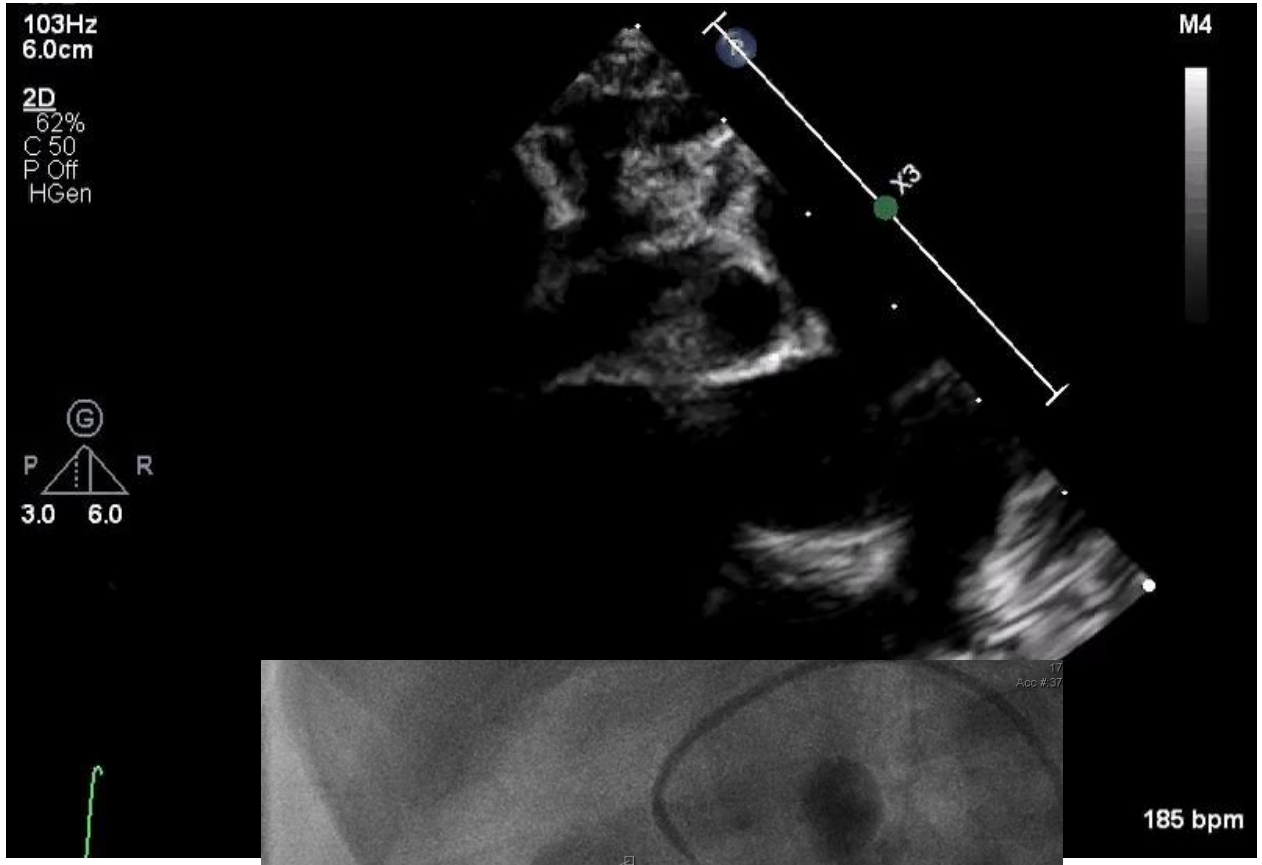


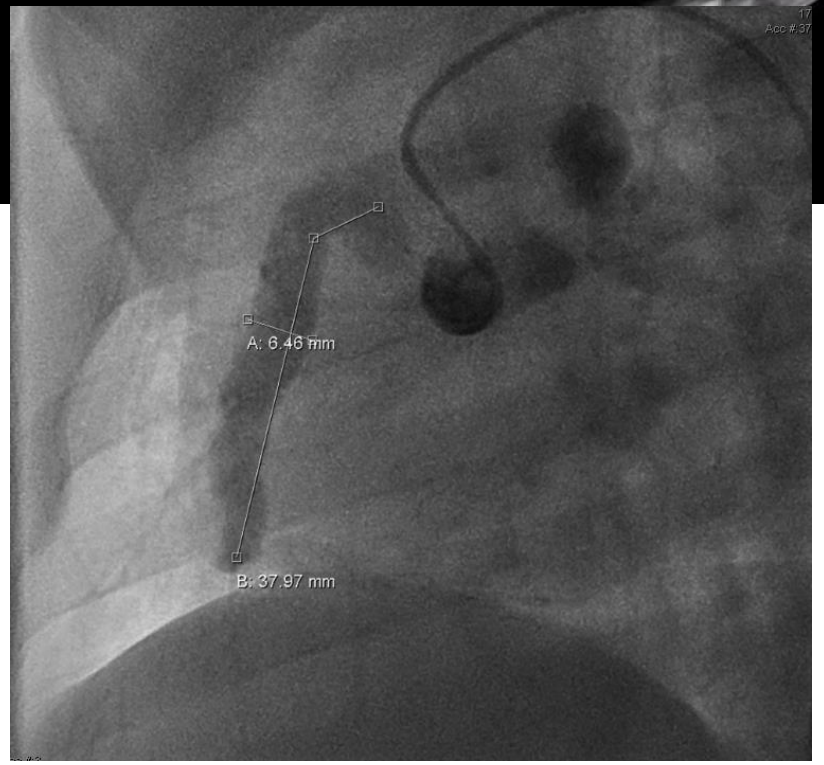

R

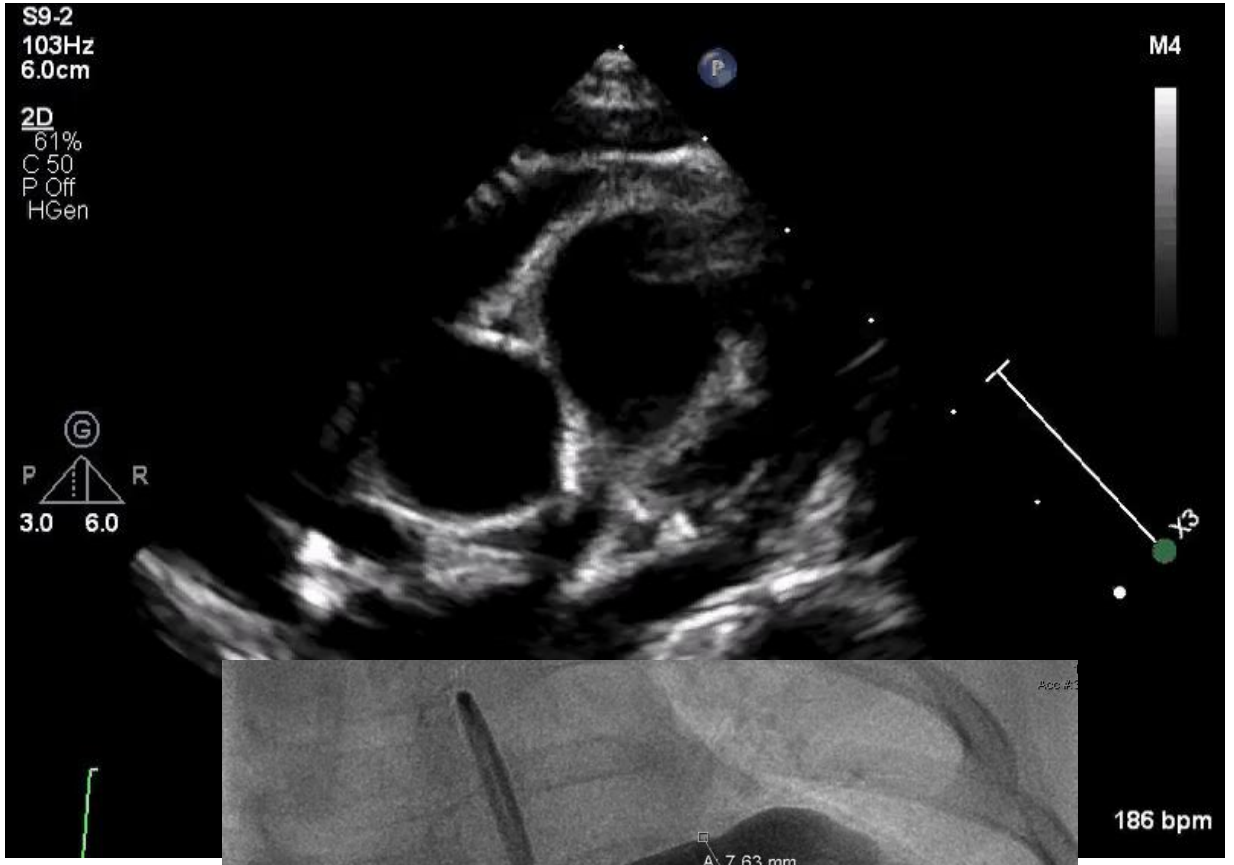
Kawasaki Disease

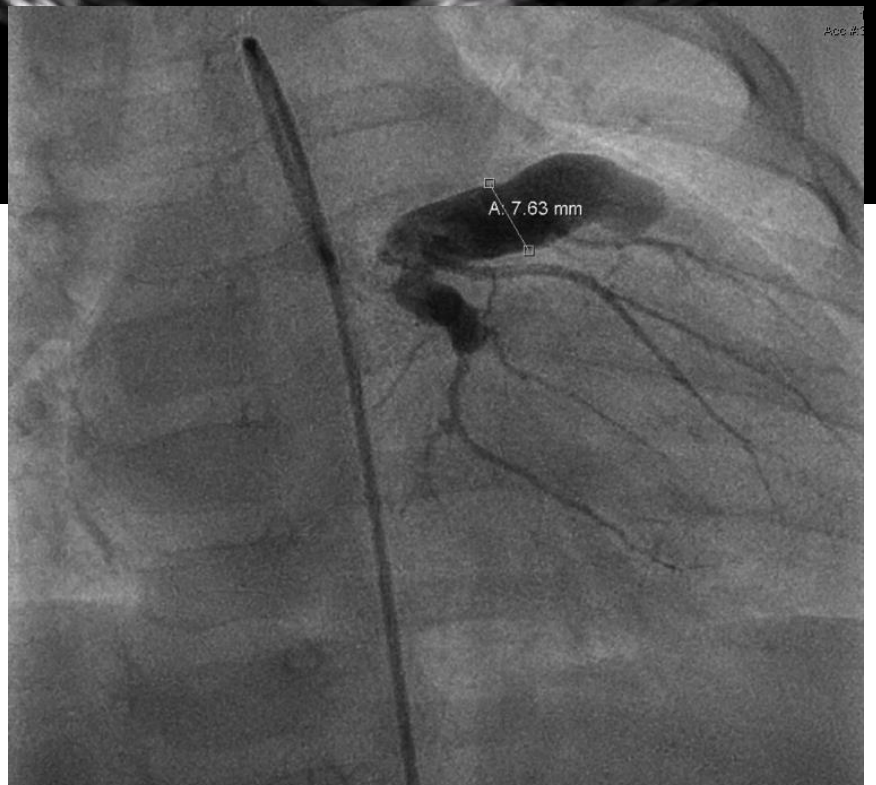
- Kawasaki disease is the most common inflammatory vasculitis in children
- Incidence of Giant coronary artery aneurysms vary by ethnicity
  - Highest among Hispanics (5.9%)
  - Lowest among Asians (1.8%)
- Early reports: SCD reported early in the course of Kawasaki disease in 2% of patients
  - Significant improvement in outcomes since IVIG: 0.17%
  - Late deaths generally in those with persistent bilateral giant aneurysms.
    - Sustained VT at the site of a prior myocardial infarction
- History is typically positive though not all events may be exertional
- Physical examination is often normal
- ECG may show signs of ischemia

Holman RC, Pediatr Infect Dis J. 2010;29(6):483 Newburger JW, N Engl J Med 1986;315:341-347. Son MB, Pediatrics.2009;124(1):1.


103Hz
6.0cm
2D
62%
C 50
P Off
HGen
M4
X3
P
R
3.0
6.0
185 bpm


A: 6.46 mm
B: 37.97 mm

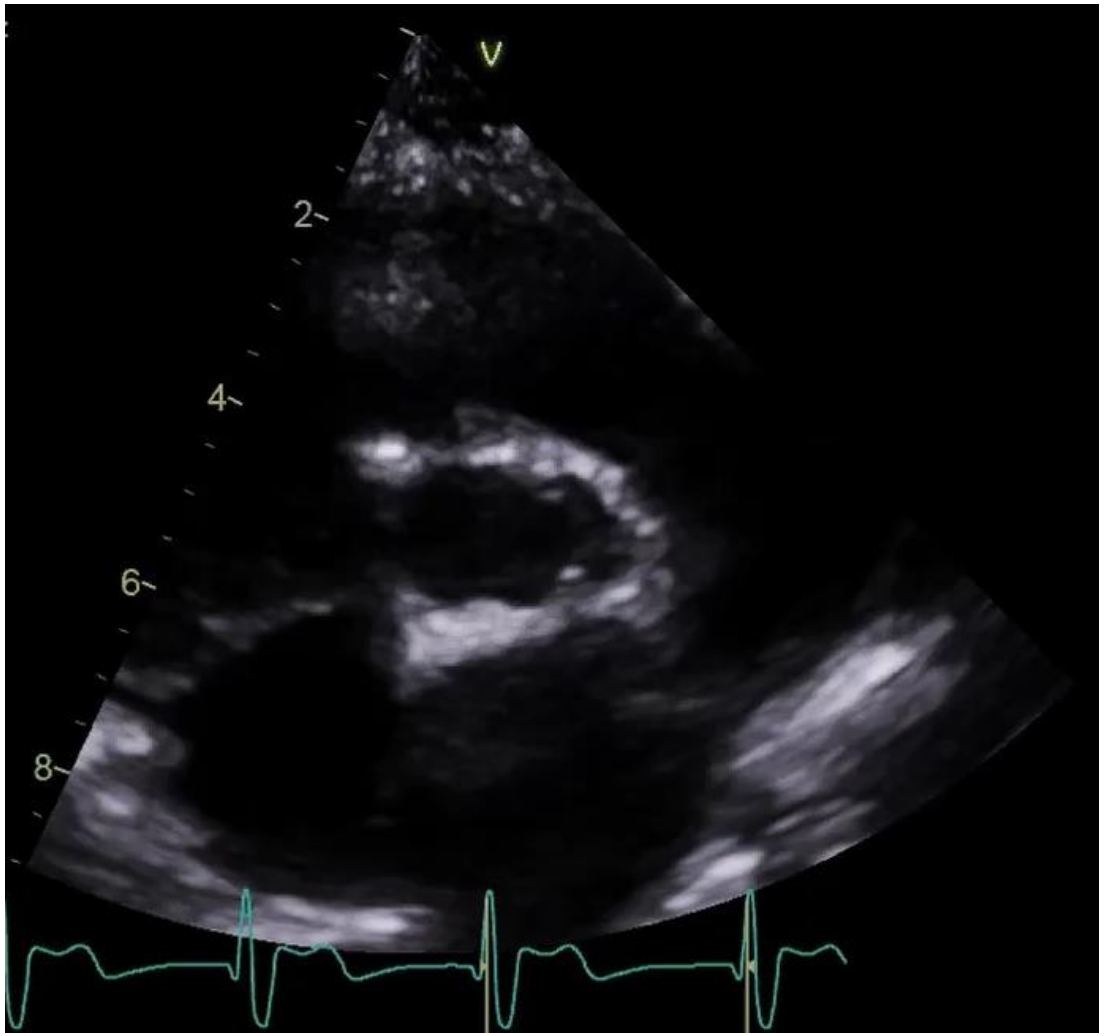

S9-2
103Hz
6.0cm
2D
61%
C 50
P Off
HGen
M4
X3
P
R
3.0
6.0
186 bpm

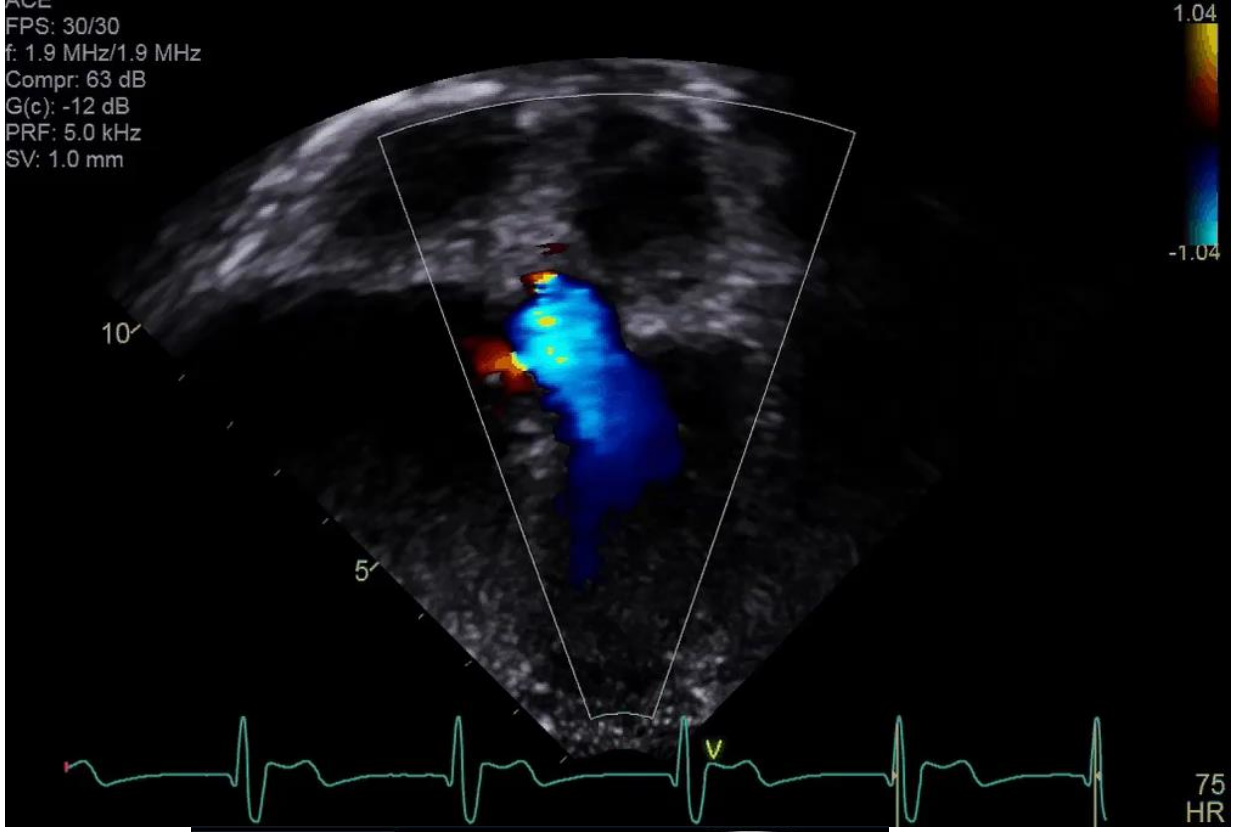

A: 7.63 mm

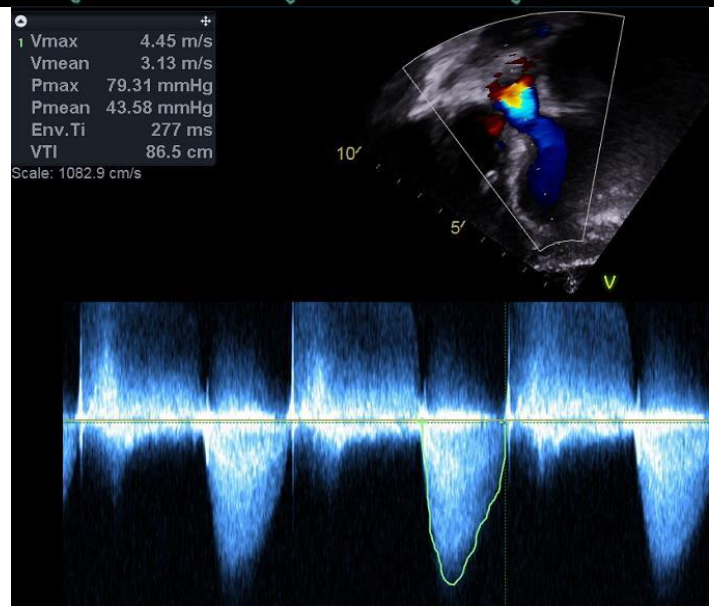
Congenital heart disease – LVOT Obstruction

- Bicuspid/Bicommisural aortic valve is very common estimated at 0.5-2% of the population! More common in males
- History positive for exertional chest pain or exertional syncope.
  - Mechanism is ischemia with exertion.
- Prominent PMI, ejection click, Loud harsh crescendo-decrescendo murmur at RUSB. May radiate to the neck. Palpable thrill in precordium or neck
- ECG: LV strain, Left axis deviation, ST changes

Circulation. 2005;111(7):920. Cardiol Young. 1999;9(5):488 Am Heart J. 2005;150(3):513.

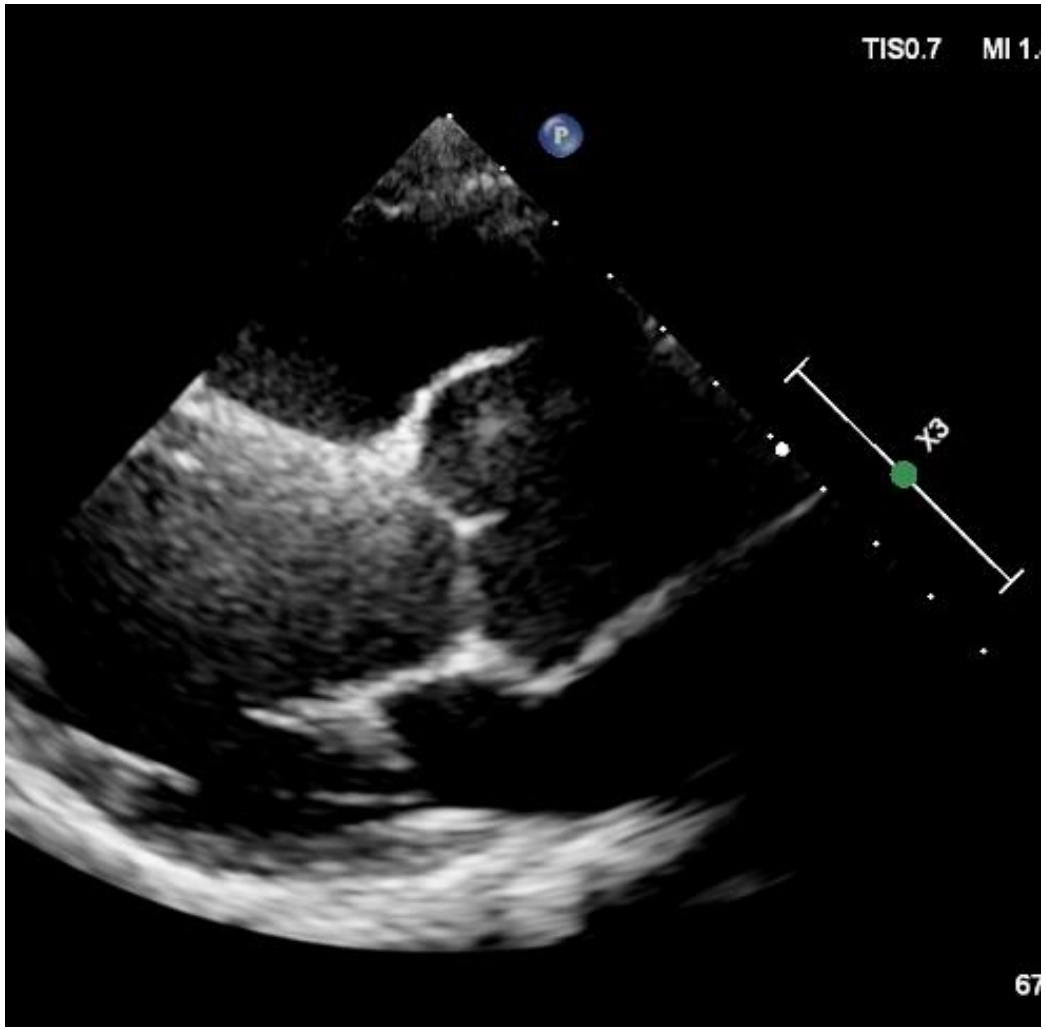

V
2
4
6
8


ACE
FPS: 30/30
f: 1.9 MHz/1.9 MHz
Compr: 63 dB
G(c): -12 dB
PRF: 5.0 kHz
SV: 1.0 mm
1.04
-1.04
10
5
V
75
HR


Vmax 4.45 m/s
Vmean 3.13 m/s
Pmax 79.31 mmHg
Pmean 43.58 mmHg
Env.Ti 277 ms
VTI 86.5 cm
Scale: 1082.9 cm/s
10
5
V

Marfan Syndrome

- Most common inherited connective tissue disorder
  - incidence 1 in 3000
- Manifestations
  - Arachnodactyly, pectus deformity, hindfoot valgus, reduced upper segment/lower segment ratio, scoliosis/kyphosis, ectopia lentis, protrusion acetabuli, dural ectasia, facial features
  - Cardiac – Aortic dilation, mitral prolapse
  - SCA is from dissection



Am J Cardiol . 1989 Feb 1;63(5):317-21. doi: 10.1016/0002-9149(89)90338-x. PMID: 2913733
Br Heart J . 1984 Feb;51(2):220-30. doi: 10.1136/hrt.51.2.220. PMID: 6691872

Marfan Syndrome

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Systemic Score

Click one or more physical characteristics to Include in the Calculation

Wrist AND Thumb Sign (+3)	Wrist OR Thumb Sign (+1)
Pectus Carinatum Deformity (+2)	Pectus Excavatum or Chest Asymmetry (+1)
Hindfoot Deformity (+2)	Plain Flat Foot (+1)
Spontaneous Pneumothorax (+2)	Dural Ectasia(+2)
Protucio Acetabulae (+2)	Scoliosis or Thoracolumbar Kyphosis (+1)
Reduced Elbow Extension (+1)	3 of 5 Facial Features (+1)
Skin Striae (+1)	Severe Myopia (+1)
Mitral Valve Prolapse (+1)	

Reduced Upper Segment / Lower Segment & Increased Arm span / Height (+1)

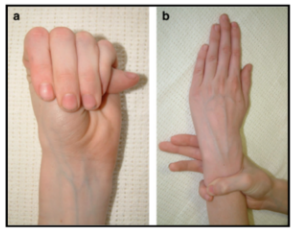
Patient is:

○ Caucasion
○ Black

Height (cm)

To learn more about each physical characteristic, click on the accordion links below:

1. Wrist and/or thumb sign



The thumb sign is positive when the entire distal phalanx of the adducted thumb extends beyond the ulnar border of the palm with or without the assistance of the patient or examiner to achieve maximal adduction. The wrist sign is positive when the tip of the thumb covers the entire fingernail of the fifth finger when wrapped around the contralateral wrist.

2. Anterior chest wall deformity

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Spontaneous Pneumothorax (+2)	Dural Ectasia(+2)
Protucio Acetabulae (+2)	Scoliosis or Thoracolumbar Kyphosis (+1)
Reduced Elbow Extension (+1)	3 of 5 Facial Features (+1)
Skin Striae (+1)	Severe Myopia (+1)
Mitral Valve Prolapse (+1)	

Reduced Upper Segment / Lower Segment & Increased Arm span / Height (+1)

Patient is:

○ Caucasion
○ Black

Height (cm)

Arm span (cm):

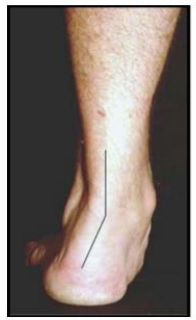
Lower segment (cm):

Results

Upper/Lower segment ratio:

3. Hindfoot deformity

Hindfoot valgus in combination with forefoot abduction and lowering of the midfoot (previously referred to as medial rotation of the medial malleolus) should be evaluated from anterior and posterior view. The examiner should distinguish this from the more common "flat foot" (one point) without significant hindfoot valgus.

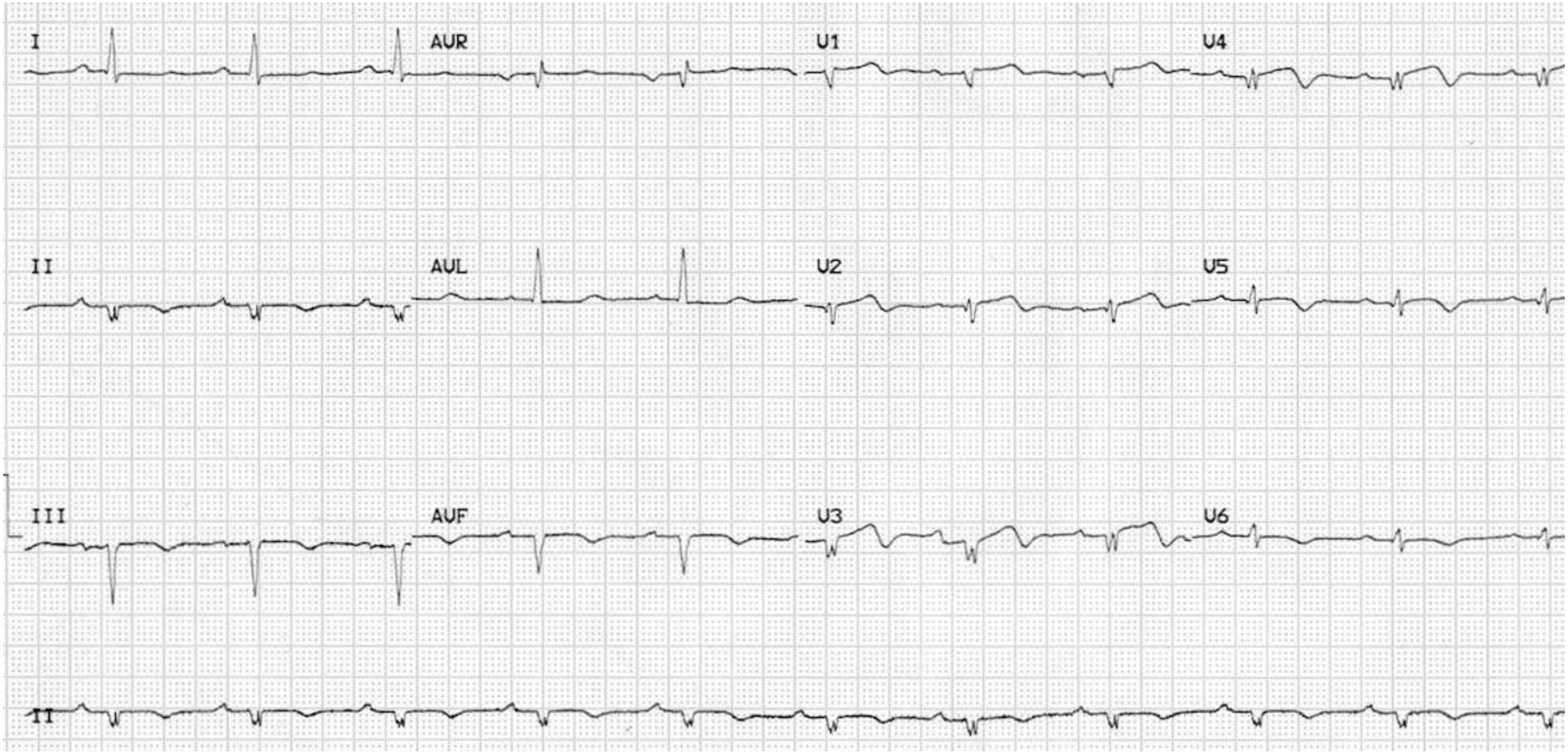


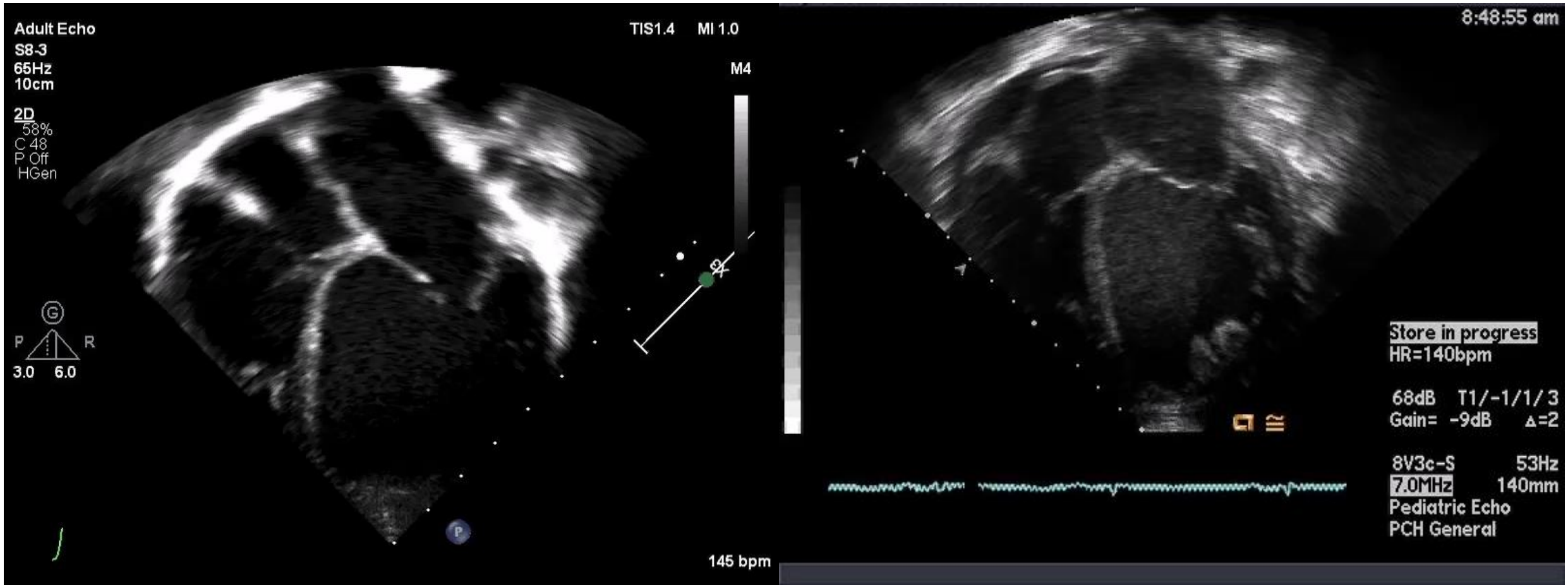
4. pneumothorax

Marfan.org/dx/score/

Dilated cardiomyopathy

- Symptoms correlate to degree of myocardial dysfunction
- Syncope/SCA often exertional from ischemia or arrhythmia
- Dyspnea, decreased exercise tolerance are most common symptoms
- Family history often positive
- Exam – Tachycardia/Tachypnea, laterally and downward displaced PMI, Mitral regurgitation murmur (high pitched, holosystolic, blowing murmur at apex with radiation to axillae)S3/S4 gallop, JVD, hepatomegaly
- ECG – very high or low QRS voltages, Left axis deviation, left atrial enlargement, LVH, LV strain, ST changes


I
AUR
U1
U4
II
AUL
U2
U5
III
AUF
U3
U6
II


Adult Echo
S8-3
65Hz
10cm
2D
58%
C 48
P Off
HGen
TIS1.4 MI 1.0
M4
G
P
R
3.0 6.0
145 bpm
8:48:55 am
Store in progress
HR=140bpm
68dB T1/-1/1/3
Gain= -9dB Δ=2
8V3c-S 53Hz
7.0MHz 140mm
Pediatric Echo
PCH General

Electrical Abnormalities

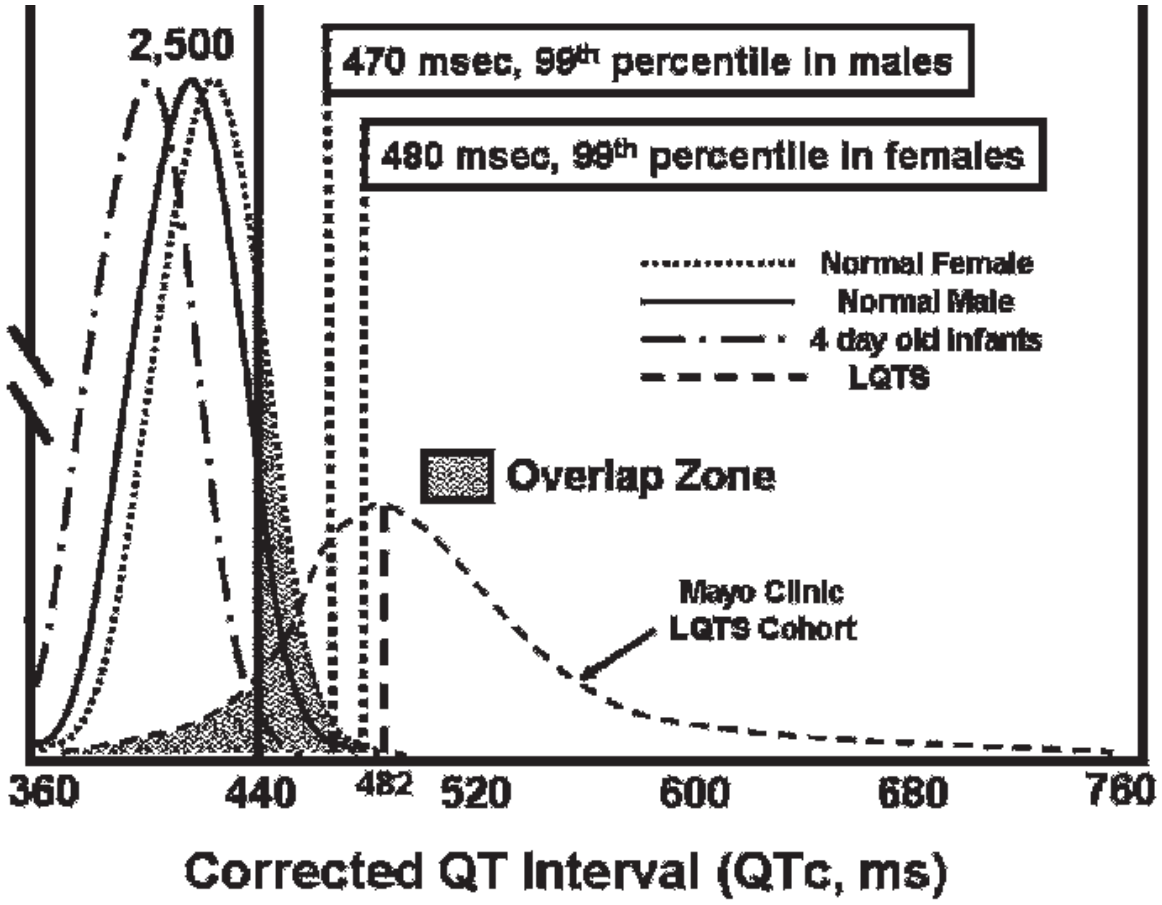
- Long QT syndrome
- Brugada syndrome
- Catecholiminergic Polymorphic Ventricular Tachycardia (CPVT)

Long QT Syndrome

- Prolongation of the QT interval associated with syncope, polymorphic VT (torsades de pointes) or SCD
  - Genetic but can be acquired from medications or electrolyte abnormalities
- Prevalence is as high as 1 in 2500
- Among those with known LQT about 1/3 have a cardiac event (syncope, cardiac arrest, SCD)
- In patients with symptomatic, untreated LQTS, the mortality rate is as high as 20% for the first year and 50% at 10 years.
- Varying forms of inheritance from autosomal dominant to recessive
  - AR form: Jervell and Lange-Nielsen Syndrome is associated with sensorineural hearing loss
  - Some LQT in Turner Syndrome (dissection more common)

Schwartz PJ, Circ. 2009 Nov;120(18):1761-7 Prioro SG, N Engl J Med 2003, 348:1866–74. Goel AK, Pediatr Cardiol 2004, 25:370–378.

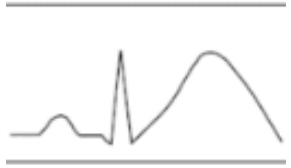
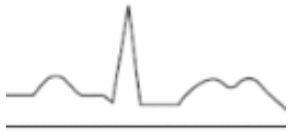
What is a normal QTc?
99%ile Males = 470 msec
99%ile Females = 480 msec
“Gray zone” above 450 msec
Why? - Mayo clinic LQT cohort
Mean QTc = 482 msec



Splawski I, Circ 2000,102:1178–1185.

Vincent GM, N Engl J Med 1992; 327:846.

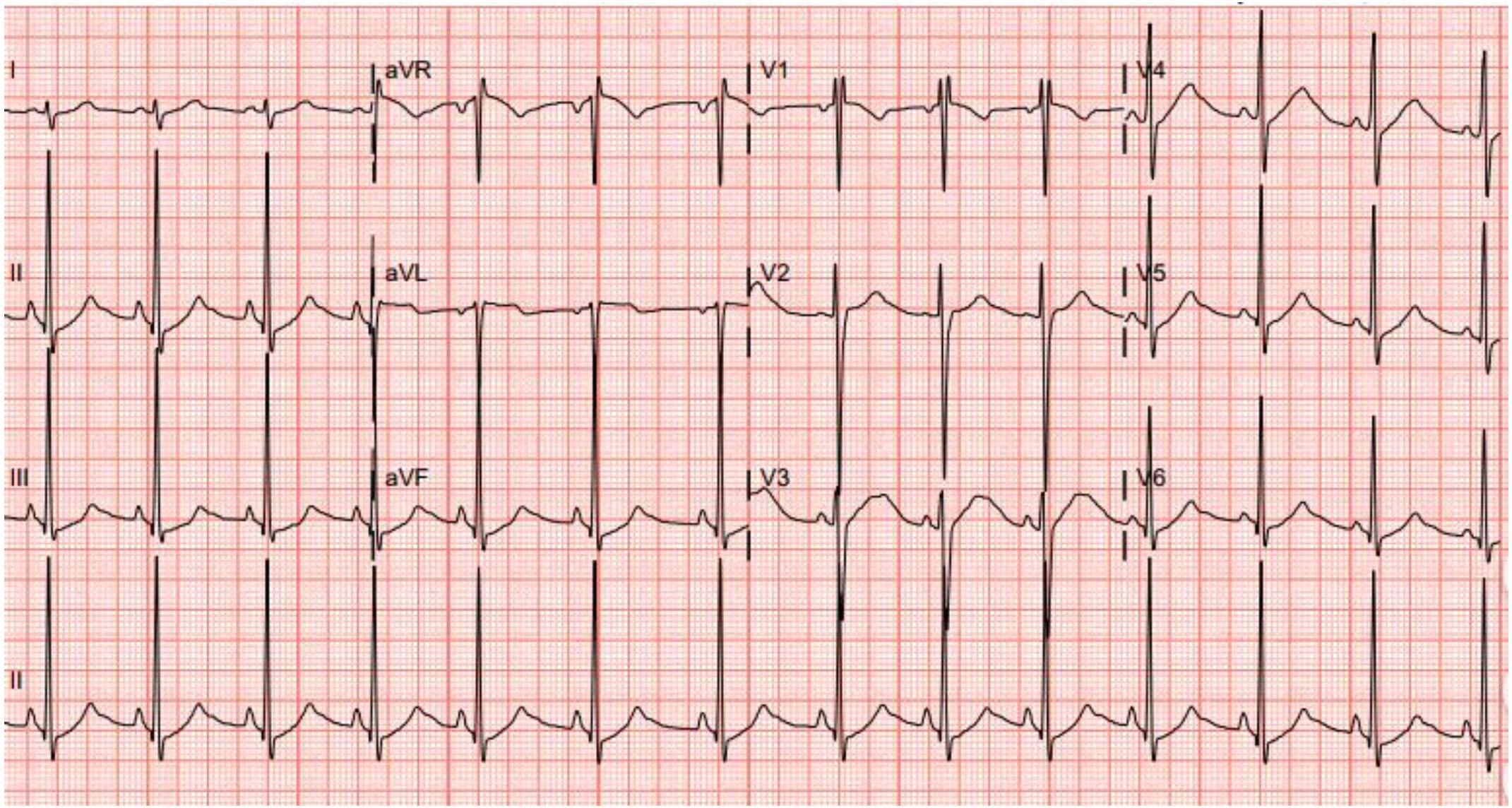
Long QT Subtypes

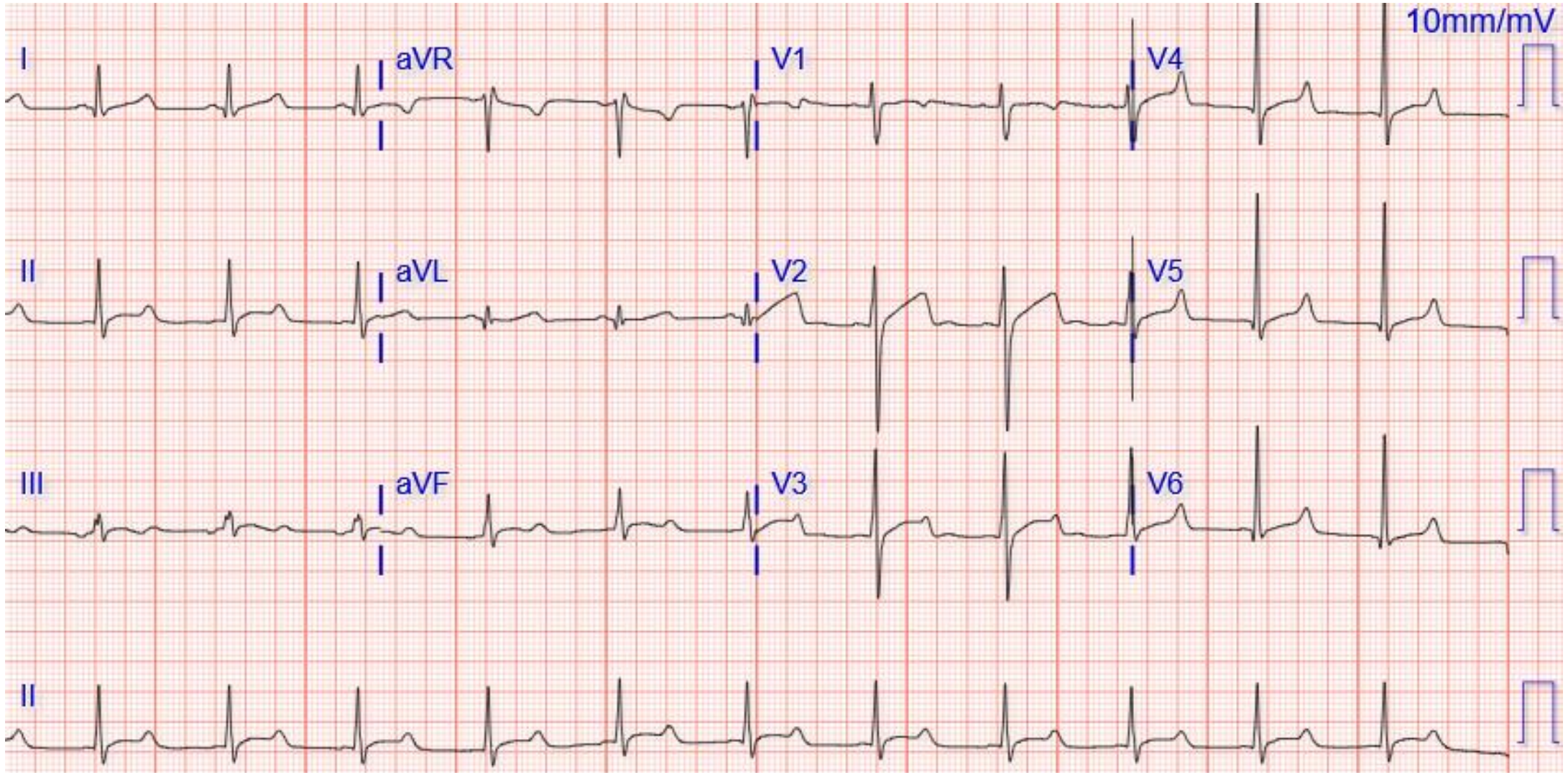
Subtype	%	Gene	Mutation		Manifestation
Long QT 1	**42%**	**KCNQ1**	**K+ Channel mutation**		**Events with exercise / swimming**
Long QT 2	**45%**	**HERG**	**K+ Channel mutation**		**Events with loud noises**
Long QT 3	**8%**	**SCN5A**	**Na+ Channel mutation**		**Events during sleep**

SCN5A mutations
Gain of function causes long QT syndrome (LQT 3)
Loss of function causes Brugada Syndrome

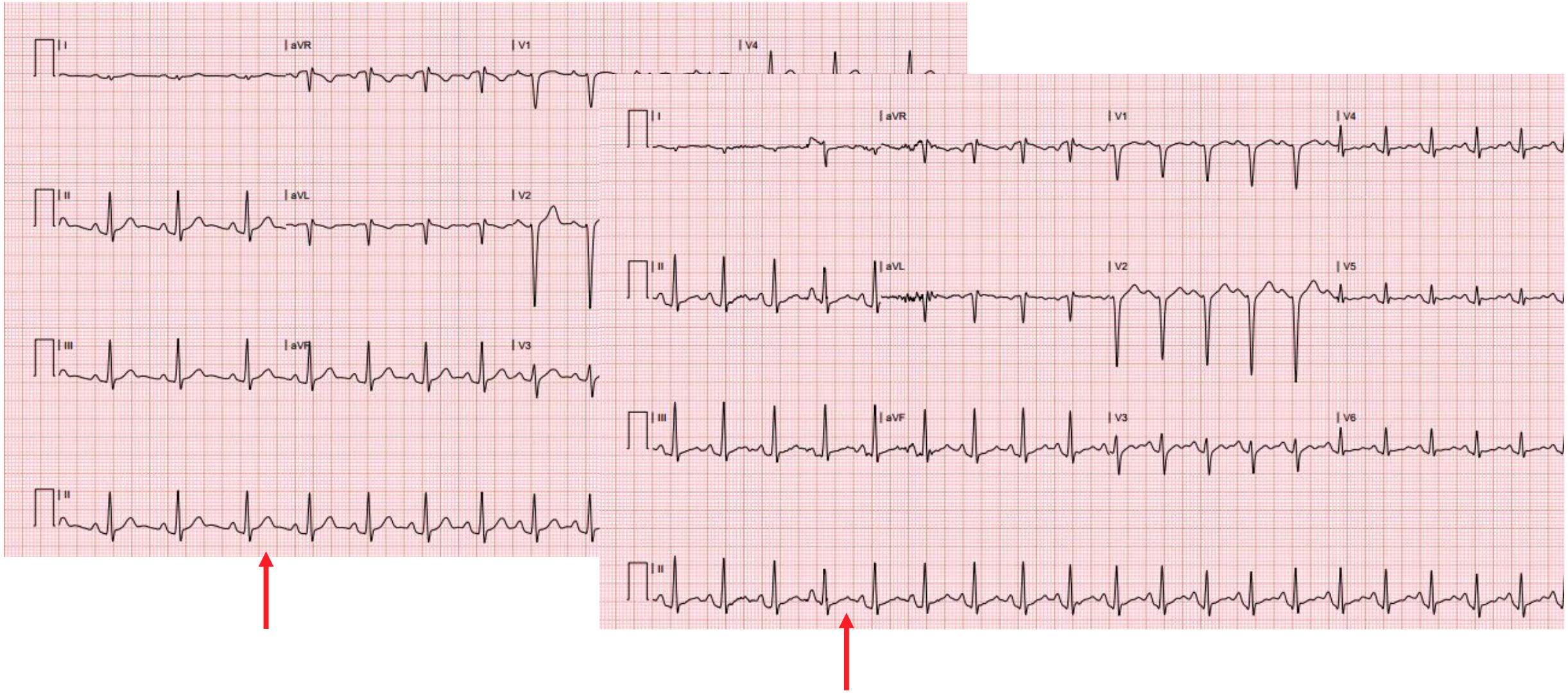
Splawski I, Circ 2000,102:1178–1185.

Vincent GM, N Engl J Med 1992; 327:846.


I
aVR
V1
V4
II
aVL
V2
V5
III
aVF
V3
V6
II

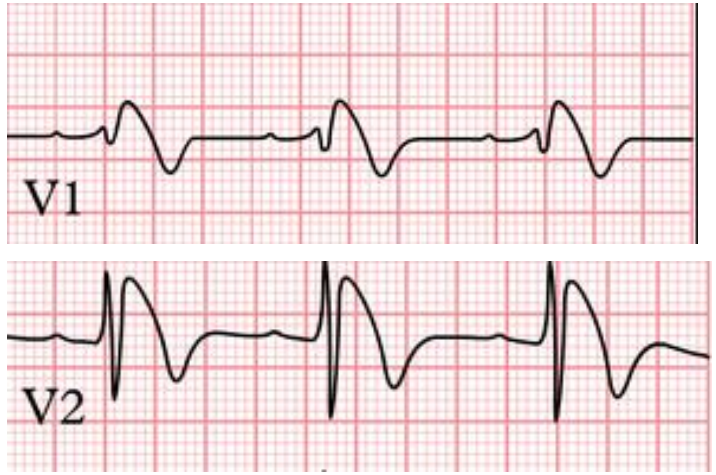

10mm/mV
I
aVR
V1
V4
II
aVL
V2
V5
III
aVF
V3
V6
II

ECG at rest and post exercise: QTc prolongation



Brugada Syndrome

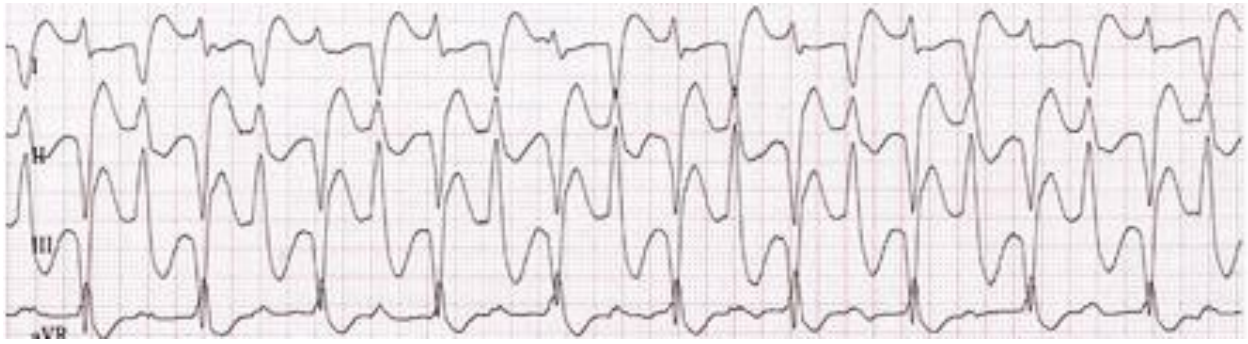
- Syncopal/SCA may not be exertional
- ST elevation in right precordial leads associated with ventricular arrhythmias and sudden death.
- Shares the SCN5a gene with LQT3
- Rhythm disturbance is typically Vtach or Vfib
- ECG changes may be masked and elicited with Na channel blockers
  - Fever can also elicit the changes
- Prevalence in US is 0.01- 0.4% of general population
  - Majority of affected individuals are of Asian descent
  - Male predominance 2x to 9x higher
- Events may occur during sleep (agonal nocturnal respiration)
- Most picked up by routine ECG or after event in relative
  - AD inheritance pattern
- Physical examination is normal
- ECG has J point elevation with downsloping ST Elevation in V1/V2



Priori SG, Heart Rhythm. 2013 Dec;10(12):1932-63 Patel SS, Pacing Clin Electrophysiol. 2009;32(6):704.

Catecholaminergic polymorphic ventricular tachycardia (CPVT)

- Hypersensitivity to inward calcium currents and abnormal release of calcium ions from the sarcoplasmic reticulum.
- Children present with frequent ventricular ectopy of Vtach with exercise leading to syncope or SCD
  - Bi-directional Vtach on exercise test is pathognomonic
- Incidence 1 in 10,000
- Genetic testing positive in 70% (RYR2, Casq2)
  - 30% have a family history of sudden death before age 40
- Resting ECG/physical exam normal



JACC 2009 Sep 22;54(13):1189

Acquired causes

- Myocarditis
- Commotio cordis

Myocarditis and SCD

- Murine Models have shown that exercise can increase viral titers and increase likelihood of death
- Unlike heart failure the risk of SCD in myocarditis does not correlate with the severity of myocardial inflammation
- SCD has been reported in myocarditis with normal ventricular systolic function
- Myocarditis can be asymptomatic or have non-specific symptoms and the first indication of myocardial involvement is a SCD event.

Arch Dis Child. 2008;93:594–8.
Eur J Epidemiol. 1989;5:348–50.
Expert Rev Cardiovasc Ther. 2013;11:193–201.

Causes of traditional myocarditis

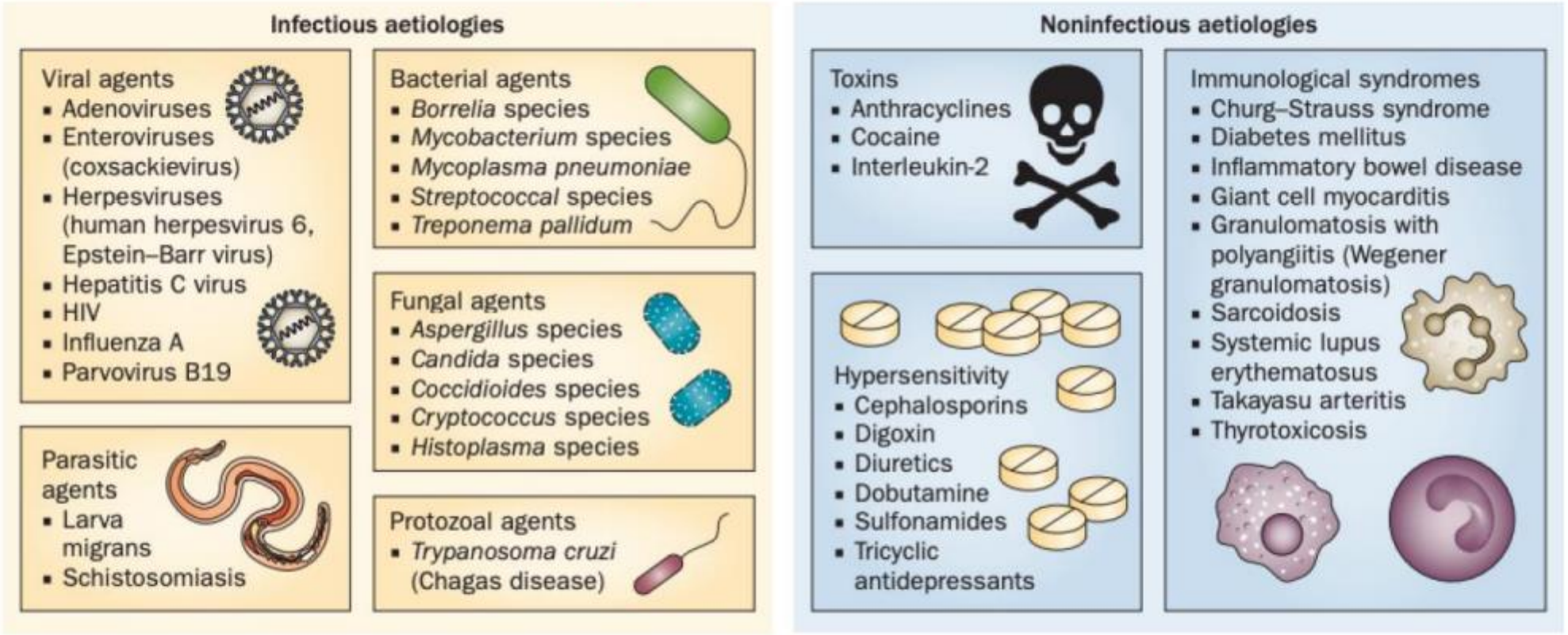


Figure 1 | Common causes of myocarditis. Viral infection is the most common aetiology, but several other aetiologies of myocarditis have also been implicated.

https://www.cdc.gov/vaccines/acip/meetings/downloads/slides-2021-06/02-COVID-Oster-508.pdf

Myocarditis diagnosis

Probable

1. Symptoms
   - Chest pain/pressure/discomfort
   - Dyspnea/shortness of breath
   - Palpitations
2. Abnormal testing
   - Elevated troponin
   - Electrocardiogram (ECG or EKG) findings
   - Decreased function on echo or MRI
   - MRI findings consistent with myocarditis
3. No other identified cause

Confirmed

1. Symptoms
   - Chest pain/pressure/discomfort
   - Dyspnea/shortness of breath
   - Palpitations
2. Abnormal testing
   - Biopsy
   - Elevated Troponin AND MRI findings consistent with myocarditis
3. No other identified cause

Cases with individuals who lack the listed symptoms but who meet other criteria may be classified as subclinical myocarditis (probable or confirmed)

https://www.cdc.gov/vaccines/acip/meetings/downloads/slides-2021-06/02-COVID-Oster-508.pdf

COVID-19 Myocarditis

- Most of **adult hospitalizations** show acute cardiac injury (Myocarditis)
  - >50% of adults with COVID-19 respiratory distress had acute troponin elevation. 87% required vasopressors.
  - MRI > 2 months after Dx showed some cardiac abnormality (myocarditis) in 20-78% of adults hospitalized with COVID-19
  - Autopsy findings combining 22 studies – cardiovascular abnormality present in 47.8%
  - Estimates for myocarditis other viruses 0.1-1%
- Fulminant myocarditis is rare but non-specific cardiac inflammation is common

Liu. *Circulation*. 2020;142:68-78. doi:10.1161/CIRCULATIONAHA.120.047549, Phelan. https://jamanetwork.com/journals/jamacardiology/fullarticle/2766124, Puntman. *JAMA Cardiol*. 2020:e203557.

Nat. Rev. Cardiol. 2021;18:169–193. Metkus. Circulation. 2021;143:553–565 doi.org/10.1161/CIRCULATIONAHA.120.050543 Halushka Cardiovasc Pathol. 2021;50:107300.

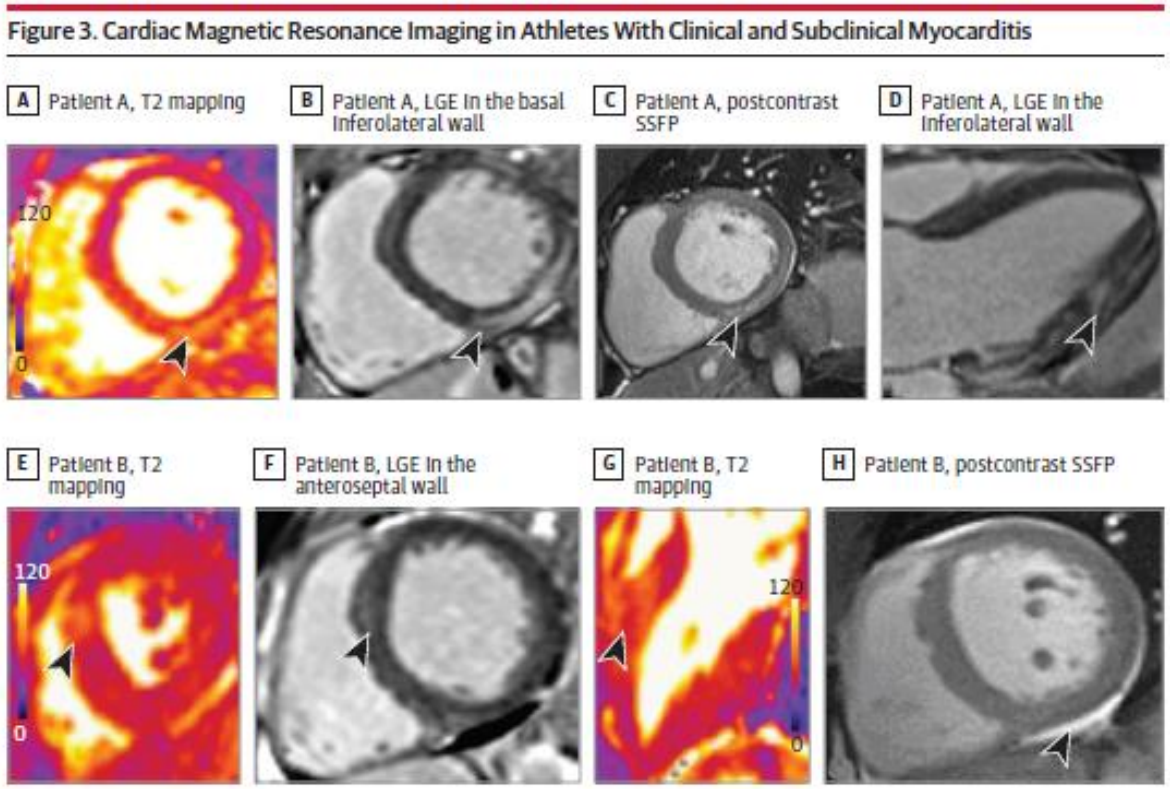
COVID-19 Myocarditis in Pediatrics

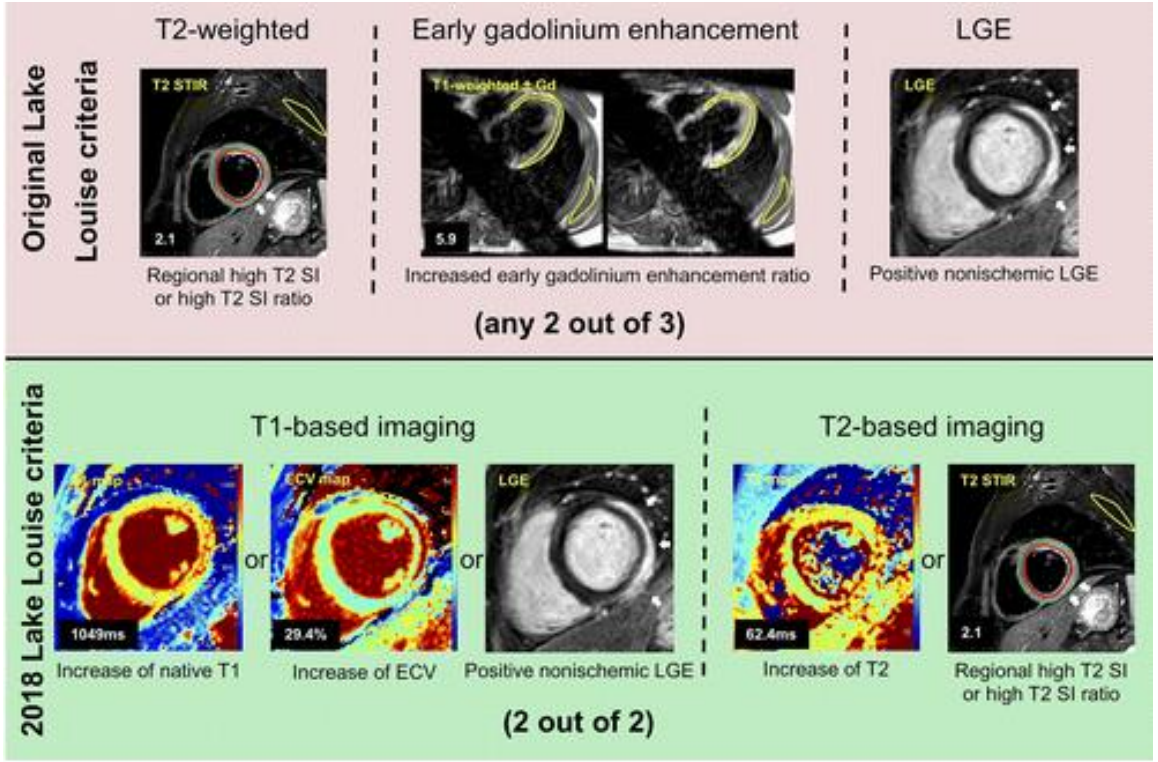
- Most children with acute SARS-CoV-2 have mild or asymptomatic disease and are not hospitalized
- Most of the data for this group of children is from young adults particularly studies of athletes.
  - Initial data in young adults suggested incidence as high as 20-30%
- Cardiovascular MRI in 26 college athletes Covid positive – 12 with mild symptoms with short course, others asymptomatic
  - 15% (n=4) with cardiac MRI changes suggestive of myocarditis
  - Additional 30% (n=8) – with cMRI changes suggestive of prior myocardial injury but not meeting Lake Louis criteria for myocarditis
  - No changes in ECG, echo or serum Troponin I levels

JAMA Cardiol. 2021;6(1):116-118. doi:10.1001/jamacardio.2020.4916

Liu. *Circulation*. 2020;142:68-78. doi:10.1161/CIRCULATIONAHA.120.047549, Phelan. https://jamanetwork.com/journals/jamacardiology/fullarticle/2766124, Puntman. *JAMA Cardiol*. 2020:e203557.

Cardiac MRI imaging of myocarditis





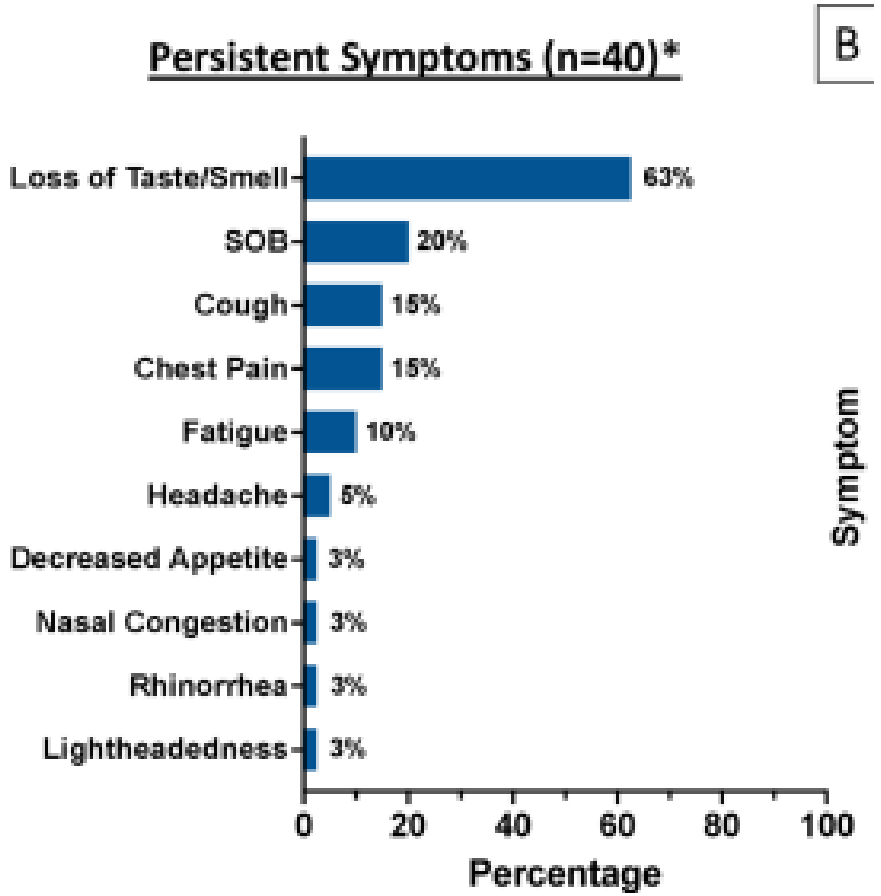
Edema, Necrosis, Hyperemia

Radiology: Cardiothoracic Imaging 2019; 1(3):e190010

JAMA Cardiol. doi:10.1001/jamacardio.2021.2065

ORCCA Registry

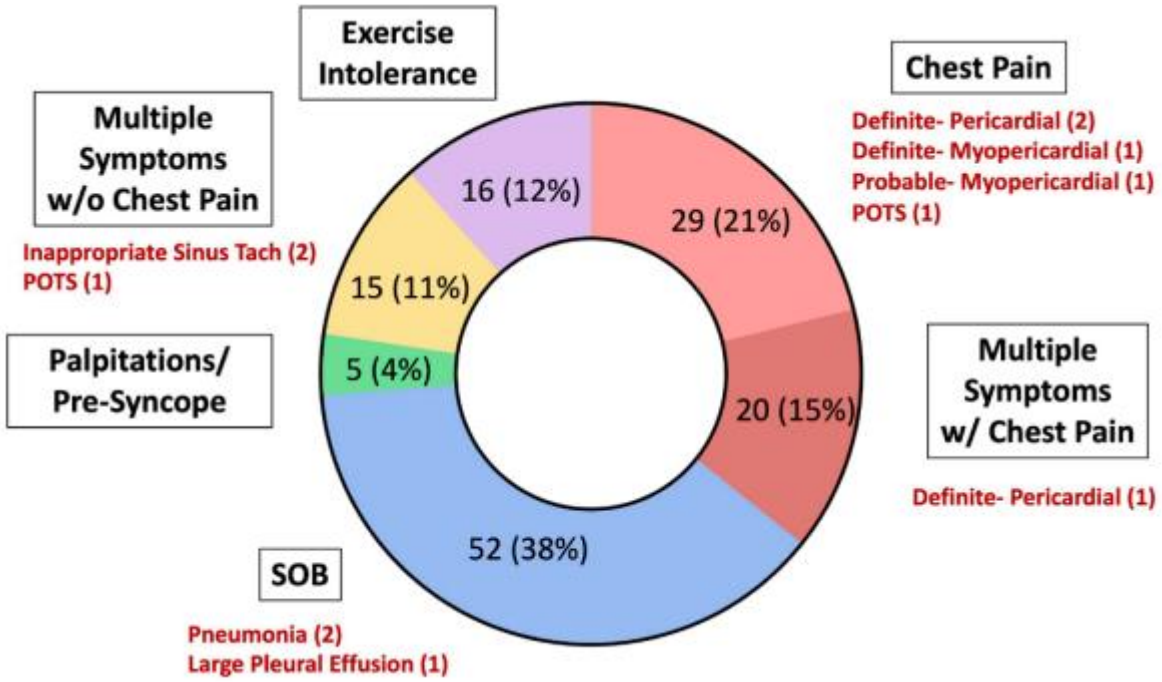
- Outcomes Registry for Cardiac Conditions in Athletes
- Observational cohort of **3597** collegiate athletes after SARS-CoV-2 infection
- Persistent symptoms (>3 weeks) in 1.2%
  - 20% had dyspnea, 15% with chest pain
  - ECG, Echo and troponin normal in all patients
  - 8 patients also had exertional symptoms …



Br J Sports Med . 2021 Nov 1;bjsports-2021-104644. doi:10.1136/bjsports-2021-104644

ORCCA Registry

- Exertional symptoms reported in 4% (n=137) of athletes.
- 12 diagnosed with SARS-CoV-2 associated sequelae
  - 5 “cardiac involvement”
  - 2 Pneumonia
  - 2 sinus tachycardia
  - 2 POTS
  - 1 pleural effusion

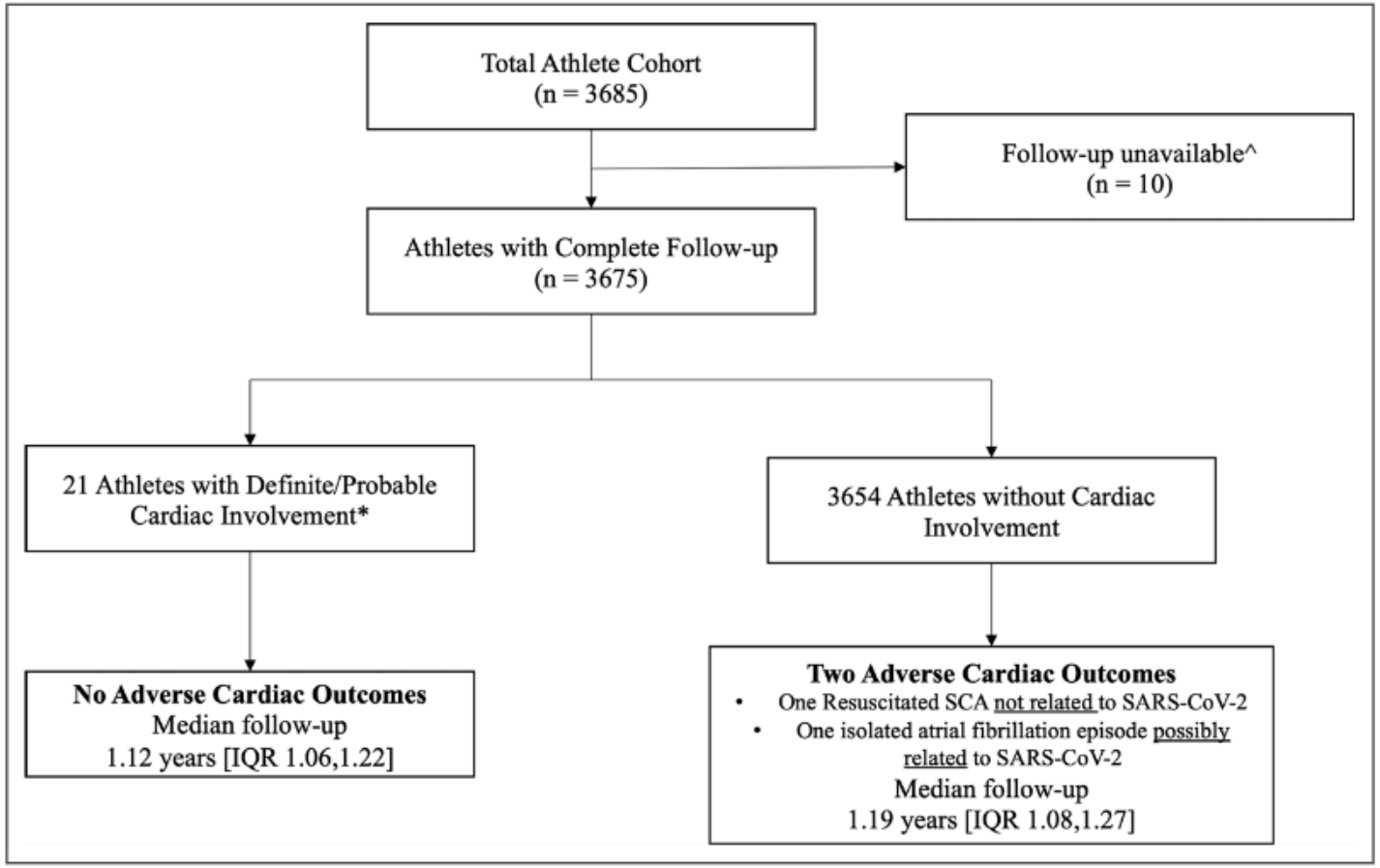


- All patients with cardiac involvement had **<u>exertional chest pain</u>**
  - 5/24 = 20.8% were abnormal.
- Overall cardiac involvement was **<u>0.14% of study population.</u>**

Br J Sports Med . 2021 Nov 1;bjsports-2021-104644. doi:10.1136/bjsports-2021-104644

ORCCA Registry – 1 year follow up

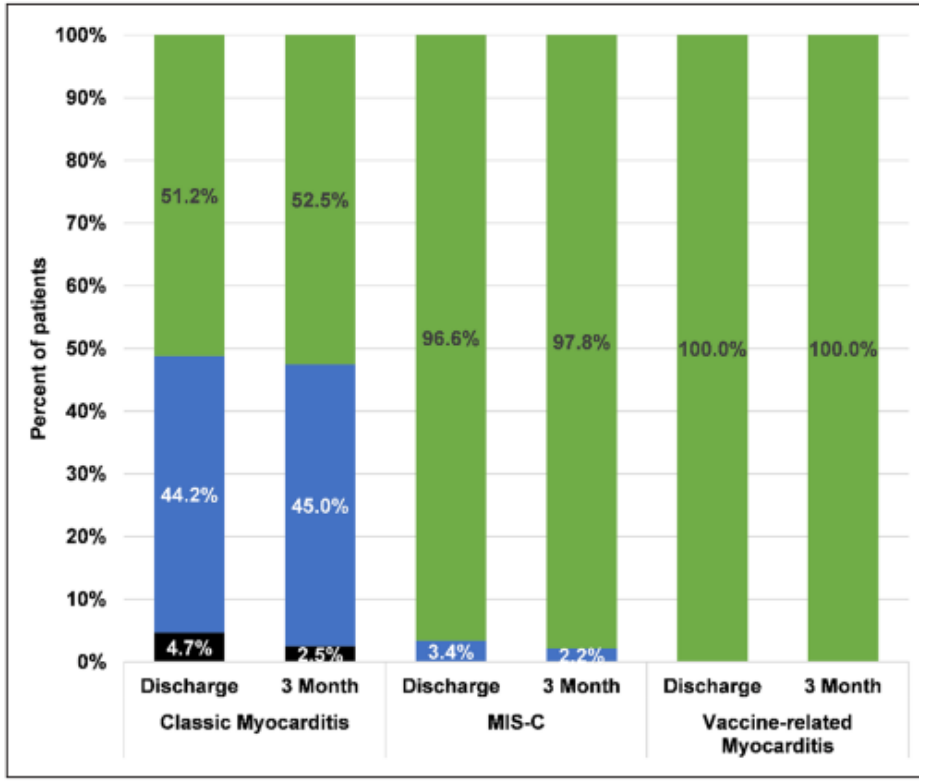
- 3675 Athletes (vs 3597)
- 21 (0.6%) with cardiac involvement
  - Vs 5 (0.14%)
- No adverse cardiac outcomes in this group
- One unrelated SCD
- One athlete with atrial fibrillation (possibly related)
  - Most common arrhythmia reported in adults with SARS-CoV-2 infection

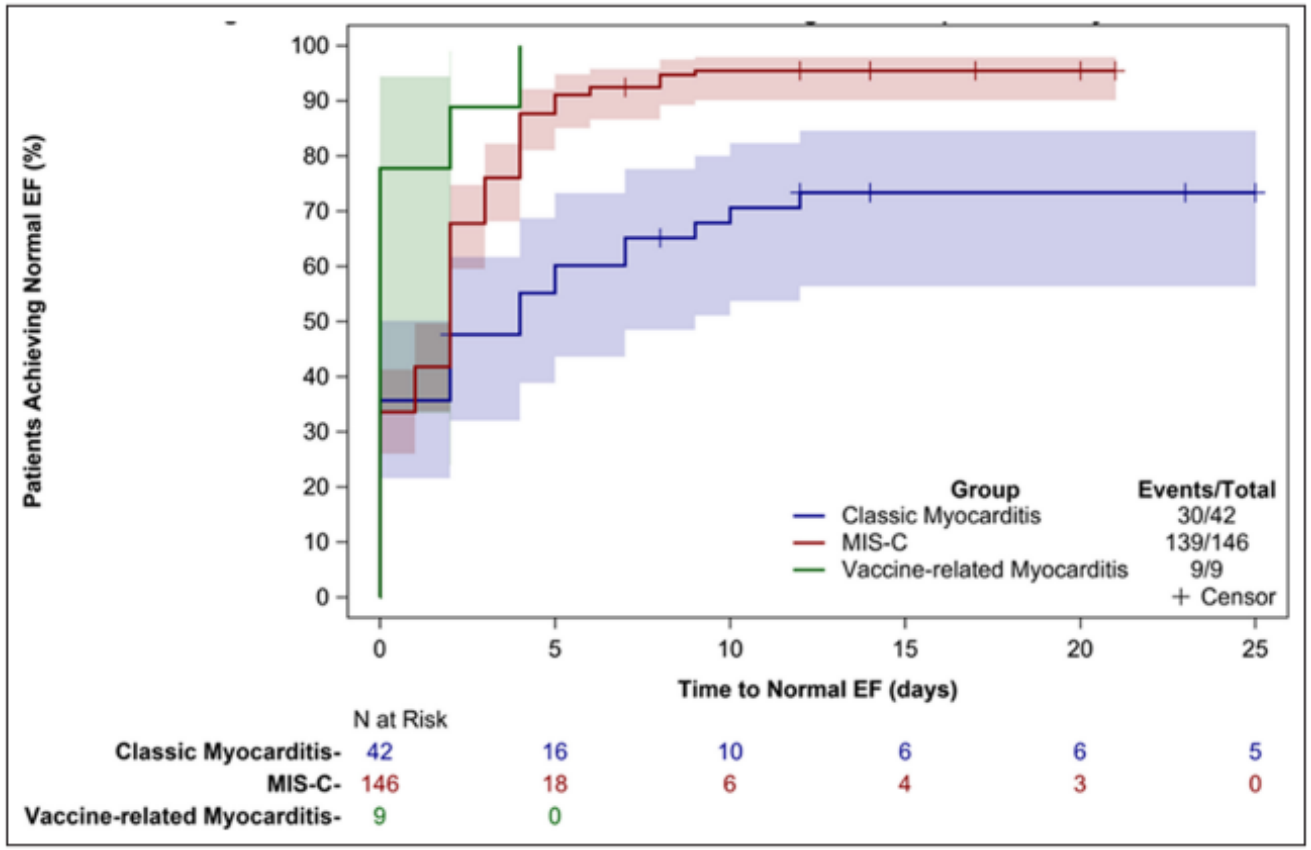


Circulation.2022;145:1690–1692. DOI: 10.1161/CIRCULATIONAHA.121.058272

MIS-C vs. Classic viral myocarditis vs. post- vaccine Myocarditis

- 149 MIS-C, 43 with classic (typically viral) myocarditis, 9 with post-vaccine myocarditis





J Am Heart Assoc. 2022;11:e024393. DOI: 10.1161/JAHA.121.024393

Guidelines for returning to competitive sports after COVID-19 infection

Asymptomatic, mild or moderate COVID-19 symptoms (no hospitalization)

During assessment:
1. Guidance re: duration of isolation
2. Light cardio only which can start 24 hours after last fever, exercise at a level that does not provoke symptoms
3. AHA 14-element screening evaluation - symptoms of myocarditis (incidence: <1%):

AHA screening negative

Cleared to begin RTP progression

AHA screening positive

COVID – 19 with cardiac symptoms – Chest pain, palpitations, syncope

In person visit with PCP and physical exam
1. No exercise during isolation
2. AHA 14-element screening evaluation
3. Complete physical exam
4. EKG
5. Troponin

Normal evaluation, no exertional symptoms

Cleared to begin RTP progression minimum of 24 hours from last fever

Abnormal exam, labs, or significant exertional symptoms

Cardiology referral

Severe COVID-19: Hospitalization or Diagnosis of MIS-C.

Refer to Cardiology/MIS-C specialist. Hospitalized patients follow up initially at ~2 and 6 weeks post discharge.

Note – these guidelines are meant to serve as a generalized approach to patients and individual circumstances may warrant deviation from these guidelines.

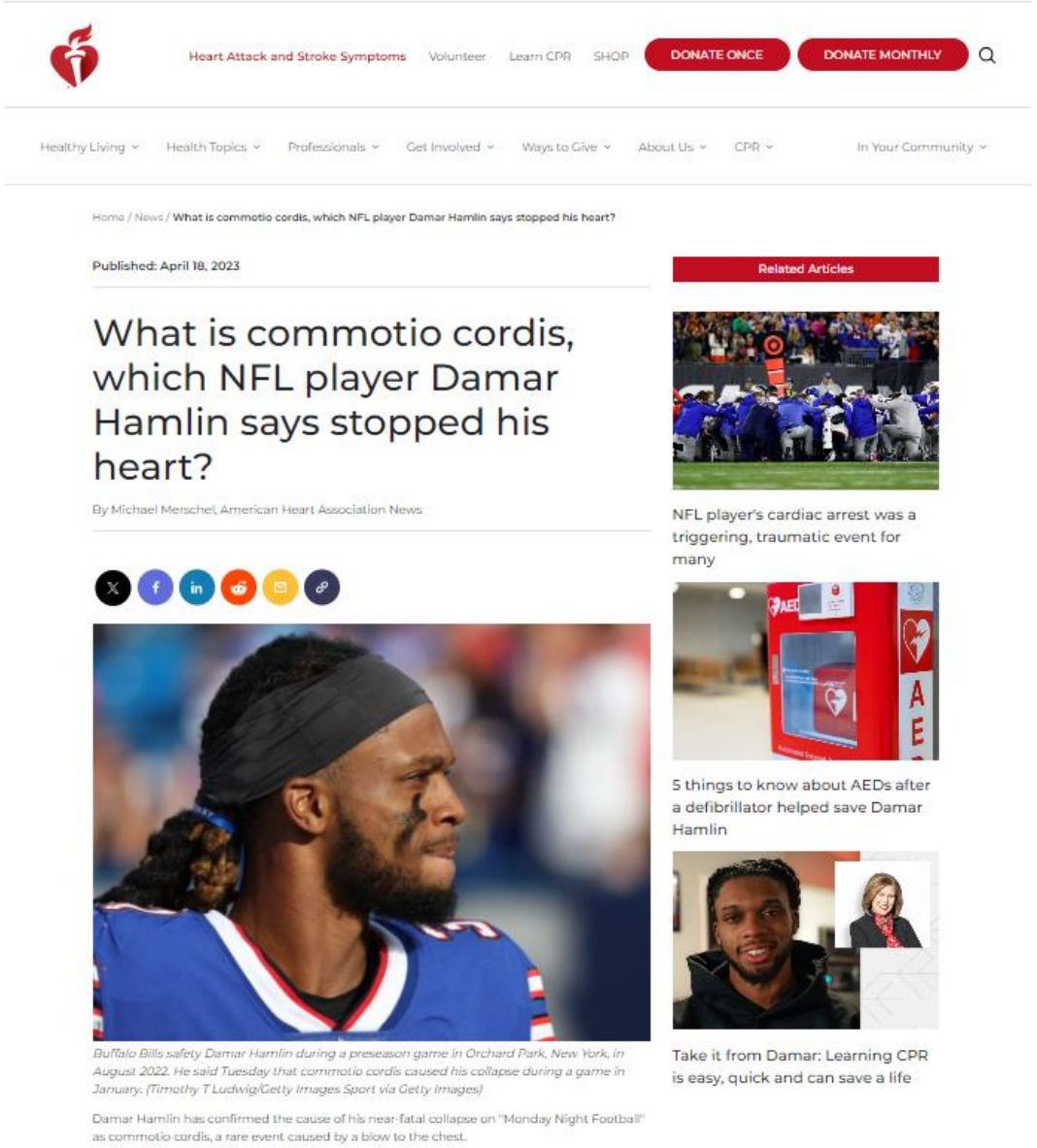
Guidelines adapted from AAP Interim Guidance and ACC expert consensus for Return to Sport

References:
2022-01-21 RTP algorithm.pdf (aap.org)
J Am Coll Cardiol. 2022 May 3;79(17):1717-1756

Updated 4/29/23

Commotio Cordis

- Traumatic impact that causes induces VF by causing dispersion of ventricular repolarization.
- Timing within 20-40 millisecond window on the upslope of T wave
- Harder, smaller diameter objects more likely to cause VF
  - Baseball, Hockey puck
- 1/3 of patients wore protective equipment and still had VF



Heart.org
N Engl J Med 1998; 338:1805.
J Cardiovasc Electrophysiol 2007; 18:115.
Pediatrics 2002; 109:873.

Emergency Action Plan for Sudden Cardiac Arrest (SCA)

Universal Response to the Collapsed Athlete

Collapsed and Unresponsive*

Assume SCA

*Unresponsive to shoulder tap and verbal stimuli

Signs of SCA:
Continued respirations/gasping
Seizure-like activity/twitching
Eyes open/rolled back
(Falsely assuming a pulse)

Call EMS

Begin hands-only CPR (chest compressions)

Retrieve and apply an AED as soon as possible

Drezner J. Prevention of sudden cardiac death in athletes: progress and pitfalls (2023 John R. Sutton Clinical Lecture). In: Proceedings of the 2023 ACSM Annual Meeting & World Congresses. 2023 May 31 to June 2; Denver (CO).

AMERICAN COLLEGE of SPORTS MEDICINE
LEADING THE WAY

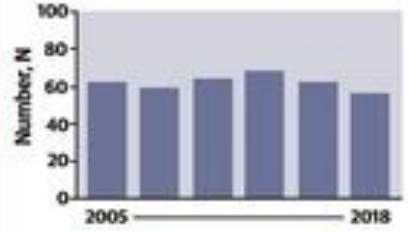
CENTRAL ILLUSTRATION: Evolution of Sports-Related Sudden Cardiac Arrest Frequency, Initial Management, and Survival Rate

Similar Frequency

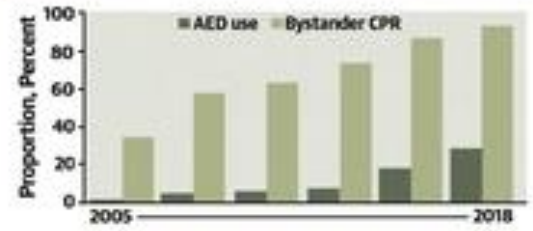
Improved Management

Higher Survival

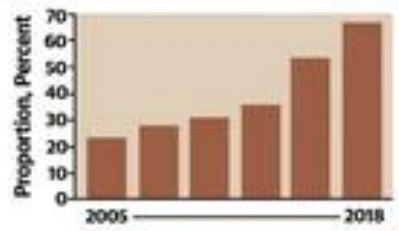
Total Number of SrSCA



AED Use and Bystander CPR



Survival to Discharge



Karam, N. et al. J Am Coll Cardiol. 2022;79(3):238-246.

Heart Safe Initiatives

Project ADAM affiliate sites strive to assist schools and communities in establishing a practiced plan to respond to a sudden cardiac arrest. We provide the foundation to develop and sustain A CPR/AED program, including planning templates, a reference manual and one-on-one consultation. Contact your local affiliate for support.

If you have questions or know someone who could benefit from Project ADAM, please contact us.
projectadam@childrenswi.org

Summary

- SCA/SCD is rare
- HCM, coronary abnormalities and myocarditis are leading causes
  - Others: Long QT, LVOTO, Aortic dissection, commotio cordis
- Screening with PPE is recommended but will not capture all cases – some will present with SCA/SCD
  - Emergency action plan

Thank you !