



## The Emergence of Pediatric Sports and Exercise Cardiology: Promoting Physical Activity in All Children and Young Adults

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# Disclosures

No financial disclosures

# Objectives

- Define the components of sports and exercise cardiology
- Appreciate sports and exercise cardiology as a sub-specialty within pediatric/congenital cardiology
- Understand the goals of pre-participation screening and how the pediatric population is distinct from adults
- Recognize the complexity and importance of exercise counseling in patients with congenital or acquired heart disease
- Appreciate the role of cardiopulmonary exercise testing and formal cardiac rehabilitation and fitness programs in pediatric and young adult patients
- Help (your friendly pediatric cardiologist) promote physical activity in all patients

## Patient Vignettes

16 year old with a family history of sudden cardiac death in his father, age 44, autopsy showed a large heart. No genetic testing. He wants to be cleared for AAU basketball. EKG and echo were normal. Can he play? Does he have any risk of SCD?

Same patient, but genetic testing done in father and he shares the same gene.

Same patient, but he has HCM, largest LV thickness was 17 mm, no fibrosis on MRI. No symptoms. No acute changes on stress testing.

Same patient, but has fibrosis on MRI. Holter monitor is normal.

13 year old with HLHS s/p Fontan at age 4. Followed yearly. No significant changes on routine testing. O2 sats 90% at rest. He says he gets out of breath easily and feels his heart racing when he runs. He sometimes looks flushed and maybe a bit blue around the lips. Mom is nervous. What would you do and recommend?

Same patient: He wants to try out for the tennis team.

# (Adult) Sports Cardiology

Historically, 2 main objectives:

- Ensuring safety by screening for conditions that predispose to sudden cardiac death (SCD)
- Optimizing the performance of the cardiovascular system during exercise (including investigating symptoms during exercise)

Primary concern is CAD, but also electrical abnormalities and cardiomyopathies may be present

# Questions

- Are pediatric and young adults different? Is pediatric physiology different? Is screening different?
- What is the definition of "athlete"? Is there a difference between recreational play, gym class, or team sports? 6 yo vs 15 yo?
- What about people with pre-existing congenital heart disease? Can they be an "athlete"? Should they be an "athlete"?

Why does this matter?

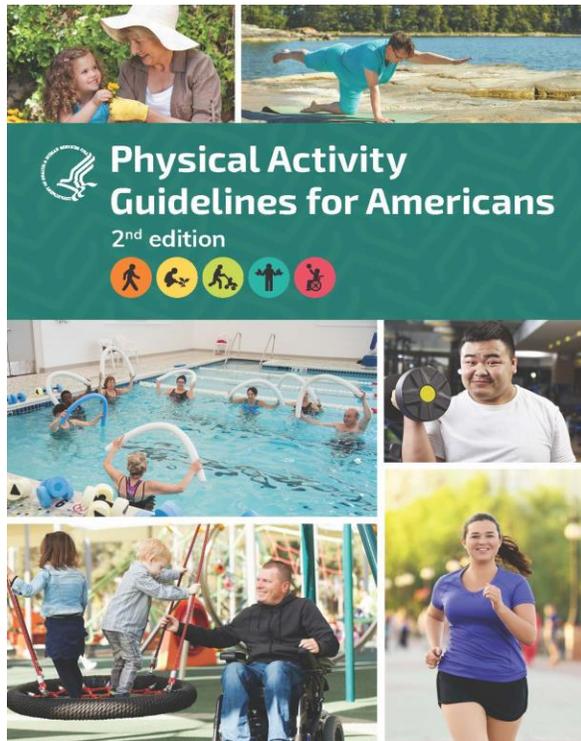


Table 2-1. Health Benefits Associated With Regular Physical Activity

**Children and Adolescents**

- Improved bone health (ages 3 through 17 years)
- Improved weight status (ages 3 through 17 years)
- Improved cardiorespiratory and muscular fitness (ages 6 through 17 years)
- Improved cardiometabolic health (ages 6 through 17 years)
- Improved cognition (ages 6 to 13 years)\*
- Reduced risk of depression (ages 6 to 13 years)

**Adults and Older Adults**

- Lower risk of all-cause mortality
- Lower risk of cardiovascular disease mortality
- Lower risk of cardiovascular disease (including heart disease and stroke)
- Lower risk of hypertension
- Lower risk of type 2 diabetes
- Lower risk of adverse blood lipid profile
- Lower risk of cancers of the bladder, breast, colon, endometrium, esophagus, kidney, lung, and stomach
- Improved cognition\*
- Reduced risk of dementia (including Alzheimer's disease)
- Improved quality of life
- Reduced anxiety
- Reduced risk of depression
- Improved sleep
- Slowed or reduced weight gain
- Weight loss, particularly when combined with reduced calorie intake
- Prevention of weight regain following initial weight loss
- Improved bone health
- Improved physical function
- Lower risk of falls (older adults)
- Lower risk of fall-related injuries (older adults)

**Note:** The Advisory Committee rated the evidence of health benefits of physical activity as strong, moderate, limited, or grade not assignable. Only outcomes with strong or moderate evidence of effect are included in this table.

\*See [Table 2-3](#) for additional components of cognition and brain health.

# Benefits of Exercise/Physical Activity

Lower risk of death

Lower risk of other chronic long-term medical problems

Better outcomes after surgery

Lower incidence of depression/anxiety

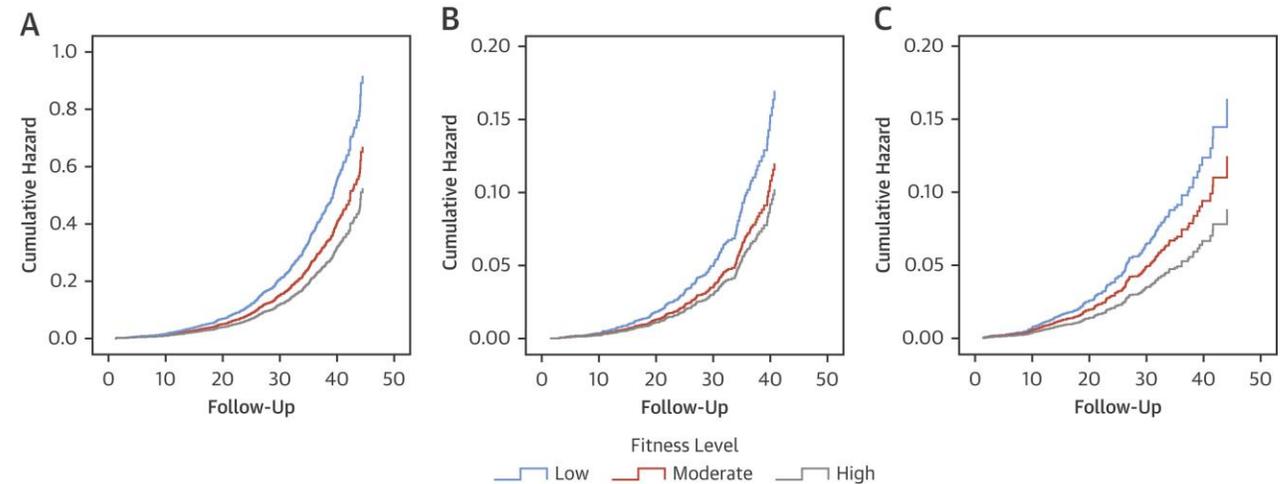
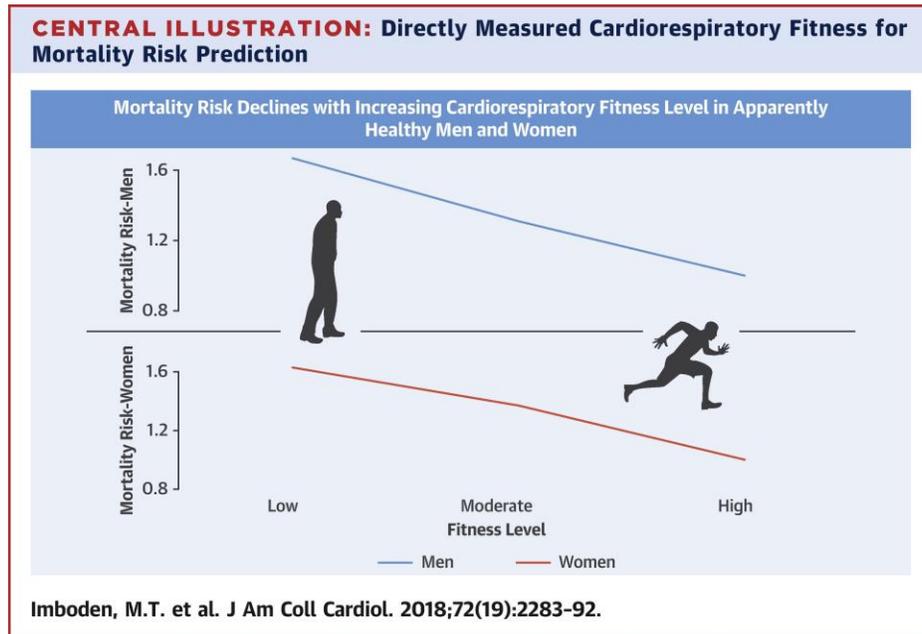
Improved bone health

Better weight/less obesity

Higher quality of life\*

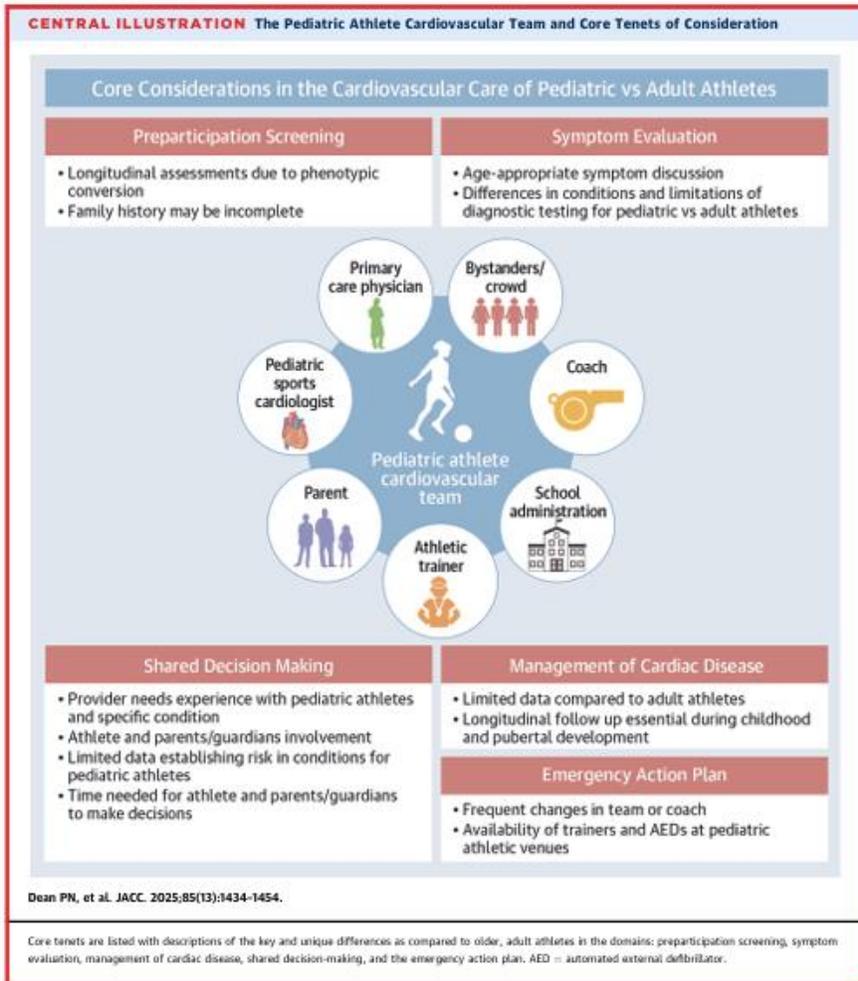
# Long-term Benefit

- Higher VO<sub>2</sub> (exercise capacity) as measured on cardiopulmonary exercise testing (CPET) is associated with lower mortality
- Every 1 met increase (3.5 ml/kg/min) was associated with a 20% lower CVD risk
- Physical activity in childhood likely correlates with VO<sub>2</sub> as adults



Hazard plots for (A) all-cause, (B) cardiovascular disease (CVD), and (C) cancer mortality according to cardiorespiratory fitness (CRF) level

# Tenets of Pediatric Sports Cardiology



This graphic does not include:

- CPET interpretation
- The use of cardiac rehab/fitness programs
- Promotion of physical activity in all young patients

# Pediatric/Congenital Sports and Exercise Cardiology

- Prescreening for exercise participation and risk assessment for sudden death
- Evaluation of cardiac symptoms and return to play evaluation
- Assessment of risk in patients with CHD, cardiomyopathies, or arrhythmias
- Cardiopulmonary exercise test (CPET) interpretation
- Guiding patients with known or suspected heart disease looking for a safe and fun way to become active and healthy
- Cardiac performance training and fitness enhancement in healthy athletes
- Management of a formal cardiac rehabilitation and fitness program

# Pre-participation Screening and Symptom Evaluation

## Preparticipation Screening: Why worry about healthy teens?

- Rare incidence of sudden cardiac death
- Uncommon, but outcome is devastating
- Usually occurs in healthy, asymptomatic children and young adults
- Attracts attention from schools, media
- Many findings start to appear with pubertal development

# Medway High lacrosse player dies

Second area death in sport at schools

By Erin Conroy, Globe Correspondent | April 29, 2007

A Medway High School sophomore collapsed yesterday during lacrosse practice and was pronounced dead at a nearby hospital. It was the second sudden death in a week of a young lacrosse player in the area.

Nick Souza, who would have turned 16 on

### ARTICLE TOOLS

COLORADO SPRINGS, Colo. (AP) - A 17-year-old player from Harrison High School collapsed during the third quarter of a game Friday night and died after being taken to Memorial Hospital.

Center Fermin Vialpando, a senior running down the field when he fell, apparently after a seizure, reported. The game was halted

His mother, Sundae Vialpando, told the Denver Post on Saturday that he had no known health problems. "Doctors sometimes with an athlete - it n

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UPDATE

47%

# player found dead had torn aorta, says

A 17-year-old student at Collins Hill High School who was found dead over the weekend had a tear in his aorta that killed him, an investigator said Tuesday.

It's not yet clear what caused the tear that killed Terrell Wilson, an Honors Student and football player at Collins Hill High School in Suwanee.

RELATED:  
• [More Gwinnett news](#)

The tear could have occurred naturally or as a result of an injury that Wilson sustained, said Ted Bailey, chief forensic investigator for the Gwinnett County Medical Examiner's office.

Gwinnett medical examiner's staff are interviewing Wilson's family and football coaches, he said.

"We're interviewing his family and his football coaches, to see if he got an injury," Bailey said.

Cpl. Illana Spellman, a spokeswoman for the Gwinnett County Police Department, said Tuesday that detectives do not believe that Wilson's death is suspicious and that they are not investigating it.

Terrell was a lineman on the freshman football team.

Counselors visited Terrell's classes at the start of each class period Monday to talk with Gwinnett County Schools district spokesman Jorge Quintana.

HOME > NEWS > LOCAL > MASS.

# Medfield boy, 14, dies at lacrosse practice

Needham school to offer counseling

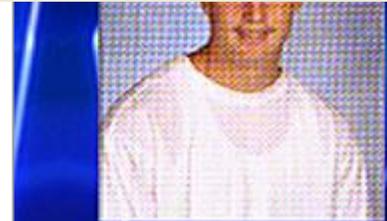
By David Abel and Michael Naughton, Globe Correspondent | April 25, 2007

An eighth-grader at St. Sebastian's School in Needham died yesterday while practicing lacrosse on a school field, officials said.

... started CPR. They said the first patrol car on the scene had a defibrillator that was used on the teenager before he was taken away.

... football team members participated in a practice in which players strap on helmets and tackle each other. That was when 16-year-old ... collapsed, stunning players and

... volunteer coach, who is also a police officer on the school's side when he collapsed and



Video: Egg Harbor High School Football Player Dies

# Incidence of SCD

**TABLE 1** Epidemiology of SCD and SCA in Athletes

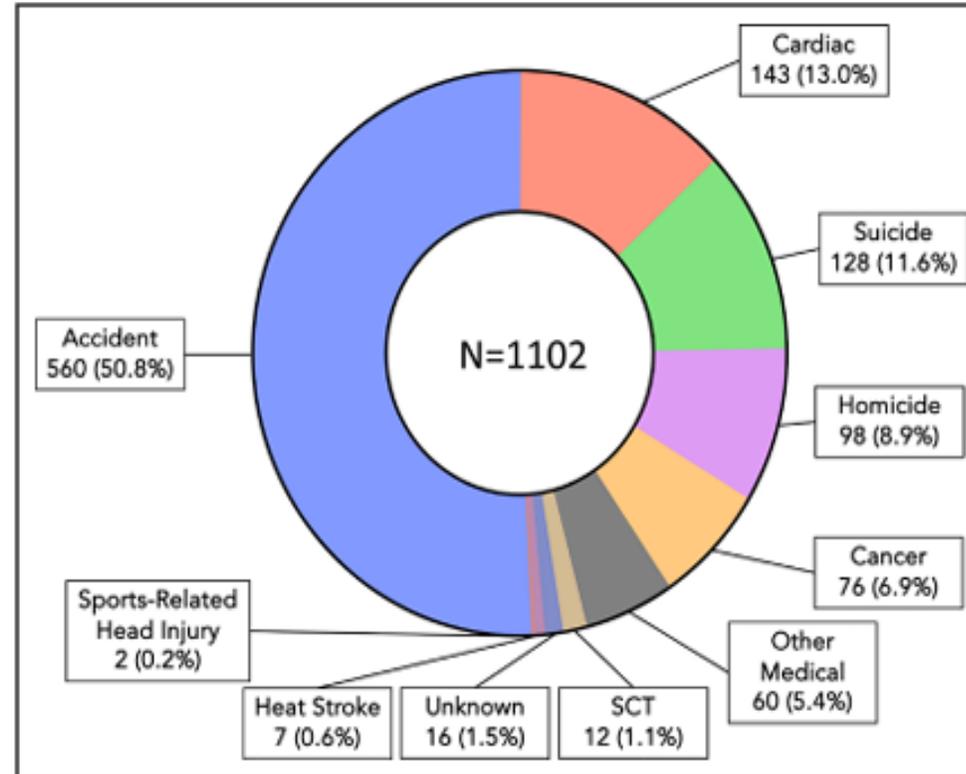
First Author et al. <sup>14</sup>	Year	Country	Case Identification	SCD/SCA	Years Studied	Annual Incidence	Number of Years	Age Range, y	Number of Deaths
Van Camp et al. <sup>14</sup>	1995	United States	National Center for Catastrophic Injury Research and media database	SCD	1983-1998	0.3/100,000	10	17-24	160
Corrado et al. <sup>23</sup>	2003	Italy	Mandatory death reporting	SCD	1979-1999	2.3/100,000	20	12-35	55
Maron et al. <sup>2</sup>	2009	United States	USA Registry for Sudden Death in Athletes	SCA + SCD	1980-2006	0.6/100,000	27	8-39	690
Drezner et al. <sup>222</sup>	2009	United States	1,710 high schools with AEDs surveyed for SCA or SCD	SCA + SCD	2005-2007	4/100,000 SCA + SCD 2.2/100,000 SCD	Within 6 months of survey	14-17	14
Harmon et al. <sup>714</sup>	2011	United States	Parent Heat Watch database, NCAA Resolutions list, insurance claims	SCD	2004-2008	2.3/100,000	4	18-26	37
Marçon et al. <sup>723</sup>	2011	France	Emergency medical response to confirmed cardiac cases	SCA + SCD	2005-2010	1/100,000	5	12-75	820
Steinil et al. <sup>100</sup>	2011	Israel	Retrospective review (2 Israeli newspapers by 2 media researchers)	SCD	1985-2009	1st 2.5/100,000 2nd 2.6/100,000	24	12-44	24
Roberts et al. <sup>71</sup>	2013	United States	Minnesota State High School League records	SCD	1993-2012	0.24/100,000	19	12-19	4
Maron et al. <sup>724</sup>	2013	United States	USA Registry for Sudden Death in Athletes and NCAA resolutions list for cardiac cases	SCD	2002-2011	1.6/100,000 presumed 1.2/100,000 confirmed	9	17-26	64
Maron et al. <sup>99</sup>	2013	United States	USA Registry of Sudden Death in Athletes	SCD	1986-2011	0.6/100,000	26	12-18	13
Torsdahl et al. <sup>717</sup>	2014	United States	2,149 school students—events occurred on school campus	SCA + SCD	2009-2011	1.1/100,000	2	14-18	18 SCA 2 SCD
Harmon et al. <sup>6</sup>	2015	United States	NCAA database	SCD	2003-2013	1.9/100,000	10	17-24	79
Marçon et al. <sup>724</sup>	2015	United States	Population the Oregon-Sudden Unexpected Death Study (Oregon-SUDS)	SCA	2002-2013	2.2/100,000 (during sports activities)	11	35-65	1,247 SCA cases, 63 SCA during sports activities
Malhotra et al. <sup>71</sup>	2018	United Kingdom	Football association registry	SCD	1996-2016	6.8/100,000	20	16	8
Peterson et al. <sup>714</sup>	2021	United States	National Center for Catastrophic Sports Injury Research in collaboration with national sports organizations	SCD + SCA	2014-2018	1.5/100,000 (high school) 1.9/100,000 (NCAA level)	4	11-29	331 SCA, 173 SCD

NCAA = National Collegiate Athletic Association; SCA = sudden cardiac arrest; SCD = sudden cardiac death.

- Rare
- Incidence varies amongst studies
- 1.2-1.9/100,000 (NCAA 17-24 yrs)
- 3.6/100,000 --> 0.9/100,000 after screening initiated (Italy)
- 0.47-1.21/100,000 (age < 35) vs. 6.64/100,000 (age > 35)

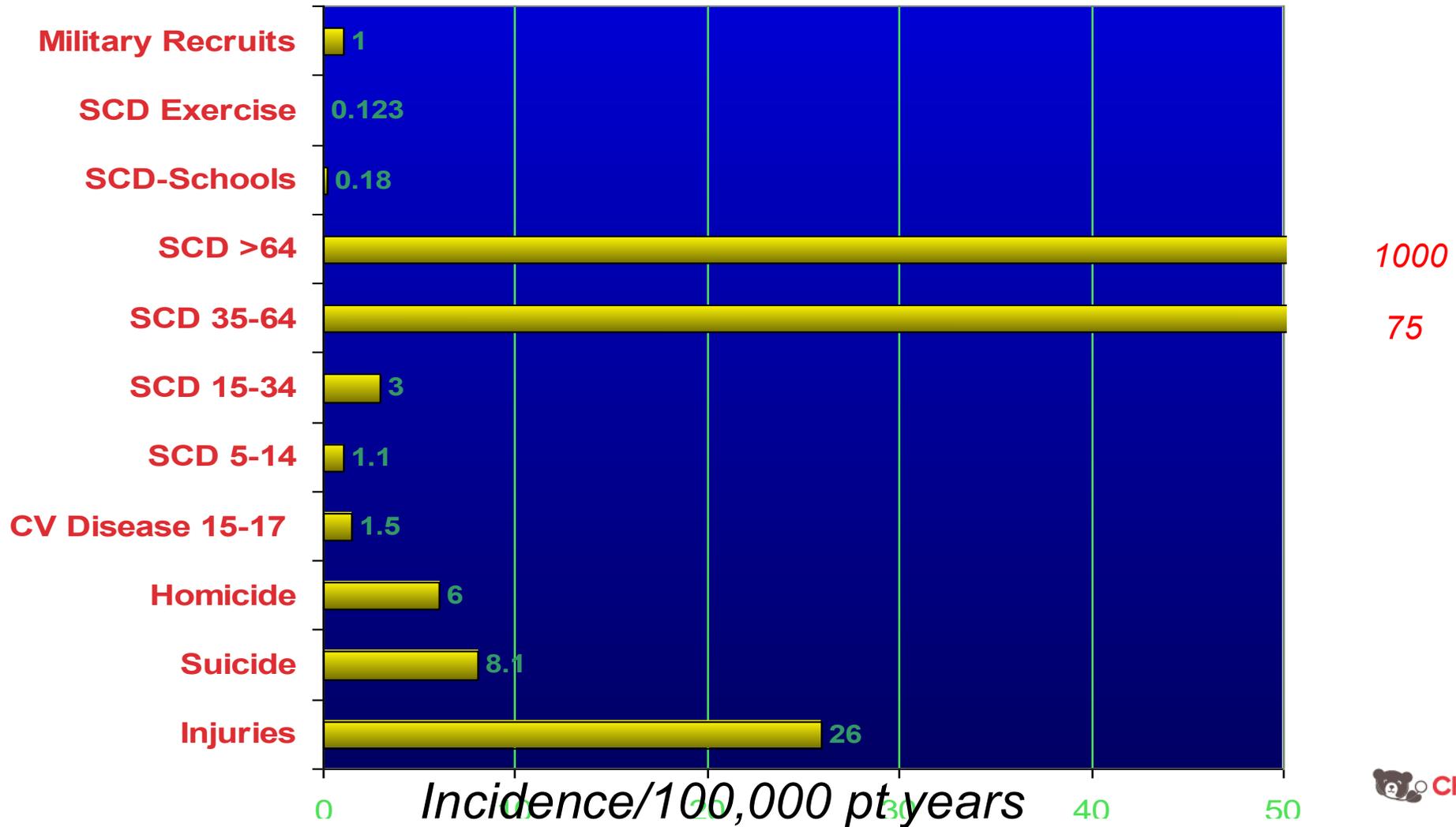
Finocchiaro et al, JACC 2023

Petek et al, Circulation 2024  
Cardiac etiology in 13% of sudden death

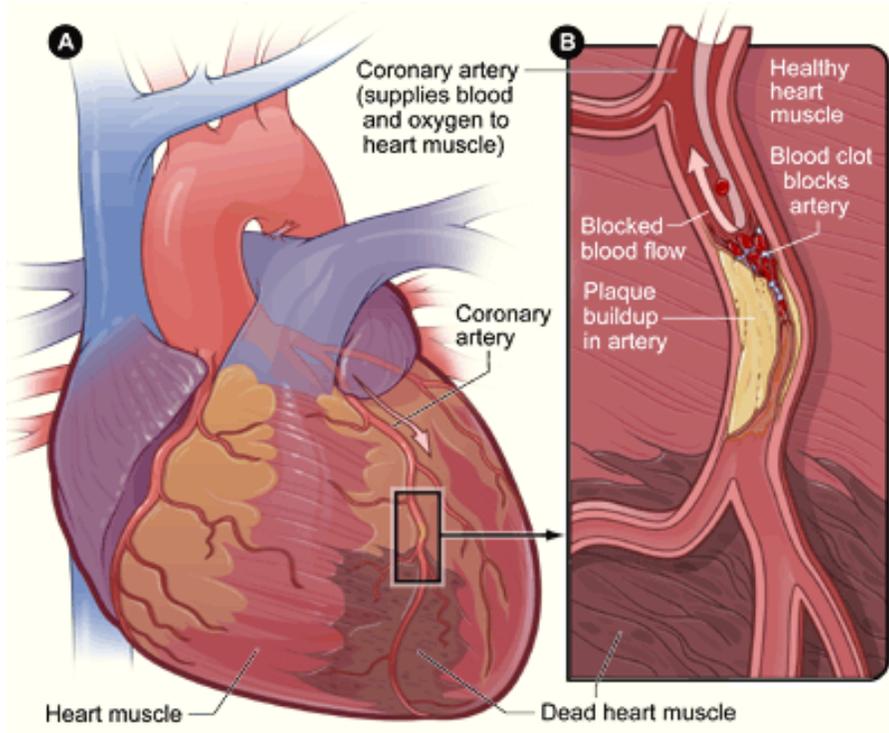


**Figure 1.** Causes of death among National Collegiate Athletic Association athletes (n=1102). SCT indicates sickle cell trait.

# Adolescent SCD in Context

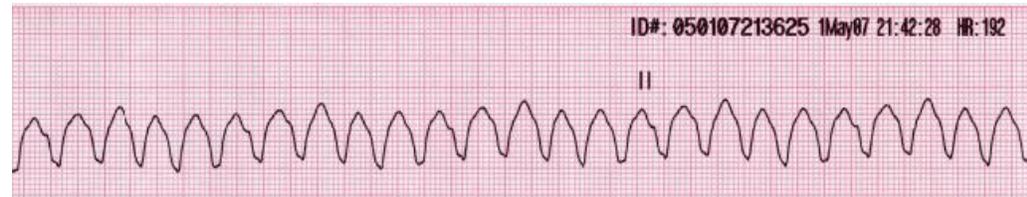


# Sudden Death in Adults vs. Children: The Biggest Difference



Kids don't get CAD

...most pediatric VT non-ischemic



# Screening: What are We Screening For?

- Arrhythmia Vulnerability Syndromes

- LQTS



- BrS/ARVC/CPVT/SQTS/PCCSD



- WPW

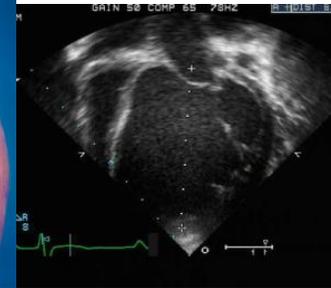
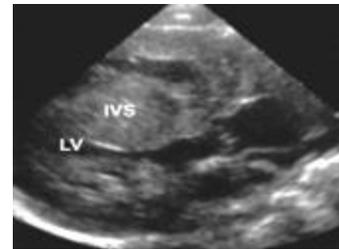


- CCHB

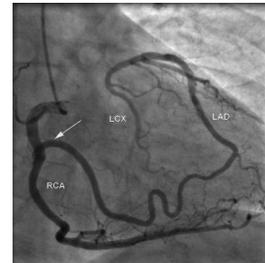


- Cardiomyopathies

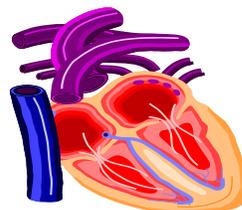
- HCM, DCM, LVNC, RCM



- Coronary artery abnormalities

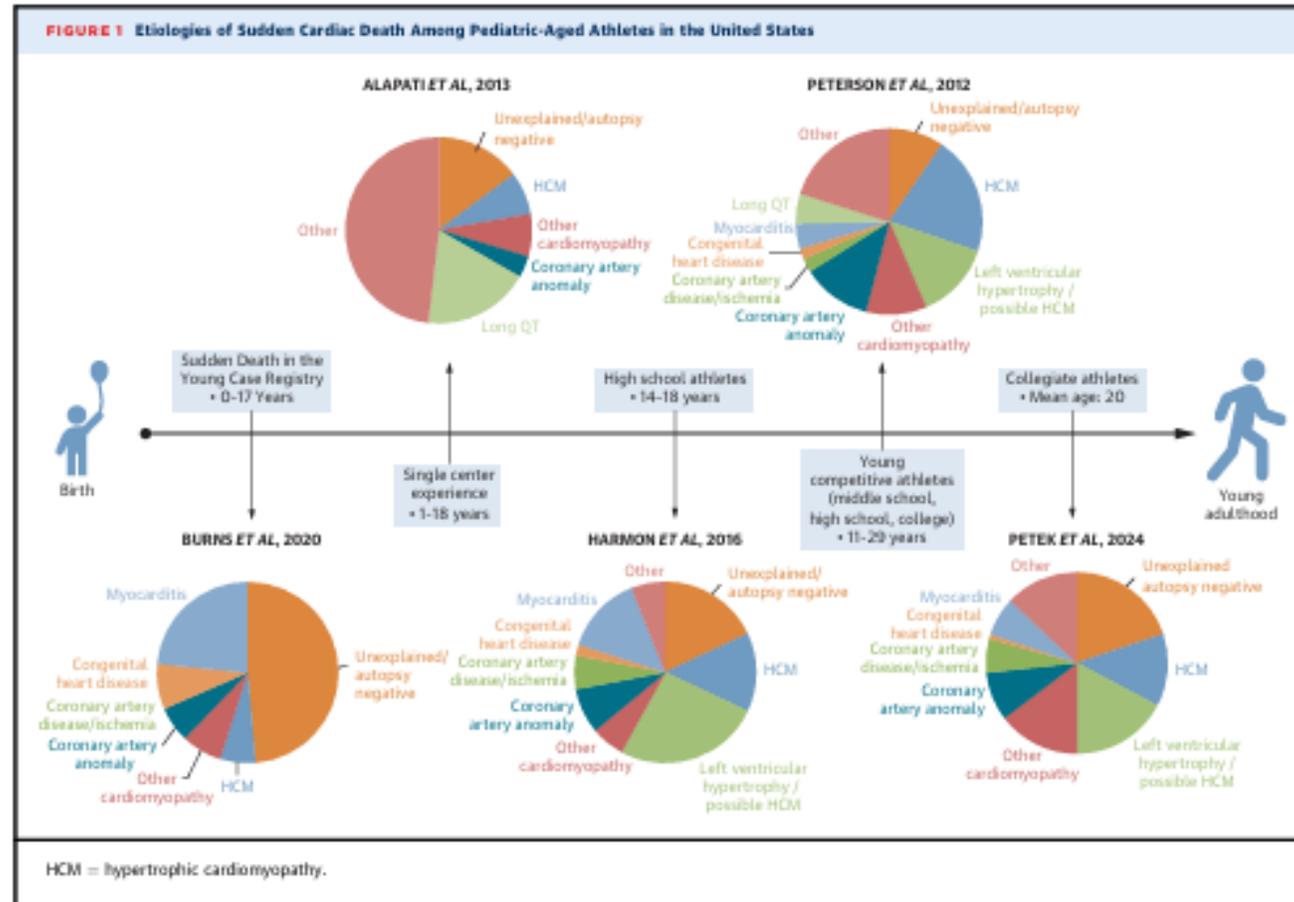


- Congenital heart disease



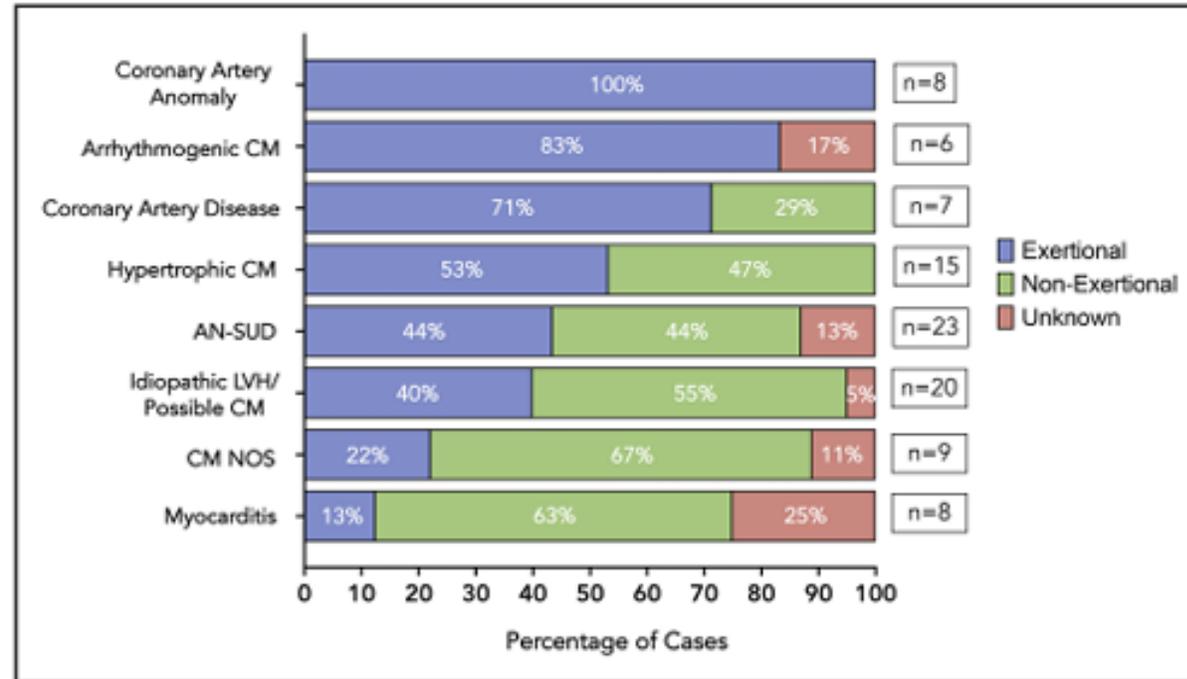
# Etiologies of SCD Over Time

Dean et al, JACC 2025



# Exertional status of SCD

Petek et al. Circulation, 2024  
Overall, ~50% exertional



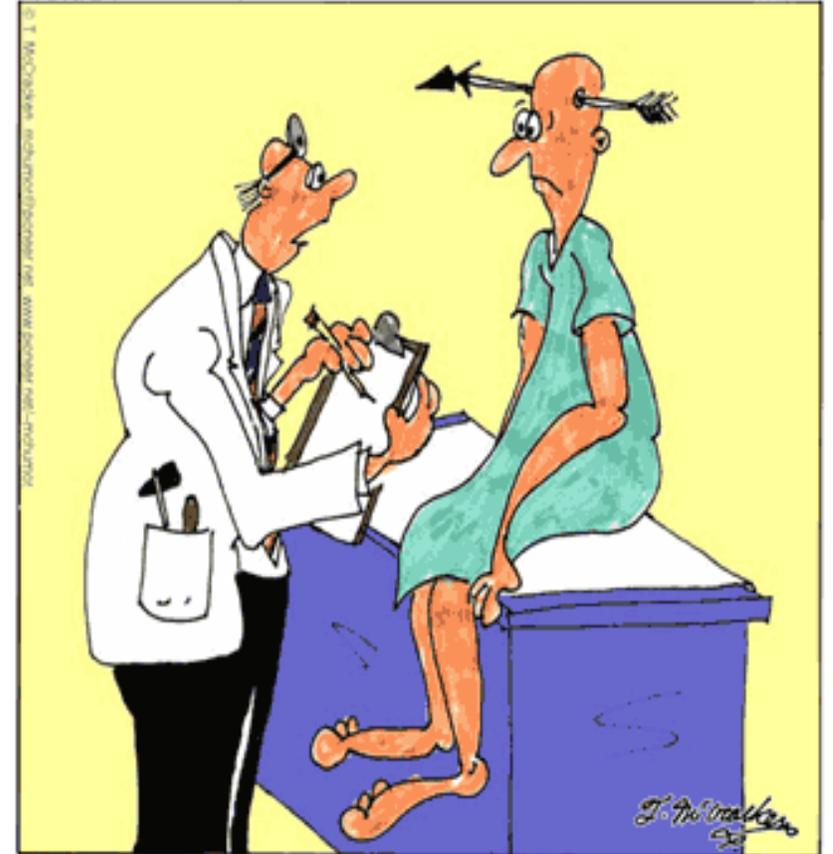
**Figure 4. Exertional status at time of death by common causes of sudden cardiac death.**

AN-SUD indicates autopsy-negative sudden unexplained death; CM, cardiomyopathy; LVH, left ventricular hypertrophy; NOS, not otherwise specified; and SCD, sudden cardiac death.

## What are we screening with?

- History & Physical
- Pre-participation Forms
  - (AHA/AAP 14-point evaluation)
- ECG?
- Echo?
- More?

**MCHUMOR** by T. McCracken



“Off hand, I'd say you're suffering from an arrow through your head, but just to play it safe, I'm ordering a bunch of tests.”

# Screening Children for Cardiac Risk

## *Primary Physician/Caretaker Screening - **Annual***

- Detailed history, inquiring about pertinent symptoms (Syncope, chest pain, palpitations, dizziness, rapid heart rate)
- Exercise history (endurance, types of exercise, exercise-associated symptoms)
- Discussions regarding illicit drug use, alcohol, caffeine, smoking, medications (including over-the-counter, “health/nutritional supplements”, anabolic steroids)
- Family history of congenital heart disease, arrhythmias, sudden death, inherited cardiac diseases (Long QT, HCM, Marfan)
- Physical exam: vital signs (BP), cardiac exam, 4-extremity pulses, perfusion, weight, general overall health and fitness

# Common Symptoms

- Palpitations
  - could be sinus tachycardia, ectopic beats, or a primary arrhythmia
- Shortness of breath/trouble breathing/"winded"
  - deconditioning or overtraining, asthma, anemia, poor sleep, poor hydration (very common!), performance anxiety, cardiac
- Fainting/near syncope
  - Exertional (bad), after exertion: usually orthostatic
- Chest pain
  - exertional vs. non-exertional; still usually chest wall pain or "normal" physiologic limit discomfort

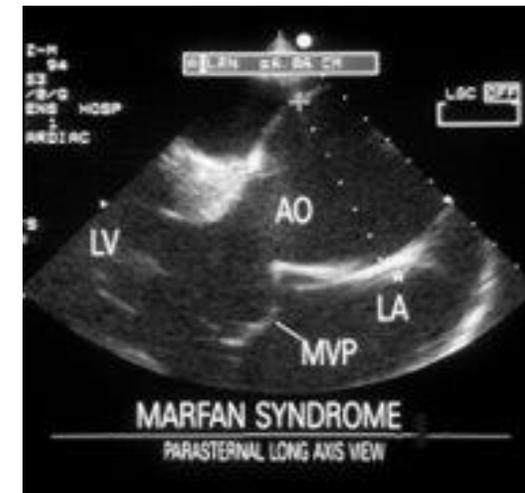
# When do I need a subspecialist?

- Referral suggested if:

- Exercise-associated symptoms or serious-sounding symptoms at rest
- If patient is limiting activity due to symptoms
- Suspicious history, physical, malignant family history or other potential risk factors

# Etiologies and screening

- Arrhythmogenic
  - EKGs may have more value (eg. LQTS, WPW)
  - CPVT: normal EKG at baseline, ventricular ectopy with exercise (burst)
- Cardiomyopathy
  - HCM: family history, usually (~90%) with EKG abnormalities (some false negatives, voltage criteria for LVH is not HCM)
  - ARVC: more common in certain regions/ethnicities
- CT disorder
  - Marfan Syndrome, Loeys-Dietz: family hx, PE findings
- SUD
  - Family history?
  - Post-mortem genetic testing?



## How good is the PPE?

- Limited data
  - Very low sensitivity and only “ok” specificity
- Sudden death (or arrest) is often 1<sup>st</sup> symptom
- Lack of uniformity
- Non-physicians?
- Pre-test probabilities, “non-cardiac” chest pain, syncope, murmur, etc.
- Family history questions (broad) vs. screening guidelines

**PPE should be done but be thoughtful and discerning!**

# Accuracy of ECG-inclusive preparticipation screening in athletes: more work to be done

*Expert Rev. Cardiovasc. Ther.* 10(6), 671–673 (2012)

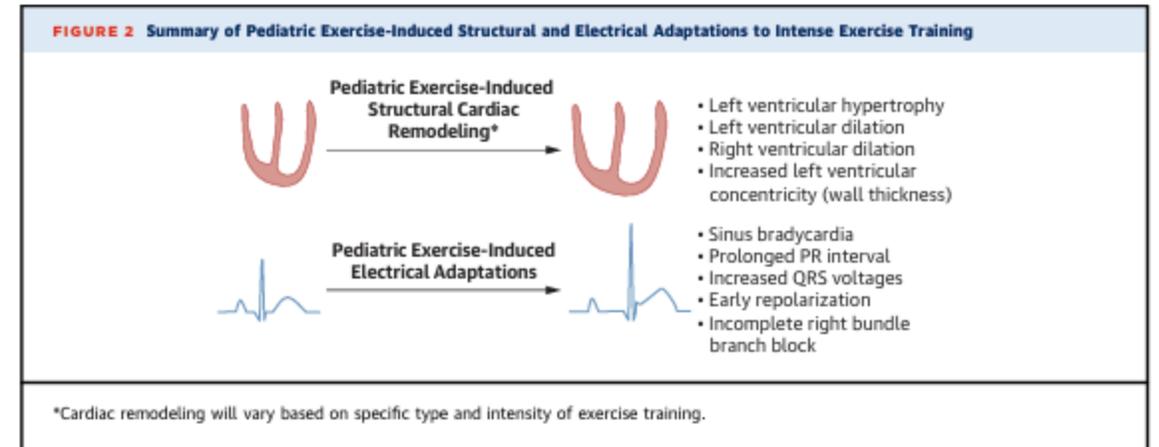
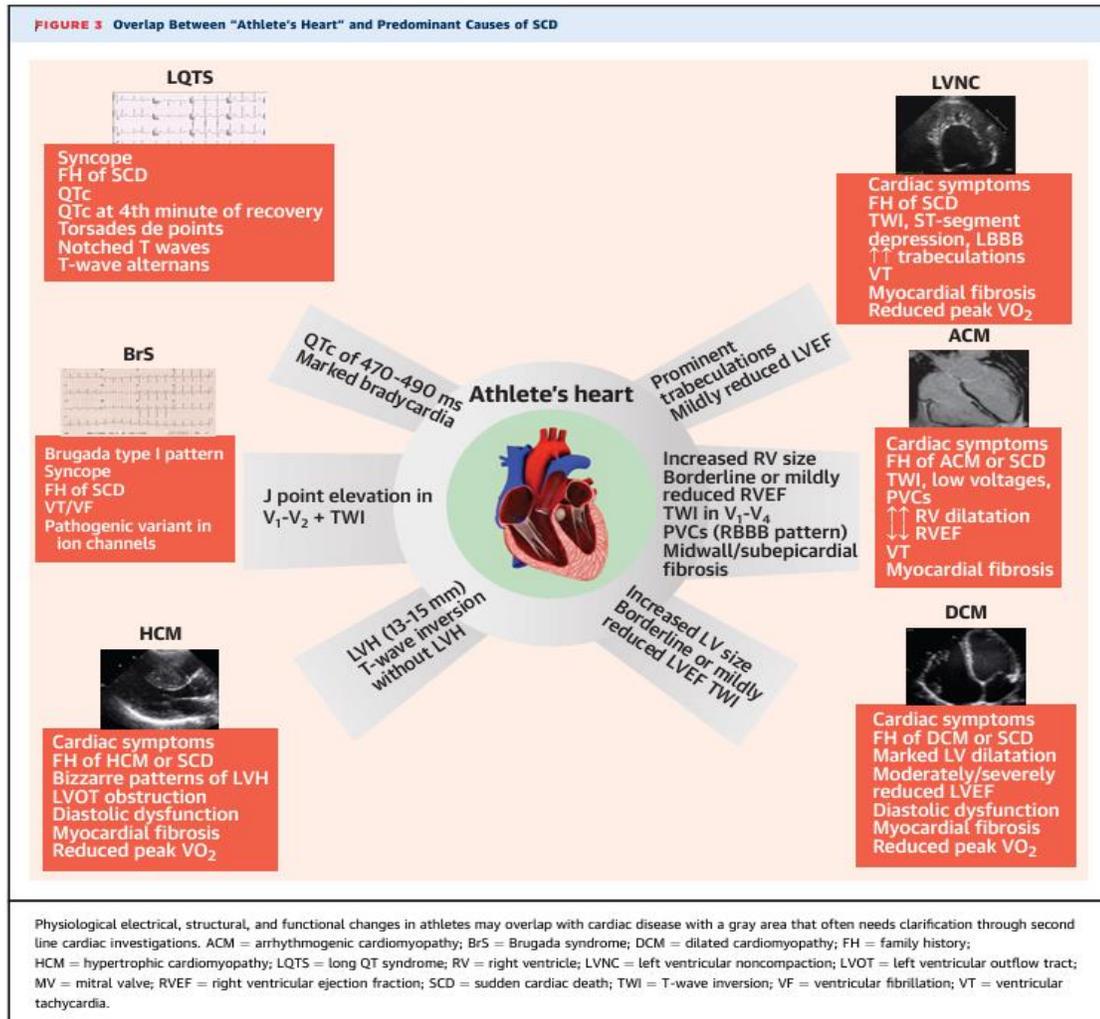
“The controversial role of ECG during preparticipation screening remains one of the most widely and passionately debated areas in cardiovascular and sports medicine.”

“Only with more data will we be able to responsibly determine the merits of adding ECG to the basic foundation of medical history and physical examination.”

## Should an EKG be done?

- Not currently recommended in the US (may change)
- Various local or national organizations may have specific requirements for other testing (ekg, echo, etc.)
- Better sensitivity and specificity than the PPE alone, but...
  - With cardiologist-read EKGs, 1-3% false positives
  - In black patients, up to 6-7% false positives
  - Computer read has a 2-5% false positive (possibly higher, eg. QTc)
  - HCM may have up to 10% false negative EKG and ARVC > 30% false negative
  - Yearly EKG? With higher SUD diagnoses (EKG findings preceding echo changes)

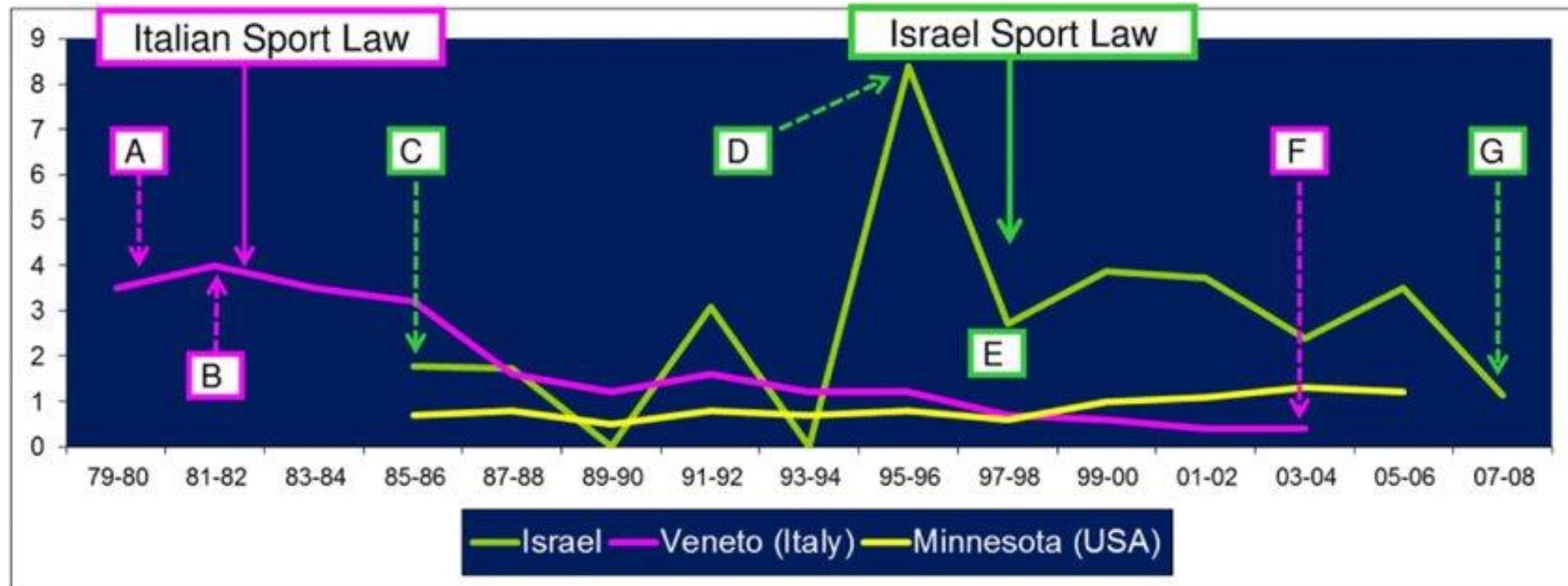
# It gets complicated...



Finocchiaro, G, Westaby, J, Sheppard, M. et al. Sudden Cardiac Death in Young Athletes: JACC State-of-the-Art Review. JACC. 2024 Jan, 83 (2) 350-370. <https://doi.org/10.1016/j.jacc.2023.10.032>

## Mandatory Electrocardiographic Screening of Athletes to Reduce Risk for Sudden Death: Proven Fact or Wishful Thinking?

Steinvil et al. JACC 2011



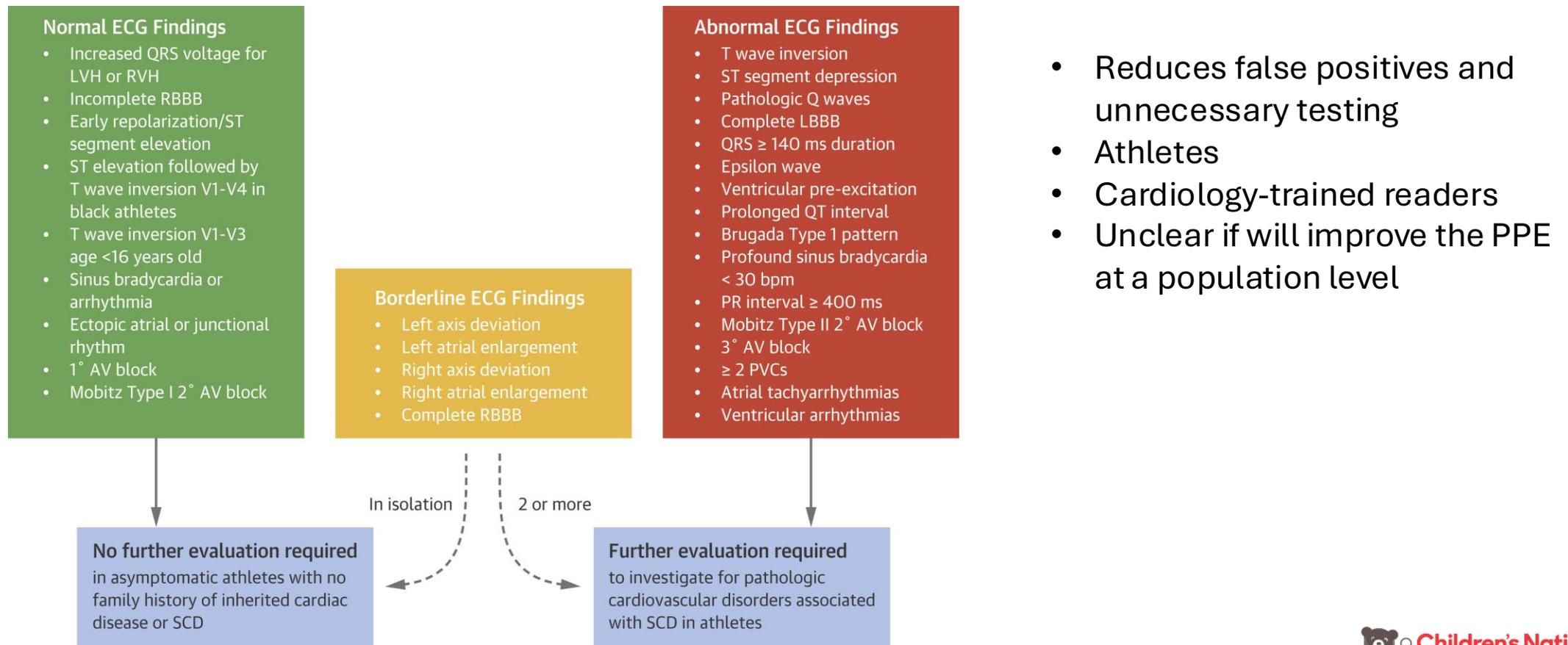
“Mandatory ECG screening of athletes had no apparent effect on risk for SCD”

# EKG Screening

- Italy vs. USA
  - What's different? (other than food and football)
  - Ethnic and racial heterogeneity; impacts test accuracy
  - Genetic and phenotypic differences (ARVD vs HCM)
  - Size of population to be screened (15M+ athletes in US)
- Cost of screening and payor (\$750 million in 2007 estimate)
- Inappropriate disqualification
- Unnecessary additional testing, with associated costs (over \$2 billion)
- Unnecessary anxiety
- Huge increase in physician manpower and other resources

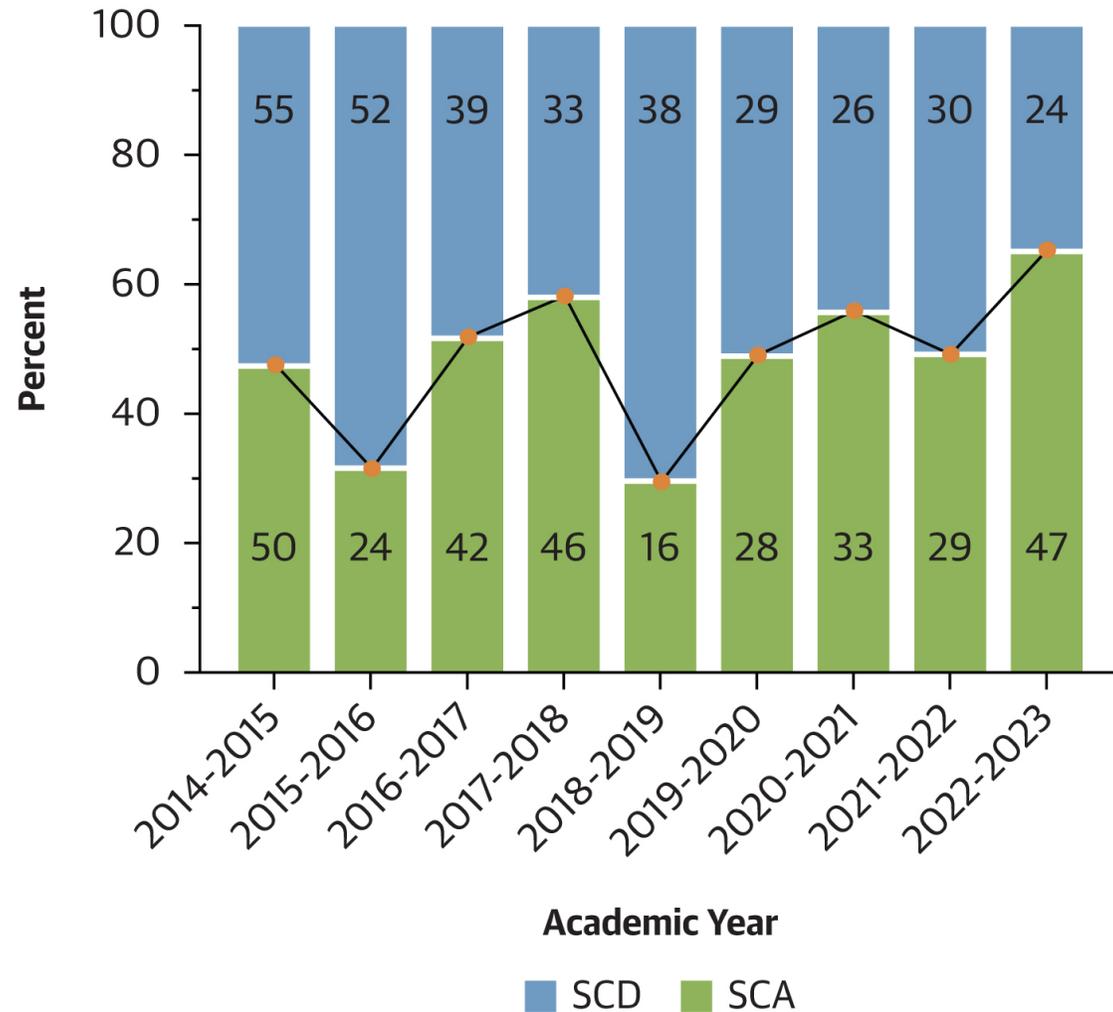
# International EKG Criteria

Sharma, S, Drezner, J, Baggish, A. et al. International Recommendations for Electrocardiographic Interpretation in Athletes. *JACC*. 2017 Feb, 69 (8) 1057–1075.



- Reduces false positives and unnecessary testing
- Athletes
- Cardiology-trained readers
- Unclear if will improve the PPE at a population level

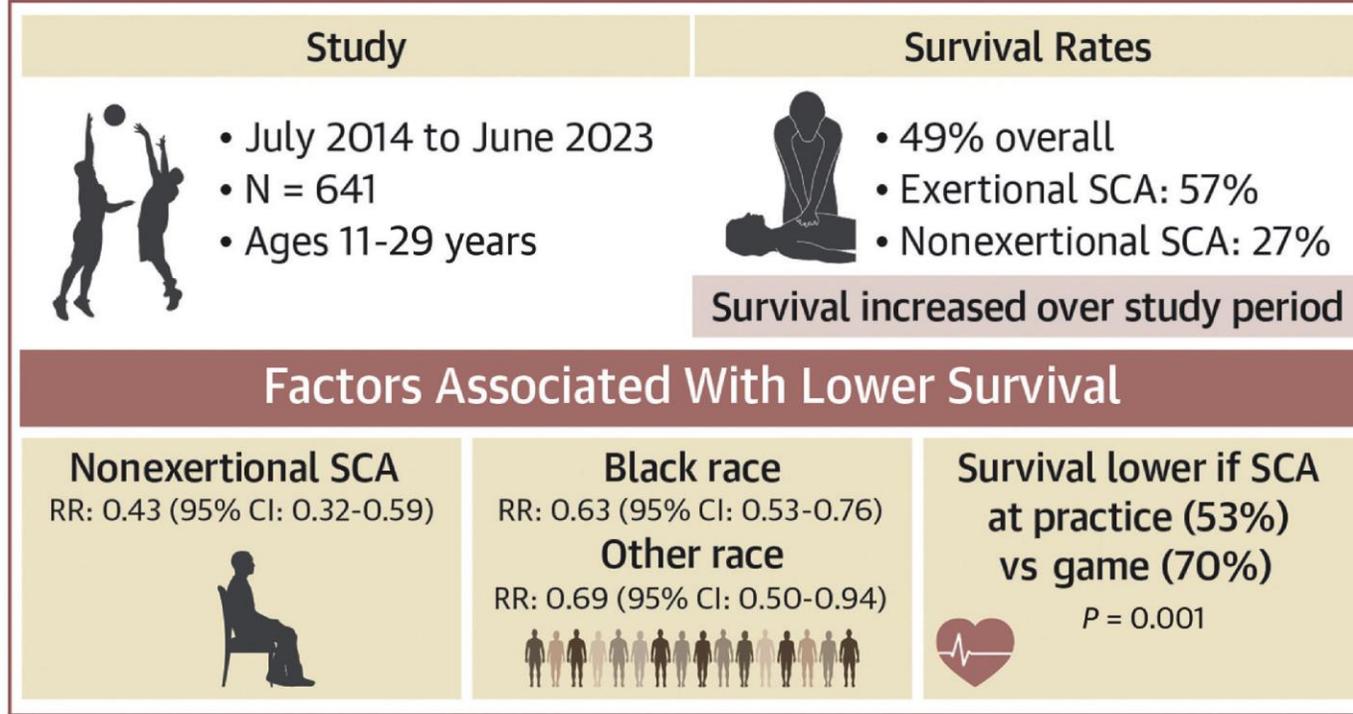
# Outcomes



Petek, B, Churchill, T, Moulson, N. et al. Survival Outcomes After Sudden Cardiac Arrest in Young Competitive Athletes From the United States. *JACC*. 2025 May, 85 (17) 1682–1692. <https://doi.org/10.1016/j.jacc.2025.03.006>

# SCA Outcomes

## CENTRAL ILLUSTRATION: Survival Outcomes After Sudden Cardiac Arrest in Young Competitive Athletes From the United States



Petek BJ, et al. JACC. 2025;85(17):1682-1692.

## Secondary Prevention of SCD (and morbidities)

- Emergency Action Plan
  - Identifying key personnel and roles
  - Ensuring access to life saving equipment (eg. AEDs)
  - Coordinating a multi-disciplinary team for a well-rehearsed response
- Coaches/staff (and teammates?) trained in BLS
  - Nearest to the player/allows for immediate resuscitation



# EKG Screening

- If EKG screening becomes recommended, likely only pediatric/adult cardiologists with expertise in sports cardiology should interpret them
- Carefully designed screening programs would be necessary (not large-scale EKG testing of everyone)
- Mortality benefit and unintended harms would need to be carefully monitored
- EAP development and implementation should be the priority

# **Management of Patients with Known Cardiac Abnormalities & Hemodynamic Assessment and Guidance for Safe Exercise**

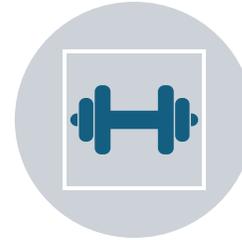
# Exercise Capacity in Congenital Heart Disease (CHD)

- Exercise capacity or peak  $\text{VO}_2$  is the how much oxygen can be delivered to the muscles to make energy necessary for exercise (also called cardiorespiratory fitness)
  - It includes the cardiovascular system, lungs, and muscles (mitochondria)
- Most patients with significant CHD have reduced  $\text{VO}_2$  at baseline than those of a similar age and weight without CHD
  - This does NOT mean they can't exercise, but rather something is limiting them (residual obstruction, hemodynamic or functional problem, post-op chest wall/lung abnormality)
  - **$\text{VO}_2$  is often modifiable; meaning the reason for the low  $\text{VO}_2$  isn't only because of the heart problem, but possibly because they aren't exercising regularly**

# Exercise Capacity in Congenital Heart Disease (CHD)



Many studies have shown a correlation between poor exercise capacity and outcomes



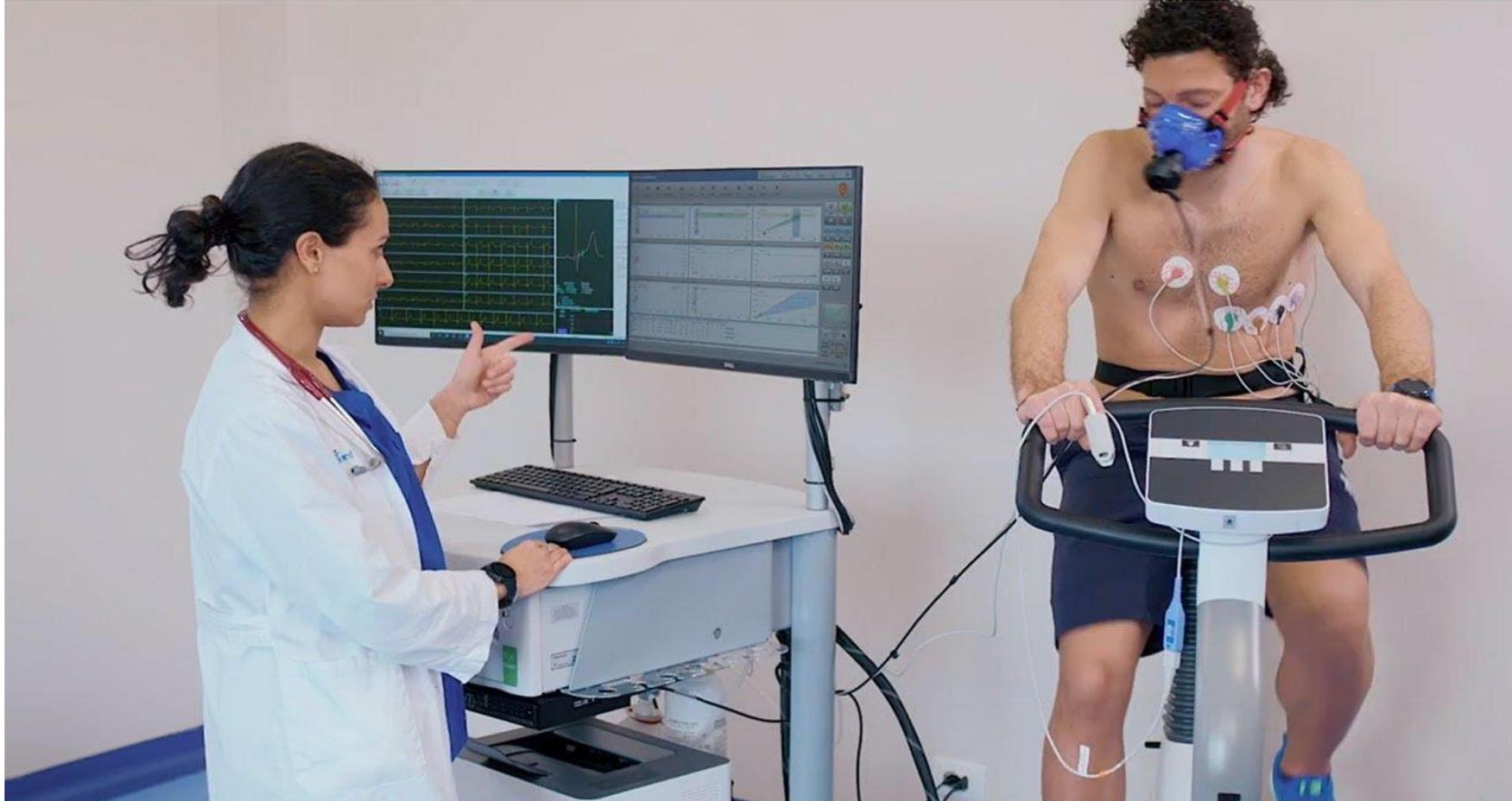
Exercise capacity *decreases* with age and many regular daily activities require a baseline capacity that those with CHD may not have

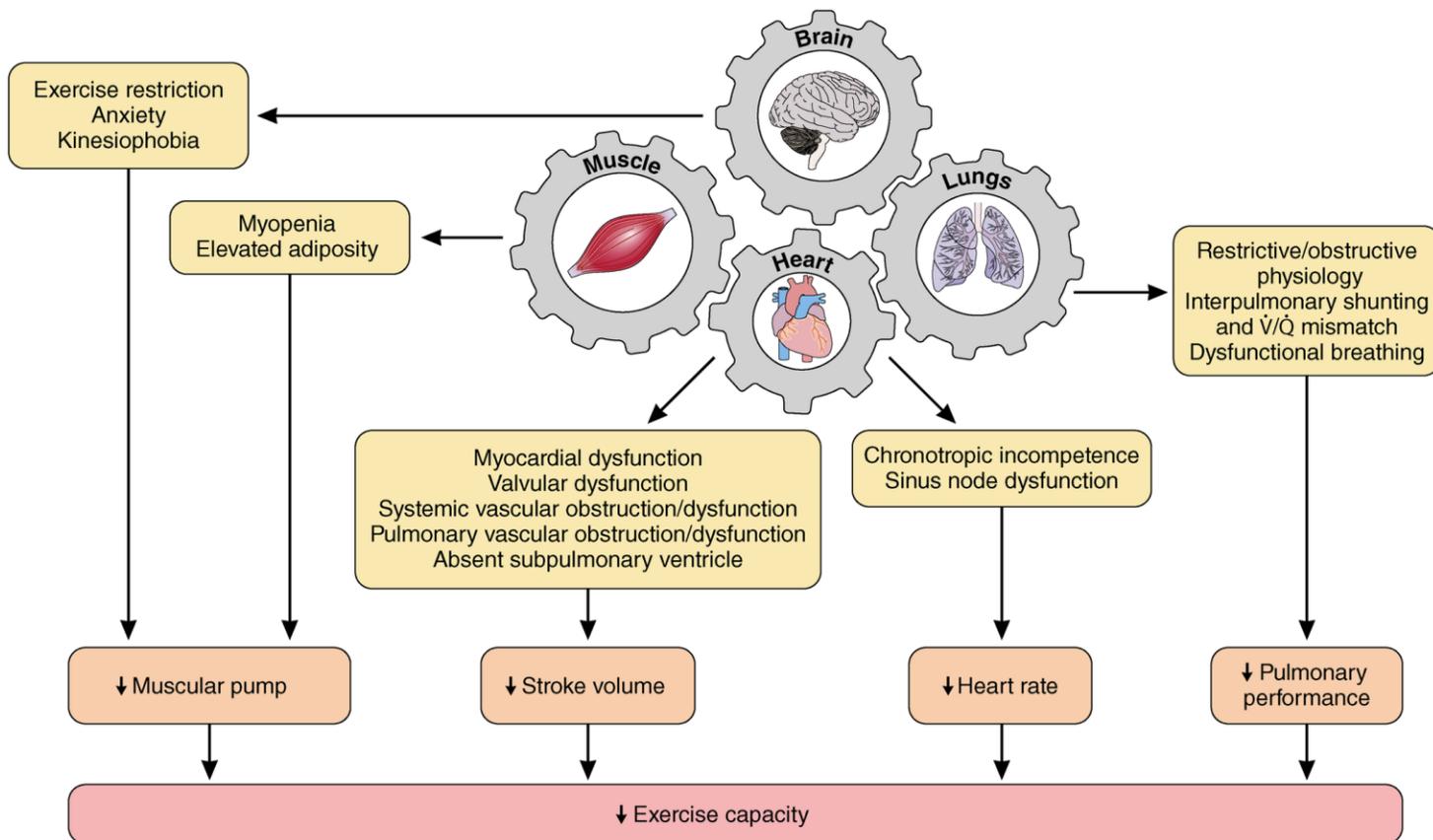


Exercise as a **therapy** may result in better outcomes and prognosis (and patients who report more activity as young children have better exercise capacity as adults)



Obesity (and being underweight) is associated with lower exercise capacity





Cifra et al. Cardiopulmonary Exercise Test Interpretation Across the Lifespan in Congenital Heart Disease: A Scientific Statement From the American Heart Association. JAHA (14):4, 9 Jan 2025.

# Indications for CPET

## Diagnosis

- Distinguishes the cause of exercise-related symptoms (heart, lungs, fitness, or other)
- Detects "unexpected" findings, often missed due to underreported symptoms
- Differentiates "athlete's heart" from cardiomyopathy
- Identifies hemodynamic issues not present at rest

## Risk Assessment

- Ensures exercise safety based on ACC/AHA guidelines

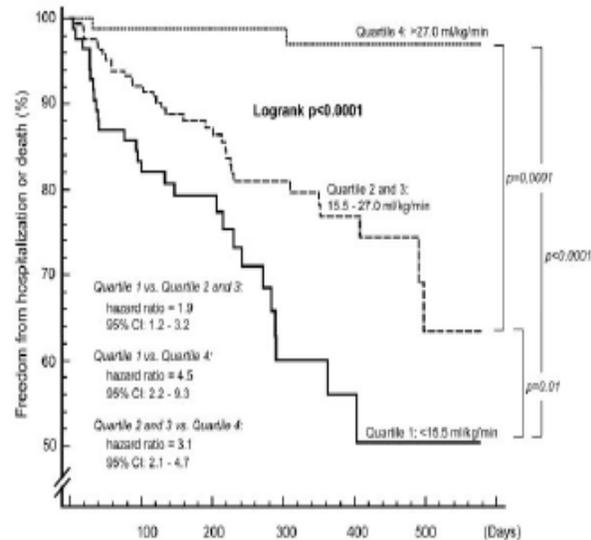
## Prognosis

- Poor CPET values correlate with worse outcomes (morbidity/mortality) and surgical risk while higher values suggest improved long-term outcomes
- Serial changes of exercise capacity often precede symptom development or abnormalities on echo/MRI and may be the best indicator for the need for reoperation

## Guidance

- Helps to create exercise prescriptions or cardiac rehab plans to improve short- and long-term outcomes using specific data from the test
- Regular testing familiarizes patients and families with normal exercise sensations, reducing anxiety about physical activity
- Helps to evaluate impact of antiarrhythmics or pacemaker function at physiological exercise state

# Peak VO<sub>2</sub> and Morbidity/mortality in CHD



Quartile 1: peak VO <sub>2</sub> <15.5 ml/kg/min						
At risk	84	68	41	28	10	3
Event-free survival	100%	82.1%	70.3%	60.1%	56.1%	50.0%

Quartile 2 and 3: peak VO <sub>2</sub> 15.5 - 27.0 ml/kg/min						
At risk	184	147	104	63	23	11
Event-free survival	100%	92.1%	87.2%	80.8%	76.8%	63.4%

Quartile 4: peak VO <sub>2</sub> >27.0 ml/kg/min						
At risk	81	70	60	55	52	10
Event-free survival	100%	98.8%	98.8%	97.8%	97.8%	97.0%

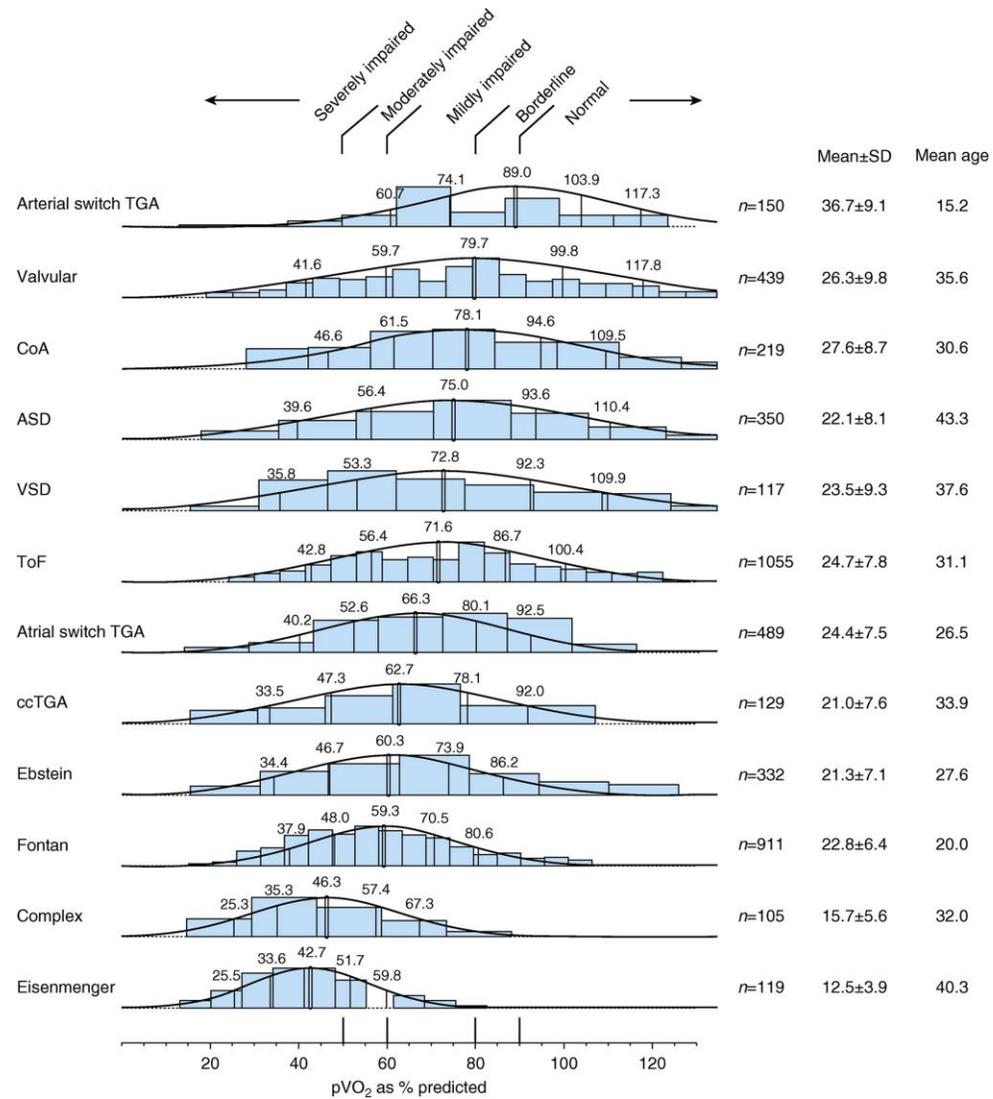
**Figure 4.** Kaplan-Meier plots for combined and point of hospitalization or death (event-free survival). Patients were classified into increasing quartiles (1 through 4) of peak  $\dot{V}O_2$  (peak  $VO_2$ ), and hazard ratios, 95% CIs, and log-rank probability values for comparisons between quartiles are shown.

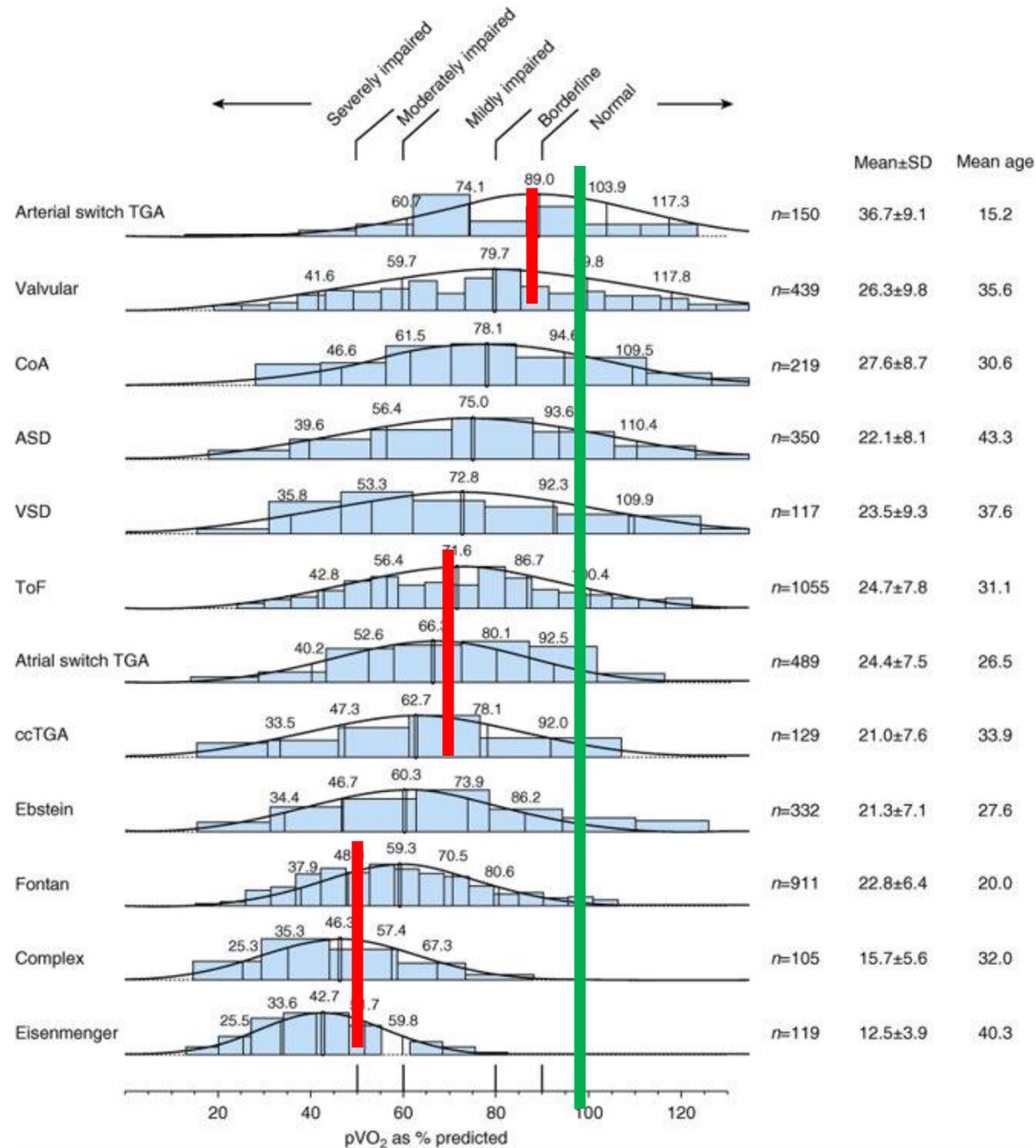
**TABLE 5. Significant Predictors of Hospitalization or Death on Cox Proportional-Hazards Analysis**

	P	Hazard Ratio	95% CI for Hazard Ratio
<b>Single-variable analysis</b>			
NYHA class	<0.001	2.556	1.790–3.652
Peak $\dot{V}O_2$	<0.001	0.908	0.873–0.943
Diagnosis	0.04	...	...
Peak heart rate	<0.001	0.985	0.976–0.991
Age at surgery	0.04	1.018	1.000–1.036
<b>Multivariable analysis</b>			
NYHA class	0.002	2.150	1.317–3.486
Peak $\dot{V}O_2$	0.01	0.937	0.890–0.986

Hazard ratio refers to unit increase in NYHA class, peak  $\dot{V}O_2$  ( $mL \cdot kg^{-1} \cdot min^{-1}$ ), heart rate (bpm), and age at surgery (years).

# Exercise capacity in CHD



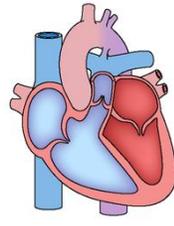




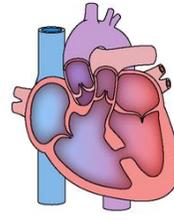
Elite athlete



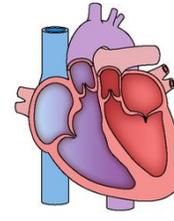
No heart disease



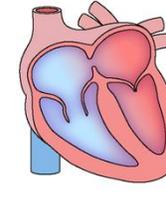
ASD  
VSD  
PDA



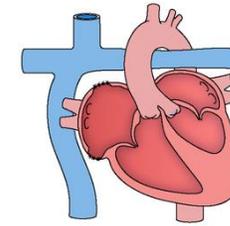
Tetralogy of Fallot



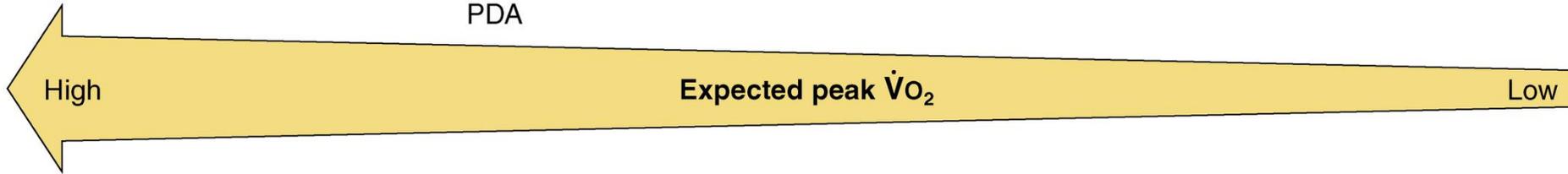
TGA s/p atrial switch



Ebstein anomaly



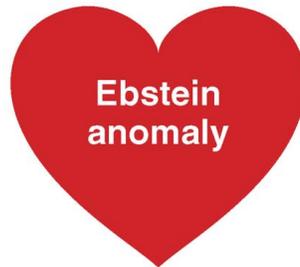
Fontan circulation



$\dot{V}O_2 < 52.3\%$   
 $VE/VCO_2$  slope  $> 35.4$



$\dot{V}O_2 < 62\% - 70\%$   
 $VE/VCO_2$  slope  $\geq 31$



$\dot{V}O_2 < 60\%$



$\dot{V}O_2 < 16.6$  mL/kg/min  
10% decrease in peak  $\dot{V}O_2$   
3% decrease/y  
VAT  $< 9$  mL/kg/min  
HR  $< 122$  beats/min

Cifra et al. Cardiopulmonary Exercise Test Interpretation Across the Lifespan in Congenital Heart Disease: A Scientific Statement From the American Heart Association. JAHA (14):4, 9 Jan 2025.



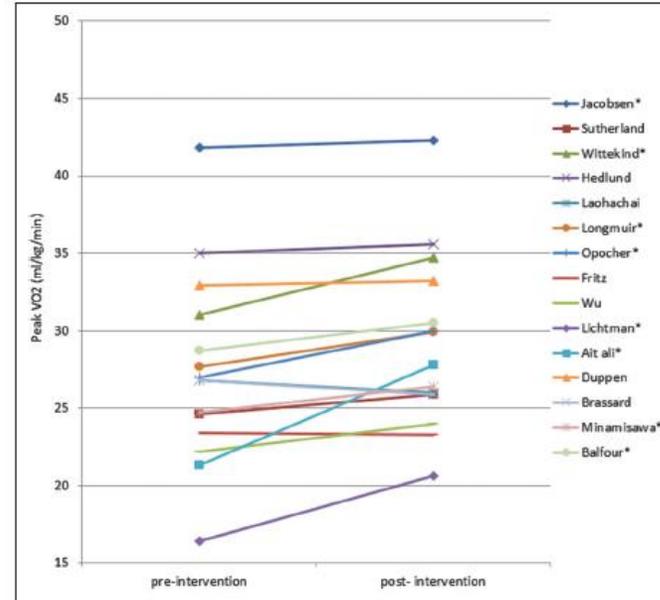
Children's National™

# Exercise training in Fontan patients

Sheffers et al, Physical exercise training in patients with a Fontan circulation: A systematic review, EJPC, 2020

Scheffers et al.

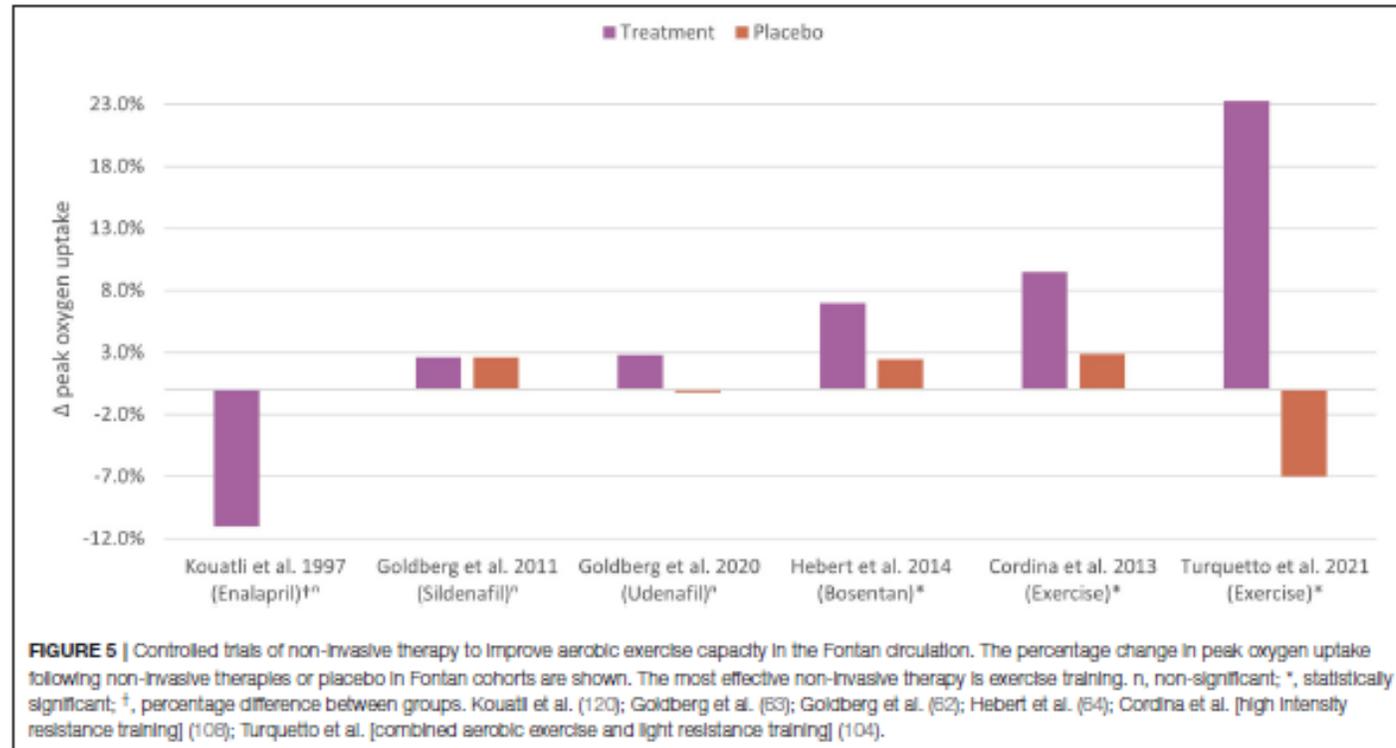
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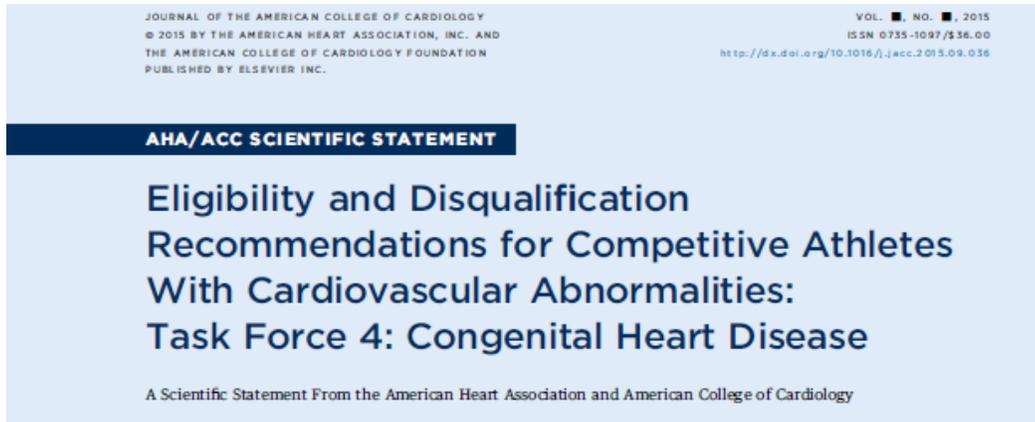
**Figure 2.** Peak oxygen uptake (VO<sub>2</sub>) changes before and after training. Cordina et al. is not shown, the study only mentioned improvement of peak VO<sub>2</sub> in ml/min and percentage of predicted. \*Significant increase.

# Fontan and exercise capacity

Tran et al. Exercise Intolerance, Benefits, and Prescription for People Living With a Fontan Circulation: The Fontan Fitness Intervention Trial (F-FIT)—Rationale and Design. *Frontiers in Pediatrics*, 2022



# Eligibility Recommendations for Athletes with CHD



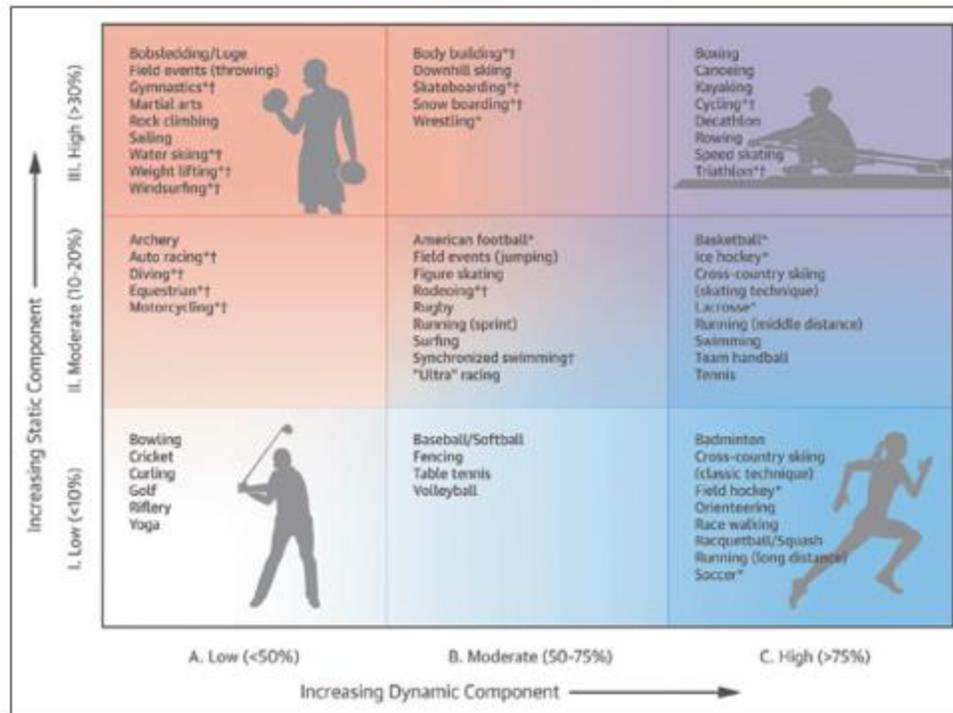
2015



2025

The level of sports participation recommended includes consideration of both the training and the competitive aspects of the activity but must be individualized to the particular patient, taking into account the patient's functional status and history of surgery. Noninvasive testing, such as formal exercise testing, Holter monitoring, echocardiography, and cardiac magnetic resonance imaging studies, is also often useful.

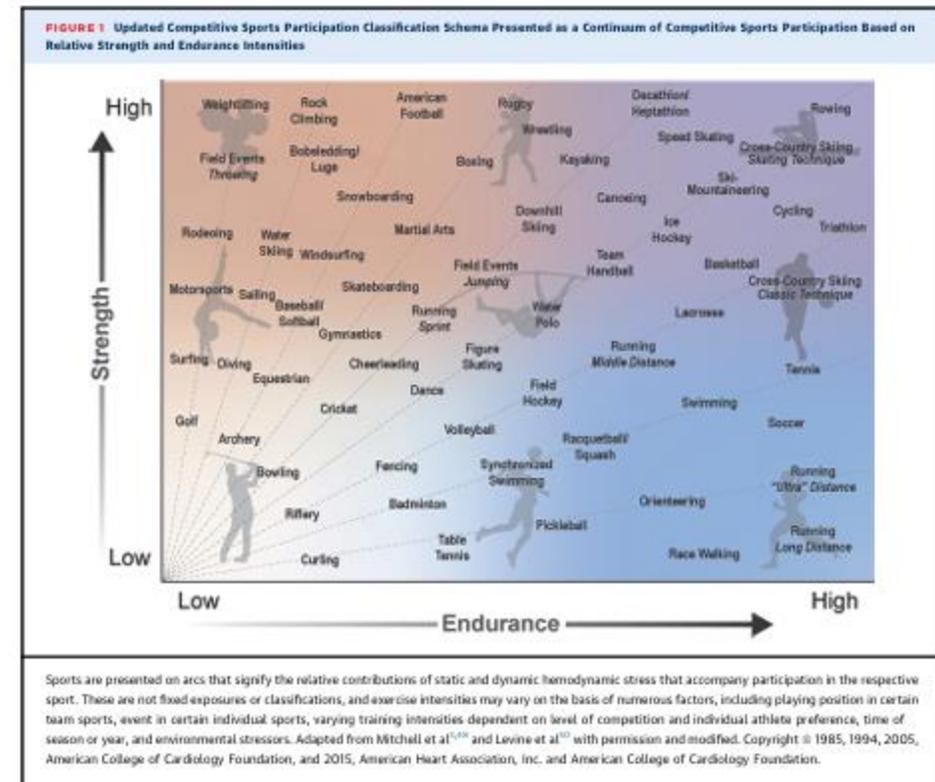
2015



**Figure 1. The classification of different sports/exercises is based on the relative contribution of static vs dynamic exercise intensity.**

Reprinted from Levine et al<sup>2</sup> with permission. Copyright ©2015, the American Heart Association, Inc, and the American College of Cardiology Foundation. Reprinted from Mitchell et al<sup>1</sup> with permission. Copyright ©2005, the American College of Cardiology Foundation.

2025



# Problems with Eligibility Guidelines for CHD

- Dealing with competitive and contact team sports
- Situations where patient and coach may push beyond comfort zones/pressure to perform: organized, emphasis on performance, high intensity
- May unnecessarily restrict patients from beneficial (and fun!) physical activity
  - These activity restrictions are based on the inherent risk of the activity rather than the intensity of a specific patient performing that activity
- Lack of published guidelines for non-competitive sports

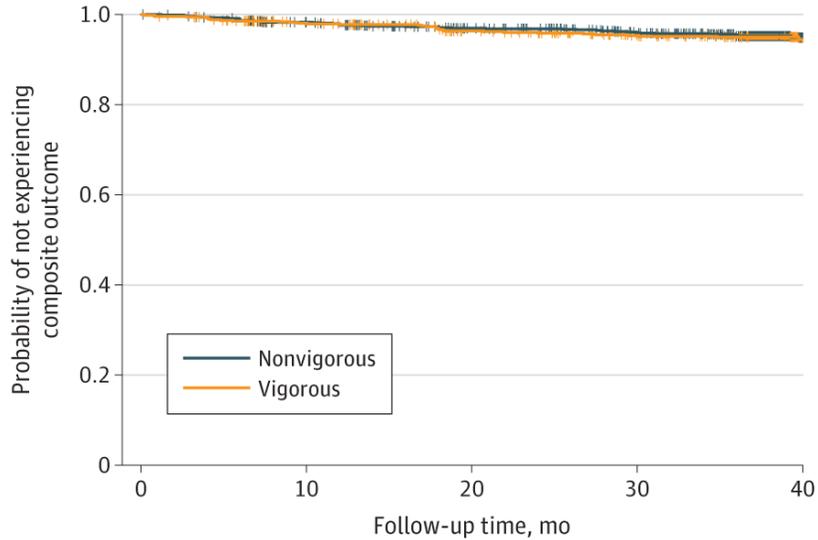
\*\*\* Do not conflate recreational/leisure physical activity with these guidelines!

- ✓ Leisure activities are typically submaximal!
- ✓ Aerobic (submaximal) exercise capacity is normal in most CHD patients even when maximal exercise capacity is decreased
- ✓ Numerous studies showing safety of exercise training in CHD, specifically at submaximal levels

## Change in Focus?

- As surgical outcomes and long-term prognosis (with longer life expectancy) improve, we need to shift our focus to long-term physical functioning and quality of life
- More rapid decline in exercise capacity over time in CHD patients than age and sex-matched controls and possibly starting out somewhat impaired exercise capacity means we need to do more earlier in life
- Recent data in (risk assessed and treated) patients with HCM (LIVE-HCM) – no difference in event rate with exercise, long QT syndrome (LIVE-LQTS) – 2 SCA, not with exertion, and nearly **all** cardiac rehab/exercise testing studies in children with known cardiac issues show a relatively low risk of SCA with physical activity

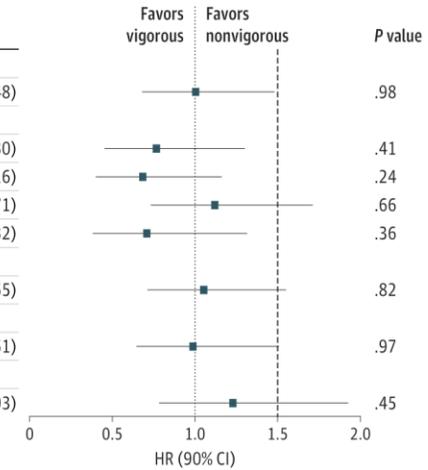
# LIVE-HCM



No. at risk			
Nonvigorous	961	852	262
Vigorous	699	608	193

Kaplan-Meier Survival Curve for Freedom From Composite End Point (Death, Cardiac Arrest, Appropriate Implantable Cardioverter Defibrillator Shock, or Arrhythmic Syncope) by Exercise Group. Vigorous and nonvigorous groups did not differ in freedom from composite end point.

Analyses	No.	HR (90% CI)
Primary analysis		
Vigorous vs nonvigorous	1660	1.01 (0.68-1.48)
Secondary/post hoc analysis		
Vigorous vs sedentary		0.77 (0.45-1.30)
Moderate vs sedentary		0.69 (0.40-1.16)
Vigorous vs moderate		1.12 (0.73-1.71)
Vigorous competitive vs nonvigorous	1220	0.71 (0.39-1.32)
Overt HCM <sup>a</sup>		
Vigorous vs nonvigorous	1534	1.06 (0.72-1.55)
Overt HCM, fully adjusted <sup>b</sup>		
Vigorous vs nonvigorous	1309	0.99 (0.65-1.51)
Overt HCM, NYHA class I <sup>c</sup>		
Vigorous vs nonvigorous	1173	1.23 (0.79-1.93)



Forest Plot for Hazard Ratio (HR) (1-Sided 95% CI) Comparing Composite Outcomes Between Exercise Groups. HRs for primary, secondary, and post hoc analyses comparing the composite outcome (death, cardiac arrest, appropriate implantable cardioverter defibrillator [ICD] shock, arrhythmic syncope) between those exercising vigorously and those exercising nonvigorously. Presented are 90% 2-sided CIs. The upper limits of these intervals correspond to a 1-sided .05 significance level used to evaluate noninferiority. Primary analysis is shown followed by 2 secondary analyses: pairwise comparisons of the 3 groups and after excluding noncompetitive vigorous individuals to compare vigorous-competitive vs nonvigorous. Post hoc analyses are shown of subgroups.

<sup>a</sup>The first subgroup included those with overt hypertrophic cardiomyopathy (HCM), ie, phenotype-positive only, and controlled for prespecified covariates age, sex, race, recruitment method (site or self), age at diagnosis, and presence of an ICD.

<sup>b</sup>The next model added sudden cardiac death risk factors that differed by an effect size of at least 12% between the groups (history of sudden cardiac arrest and septal thickness).

<sup>c</sup>The final subgroup excluded those with exercise-related symptoms (ie, asymptomatic, phenotype-positive only).

# LIVE-LQTS

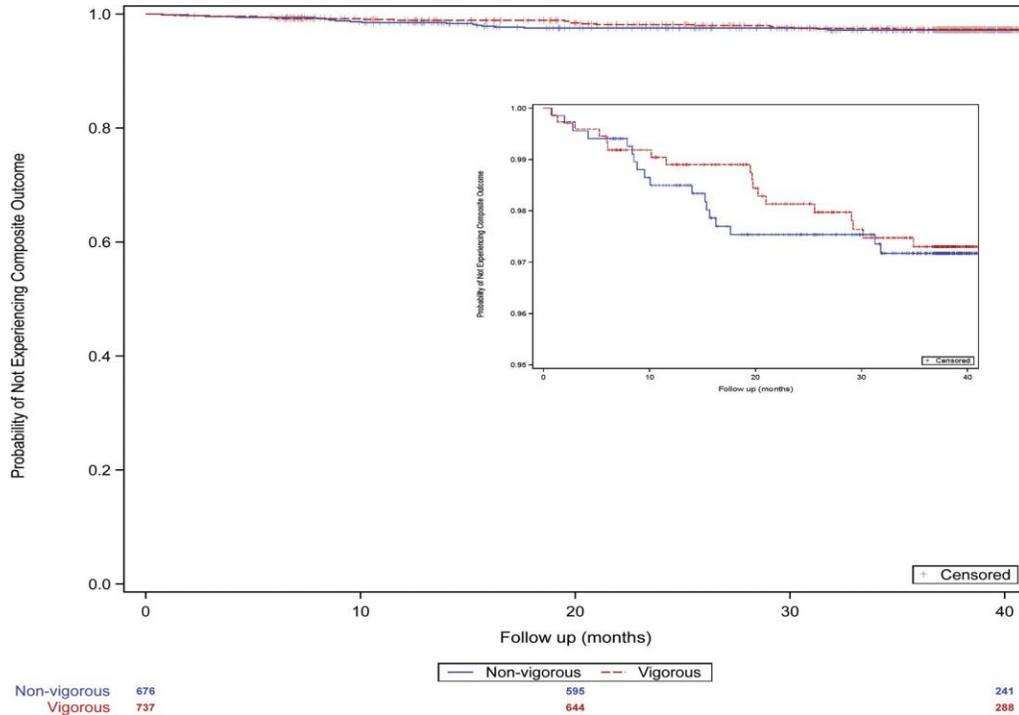


Figure 2. Kaplan-Meier survival curve for freedom from composite end point (death, sudden cardiac arrest, appropriate implantable cardioverter defibrillator shock, or arrhythmic syncope) by exercise group. There was no statistically significant difference in freedom from composite end points between those exercising vigorously and those exercising nonvigorously. Inset shows a magnified y axis.

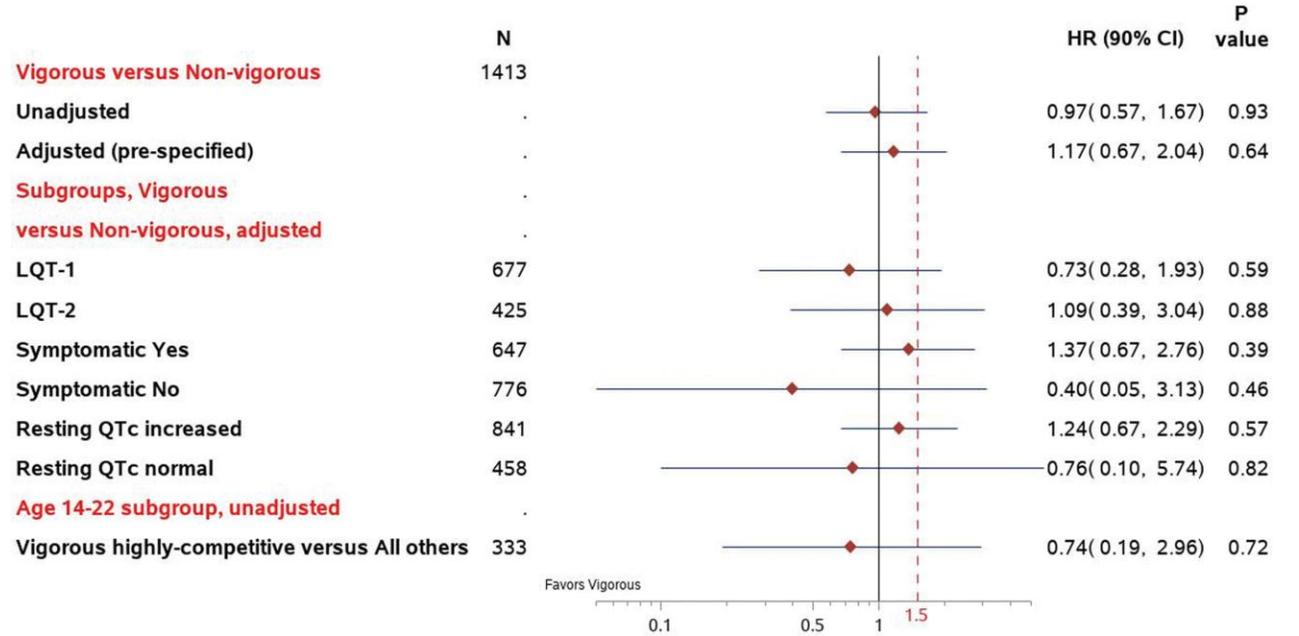
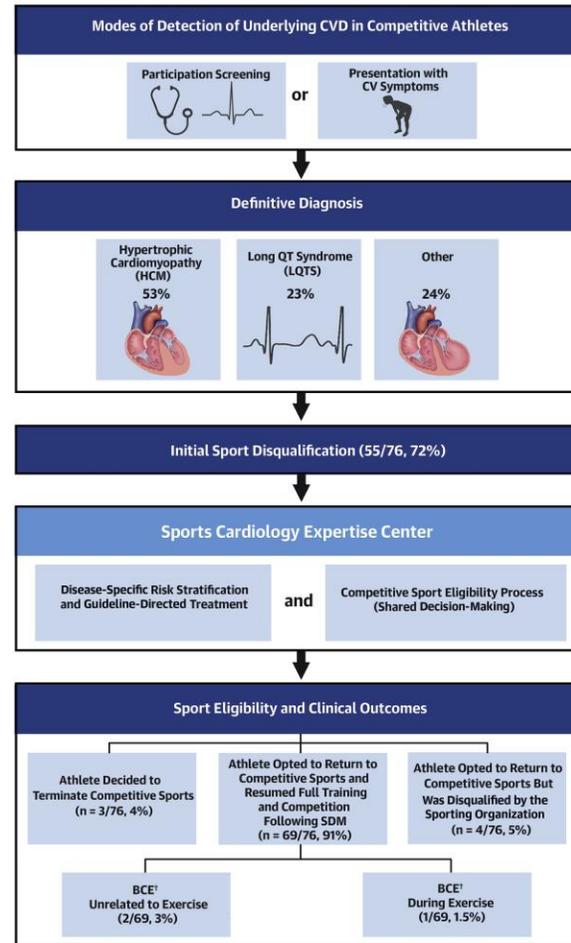


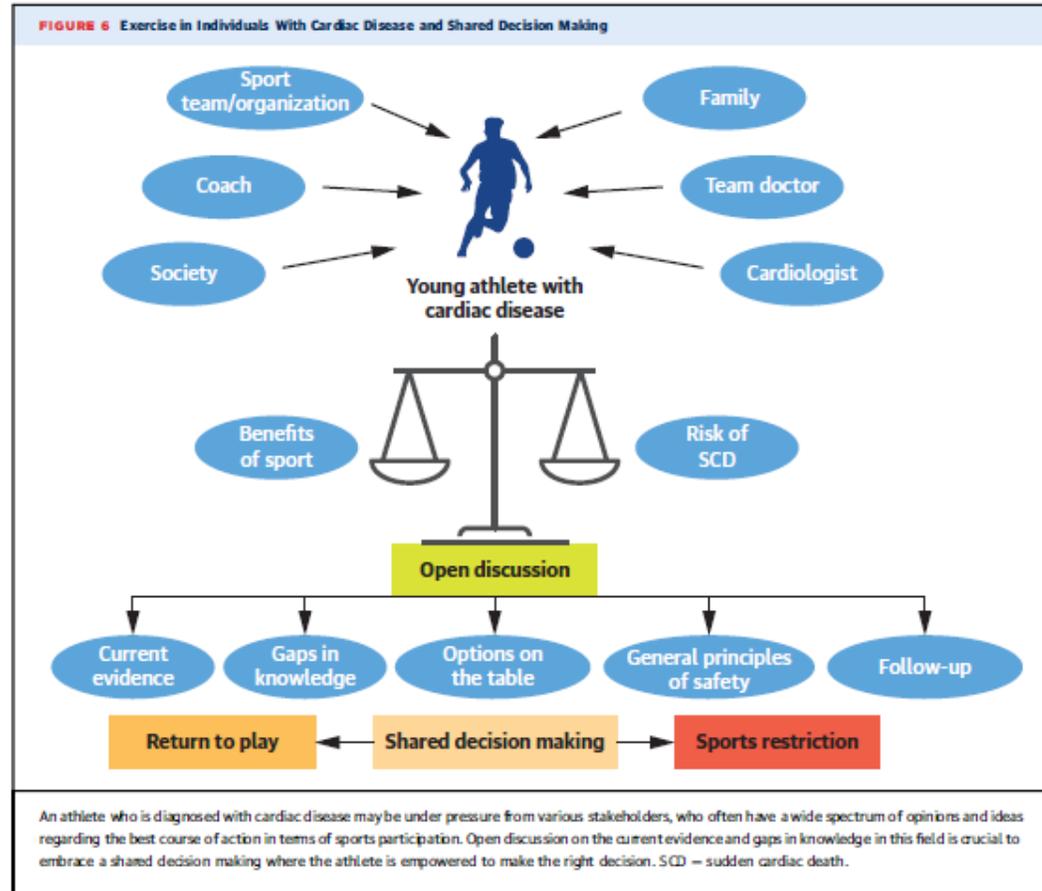
Figure 3. Forest plot for HR (one-sided 95% CI) comparing composite outcomes between exercise groups. Hazard ratios (HRs) for primary, secondary, and post hoc analyses comparing the composite outcome (death, sudden cardiac arrest, appropriate implantable cardioverter defibrillator shock, arrhythmic syncope) between those exercising vigorously and those exercising nonvigorously. The 90% 2-sided CIs are presented. Upper limits of these intervals correspond to a one-sided 0.05 significance level used to evaluate noninferiority. Primary analysis is shown followed by post hoc analysis including clinical factors (QTc and presence of previous symptoms) and then post hoc analyses of subgroups known to differ in outcome rates: first, those with the 2 most common long QT (LQT) genotypes, LQT1 and LQT2; next, those with and without previous symptoms (syncope or cardiac arrest); and then those with and without prolonged resting QTc ( $\geq 470$  ms for male participants or 480 ms for female participants). Finally, the subgroup of individuals from 14 to 22 years of age participating in vigorous-competitive exercise (varsity/junior varsity/traveling team) is compared with others in this age group.

## CENTRAL ILLUSTRATION: Return-to-Play Among Elite Athletes With Genetic CVD

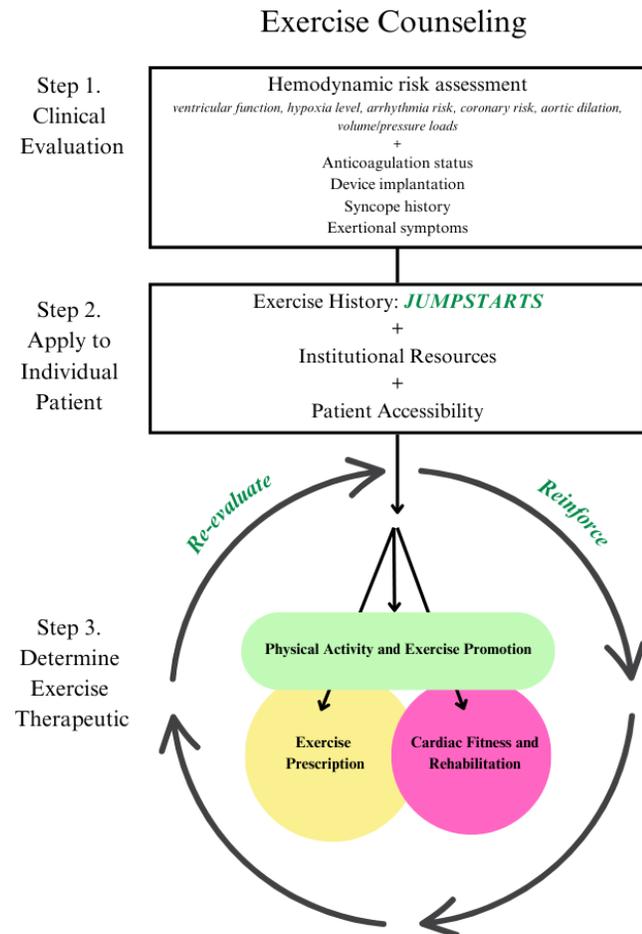


Martinez KA, et al. J Am Coll Cardiol. 2023;82(8):661-670.

# Shared Decision Making



# Exercise Counseling (applicable to all)



Pre-published: Taken from "Exercise Counseling in Congenital Heart Disease: A Guide for the Congenital Cardiologist"

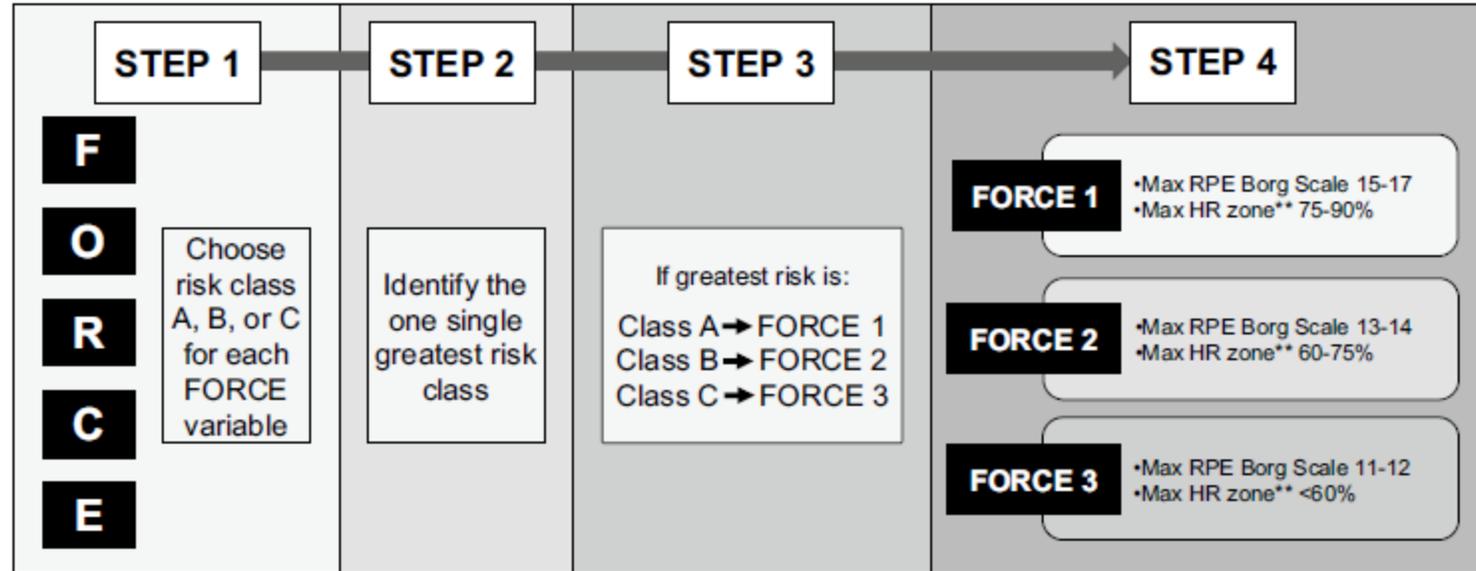
# FORCE

Table 1 FORCE definitions

FORCE Variable		RISK CLASS		
		CLASS A	CLASS B	CLASS C
<b>F</b>	Function -biventricular system or single RV/LV	Normal - Mild (ejection fraction $\geq 45\%$ )	Moderate (ejection fraction 30-44%)	Severe (ejection fraction $< 30\%$ )
<b>O</b>	Oxygen level - pulse oximetry rest/exercise	$\geq 90\%$	84 - 89%	$\leq 83\%$
<b>R</b>	Rhythm*	No/Low grade arrhythmia*	Moderate but stable arrhythmia*	Higher grade arrhythmia*
<b>C</b>	Complex physiology (inc. aortic size)/Coronary risk	Clinical judgement	Clinical judgement	Clinical judgement
	Aortic dilation	Absolute size $< 35\text{mm}$ (Z score $< 4$ )	Absolute size 36-49mm (Z score $\geq 4$ to $\leq 6$ )	Absolute size $> 50\text{mm}$ (Z score $> 6$ )
<b>E</b>	Elevated load: Hypertrophy (wall thickness)	Mild (Z score $< 4$ )	Moderate (Z score $\geq 4$ to $\leq 6$ )	Severe (Z score $> 6$ )
	Pressure load (outflow) (peak echo gradient)	Mild (gradient $\leq 35$ mmHg)	Moderate (gradient 36-65 mmHg)	Severe (gradient $> 65$ mmHg)
	Pulmonary hypertension (PA pressure)	Mild ( $\leq 50\%$ systemic)	Moderate (51-75% systemic)	Severe ( $> 75\%$ systemic)
	Ventricular volume load (end diastolic Z score)	Mild (Z score $< 4$ )	Moderate (Z score $\geq 4$ to $\leq 6$ )	Severe (Z score $> 6$ )
*Rhythm Designations				
No/Low grade arrhythmia	No arrhythmia to mild arrhythmia; no exercise restrictions			
Moderate but stable arrhythmia	Known but stable, well tolerated, or at least partially treated; some exercise precautions			
Higher grade arrhythmia	Potentially unstable, poorly controlled, or incompletely evaluated; typically with exercise restrictions			

Gauthier N, Reynolds L, Curran T, O'Neill J, Gauvreau K, Alexander ME. FORCE Risk Stratification Tool for Pediatric Cardiac Rehabilitation and Fitness Programs. *Pediatr Cardiol.* 2023 Aug;44(6):1302–10.

# FORCE



STEP 1: Evaluate each FORCE variable\* and identify each individual risk class\* (A= lowest, B= medium, C= highest)

STEP 2: Identify the single greatest risk class among all variables

STEP 3: Assign final FORCE Level based on single greatest risk category

STEP 4: Assign associated FORCE exercise intensity

\*see reference table for variable and class definitions

\*\*max intensity HR zone= $[(\text{max HR}-\text{resting HR})(\text{desired } \%) + \text{resting HR}]$   
HR= heart rate

RPE= rate of perceived exertion

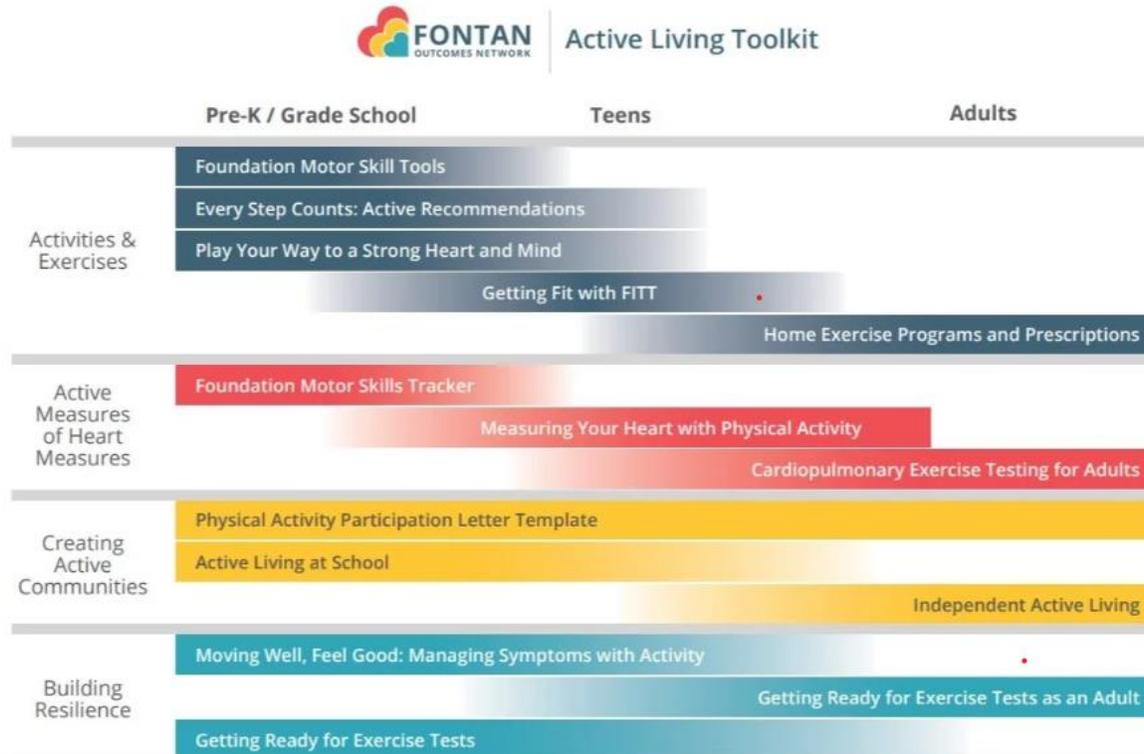


# JUMPSTARTS Exercise History

<b>J</b> oy	Activities, hobbies, extra-curriculars that bring the patient happiness
<b>U</b> sual habits	Activities done in a typical day or week such as walking to school, playing on the playground, physical education, extracurriculars
<b>M</b> indset	Current mindset toward physical activity and exercise (48)
<b>P</b> rior experience	Prior physical activity/exercise/sport experience earlier in life
<b>S</b> ymptoms	Symptoms with exercise including normal physiologic responses to exercise which may be interpreted as abnormal by the patient (e.g. breathing hard, heart rate increases), as well as red flag symptoms (e.g. symptoms of ischemic chest pain, exertional syncope)
<b>T</b> arget interests	What is the patient interested in doing more of in the future; do they have personal goals?
<b>A</b> pprehension	Fears or concerns related to exercise for the patient, the family, potentially the school or coach
<b>R</b> esources	What resources are available to the patient/family such as home exercise equipment, school gym, safe and walkable neighborhood
<b>T</b> ime	What time commitment is reasonable
<b>S</b> ustainable	The PAE intervention must be sustainable!

Pre-published: Taken from "Exercise Counseling in Congenital Heart Disease: A Guide for the Congenital Cardiologist"

# Toolkit to guide physical activity in Fontan patients



<https://www.svone.org/pae-tools/#clinician>

Use the buttons below to jump to a specific section of the toolkit.

- Activities & Exercises
- Building Resilience
- Active Measures Of Heart Measures
- Creating Active Communities

# Cardiac Rehabilitation in CHD

McBride et al, Cardiopulmonary Rehabilitation in Pediatric Patients With Congenital and Acquired Heart Disease, JCRP, 2020

**Table 2**  
**Cardiac Rehabilitation and Exercise Training in Pediatric Congenital Heart Disease**

Reference	Diagnosis	n	Age, yr	Program Training	Outcome
McBride et al (2007) <sup>4</sup>	Heart failure awaiting TX	20	13 ± 3.2	3 sessions/wk aerobic and resistance	Increased work efficiency (10%)
Duppen et al (2015) <sup>12</sup>	CHD and acquired heart disease	7	13-19	4-5 sessions/wk aerobic	Increased $\dot{V}O_2$ (20%) and TM time (21%)
Brassard et al (2006) <sup>8</sup>	Fontan	7	11-26	3 sessions/wk aerobic and resistance (↓ resting SBP 9 mm Hg)	Improved skeletal muscle function
Hedlund et al (2016) <sup>16</sup>	TOF/Fontan	93	10-25	3 sessions/wk aerobic	Improved $\dot{V}O_2$ (5%)
Longmuir et al (1991) <sup>11,a</sup>	CHD	129	10-16	2 sessions/wk	Improved $\dot{V}O_2$
Opocher et al (2005) <sup>9</sup>	Fontan	10	7-12	2 sessions/wk home training 2 times/wk	Improved $\dot{V}O_2$ (15%) Increased $O_2P$ (19%)
Rhodes et al (2005, 2006) <sup>5,6</sup>	CHD	30	8-17	2 sessions/wk aerobic and resistance	Improved $\dot{V}O_2$ (14%) Improved work (12%) Improved VAT (18%)
Avitabile et al (2014) <sup>17</sup> and Kirk et al (2014) <sup>18</sup>	Fontan, DCM	18	8-31	2 sessions/wk aerobic	Improved $\dot{V}O_2$ (11%) Increased $O_2P$ (12%)

Abbreviations: 6MWT, 6-min walk test; CHD, congenital heart disease; DCM, dilated cardiomyopathy;  $O_2P$ , oxygen pulse; SBP, systolic blood pressure; TOF, tetralogy of Fallot; TM, treadmill; TX, transplant; VAT, ventilatory anaerobic threshold;  $\dot{V}O_2$ , oxygen uptake.

<sup>a</sup>Data unavailable in online text.

Home based CR strategies have also mostly shown some improvement in exercise capacity

# CNH Cardiac Rehabilitation and Fitness Program

## Patient selection

- Patients ages 8-35 with CHD (repaired/unrepaired), pulmonary HTN, inherited arrhythmia, cardiomyopathy, or s/p heart transplant
- Who also meet one of the following criteria:
  - Sedentary lifestyle
  - Physical debilitation or recovering from intervention/surgery
  - Could benefit from a better understanding of and confidence in safe and effective exercise/PA

# Structure of the Cardiac Rehabilitation and Fitness Program at CNH

## 3-4 month program

- 3 models: in-person, virtual/home-based, and hybrid
- Sessions are one hour in length including exercise time with a warm-up and cool-down. 1:1 sessions.
- 2-3 sessions/week
- General fitness goals: 60% aerobic, 25% strength, 15% flexibility; will modify for disease process/goals
- Will incorporate educational components during the session as well as modules/reading materials as “homework”
- Home exercise plan 2-4 days/week

## Individual Treatment Plans (ITPs)

- An initial ITP is completed on first scheduled visit, and then **at least every** 30 calendar days from the initial and subsequent ITPs. ITPs focus on outcomes and patient goals.
- Discharge ITP is completed on last day of patient’s program. By this point, the patient should be independent to carry out a productive exercise program at home at least 3 times/week.
- Elements:
  - Exercise assessment/plan, cardiac risk, psychosocial, nutrition, and outcomes/goals**
    - **Examples for exercise assessment/tracking: Modified push-ups, plank, squat test, sit and reach**
  - Nutrition:** refer to nutritionist if necessary
  - Psychosocial:** mood, depression/anxiety, concerns regarding PA/exercise

**Mindset Training** – 3-4 sessions with Vicki Freedenburg (Certified mindfulness-based stress reduction instructor (MBSR))

**Neurocognitive testing**

Levels	Entry	Intermediate	Advanced
Aim	Familiarizing the patient with aerobic training and exercises that improve strength and endurance	Improving aerobic exercise capacity, muscular strength, and endurance	Improving and maintaining aerobic exercise capacity, muscular strength, and endurance
<b>Aerobic Training</b>			
Time/session (up to 30-60 min)	> 15 min	> 20 min	> 25 min
Frequency (x per week)	2	2-4	3-5
Intensity			
FORCE 1	HR 60-75% (RPE 12-15)	70-85% (12-16)	70-90% (13-17)
FORCE 2	50-75% (12-14)	60-75% (12-14)	65-75% (13-14)
FORCE 3	< 60% (11-12)	< 60% (11-12)	< 60% (11-12)
<b>Strength Training</b>			
Frequency (x per week)	2	2-3	2-3
# of exercises	3-6	4-6	6+
# of sets	1-2	2-3	2-3
# of reps*	10-15	10-15	8-15
<b>Flexibility Training</b> (should be done with all training sessions)			
Time/session	5 min	5 min	5 min

## Putting everything together...

### Case 1:

18 y/o F, HS crew, scholarship for D1 program, complaining of worsening times, some shortness of breath and something catching in her neck (she will frequently try to clear her throat) when exercising. Had a flu-like illness about a year ago, and then starting noticing these symptoms. No other medical problems.

Vitals/PE: normal

EKG: normal

Echo: Normal function and coronaries. Suspicion of a double aortic arch.

MRA: confirmed double aortic arch.

Next step?

# Case 1

CPET!

Peak VO<sub>2</sub> was 133% predicted, breathing reserve was 5%

Pre- and post-exercise spirometry was normal.

Diagnosis: Double aortic arch with some tracheal compression during increased cardiac output

Plan: Vascular ring repair

## Case 2

14 yo referred for high blood pressure on 3 separate occasions. Healthy, plays football (D-line). No symptoms. Mother and maternal grandparents with HTN. Father's hx not known.

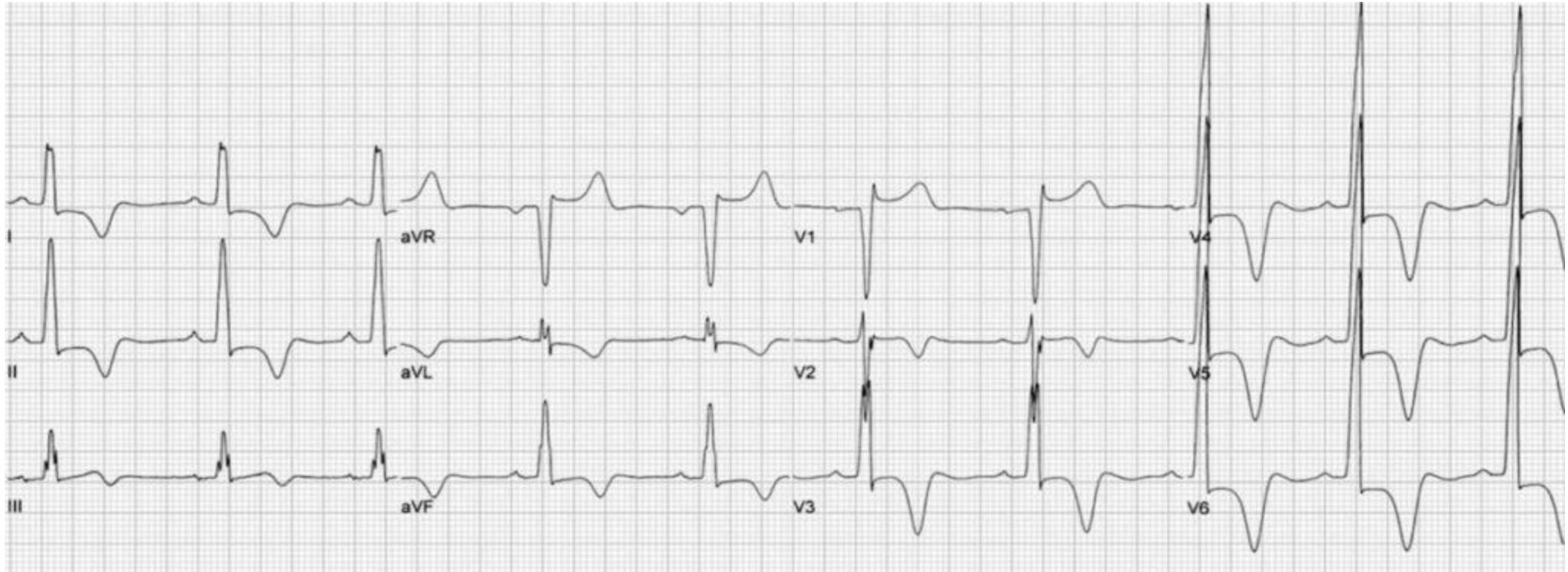
Vitals: BMI 31, RA BP 142/86, RL BP 150/88, normal cardiac exam

EKG: NSR, LVH with T wave inversion inferior and lateral leads

Echo: circumferential LVH

## Case 2

EKG





## Case 2

Next steps:

Concern for HCM

MRI: circumferential LVH, no asymmetric hypertrophy or fibrosis, largest wall thickness was 13 mm

CPET: VO<sub>2</sub> was 110% predicted. EKGs normalized at peak/early recovery. Peak exercise BP was 220/60.

## Case 2

Diagnosis: resting HTN and exercise induced HTN with secondary hypertrophy of the LV

Plan: anti-hypertensives

## Case 3

Healthy 16 yo AAU basketball player referred for syncope that occurred 3 times, all after warm-ups and prior to starting basketball games. Some dizziness when he stands up quickly. No exertional (during) symptoms. Drinks about 60 oz of water daily. Negative family history. No history of COVID or other known viral illnesses.

PE: vitals/exam normal

Ekg: (same as case 2), NSR, LVH with T wave inversion inferior and lateral leads

Echo: Normal function/coronaries. No MR. Mildly increased ventricular septal and apical thickness, measuring ~10-11 mm.

## Case 3

CPET: Peak VO<sub>2</sub> 98% predicted. Normal BP response. EKGs did not normalize with exercise.

MRI: mild LVH, slightly increased thickness of the apex, largest measurement 11 mm. A few patchy areas of fibrosis within the apical segment.

Diagnosis: Early HCM

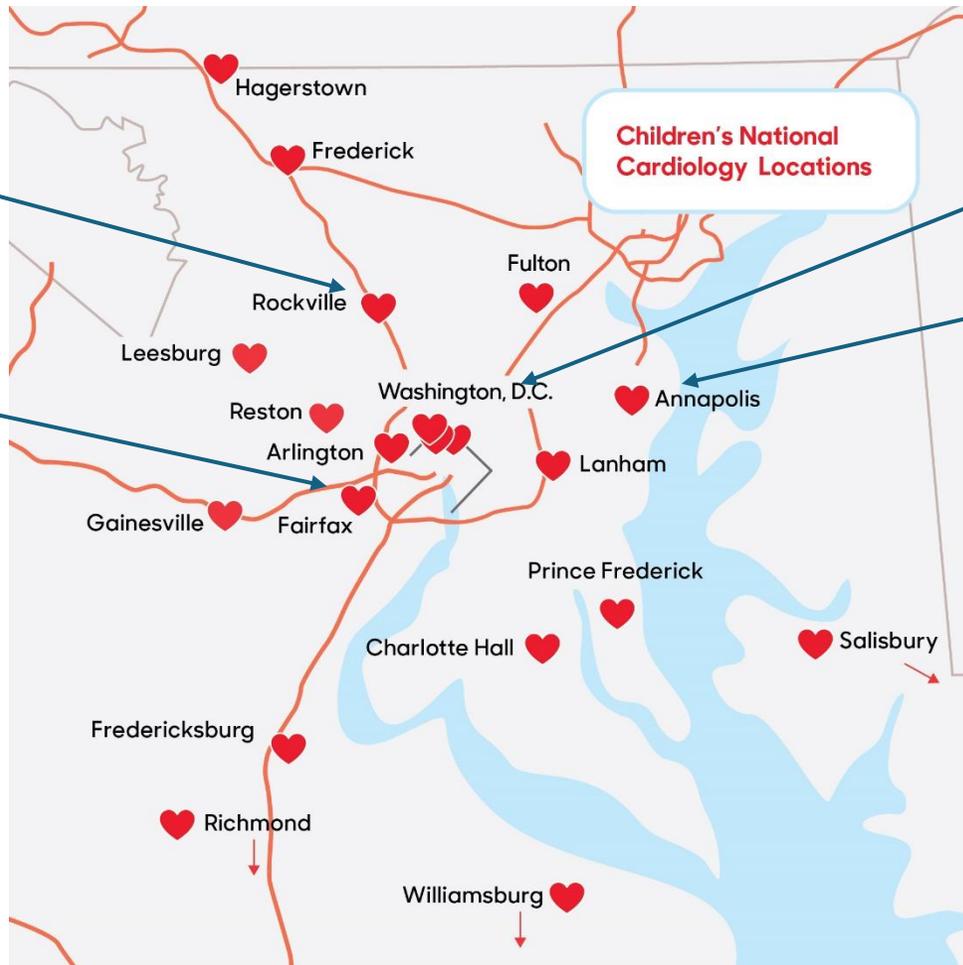
Plan: Genetic testing

Exercise: Given the early fibrosis, likely a high risk lesion. SDM with family; he and parents thought he should not continue basketball.

# Sports Cardiology Clinic Locations

Montgomery County  
Outpatient Center

Northern Virginia  
Outpatient Center



Main Hospital

Annapolis Outpatient Center

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Thank You!

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