

# Pediatric Chest Pain & Sports Participation in Young Athletes

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Wenatchee  
Cardiac Symposium



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# Pediatric Chest Pain and Sports Participation

## Objectives

- Understand etiologies of chest pain in the pediatric population
- Review CV sports screening guidelines for young athletes
- Identify risk factors for sports participation

# Pediatric Chest Pain

- Etiologies
- Clinical Presentation
- Diagnostic testing
- Clinical Management

# Pediatric Chest Pain – Etiologies

- Congenital Heart Disease (CHD)
- Acquired Heart Disease (AHD)
- Genetic Heart Disease (GHD)

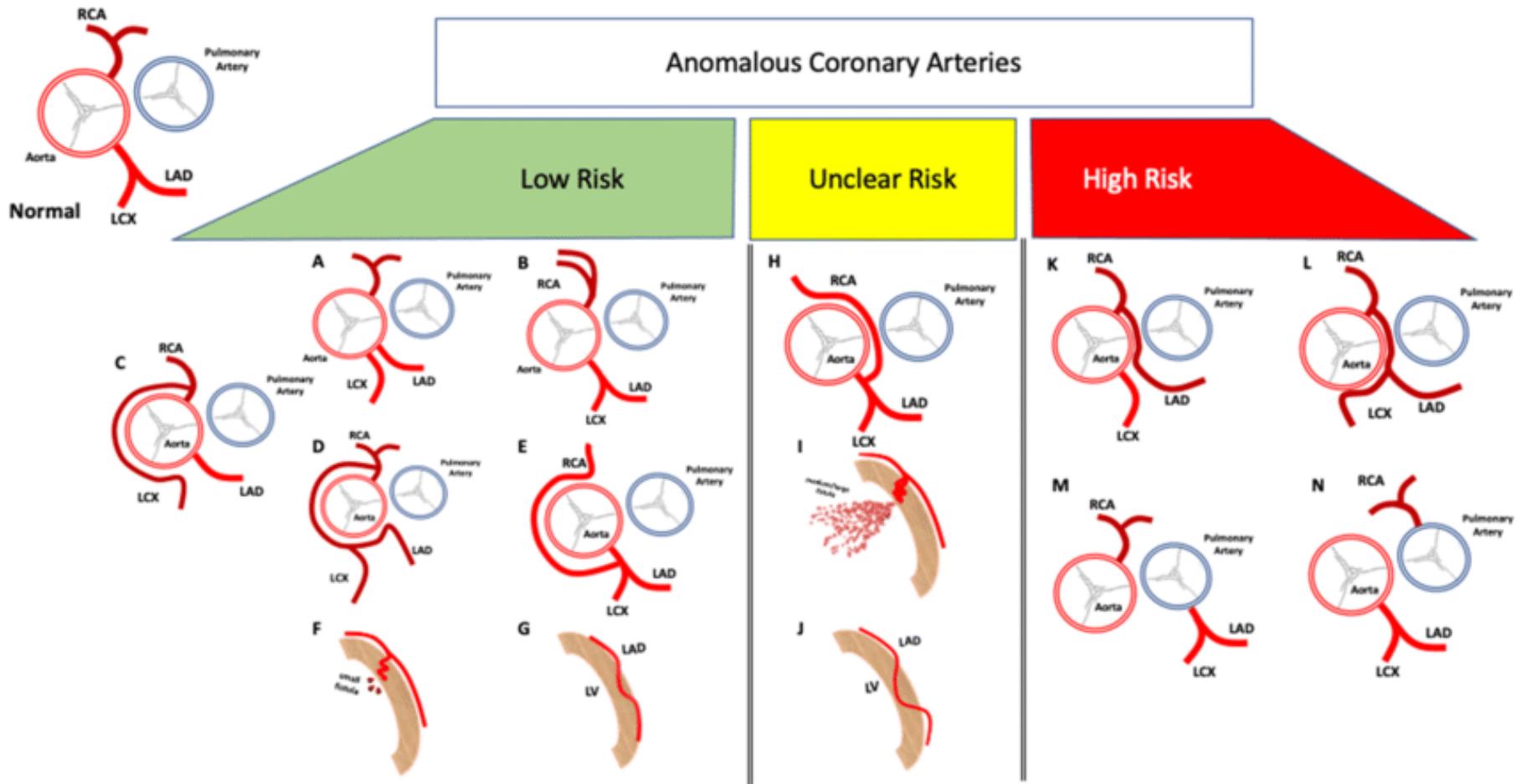
# Pediatric Chest Pain – CHD

- Congenital Heart Disease
  - **Most**(\*) forms of CHD (i.e. intracardiac shunts, valvulopathy, coarctation, patent ductus arteriosus, etc.) do *NOT* present a substrate for sensation of chest pain
    - Any associated physiologic imbalance does *NOT* result in myocardial ischemia
  - Coronary artery origin abnormalities
  - Left heart obstructive lesions
    - Aortic stenosis (\*)
    - Supravalvar aortic stenosis (\*)
      - William's syndrome, Noonan syndrome
    - Hypoplastic left heart syndrome

# Pediatric Chest Pain – Coronary Origin Anomalies

- Coronary artery origin abnormalities
  - Anomalous origin of the left coronary from the right coronary cusp (AOLCA)
  - Anomalous origin of the right coronary from the left coronary cusp (AORCA)
  - Single coronary artery origin

# Pediatric Chest Pain – Coronary Origin Anomalies



Adam, Eduardo, Generoso, Giuliano, Bittencourt, Marcio. Anomalous Coronary Arteries: When to Follow-up, Risk Stratify, and Plan Intervention  
 VL - 23. DO - 10.1007/s11886-021-01535-x. JO - Current Cardiology Reports

# Pediatric Chest Pain – Coronary Origin Anomalies

- Clinical Presentation
  - AOLCA: can be asymptomatic, can present with s/sx of myocardial ischemia/infarction, especially with exertion (or high CO states)
  - AORCA: most often are asymptomatic, though can present with s/sx of myocardial ischemia/infarction, especially with exertion (or high CO states)
    - Dependent upon anatomic characteristics: inter-arterial and/or intramural course
  - Single coronary artery: most often asymptomatic

# Pediatric Chest Pain – Coronary Origin Anomalies

- Diagnostic Testing
  - ECG: ischemic change, ventricular strain, repolarization abnormalities
  - Echocardiography
  - CT angiography
  - Cardiopulmonary Exercise Stress Test: functional assessment; ischemic change with exertion, arrhythmias, etc.
  - Stress Perfusion Imaging

# Pediatric Chest Pain – Coronary Origin Anomalies

- Management
  - AOLCA
    - Surgical reimplantation
  - AORCA
    - Variable / controversial
    - Conservative management/observation for most
    - Surgical reimplantation or unroofing IF clinical history and diagnostic testing supportive of a high-risk lesion (symptoms correlate with electrophysiologic and/or hemodynamic abnormalities)
  - Single coronary artery
    - Conservative management/observation

# Pediatric Chest Pain – Acquired Heart Disease

- Acquired Heart Disease
  - Myocarditis / Endocarditis / Pericarditis / Pan-carditis
- Clinical Presentation
  - Typically occur in the setting of infectious symptoms
  - Chest pain – sharp or dull
    - Myocarditis – non-specific
    - Pericarditis – worsens with deep inspiration, worsens in supine position, friction rub noted on exam
  - Tachycardia, +/- hypertension
  - Shortness of breath
  - Irritability/lethargy in infants
  - Pericarditis can be subclinical
  - Infants/children with myocarditis/endocarditis are often toxic appearing

# Pediatric Chest Pain – Acquired Heart Disease

- Clinical Presentation
  - Special considerations
    - Post-pericardiotomy syndrome following CHD surgery
    - Endocarditis in a patient with a bioprosthetic valve
    - Marfan Syndrome with aortic dilation
    - SLE / autoimmune pericarditis
    - Rheumatic fever / RHD
    - Note: Kawasaki disease, even with coronary artery pathology, typically does NOT present with chest pain.

# Pediatric Chest Pain – Acquired Heart Disease

- Diagnostic Testing
  - CXR
  - Complete respiratory viral panel
  - Serology (CBC, ESR/CRP, CMP), particularly for myocarditis
  - ECG (ischemic change, repolarization abnormalities, ectopy/arrhythmia)
  - +/- Echocardiography (valve insufficiency, ventricular function, pericardial effusion)
  - +/- blood cultures if concern for endocarditis
  - cMRI (myocarditis): LGE, scarring/fibrosis

# Pediatric Chest Pain – Acquired Heart Disease

- Management
  - Sports restriction for 4-6 months
    - Myocarditis
      - Repeat cMRI prior to full clearance

# Pediatric Chest Pain – Acquired Heart Disease

- Management
  - Endocarditis
  - Myocarditis
    - Non-steroidal anti-inflammatory
    - IVIG / steroids
    - May need inotropic support
  - Pericarditis
    - NSAID's
    - Colchicine

# Pediatric Chest Pain – Genetic Heart Disease

- Genetic Heart Disease
  - Hypertrophic (obstructive) cardiomyopathy
  - Supravalvar aortic stenosis
    - William's syndrome, Noonan syndrome

# Pediatric Chest Pain – Genetic Heart Disease

- Clinical Presentation
  - Rare causes of chest pain in the pediatric population
  - Hypertrophic (Obstructive) Cardiomyopathy
    - Most are entirely asymptomatic
    - CP typically exertional
    - Consider family history
    - Systolic ejection murmur on exam IF obstructive
  - Aortic Stenosis / Supravalvar aortic stenosis
    - Most are entirely asymptomatic
    - CP typically exertional
    - Consider genetic syndromes (William's, Noonan's)
    - Systolic ejection murmur on exam

# Pediatric Chest Pain – Genetic Heart Disease

- Diagnostic Testing
  - ECG
    - Ventricular hypertrophy, repolarization abnormalities, strain
  - Echocardiography (ventricular hypertrophy, AS gradients, ventricular function)

# Pediatric Chest Pain – Genetic Heart Disease

- Management
  - Hypertrophic (Obstructive) Cardiomyopathy
    - Dependent upon degree of obstruction
  - Aortic Stenosis / Supravalvar aortic stenosis
    - Dependent upon trans-valvar, supra-valvar gradients



# Pediatric Sports Screening & Participation

- General Screening
  - Otherwise healthy youth, previously undiagnosed
- Risk factor assessment for young athletes with heart disease
  - Congenital heart disease
  - Arrhythmias
  - Cardiomyopathy
  - History of myocarditis

# Pediatric Sports Screening & Participation

## General Screening (i.e. otherwise healthy, undiagnosed)

- **Purpose (Why)**
  - *Decreasing the incidence* of sudden cardiac death (SCD) in young athletes
  - There IS consensus that young athletes should be screened prior to participation in competitive sports. HOWEVER ...
  - There are ongoing debates regarding: the *true incidence* of SCD and the optimal *methods* for screening
- **Incidence**
- **Etiologies (What?)**
- **Screening Tools (How?)**
  - History and physical examination
  - ECG
  - Echocardiography
  - Cardiac MRI

# Elements of an Ideal Screening program\*

- Focuses on **common**, treatable conditions
- Implements a suitable, cost-effective test that can **accurately detect** the condition
- Leads to a **widespread** intervention (universal application) that can reduce complications of the condition **without causing adverse effects**

\* (Wilson and Jungner)

# Pediatric Sports Screening & Participation – AHA

## Incidence

- Sudden cardiac death (SCD) is the leading non-traumatic cause of death among young athletes.
  - The precise incidence of SCD among high school athletes, for example, is unknown, but estimates range from 1 in 23,000 to 1 in 300,000.
  - SCD of athletes can be caused by a variety of cardiovascular diseases, but is most commonly associated with **congenital or acquired malformations**.

# Pediatric Sports Screening & Participation

- Incidence ranges\*
  - High school athletes
    - From 1/46,000 to 1/917,000 athlete years
  - College athletes
    - From 1/43,000 to 1/83,000 athlete years
- Ten fold higher risk in male athletes than female counterparts
- African American athletes at higher risk
- Basketball and American football emerged as independent risk factors

• \* Petek and Baggish

# Pediatric Sports Screening & Participation – most common causes (Schmehil et al)

## Etiologies

- Autopsy data from United States National Registry (1980-2011)
  - 842 athletes
    - Males predominantly affected, 6.5:1
      - African Americans and other minorities most represented (5x greater)
    - HCM was most common (302/842, 36%)
    - Congenital coronary anomalies, ARVC, LQTS

# Sudden deaths in young competitive athletes: analysis of 1866 deaths in the United States, 1980-2006

CIRCULATION, 2009 Mar 3;119(8):1085-92.

- [Barry J Maron<sup>1</sup>](#), [Joseph J Doerer](#), [Tammy S Haas](#), [David M Tierney](#), [Frederick O Mueller](#)
- **Abstract**
- **Background:** Sudden deaths in young competitive athletes are highly visible events with substantial impact on the physician and lay communities. However, **the magnitude of this public health issue has become a source of controversy.**
- **Methods and results:** To estimate the absolute number of sudden deaths in US competitive athletes, we have assembled a large registry over a **27-year period** using systematic identification and tracking strategies. A total of **1866 athletes who died suddenly** (or survived cardiac arrest), **19+/-6 years of age**, were identified throughout the United States from 1980 to 2006 in **38 diverse sports**. Reports were less common during 1980 to 1993 (576 [31%]) than during 1994 to 2006 (1290 [69%],  $P < 0.001$ ) and increased at a rate of 6% per year. **Sudden deaths were predominantly due to cardiovascular disease (1049 [56%]), but causes also included blunt trauma that caused structural damage (416 [22%]), commotio cordis (65 [3%]), and heat stroke (46 [2%]).** Among the **1049 cardiovascular deaths**, the highest number of events in a single year was 76 (2005 and 2006), with an **average of 66 deaths per year** (range 50 to 76) over the last 6 years; 29% occurred in blacks, 54% in high school students, and **82% with physical exertion during competition/training**, whereas only **11% occurred in females** (although this increased with time;  $P = 0.023$ ). **The most common cardiovascular causes were hypertrophic cardiomyopathy (36%) and congenital coronary artery anomalies (17%).**
- **Conclusions:** In this national registry, the absolute number of cardiovascular sudden deaths in young US athletes was somewhat higher than previous estimates but relatively low nevertheless, with a rate of **<100 per year**. These data are relevant to the current debate surrounding preparticipation screening programs with ECGs and also suggest the need for systematic and mandatory reporting of athlete sudden deaths to a national registry.

# Pediatric Sports Screening & Participation – most common causes (Petek, Baggish)

- Historically, the most frequent cause of SCD in athletes < 35yrs was thought to be HCM
- Some studies of incidence in NCAA athletes report conflicting results
  - Harmon et al, 2003-2013, unexplained **autopsy negative SCD accounted for 25%** of cases, followed by **coronary artery anomalies (11%)**, **myocarditis (9%)** and coronary atherosclerosis (9%) – **HCM (8%)**
  - Maron et al, 2002-2011, 45% of CVD related deaths due to **HCM**
  - Many athletes with LVH on past autopsy may be attributed to HCM, but idiopathic LVH has emerged as a distinct entity from HCM.
- Although HCM is a likely common cause, **other more common etiologies may exist in parallel.**

# Pediatric Sports Screening & Participation – most common causes (Schmehil et al)

**Table 1** Cardiac conditions that increase young athletes' risk for SCD

Anomalous origin of coronary artery (second most common cause in the United States)

Aortic valve stenosis

Aortic dissection (usually complication in Marfan syndrome)

Arrhythmogenic right ventricular cardiomyopathy (ARVC) (most common cause in Italy)

Brugada syndrome (more prevalent in those of Asian descent)

HCM

Dilated cardiomyopathy

Coarctation of aorta

Congenital heart block (Mobitz type II, complete, or third degree)

Congenital or acquired long QT syndromes

Short QT syndrome

Coronary artery disease (rare in those younger than 35)

Restrictive cardiomyopathy

Endocarditis

Ehlers-Danlos syndrome

Mitral valve prolapse

Myocarditis

Pericarditis

Postoperative congenital heart disease

Status post heart transplant

Kawasaki disease (coronary artery abnormalities)

Wolff-Parkinson-White syndrome

SCD, sudden cardiac death; HCM, hypertrophic cardiomyopathy.

# Prevalence of Cardiomyopathy

**Table 1. Prevalence of the Most Common Inherited Cardiac Conditions**

	Children (1–puberty)	Adults (19–64 y)
HCM	Uncommon	1:250/500*
DCM	Uncommon†	1:250/500‡
ARVC	Uncommon	1:2000/5000
RCM	Uncommon	Uncommon
LQT	1:2000	1:2000
Brugada (type 1 ECG)	Uncommon	1:2000/5000§
CPVT	1:5000/10 000	1:5000/10 000

ARVC indicates arrhythmogenic right ventricular cardiomyopathy; CPVT, catecholaminergic polymorphic ventricular tachycardia; DCM, dilated cardiomyopathy; HCM, hypertrophic cardiomyopathy; LQT, long QT; and RCM, restrictive cardiomyopathy.

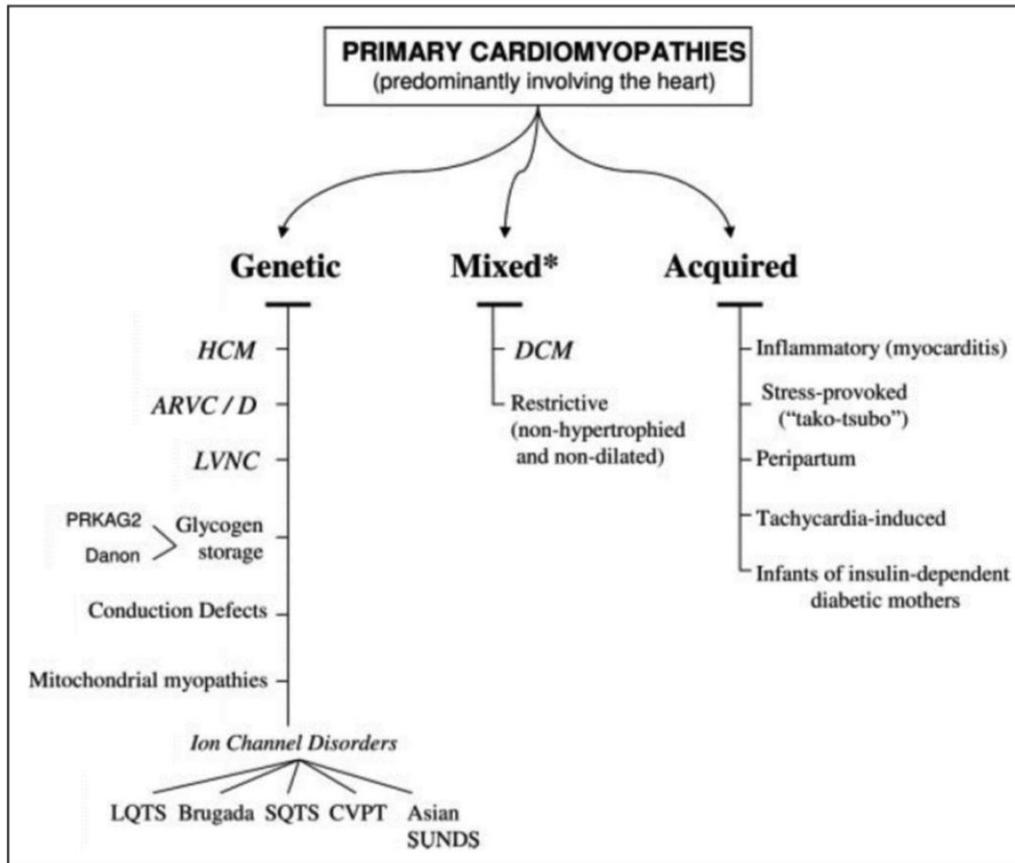
\*General population including gene carriers.<sup>23,24</sup>

†Presentation in first year of life 14× more common than during childhood and adolescence.<sup>20</sup>

‡Estimate generated from HCM/DCM incidence data.<sup>32</sup>

§Prevalence of 1:1000 from Japan in adults aged 40–64 y.<sup>36</sup>

# Prevalence of Cardiomyopathy



**Figure 10. American Heart Association Classification of Cardiomyopathies.** Primary cardiomyopathies in which the clinically relevant disease processes solely or predominantly involve the myocardium. The conditions have been segregated according to their genetic or nongenetic causes. \*Predominantly nongenetic; familial disease with a genetic origin has been reported in a minority of cases. ARVC/D indicates arrhythmogenic right ventricular cardiomyopathy/dysplasia; CPVT, catecholaminergic polymorphic ventricular tachycardia; DCM, dilated cardiomyopathy; HCM, hypertrophic cardiomyopathy; LQTS, long-QT syndrome; LVNC, left ventricular noncompaction; SQTS, short-QT syndrome; and SUNDs, sudden unexpected nocturnal death syndrome. Reproduced from Maron et al<sup>17</sup> with permission of the publisher. Copyright ©2006, American Heart Association, Inc.

# Prevalence of Coronary Artery Anomalies (CAA's)

- In the general population, both the course of each epicardial artery and the distribution of the collateral branches may show significant interindividual variability, so various phenotypes have been identified as normal variants, which are distinguished from CAAs by their higher prevalence (ie, >1% among unselected individuals) and not having been associated with major adverse events.
- The term **CAA** has historically been restricted to those occurring in **< 1% of the general population**

# Pediatric Sports Screening & Participation

## Screening Tools

- History and physical examination
- ECG
- Echocardiography
- Cardiac MRI

# Pediatric Sports Screening & Participation

- American Heart Association Screening Algorithm
- Viewpoints and Editorials
  - **Cardiac screening to prevent sudden death in young athletes.** Christopher Schmehil, et. al. Transl Pediatr 2017;6(3):199-206
  - **We Have Plenty of Reasons to Propose New, Updated Policies for Preventing Sudden Cardiac Death in Young Athletes.** Paolo Angelini, MD, et. al. J Am Heart Assoc. 2020;9:e014368. DOI:10.1161/JAHA.119.014368
  - **Policies to Prevent Sudden Cardiac Death in Young Athletes: Challenging, But More Testing Is Not the Answer.** Paul Dorian, MD, et al. J Am Heart Assoc. 2020;9:e016332. DOI: 10.1161/JAHA.120.016332.
  - **Current controversies in pre-participation cardiovascular screening for young competitive athletes.** Petek, Baggish. Expert Rev Cardiovasc Ther. 2020 July ; 18(7): 435–442. doi:10.1080/14779072.2020.1787154.

# Pediatric Sports Screening & Participation – AHA

American Heart Association: 14 Point Screen – elements collectively obtained from ...

- **Personal History**
- **Family history**
- **Physical exam**

# Pediatric Sports Screening & Participation – AHA

- **Personal History**

- Exertional chest pain/discomfort
- Exertional syncope or near-syncope
- Excessive exertional and unexplained fatigue/fatigue associated with exercise
- Prior recognition of a heart murmur
- Elevated systemic blood pressure
- Prior restriction from participation in sports
- Prior testing for the heart ordered by a physician

# Pediatric Sports Screening & Participation – AHA

- **Family history**

- Premature death – sudden and unexpected before age 50 years due to heart disease in one or more relatives
- Disability from heart disease in a close relative under 50 years old
- Specific knowledge of certain cardiac conditions in family members: *hypertrophic or dilated cardiomyopathy*, *LQTS* or other ion channelopathies, *Marfan syndrome* or clinically important arrhythmias

# Pediatric Sports Screening & Participation – AHA

- **Physical exam**

- Heart murmur\* – exam supine and standing or with Valsalva, specifically to identify murmurs of dynamic left ventricular outflow tract obstruction
- Femoral pulses to exclude aortic coarctation
- Physical stigmata of Marfan syndrome
- Brachial artery blood pressure (sitting, preferably taken in both arms)

# Pediatric Sports Screening & Participation – AHA

- What is the **role of electrocardiography (ECG)** in pre-participation examination screening?
  - If a resting 12-lead ECG can improve detection of underlying cardiac conditions in asymptomatic competitive athletes compared with medical history and physical examination alone, why not implement universal ECG screening?

# Pediatric Sports Screening & Participation – AHA

## Arguments against universal ECG screening per AHA

- Strain the health care system (staff required to perform & interpret, operator dependency)
- There were over **8.2 million high school athletes** in the U.S. during the 2024-25 school year, according to the National Federation of State High School Associations (NFHS). This is an all-time high, with 4,723,907 boys and 3,536,984 girls participating. Participation has been increasing for the past three consecutive years
  - **Estimates state that ECG screening would cost \$2.5-3.5 billion per year**

# Pediatric Sports Screening & Participation – AHA

- False positives/negatives.
  - A recent systematic review found false positive rates for physician-read ECGs of athletes generally ranges from 1.3-2.8%, but can be as high as 6.8% in Arab and Black athletes.
  - Computer-read ECGs have false positive rates from 2.3% to 5%.
  - False-negative rates of 12-lead ECG have been demonstrated to be 10% for hypertrophic cardiomyopathy (the leading cause of SCD in athletes).
- Detection bias
  - ECG's have limited utility in detection of anomalous coronary artery origins
  - Age-dependent penetrance

# Pediatric Sports Screening & Participation – AHA

- Equity/Access
  - Requiring all athletes to have ECGs may also be inaccessible to children from low-income families unless there is accompanying financial support. This is particularly important to recognize, as studies suggest certain racial groups may be at greater risk of sudden death.
- As a result, prescreening programs that require the use of expensive tests would need to find a way to overcome these critical access disparities.

# Pediatric Sports Screening & Participation – AHA

- According to a survey of pediatricians, family doctors, and high school athletic directors in Washington State, less than 6% of doctors follow national sudden cardiac death screening guidelines when performing physicals on high school athletes
- Challenges to the AHA's guidelines regarding the role of ECG's ...

# Pediatric Sports Screening & Participation

- Viewpoints and Editorials
  - **Cardiac screening to prevent sudden death in young athletes.** Christopher Schmehil, et. al. Transl Pediatr 2017;6(3):199-206

# Pediatric Sports Screening & Participation (Schmehil et al)

- The 14 element CV screening checklist released by the AHA in 2014 is recommended to be performed during all pre-participation physical examinations (PPE).
- At the discretion of the examiner, **a positive response or finding in any 1 or more of the 14 items** may be judged sufficient to trigger a comprehensive CV evaluation that may include ECG, echocardiography, or stress testing
- Currently, **AHA guidelines do NOT recommend the use of routine screening ECG** in the physical evaluation of young athletes

# Pediatric Sports Screening & Participation (Schmehil et al)

- Other countries, like Italy and Japan, routinely use ECG's as part of their PPE screenings
  - The Italian government passed a law in 1971 (and revised in 1982) stating that Italian athletes participating in competitive sports must perform PPE screening consisting of H&P, ECG and a 3 minute exercise step test.
  - Later study in Italy by Corrado et al found that using routine ECG reduced the annual incidence of SCD by 90%
  - Consequently, the International Olympic Committee began recommending ECG screening in 2004. FIFA (Federation Internationale de Football Association) later followed

# Pediatric Sports Screening & Participation (Schmehil et al).

- Why ECG alone is not sufficient
  - ECG is unable to accurately characterize **physiologic cardiovascular adaptations** that occur with athletic conditioning

# Pediatric Sports Screening & Participation (Schmehil et al).

- Unique physiology of the **athlete's heart** (Schmehil et al).
  - The cardiovascular system undergoes adaptation to athletic conditioning dependent upon the nature, duration, extent, intensity of exercise
  - Exercise will induce some sort of cardiac remodeling in approximately 50% of athletes
  - Long term, **endurance** exercise primarily produces volume overload induced LV dilation
  - Long term, **resistance** training primarily produces a pressure overload induced LV hypertrophy
  - *Left atrial enlargement* has been noted in 20% of athletes involved in endurance training.

# Pediatric Sports Screening & Participation (Schmehil et al).

- Why ECG alone is not sufficient
  - Approximately 40% of trained athletes have ECG changes
    - Repolarization patterns
    - Increased QRS voltages
    - Diffuse T wave inversion
    - Deep Q waves

# Pediatric Sports Screening & Participation (Schmehil et al).

- Several studies have sought to establish ECG criteria to minimize the false positive rate, especially in athletes with a physiologically normal heart.
  - **European Society of Cardiology (ESC)**
    - Initial ESC criteria in 2005, later revised in 2010 to classify ECG changes in two groups: “common and training related” and “uncommon and training un-related”
    - The 2010 ESC criteria improved specificity and decreased false positives from 16.3% to 9.6% while preserving sensitivity.
  - **Stanford Criteria (2011)**
  - **Seattle Criteria (2013)**
- A 2014 study comparing the above 3 demonstrated that the rate of false positive ECG abnormalities (requiring further workup) significantly decreased when the Stanford (8%) and Seattle (6%) criteria were applied (vs ESC at 26%)

# Pediatric Sports Screening & Participation (Schmehil et al).

**Table 3** Abnormal ECG findings as defined by the Seattle criteria

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T-wave inversion  
ST segment depression  
Pathologic Q waves  
Complete left bundle branch block  
Intraventricular conduction delay  
Left axis deviation  
Left atrial enlargement  
Right ventricular hypertrophy pattern  
Ventricular pre-excitation  
Long QT interval  
Short QT interval  
Brugada-like ECG pattern  
Profound sinus bradycardia  
Atrial tachyarrhythmias  
Premature ventricular contractions  
Ventricular arrhythmias

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ECG, electrocardiogram.

**Table 4** Normal ECG findings as defined by the Seattle criteria

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Sinus bradycardia  
Sinus arrhythmia  
Ectopic atrial rhythm  
Junctional escape rhythm  
1° AV block (PR interval >200 ms)  
Mobitz Type I (Wenckebach) 2° AV block  
Incomplete right bundle branch block  
Isolated QRS voltage criteria for LVH  
Early repolarization  
Convex ST segment elevation combined with T-wave inversion in V1-V4

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ECG, electrocardiogram; LVH, left ventricular hypertrophy.

# Pediatric Sports Screening & Participation (Schmehil et al).

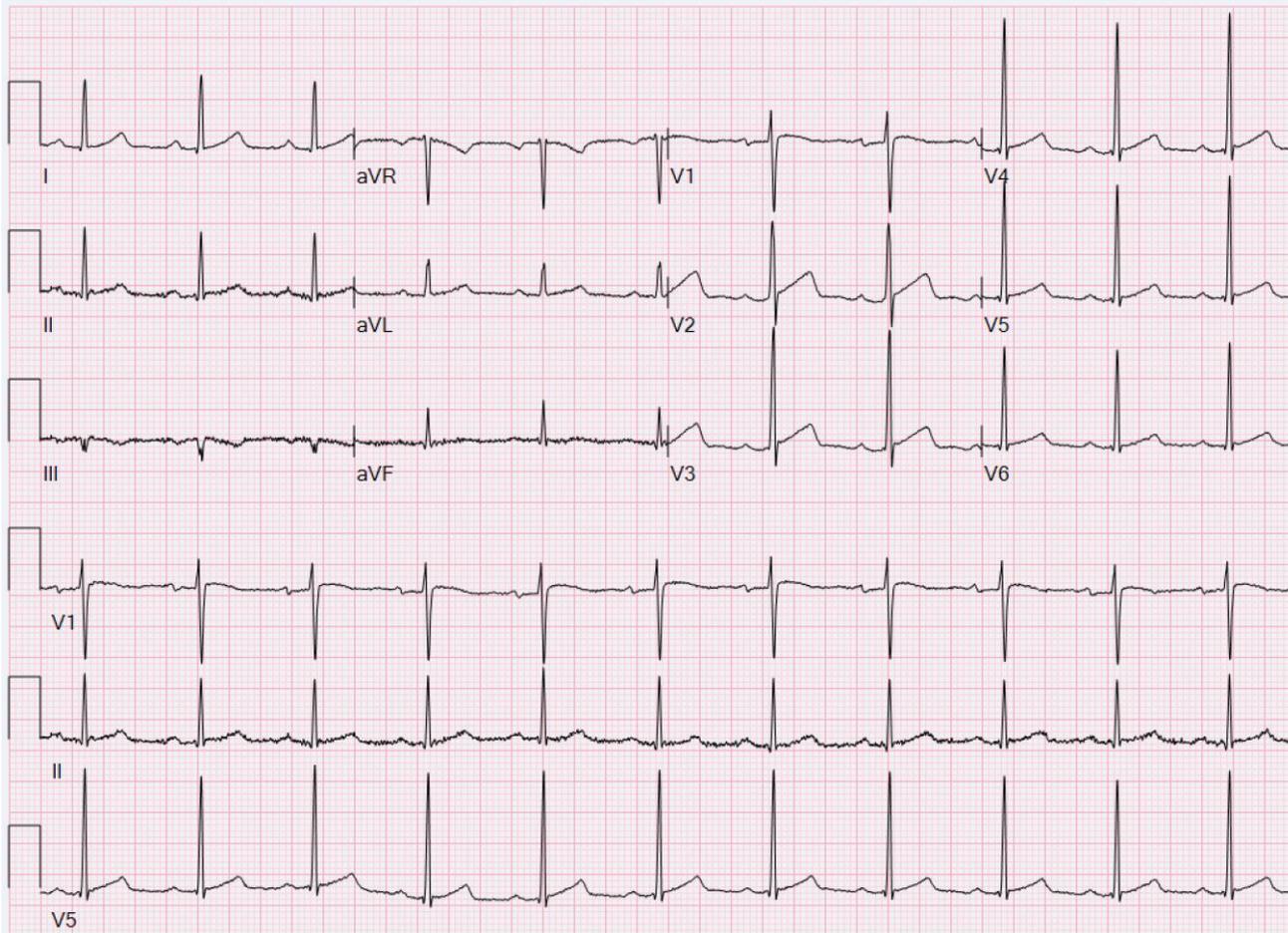
- In 2015, Stanford examined the prevalence of athletes who screened positive with the PPE from the original AHA checklist in combination with screening ECG's
  - 1596 participants (high school, college, professional)
  - Positive (“yes”) response to at least one checklist question in 25%
  - Abnormal ECG's found in ..
    - 6% (Seattle criteria), 8% (Stanford criteria), 26% (ESC criteria)
  - Abnormal ECG **and** at least one positive response on checklist
    - 1.4% (Seattle criteria), 2.4% (Stanford criteria), 6.1% (ESC criteria)
  - No athlete was found to have a clinical condition excluding them from sports participation

# Pediatric Sports Screening & Participation (Schmehil et al).

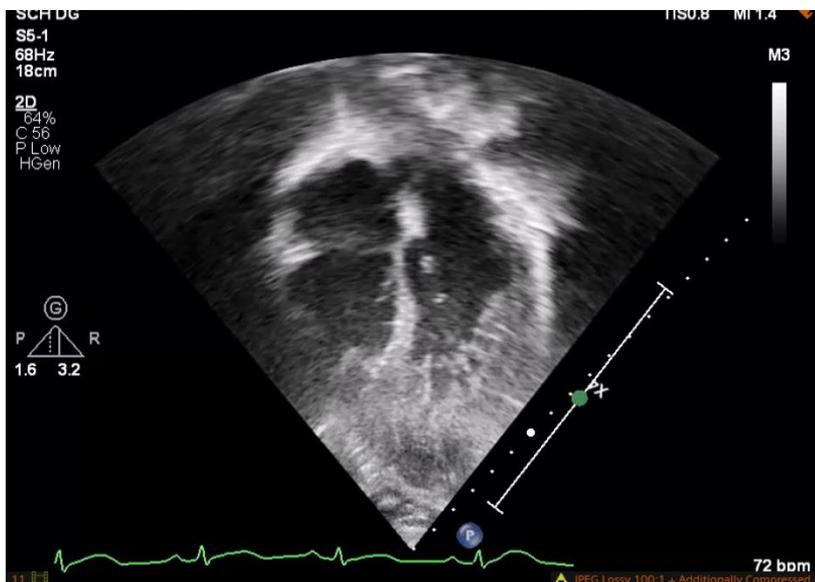
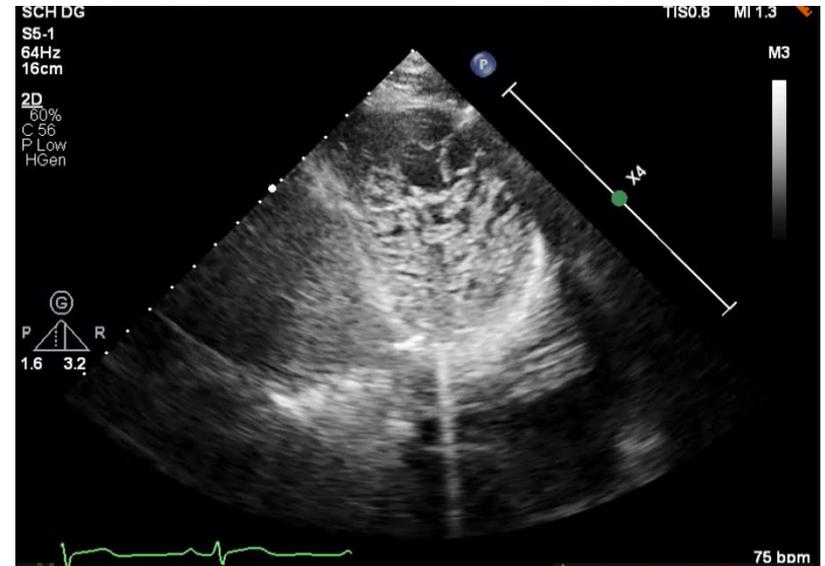
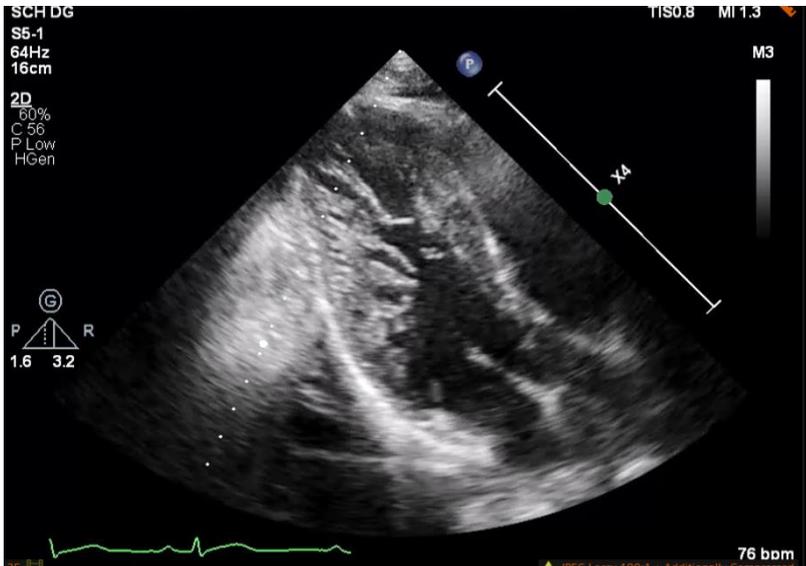
- Study conclusions ...
  - **AHA guidelines** lead to increased false positive results and unnecessary testing
    - Subjectivity in some of the checklist questions
      - What is deemed a positive response may vary amongst providers
      - Differences in athletic populations (i.e. level of competition)
      - How the questions are asked and understood
      - Quality and detail of history and physical exam
  - SCD is the presenting symptom in up to 80% of cases.
    - The 20% who do have warning signs before SCD may have non-specific symptoms

# Pediatric Sports Screening & Participation

- Why ECG alone is not sufficient (false negatives)



# Pediatric Sports Screening & Participation



# Pediatric Sports Screening & Participation

- What is the potential role of echocardiography?

# Pediatric Sports Screening & Participation

- Why **Echocardiography** alone is not always sufficient
  - Routine use of echo is expensive
  - Acoustic limitations can limit accuracy of definitively delineating coronary artery origins
  - Acoustic limitations can limit full, three dimensional visualization of the RV due to it's unique geometry (i.e. ARVC)
  - Phenotypic expression of cardiomyopathy is variable and may not manifest on initial echo screening (Age-dependent penetrance)
  - No identification of electrophysiologic based hrCVC's (i.e. LQTS, WPW, Brugada, etc.)

# Pediatric Sports Screening & Participation

- Why Echocardiography alone is not always sufficient
  - The Early Screening for Cardiac Abnormalities with Pre-participation Echocardiography (ESCAPE) protocol by Northwestern
  - **Portable echo** by a frontline physician (PEFP) used to directly visualize specific anatomic structures of the heart that are involved in SCD in specific echo windows with specific measurements
    - PEFP was significantly faster than taking a detailed AHA 12 element H&P and ECG screening
    - Decreased referrals to cardiology by 33%
    - No statistical differences in echo measurements obtained by a cardiologist
    - ? More accurate screening for structural abnormalities and ?  
More cost effective (than full echo)

# Pediatric Sports Screening & Participation

- Viewpoints and Editorials
  - **We Have Plenty of Reasons to Propose New, Updated Policies for Preventing Sudden Cardiac Death in Young Athletes.** *Paolo Angelini, MD*, et. al. J Am Heart Assoc. 2020;9:e014368. DOI:10.1161/JAHA.119.014368

# Pediatric Sports Screening & Participation – Angelini et al

Journal of the American Heart Association

## VIEWPOINTS

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### We Have Plenty of Reasons to Propose New, Updated Policies for Preventing Sudden Cardiac Death in Young Athletes

Paolo Angelini , MD; Raja Muthupillai, PhD; Benjamin Cheong, MD; Robert Paisley, MD

“The world as we have created it is a process of our thinking. It cannot be changed without changing our thinking.” (Albert Einstein)

Heart Association/American College of Cardiology for detecting cardiovascular diseases in the young. This assumes that we should be satisfied with the supposed preventive achievements of H&P, but this

normal, abnormal, and high-risk ranges for various sex, age, body mass index, and race cohorts. CMR image acquisition is completed in a single 10-to-15-minute outpatient session that does not require intravenous contrast or medication.

# Pediatric Sports Screening & Participation – Angelini et al

- In 2010, we hypothesized that a more sensitive, accurate pre-participation testing modality for evaluating young athletes was needed, especially because **identifying CAAs** (among the top causes of SCD in adolescents) by alternative methods (H&P, electrocardiography, echocardiography) is difficult and unreliable in adult-sized people. **We studied the value of cardiac magnetic resonance imaging** (CMR, or magnetic resonance imaging of the heart), **jointly with a focused questionnaire and resting ECG** (neither of which required administration by a specialized physician), in a general population of adolescent (**aged 11–18 years**) candidates for sports participation in Houston public schools (**n=5169**).
- **CMR enhances quantitative evaluation of the left ventricle**, thereby allowing the precise description of as-yet-unspecified normal, abnormal, and high-risk ranges for various sex, age, body mass index, and race cohorts. CMR image acquisition is completed in a single **10-to-15-minute** outpatient session that does not require intravenous contrast or medication.

## Pediatric Sports Screening & Participation – Angelini et al

- Cost: In our study, and assuming a dedicated screening facility that processes 20 cases/day, we found the cost of CMR to dedicated centers or providers is  $\approx$ \$250/patient. A CMR-based protocol can establish a reliable diagnosis of **1 hr-CVC for every \$16,500 expended (CMR screening cost plus overhead cost, divided by prevalence) ...** Recent advances in rapid CMR could substantially reduce scan times and hence lower costs

# Pediatric Sports Screening & Participation – Angelini et al

- CMR was successfully completed and was diagnostic for structural hr-CVCs (with highly accurate positive and negative results) in >98% of those screened.

**Table.** Comparison of Results From Cited Screening Reports

	Malhotra et al <sup>11</sup> (H&P, Electrocardiography, Echocardiography) n (%)	Angelini et al <sup>7</sup> (H&P, Electrocardiography, CMR) n (%)	Williams et al <sup>1</sup> (H&P, Electrocardiography, +/- Echocardiography) n (%)
Total	11 168	5169	3620
hr-CVC	42 (0.38)	76 (1.47)	15 (0.41)
hr-CMP	6 (0.05)	14 (0.27)	2 (0.06)
HCM	5 (0.04)	3 (0.06)	2 (0.06)
DCM	1 (0.01)	11 (0.21)	0 (0.00)
ARVC (by autopsy)	2 (0.02)	0 (0.00)	0 (0.00)
hr-ACAOS-IM	2 (0.02)	23 (0.44)	1 (0.03)
R-ACAOS-IM	1 (0.01)	17 (0.33)	1 (0.03)
L-ACAOS-IM	1 (0.01)	6 (0.12)	0 (0.00)
WPW	26 (0.23)	4 (0.08)	9 (0.25)

ARVC indicates arrhythmogenic right ventricular cardiomyopathy; CMR, cardiac magnetic resonance imaging; DCM, dilated cardiomyopathy; H&P, history and physical examination; HCM, hypertrophic cardiomyopathy; hr-ACAOS-IM, high-risk anomalous origin of coronary artery from the opposite sinus of Valsalva with intramural course; hr-CVC, high-risk cardiovascular condition; hr-CMP, high-risk cardiomyopathy; L-ACAOS-IM, left ACAOS from the right sinus with intramural course; R-ACAOS-IM, right ACAOS from the left sinus with intermural course; and WPW, Wolff-Parkinson-White syndrome.

# Pediatric Sports Screening & Participation – Angelini et al

- Limitations and issues for further consideration
  - Which hrCVCs could lead to SCD
  - What about hypoplastic coronary arteries that don't have an intramural course?
  - What is the quantified risk of SCD for each hrCVC?
  - Refinement of autopsy evaluation of SCD
    - Timing of autopsy: Autopsy reports of left ventricle hypertrophy may sometimes be inaccurate because of a “postmortem pseudo-systolic state” in which fluid is absorbed by the moribund myocardium in the first hours after death, falsely creating a hypertrophied appearance
    - Whole-heart weight versus myocardial wall thickness/scarring/myofibrillar disarray

# Pediatric Sports Screening & Participation

- Viewpoints and Editorials
  - **Policies to Prevent Sudden Cardiac Death in Young Athletes: Challenging, But More Testing Is Not the Answer.** *Paul Dorian, MD*, et al. J Am Heart Assoc. 2020;9:e016332. DOI: 10.1161/JAHA.120.016332.

# Pediatric Sports Screening & Participation – Dorian et al

Journal of the American Heart Association

## VIEWPOINTS

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### Policies to Prevent Sudden Cardiac Death in Young Athletes: Challenging, But More Testing Is Not the Answer

Paul Dorian , MD, MSc; Jack M. Goodman, PhD; Kim A. Connelly, PhD, MBBS

# Pediatric Sports Screening & Participation – Dorian et al

We need to understand:

- First, the **incidence of unexpected cardiac arrest** in young people, potentially related to competitive sport or exertion.
- Second, we need to consider **what proportion of cardiac arrests in young people may be predictable** with current testing.
- Third, we need to consider **whether arbitrary restriction from sport (disqualification) is likely to have a substantial impact** on the number of observed sudden deaths over the subsequent decades.
- Finally, we must consider the potential consequences of identifying structural heart disease that may pose a risk for sudden death, with respect to the **social, psychological, health-related, and economic outcomes for the individuals thus identified and labeled as “potentially at risk.”**

## Pediatric Sports Screening & Participation – Dorian et al

- **<50% of patients who ultimately have fatal cardiac arrest are identified by screening.** This is because  $\approx 30\%$  to  $40\%$  of all sudden deaths occur in young individuals with no identifiable electrical or structural abnormalities.
- In addition, other individuals have normal testing initially but **may eventually develop** identifiable structural abnormalities such as myocarditis, cardiomyopathy, or coronary disease. Some may have “subclinical” arrhythmogenic right ventricular cardiomyopathy or hypertrophic cardiomyopathy, but these conditions are difficult to diagnose in adolescents because of the variable clinical phenotype during the first 2 decades of life (age-dependent penetrance).

## Pediatric Sports Screening & Participation – Dorian et al

- Among individuals identified with structural heart disease, the risk of sudden death is extremely low. In a detailed autopsy study, the annual risk of sudden death, in people with hypertrophic cardiomyopathy under the age of 45 years, is estimated to be 0.3 to 0.4/1000 patient years, with most of these sudden deaths occurring at rest

## Pediatric Sports Screening & Participation – Dorian et al

- They may have been advised to have procedures such as open heart surgery with its nontrivial risk, and the **disease label** may have had a very substantial impact on their economic outlook, medical insurability, self-concept, social desirability, and potentially important consequences for their life habits with respect to a sedentary versus active lifestyle.
- Most importantly, if one had “disqualified” these individuals from the armed forces, and from competitive sport, it is completely unknown what proportion would have had sudden death from other forms of strenuous activity that form part of everyday living, including recreational sport, occupational demands, and other strenuous activities associated with life in general.

## Pediatric Sports Screening & Participation – Dorian et al

- There is a major distinction between the sensitivity of screening history and physical in young individuals and potential athletes to “*diagnose conditions associated with sudden death*” versus “*identifying individuals who actually suffer sudden death.*”
- Retrospective studies of individuals who have in fact suffered cardiac arrest suggest that the incidence of premonitory symptoms, especially syncope, may be 20% to 30%

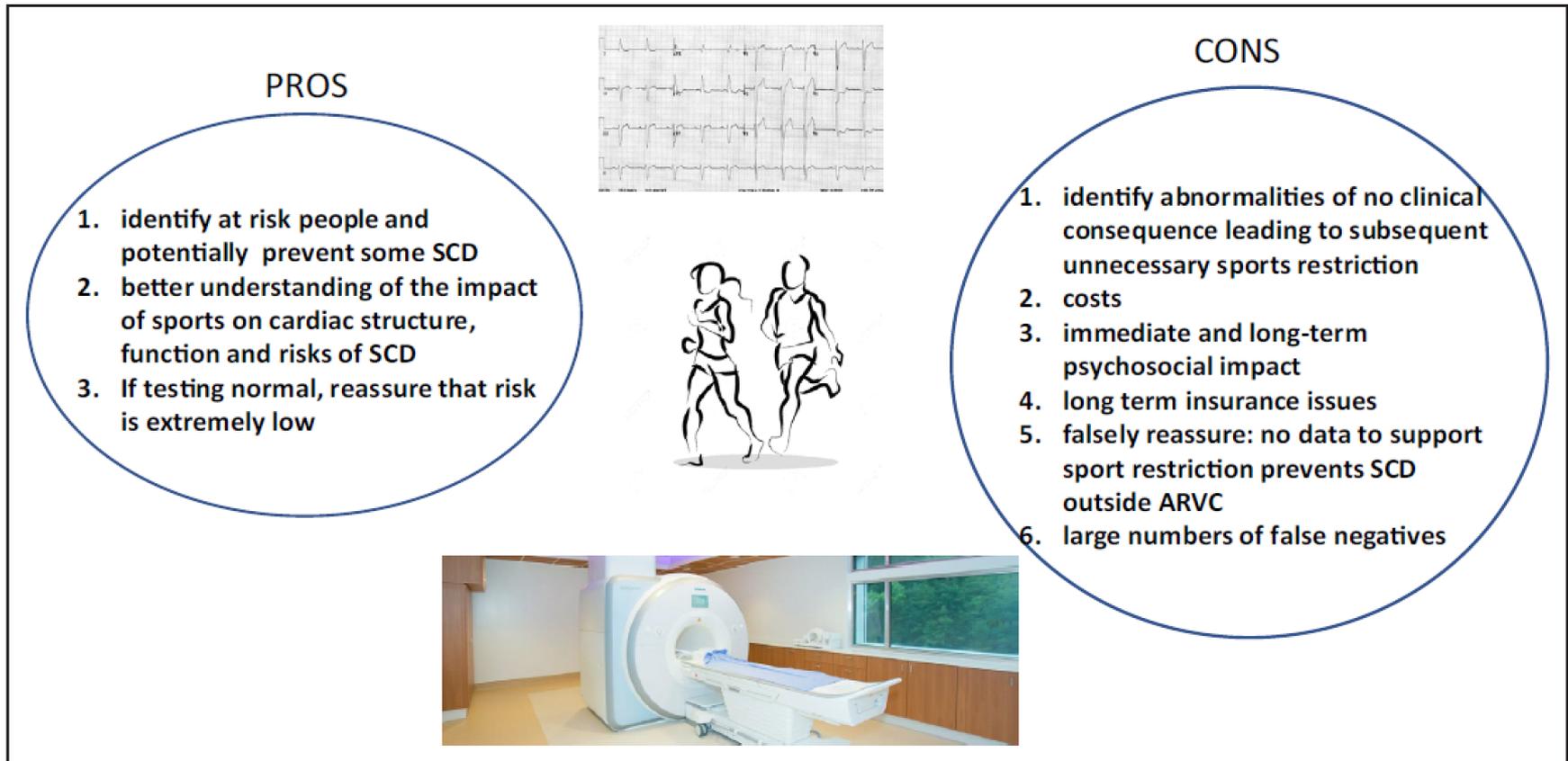
## Pediatric Sports Screening & Participation – Dorian et al

- For the vast majority of young individuals not destined to participate in competitive endurance sport, “sports restriction” would not apply, yet many would undoubtedly engage in vigorous exercise or even sport that falls outside of the scope of mandated screening often proposed; **sudden death rates during competitive versus recreational sports are identical.**
- For amateur, nonprofessional, and noncollegiate sports, physicians are not in the position to “disqualify” athletes, but should be in a position to, using the concepts of shared decision making, give their best possible advice to athletes and parents (if appropriate).

# Pediatric Sports Screening & Participation – Dorian et al

- It is very important to recognize that screening young people, especially if they are not yet athletes, has considerable individual and societal costs... it is essential that we ask (and answer) the questions:
  - What happens if I test positive?
  - Is the treatment that is likely to be offered effective?
  - Is the treatment worse than the disease?
- Developing **policies and procedures that improve the likelihood of successful resuscitation** in the case of cardiac arrest are demonstrably useful and can be deployed at a substantially lower cost than mass screening.

# Pediatric Sports Screening & Participation – Dorian et al



**Figure.** Preparticipation screening using history, physical examination, ECG, and cardiac magnetic resonance imaging as first line. ARVC indicates arrhythmogenic right ventricular cardiomyopathy; and SCD, sudden cardiac death.

# Pediatric Sports Screening & Participation

- Viewpoints and Editorials
  - **Current controversies in pre-participation cardiovascular screening for young competitive athletes.** *Petek, Baggish.* Expert Rev Cardiovasc Ther. 2020 July ; 18(7): 435–442.  
doi:10.1080/14779072.2020.1787154.

# Pediatric Sports Screening & Participation

- There were over 8.2 million high school athletes in the U.S. during the 2024-25 school year, according to the National Federation of State High School Associations (NFHS). This is an all-time high, with 4,723,907 boys and 3,536,984 girls participating. Participation has been increasing for the past three consecutive years

## Current Controversies in Pre-Participation Cardiac Screening (PPCS) \*

- Arguments in favor of PPCS
  - Most etiologies of SCD are readily detectable and treatment or restriction can save lives
- Arguments against PPCS
  - No RCT's showing that PPCS reduces mortality or has any definitive positive impact on the health and wellness of young athletes
  - Low overall incidence and unnecessary costs outweigh benefits
  - Cost
  - Age-dependent penetrance
    - What's the optimal age for PPCS? Is there a role for serial screening?

• \* (Petek & Baggish. Exper Rev Cardiovasc Ther)

## Current Controversies in Pre-Participation Cardiac Screening (PPCS) \*

- Arguments in favor of H&P
  - Complete H&P remains the cornerstone of screening – core competency, minimal additional training
  - Standardized questionnaires developed
  - Examination may reveal signs of pathology
- Arguments against H&P
  - High false positive rates, unacceptable false negative rates
  - Subjectivity – requires honest reporting by athletes (under-reporting). May not account for demographic, socioeconomic, cultural factors
  - Questionnaires developed by expert consensus and not by definitive scientific evaluation
  - Subjectivity of physical examination

• \* (Petek & Baggish. Exper Rev Cardiovasc Ther)

## Current Controversies in Pre-Participation Cardiac Screening (PPCS) \*

- Arguments in favor of ECG
  - Quick to perform (approx. 5 minutes)
  - Largely available
  - Increased detection rates (the ECG is abnormal in ...
    - 80% of the top 2/3 of conditions leading to SCD, > 90% of patients with HCM, > 80% of patients with ARVC
  - Requires advanced training for interpretation
    - Requires further advanced training for identifying physiologic ECG changes in trained athletes

• \* (Petek & Baggish. Exper Rev Cardiovasc Ther)

# Current Controversies in Pre-Participation Cardiac Screening (PPCS) \*

- Arguments against ECG
  - High false positive rates, especially when screening for an event with an overall low prevalence rate
  - False negatives (i.e. CAA's)
  - Poor interobserver reliability (variable provider training/experience)
  - Age-related penetrance

• \* (Petek & Baggish. Exper Rev Cardiovasc Ther)

# Pediatric Sports Screening & Participation

- Takeaway points for General Screening for prevention of SCD
  - Exact incidence is unknown
  - The most common cause of SCD is debated, but thought to be from autopsy-negative pathology (i.e. channelopathy/arrhythmia, CAA)
  - A focused H&P is the cornerstone of current PPCS
    - Low sensitivity, high false positive rates
    - Questionnaires are based on expert opinion
    - Subjectivity (? Encompass all demographics, socioeconomic factors), reliant on accurate patient reporting
    - Heterogeneity in H&P skills
  - AHA/ACC algorithm does not endorse additional testing

# Pediatric Sports Screening & Participation

- Takeaway points for General Screening for prevention of SCD
  - ECG's – false positive/negative rates, limited utility for detecting anomalous coronary origins (1/3 of cases of SCD)
  - cMRI – limited by availability, experience, expense
  - Correlation between intensity of exercise and risk of SCD
  - Does exercise restriction reduce the incidence of SCD
    - Physical and psychological ramifications of restriction

# The Nick of Time Foundation

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# The Nick of Time Foundation

In honor of Nick Varrenti, a previously healthy multi-sport athlete who died suddenly while playing football at 16 years.

- Youth heart screening program
- CPR Awareness and Education
- AED program for schools



<https://www.youtube.com/watch?v=ytr5I0xYBhk>



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Thank You



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