Cardiovascular Screening and the Preparticipation Sports Evaluation: To play or not to play

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Objectives

- Review the causes of sudden cardiac death in athletes
- Review current history and physical exam guidelines for preparticipation sports evaluations
- Discuss red flags on the history and physical
- Discuss Bethesda guidelines – when an athlete should be restricted
- Review the use of AEDs as secondary prevention
Sudden Cardiac Death in Athletes

- Rare event
- Usually due to a previously unrecognized cardiovascular disease
- True prevalence is not known, but estimated to be 0.5 – 2:100,000 athletes of high school age/ year
- 0.3% of athletes have a cardiac disease that puts them at risk of sudden death
Distribution of cardiovascular causes of sudden death in 1435 young competitive athletes

- HCM (36%)
- Coronary artery anomalies (17%)
- Indeterminate LVH - possible HCM (8%)
- Myocarditis (6%)
- ARVC (4%)
- MVP (4%)
- Tunneled LAD (3%)
- CAD (3%)
- AS (3%)
- Dilated C-M (2%)
- Sarcoidosis (1%)
- Aortic rupture (2%)
- Ion channelopathies (3%)
- Other congenital HD (2%)
- Other (3%)
- Normal heart (3%)

Sudden Cardiac Death (SCD): Differential Diagnosis

**Structural/Functional**
1) Hypertrophic Cardiomyopathy (HCM)*
2) Coronary Artery Anomalies
3) Aortic Rupture/Marfan*
4) Dilated Cardiomyopathy (DCM)*
5) Myocarditis
6) Left Ventricular Outflow Tract Obstruction
7) Mitral Valve Prolapse (MVP)
8) Coronary Artery Atherosclerotic Disease*
9) Arrhythmogenic Right Ventricular Cardiomyopathy (ARVC)*
10) Post-operative Congenital Heart Disease

**Electrical**
11) Long QT Syndrome (LQTS)
12) Wolff-Parkinson-White Syndrome (WPW)
13) Brugada Syndrome*
14) Catecholaminergic Polymorphic Ventricular Tachycardia (CPVT)*
15) Short QT Syndrome

**Other**
17) Drugs and Stimulants
18) Primary Pulmonary Hypertension (PPH)*
19) Commotio Cordis

* Familial / Genetic
Hypertrophic cardiomyopathy

- Most common cause of sudden cardiac death in young athletes (1/3 of cases)
- 1:500 in the general population
- Genetic - Autosomal dominant, however, sporadic mutations occur
  - Variable penetrance
- Due to a defect in the genes that encode the sarcomere protein
  - Defect results in myofibrillar disarray and fibrosis
  - Myofibrillar disarray creates a substrate for electrical instability which results in ventricular arrhythmias
Hypertrophic cardiomyopathy

- Patients with HCM may have symptoms including chest pain with exertion, dyspnea with exertion, palpitations, or syncope with exercise but most patients are asymptomatic
- Sudden death may be the first clinical manifestation
- On physical exam there may be a murmur due to dynamic left ventricular outflow tract obstruction
  - Murmur increases with standing or Valsalva, decreases with squatting
EKG findings associated with HCM

- Abnormal EKG in 70-90% of cases, but no typical EKG pattern
  - Hypertrophy pattern influences the EKG findings
- May have LVH, biventricular hypertrophy, increased R wave amplitude in right precordial leads, nonspecific ST or T wave abnormalities, abnormal Q waves
Anomalous origin of the coronary artery

- 2nd leading cause of sudden death in athletes
- Coronary artery (right or left) arises from the wrong aortic sinus
- Left main CA arising from the right sinus of Valsalva and traveling between the aorta and main pulmonary artery (intramural course) is the most common and has the highest risk of sudden death, however, the right CA arising from the left sinus has also been associated with sudden death
Anomalous origin of the coronary artery
Anomalous Coronary Arteries

- May have symptoms including chest pain or syncope with exertion but often asymptomatic
- Physical exam is typically normal
- EKG is also normal
Arrhythmia

- Long QT syndrome, Brugada syndrome, catecholaminergic polymorphic ventricular tachycardia (CPVT), Wolf-Parkinson-White (WPW)
- Likely underestimated as cause of sudden death due to negative autopsy findings
- Long QT, Brugada, and CPVT are inherited channelopathies
  - Important to determine if there is a family history of sudden death, syncope, or seizures
Arrhythmias

- Long QT
  - Involves defect in ion channels of the cell membrane resulting in abnormal myocardial action potential and prolonged QT interval on surface EKG
  - Often asymptomatic, but can have palpitations, presyncope, syncope, or seizures
    - Syncope is due to self limited episodes of polymorphic ventricular tachycardia (Torsades)
  - Sudden death occurs when the arrhythmia degenerates into vent fibrillation
  - Long QT type 1 associated with swimming
  - May be associated with congenital deafness
  - Autosomal dominant and recessive as well as acquired
Arrhythmia

- CPVT
  - Due to a mutation in the ryanodine receptor (RyR2) or Calsequestrin-2 (CASQ-2) genes – results in abnormal calcium regulation
  - Autosomal dominant or recessive inheritance or sporadic
  - History of episodic exercise or emotion induced syncope
  - Typically begins in late childhood and adolescence
  - Results in ventricular tachycardia that can degenerate into vent fibrillation
  - EKG and echo are normal, but can be induced with exercise testing
Arrhythmias

- Brugada
  - Also an inherited channelopathy – defect in sodium channel
  - Not as common in younger patients, but can result in SCD as initial symptom – fever may be a trigger
  - Characteristic EKG pattern
  - May have history of syncope

- WPW
  - More common that other types of arrhythmias but less likely to result in SCD
  - Often patients will have symptoms of episodic palpitations usually at rest but can be associated with exercise
  - Cause of SCD due to rapid conduction of atrial fibrillation to the ventricle with resultant ventricular fibrillation
Marfan

- Inherited in an autosomal dominant manner, but there may be sporadic mutations
  - Incidence of 1 in 3000 to 5000
  - Due to abnormality of the gene that encodes fibrillin-1 protein
- Characteristic body habitus – tall and thin with arm span > height
- Associated with musculoskeletal abnormalities (pectus deformity, kyphoscoliosis), joint hypermobility/ flexibility, and eye abnormalities (myopia, lens dislocation)
- Cardiac abnormalities include aortic dilation and mitral valve prolapse
  - Aorta may be prone to rupture/ dissection especially with high intensity and contact sports
- Specific diagnostic criteria
Myocarditis and Cardiomyopathies

- Myocarditis is the result of inflammation of the myocardium after a viral illness
  - Usually there is a viral prodrome
  - Myocellular damage results in ventricular dilation and dysfunction leading to heart failure
  - Risk of malignant arrhythmia during acute inflammatory phase

- Dilated cardiomyopathy
  - Often idiopathic, but can be inherited
  - Results in heart failure
  - Prone to ventricular arrhythmias and SCD
Cardiomyopathies

- Arrhythmogenic right ventricular dysplasia (ARVD)
  - Inherited – more common in Italians from Northern Italy
    - Estimated prevalence is 1:1000
  - Scarred appearance of the RV free wall – fibrofatty replacement of myocardium
  - Results in ventricular arrhythmia
  - May have history of palpitations, pre-syncope, syncope, atypical chest pain
Congenital Heart Disease

- With the exception of coronary artery abnormalities, CHD is rarely a cause of sudden death in athletes
- Aortic stenosis occasionally may result in sudden death even in patients who do not have severe stenosis
- Most patients with CHD have a known history
- The 36th Bethesda conference offers guidelines for athletic participation in athletes with congenital heart disease
Primary Prevention: Pre-participation Evaluation

- Goal: Appropriately restrict; appropriately clear
- Be thorough and conscientious
- Are there any warning signs or family history?
AHA 2007 screening guidelines

• Felt to be an effective strategy to raise the suspicion of cardiovascular disease

• Consists of 12 items
  • 8 personal and family history and 4 physical exam

• A positive response or finding in any 1 or more of the 12 items is sufficient to trigger further work-up and/or referral for cardiovascular evaluation
AHA Statement 2007

Medical History

Personal history
1. Exertional chest pain/discomfort
2. Unexplained syncope/near syncope
3. Excessive exertional and unexplained dyspnea/fatigue, associated with exercise
4. Prior recognition of a heart murmur
5. Elevated systemic blood pressure

Family History
6. Premature death (sudden and unexpected, or otherwise) before age 50 years due to heart disease, in ≥1 relative
7. Disability from heart disease in a close relative <50 years of age
8. Specific knowledge of certain conditions in family members: hypertrophic or dilated cardiomyopathy, long-QT syndrome or other ion channelopathies, Marfan syndrome, or clinically important arrhythmias

Physical examination
9. Heart murmur
10. Femoral pulses to exclude aortic coarctation
11. Physical stigmata of Marfan syndrome
12. Brachial artery blood pressure (sitting position)
2010 PPE endorsed by AAP, AAFP, AMSSM, ACSM, AOSSM, AOASM

HEART HEALTH QUESTIONS ABOUT YOU

• Have you ever passed out or nearly passed out DURING or AFTER exercise?
• Have you ever had discomfort, pain, tightness, or pressure in your chest during exercise?
• Does your heart ever race or skip beats (irregular beats) during exercise?
• Has a doctor ever told you that you have any heart problems? If so, check all that apply: □ High blood pressure □ A heart murmur □ High cholesterol □ A heart infection □ Kawasaki disease Other: ________________________
• Has a doctor ever ordered a test for your heart? (For example, ECG/EKG, echocardiogram)
• Do you get lightheaded or feel more short of breath than expected during exercise?
• Have you ever had an unexplained seizure?
• Do you get more tired or short of breath more quickly than your friends during exercise?

HEART HEALTH QUESTIONS ABOUT YOUR FAMILY

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

- **Medical history** is the most important part of the cardiovascular PPE
- Best to personally interview the athlete **and** the parent
  - Family concerns or observations compliment the picture of the adolescent’s health
  - Parents should verify elements of the history
- Ask open ended questions
- Have a checklist that helps probe for potential cardiac disease
What questions should be asked?

- Personal history
  - Focuses on symptoms such as chest pain, chest tightness, shortness of breath, dyspnea, near syncope, dizziness, syncope, exercise intolerance, and fatigue
  - Symptoms in the context of physical activity are typically more concerning and may be harbinger of cardiovascular disease
  - Detailed medication history including both prescribed medications and supplements
    - Questions about illicit drug use including performance enhancing drugs (steroids, human growth hormone, amphetamines)
What questions should be asked?

- Past medical history
  - History of rheumatic fever, Kawasaki disease, myocarditis, pericarditis, congenital heart disease, history of heart murmurs, hypertension

- Family history
  - Often underestimated, but very important given that many causes of sudden death are inherited
  - Searching for silent cardiac disease entails asking specific questions about unexplained or sudden deaths and accidents (such as drownings or car accidents), SIDS, or death in someone under the age of 50 years
Key points with the exam

- Vital signs are important – especially blood pressure
  - BP should be assessed and compared to normal values by gender and height

- Features of Marfans should be assessed
  - Kyphoscoliosis, pectus deformity, arm span > height, joint hypermobility, arachnodactyly

- Cardiac auscultation
  - Focuses on heart sounds, murmurs, and clicks
  - Examination should be performed in the supine and standing position

- Assessment of femoral pulses
Red Flags

- AHA recommendations state that a positive response to any of the 12 recommended items is judged sufficient to trigger a referral to a cardiologist
- These include:
  - Syncope or near syncope with exertion
  - Chest pain/dyscomfort with exertion
  - Palpitations at rest
  - Excessive SOB or fatigue with activities
  - Family history of Marfan, long QT syndrome, HCM, or clinically significant arrhythmia
  - Family history of sudden death especially in a first degree relative
  - Systolic murmur or diastolic murmur
  - Stigmata of Marfan syndrome
Are we doing a good job with screening?

- No universally accepted or mandated standards for screening high school and college athletes
  - Every state, athletic association, and school district has different requirements for the PPE
  - If there is a requirement it usually only applies to organized athletics through the schools and not to other athletic or physical activities (club sports, individual sports, dance, etc)
- No certification guidelines for health care professionals who perform such screening
  - Many types of practitioners are doing the evaluation including physicians, NPs, PAs, naturopaths, chiropractors, and athletic trainers
Are we doing a good job with screening?

- There has been an improvement in the last decade.
- In 1997 40% of states had either no formal screening requirement or the PPE was judged to be incomplete or inadequate.
- In 2005 81% of states were judged to have an adequate PPE (>9 of the 12 of the AHA recommended items). Only 2% of states were felt to have an inadequate PPE (<4 of the 12 items).
Are we doing a good job with screening?

- Oregon Schools Activities Association (OSAA) has its own policy regarding PPE of athletes

- **ORS 336.479, Section 1 (3)**: "A school district shall require students who continue to participate in extracurricular sports **in grades 7 through 12** to have a physical examination **once every two years**." Section 1(5) “Any physical examination required by this section shall be conducted by a (a) physician possessing an unrestricted license to practice medicine; (b) licensed naturopathic physician; (c) licensed physician assistant; (d) certified nurse practitioner; or a (e) licensed chiropractic physician who has clinical training and experience in detecting cardiopulmonary diseases and defects.”

- Current 2010 OSAA PPE form contains > 9 of the 12 AHA recommended items

- A survey distributed in 2000 to OSAA participating high school athletic directors showed that 53% of schools who responded to the survey had PPEs which contained fewer than 5 of the AHA recommended items while only 27% were implementing the PPE form recommended by the OSAA.
Does screening work?

- PPE screening with H & P alone does not have sufficient sensitivity to guarantee detection of all CV abnormalities linked to sudden death
- Sensitivity and specificity not completely known
Does screening work?

• A study published in 2010 screened 510 college athletes. All participants underwent an echocardiogram and EKG in addition to a standard PPE
  • Cardiac abnormalities with relevant sports participation risk were observed in 11/510 (2.2%), however, after further diagnostic testing only 3/11 were restricted
  • Screening with H & P alone detected 5/11 athletes
    • Sens 45.5%, spec 94.4%
  • EKG in addition to H & P detected another 5 athletes (10/11)
    • Sens 90.9%, spec 82.7%

Cardiac abnormalities detected w/ exam and EKG

- Bicuspid aortic valve – Murmur; normal EKG, no restriction
- Bicuspid aortic valve - Murmur and click; normal EKG; no restriction
- MVP - Murmur; normal EKG, no restriction
- MVP - Murmur; normal EKG, no restriction
- MVP - Normal exam, normal EKG, no restriction (echo only diagnosis)
- Pulmonic stenosis - Murmur; normal EKG, restricted due to moderate pulmonic stenosis
- LV hypertrophy – normal exam, EKG w/ increased QRS voltage, LAE; no restriction
- **LV hypertrophy** – normal exam, EKG with increased QRS voltage and t-wave abnormality, Restricted due to hypertrophic cardiomyopathy
- LV dilation – normal exam, Ekg w/ LBBB; no restriction
- **LV dilation** – normal exam, EKG w/ LBBB; restricted due to viral myocarditis
- RV dilation – normal exam, EKG w/ RBBB; no restriction
Should EKG be part of the PPE?

- Controversial topic
- Europe Society of Cardiology and the Olympic committee endorses the use of EKG as part of the PPE
  - Implemented after sudden death rate in Italian athletes decreased from 3.6 to 0.4: 100,000 after using EKG as part of the PPE
- Currently the AHA does not support the use of EKG as part of its PPE recommendations
  - False positive rate
  - Unnecessary restriction
  - Cost of implementing this type of program (est of $2 bil/year ?)
  - Infrastructure involved – is it practical and feasible with the current health care system?
Who, when, and where?

- Competitive athletes only?
- What age should screening begin?
- How often should it occur?
- Who should do the screening and where should it take place?
Bethesda Guidelines

- Group consensus regarding the medical risks imposed by competition on an athlete with a cardiovascular abnormality
- Meant to guide participation in competitive sports
  - Important component of a competitive sports activity concerns whether athletes are able to properly judge when it is prudent to terminate physical activity
- Definition most easily applied to high school, college, and professional athletics
  - Individual clinical judgment regarding competitive youth sports for children < 12 years
- The recommendations do not apply to non-competitive, recreational activities
Bethesda Guidelines

- Firm disqualification should be confined to individual athletes with probable or conclusive evidence of disease rather than those with borderline findings
  - Minimize unnecessary restrictions from sports or stigma of a cardiac diagnosis
  - May permit the occasional athlete to participate who might otherwise be at some risk
- Not all sports involve identical types of intensity
- However, intensity of conditioning regimens often exceed competition itself
## Classification of Sport

<table>
<thead>
<tr>
<th>Increasing Static Component</th>
<th>Increasing Dynamic Component</th>
<th>A. Low (&lt;40% Max O₂)</th>
<th>B. Moderate (40-70% Max O₂)</th>
<th>C. High (&gt;70% Max O₂)</th>
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</thead>
<tbody>
<tr>
<td>I. Low (&lt;20% MVC)</td>
<td></td>
<td>Billiards, Bowling, Cricket, Curling, Golf, Riflery</td>
<td>Baseball/Softball*, Fencing, Table tennis, Volleyball</td>
<td>Badminton, Cross-country skiing (classic technique), Field hockey*, Orienteering, Race walking, Racquetball/Squash, Running (long distance), Soccer*, Tennis</td>
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<tr>
<td>II. Moderate (20-50% MVC)</td>
<td></td>
<td>Archery, Auto racing*†, Diving*†, Equestrian*†, Motorcycling*†</td>
<td>American football*, Field events (jumping), Figure skating*, Rodeoing*†, Rugby*, Running (sprint), Surfing*†, Synchronized swimming†</td>
<td>Basketball*, Ice hockey*, Cross-country skiing (skating technique), Lacrosse*, Running (middle distance), Swimming, Team handball</td>
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<tr>
<td>III. High (&gt;50% MVC)</td>
<td></td>
<td>Bobsledding/Luge*†, Field events (throwing), Gymnastics*†, Martial arts*, Sailing, Sport climbing, Water skiing*†, Weight lifting*†, Windsurfing*†</td>
<td>Body building*†, Downhill skiing*†, Skateboarding*†, Snowboarding*†, Wrestling*</td>
<td>Boxing*, Canoeing/Kayaking, Cycling*†, Decathlon, Rowing, Speed-skating*†, Triathlon*†</td>
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* Indicates aerobic exercise, † indicates anaerobic exercise.
Absolute Contraindications

- Eisenmenger syndrome
- Severe pulmonary hypertension
- Severe aortic stenosis and/or regurgitation
- Severe mitral stenosis and/or regurgitation
- Cardiomyopathies
- Vascular form of Ehlers-Danlos syndrome
- Coronary artery abnormalities
- CPVT
- Acute phase of myocarditis and/or pericarditis
- Acute phase of Kawasaki disease.
Recommendations for physical activity and recreational sports

- AHA scientific statement in 2004 makes recommendations for patients with Genetic Cardiovascular Disease (ie HCM, channelopathies, ARVD, and Marfan syndrome) who wish to continue with recreational activities.
- Important that patients with these conditions continue to have an active lifestyle even though they may be restricted from competitive athletics.
- Similar to Bethesda guidelines in that it breaks activities into high, moderate, and low intensity.
- Uses a graded scale for each activity regarding permissibility of the activity for each condition.
Secondary Prevention

- Not all causes of SCD can be prevented even with every diagnostic tool currently available
- Automated external defibrillators (AEDs) should be present in public places especially where athletic activities take place
- An emergency action plan for SCA should be implemented when possible (ie first responders trained in CPR and AED use)
- Goal is to quickly activate EMS and provide access to AED within 3 to 5 minutes
Secondary prevention

- Rapid response with CPR and AED use saves lives!!
- The single greatest factor affecting survival after out of hospital SCA is the time from arrest to defibrillation
- Chance of survival declines by 7-10% for every minute that defibrillation is delayed
- Recent study evaluated survival of young athletes with SCA if early defibrillation was achieved
  - Cohort of 1710 high schools with on site AED
  - 36 cases of SCA (14 athletes)
  - 94% received bystander CPR and an AED deployed a shock in 30/36
  - 64% survived to hospital discharge (including 9/14 athletes)

Summary (1)

- Sudden cardiac arrest/ death in athletes is a rare event and often due to an undetected cardiac abnormality
- Hypertrophic cardiomyopathy is the most common cause, followed by coronary artery abnormalities, cardiomyopathies, and arrhythmias
- The preparticipation evaluation may be able to detect cardiovascular abnormalities if it is done properly, however, it has a low sensitivity
- AHA and AAP have both provided specific guidelines for the PPE
- PPE requires asking specific questions about the athlete’s own history as well as a detailed family history
Summary (2)

- A positive response to any of the 12 items on the AHA guidelines may warrant further work-up or referral to a cardiologist
  - Important to have a low index of suspicion
  - However, must be careful to not inappropriately restrict
- For those patients who have a known cardiovascular disease, the Bethesda guidelines offer recommendations for competitive athletics
- AEDs for secondary prevention do save lives in those athletes who suffer a cardiac arrest despite appropriate screening
Food for Thought

• Although sudden cardiac death in a young athlete is catastrophic and devastating it is important to keep perspective
  • There are ~ 75 cardiac deaths in young athletes/ year, but 115 deaths due to other causes (blunt force trauma, commotio cordis, heat stroke, etc)
  • Preventable causes of death such as accidents, homicides, and suicide are much more common in adolescents including athletes
    • MVA is 2500 x more common than a cardiac event during sports
References and resources

- American Academy of Pediatrics
  - [www.aap.org](http://www.aap.org)
- American Heart Association
  - [www.heart.org](http://www.heart.org)