Sudden Cardiac Death

Michelle A. Grenier, MD
Associate in Pediatric Cardiology
Texas Children’s Hospital

Financial Disclosures: None, but I do believe Prevention is a good thing.

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HARVEY, La. -- An autopsy determined that LSU baseball player Wally Pontiff, found dead in his parents' home last week, died of natural causes and no drugs were in his system, the Jefferson Parish coroner said Monday. The autopsy found an abnormality in the heart, which is still being tested, so final determination of exact cause of death may take several more days, the coroner's report said.
Sudden Cardiac Death

Sudden cardiac death (SCD)
Nontraumatic, nonviolent, unexpected event resulting from sudden cardiac arrest within 6 hours of a previously witnessed state of normal health.

Maron BJ, Epstein SE, Roberts WC. JACC 1986;7:204-14
Sudden Cardiac Death

**Warning signs during exercise**
- Palpitations
- Dizziness
- Chest Pain/ Chest Tightness
- Shortness of Breath
- Syncope

If there were warning signs, it may not be “Sudden”
How frequently does it occur?

- **Annual incidence in the general population**
  1/100,000 all comers  
  CDC, 2004, Sen-Chowdhry, McKenna, Cardiology 2006

- **2.3/100,000 athletes per year**

- **4/100,000 hikers annually**

- **Incidence higher in athletes 2.5:1**

- **Incidence higher in males 9:1**
More Numbers...Looking at Athletes under 35 years of Age


• 4% 16-64 years no cause ID: arrhythmia?  Sen-Chowdhry, McKenna Cardiol 2006
The Invincible Adolescent

- 40% occurred in athletes < 18 yrs  
  Bille, et al.  
- 33% occurred in athletes < 16 yrs  
- 9:1 Male: Female  
- 30% soccer (football), 25% Basketball,  
  15% track/running  
What is the relative cost of SCD?

- “The term ‘sudden cardiac death’ (SCD) implies the sudden and unexpected loss of a productive member of the community.”
- “The cost of SCD to society is incalculable.”
- If it is your family member, it affects you 100%.
SCD in Young Athletes

Risk profile for sudden cardiac death

- 17 years median age
- 90% male
- 52% white, 44% black
- 68% playing football or basketball
- 90% collapsed during or shortly after training session/competitive event
- 85% cardiovascular cause

Maron BJ et al. JAMA 1996; 276: 199-204
SCD in Young Athletes

Incidence of Sudden Cardiac Death by Sport

http://www.suddendeathathletes.org
Incidence of Sudden Cardiac Death by Month

http://www.suddendeathathletes.org
SCD in Young Athletes

Incidence of Sudden Cardiac Death by Time of Day

http://www.suddendeathathletes.org
Sudden cardiac death in Athletes

Distribution of cardiovascular causes of sudden death in 1435 young competitive athletes

Athlete’s Heart

• At age 18, just one month shy of his high school graduation, Lil Vic went into sudden cardiac arrest while doing what he loved best - playing basketball. It turned out that Vic had hypertrophic cardiomyopathy (HCM) or an enlarged heart, which had been undetected.
Sudden Cardiac Death

Hypertrophic Cardiomyopathy
SCD in Young Athletes

- **Etiologies of SCD**
  - Hypertrophic Cardiomyopathy
    - Leading cause of sudden cardiac death in young athletes
    - Estimated prevalence of the disease 1:500 (not all high risk)
SCD in Young Athletes

- **Hypertrophic Cardiomyopathy**
  - Disorder caused by mutations of the cardiac sarcomere unit
  - Autosomal dominant inheritance
SCD in Young Athletes

- Detection of HCM
  - History
    - Family history
      - autosomal dominant
    - Symptoms
      - exertional dyspnea, orthopnea, fatigue: diastolic dysfunction
      - chest pain: ischemia
  - Exam
    - murmur (present in only 25%)
SCD In Young Athletes

Hypertrophic cardiomyopathy

No murmur on exam

Possible murmur on exam
SCD in Young Athletes

- **Detection of HCM**
  - Murmur
    - SEM at the left lower to mid sternal edge, radiates along the LVOT
    - Harsh in quality, but may be vibratory
    - Increases with exercise
      - increased contractility
    - Increases with Valsalva, standing
      - decreased volume
SCD in Young Athletes

• **Detection of HCM: Inherent difficulties**
  - Children may not yet show LVH
  - LVH may appear at any age, and change.
  - It may not appear until adolescence, as the normal LV remodeling process appears to be most active then
SCD in Young Athletes

• **Sudden Death in HCM**
  - Lethal ventricular arrhythmia
  - May be associated with extreme physical exertion
  - May also be precipitated by volume depletion (dehydration).
SCD in Young Athletes

- **Risk Factors for SCD in HCM**
  - Prior cardiac arrest
  - Sustained or repetitive non sustained VT
  - Hypotensive BP response to exercise
  - Family history of SCD
  - Syncope (particularly exertional)
  - Extreme LVH (>30mm)
SCD in Young Athletes

**Prevention of SCD in HCM**

- Medications have not been shown to affect outcome
- One study from Italy, where there is nationwide screening, showed a diminished incidence in SCD (ARVD), with exercise restriction
Sudden Cardiac Death

- **Sudden Death in HCM**
  - Lethal heart rhythm abnormality
  - Association with extreme physical exertion
  - Autosomal Dominant/Spontaneous mutations
  - Warrants disqualification from competitive sports
HCM versus Athlete’s Heart

- Pathologic descriptions of myocardial hypertrophy in trained individuals from Kirch and Linzbach in 1958
- The heart of the trained athlete can be twice the normal size, but histologic structure remains intact
- The weight of the trained heart does not usually surpass the limit of 500g, defined as the critical heart weight
Athlete’s Heart

- Cardiac enlargement in athletes may represent a physiologic phenomenon (controversial for many years)
- Concerns about long-term consequences of morphologic left ventricular remodeling
- Concern of sudden cardiac deaths in athletes with hypertrophic cardiomyopathy (HCM) accounting for majority of catastrophes
**Figure 1.** Echocardiograms in parasternal long-axis view from an elite athlete (Olympic rower) (left) and a young asymptomatic patient with hypertrophic cardiomyopathy (HCM) (right). Magnitude of anterior ventricular septal (VS) hypertrophy is similar in each, demonstrating the morphological gray zone into which a highly trained athlete may fall and the diagnostic ambiguity that may ensue. Calibration dots are 1 cm apart. Left panel is reprinted by permission of the *New England Journal of Medicine* (1991;324:295-301).
Differentiating from HCM

- LV cavity enlarged in athlete’s heart
- Hypertrophy symmetric and homogenous
- LV cavity maintains ellipsoid shape
- LV mass regresses with deconditioning
- LV filling pattern remains normal
- Acoustic pattern of myocardium remains normal
- Restricted to certain sports
Cardiac Function

- Echo exams on 286 cyclists in Tour de France and 52 matched sedentary volunteers
- 148 in 1995 race, 138 in 1998 race, and 37 in both
- 51% had substantial LV enlargement (LVIDd > 60mm, up to 73mm)
- 9% had increased LVWT (>13 mm)
A Heavy Heart

- Left Ventricular Mass (Penn-cube formula)

<table>
<thead>
<tr>
<th>Group</th>
<th>Mass</th>
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<tbody>
<tr>
<td>Controls</td>
<td>174 g</td>
</tr>
<tr>
<td>Endurance-trained</td>
<td>249 g</td>
</tr>
<tr>
<td>Combined</td>
<td>288 g</td>
</tr>
<tr>
<td>Strength-trained</td>
<td>267 g</td>
</tr>
</tbody>
</table>
A Summary Table to Clarify

<table>
<thead>
<tr>
<th>Item</th>
<th>HCM*</th>
<th>Athlete's Heart</th>
</tr>
</thead>
<tbody>
<tr>
<td>Unusual Patterns of LVH</td>
<td>+</td>
<td>-</td>
</tr>
<tr>
<td>LV Cavity &lt; 45 mm</td>
<td>+</td>
<td>-</td>
</tr>
<tr>
<td>LV Cavity &gt; 55 mm</td>
<td>-</td>
<td>+</td>
</tr>
<tr>
<td>L.A. Enlargement</td>
<td>+</td>
<td>-</td>
</tr>
<tr>
<td>Bizarre ECG Patterns</td>
<td>+</td>
<td>-</td>
</tr>
<tr>
<td>Abnormal LV Filling</td>
<td>+</td>
<td>-</td>
</tr>
<tr>
<td>Female Gender</td>
<td>+</td>
<td>-</td>
</tr>
<tr>
<td>Thickness with Deconditioning</td>
<td>+</td>
<td>-</td>
</tr>
<tr>
<td>Family History HCM</td>
<td>+</td>
<td>-</td>
</tr>
</tbody>
</table>

“Gray zone” of LV Wall Thickness
SCD in Young Athletes

Commotio cordis

• Direct, nonpenetrating trauma to the chest wall
  – baseball, hockey puck, opponent
• Occurs during repolarization
  – electrically vulnerable
• 10% survival
• Softer baseballs shown to reduce risk
SCD In Young Athletes

Commotio Cordis (20%)

http://www.suddendeathathletes.org
SCD in Young Athletes

Commotio Cordis: Vulnerable microseconds in the cardiac cycle

Ventricular Fibrillation
SCD in Young Athletes

- **Long QT Syndrome**
  - Congenital, acquired forms
  - Classically defined by:
    - QTc > 440 msec
    - Bizarre T wave morphology
    - T wave alternans
    - Predisposition to lethal cardiac arrhythmias (*torsade de pointes*)

![Electrocardiogram](image)
SCD in Young Athletes

- **Long QT syndrome and SCD**
  - 287 patients
  - Mean age at death 6.8 +/- 5.6 yrs
  - Warning signs:
    - syncope (26%), seizures (10%), cardiac arrest (9%) preceded by emotion or exercise
  - Family history
    - 39% positive for LQTS, 31% for SCD

Sudden Cardiac Death

• **Coronary Artery Anomalies**
  - Second leading cause of SCD
  - Mechanism of sudden death
  
  - Abrupt lack of oxygen to the heart muscle
  - Heart rhythm abnormality

- No known inheritance pattern
SCD in Young Athletes

Coronary Artery Abnormalities (14%)

The right coronary artery arises from the right cusp, the left from the left cusp
Sudden Cardiac Death

Acute angle takeoff and course between the aorta and pulmonary artery. Both exacerbated by aortic root dilation with exercise.
SCD in Young Athletes

- **Coronary Artery Anomalies**
  - Second leading disease of the heart resulting in SCD
  - Mechanism of sudden death: lethal ventricular arrhythmia
    - sudden onset critical ischemia
    - recurrent infarctions
    - neurocardiogenic syncope
SCD in Young Athletes

- **Marfan Syndrome**
  - Incidence of SCD 5-8/1000
  - Ruptured aortic aneurysm
  - Disproportionate representation among basketball & volleyball players (Flo Hyman)
Marfan Syndrome

Tall stature, long arms

Long, spider-like fingers

Hyperflexible joints

Photos courtesy of Harold Chen, MD. Emedicine: Marfan Syndrome
SCD in Young Athletes

- **Marfan Syndrome**
  - Autosomal dominant
  - Prevalence 1:10,000
  - Defect in the fibrillin gene
  - Tall habitus, arachnodactyly, pectus excavatum, high arched palate, pes planus, scoliosis, ectopia lentis, hyperextensibility, dilated aortic root, mitral valve prolapse
  - Loeys-Dietz: TGF b abnormality
Sudden Cardiac Death

- **Myocarditis**
  - Viral infection of the heart muscle
  - Heritable predisposition

- **Long QT syndrome**
  - Abnormality of the electrical system
  - 60% heritable (may be higher)

- **Aortic stenosis**
  - Birth defect of the heart resulting in progressive obstruction of blood flow from the heart to the body
  - May be inherited 30-50%, NODAL and NOTCH 1
SCD in Young Athletes

- **MYOCARDITIS**
- 18-29% of childhood SCD
- Inflammation predisposes to arrhythmia up to months later
SCD in Young Athletes

- **Arrhythmogenic right ventricular cardiomyopathy**
  - Fatty infiltration and fibrosis of the right ventricle
  - Predisposed to exercise-induced ventricular arrhythmias
  - Genetic basis
SCD in Young Athletes

- **Conduction Abnormalities**
  - Wolff-Parkinson-White syndrome
    - Ventricular pre-excitation
    - Supraventricular tachycardia
  - incidence same in athletes and non-athletes
  - Sudden death
  - atrial fibrillation with rapid conduction along the bypass tract (VF)
SCD in Young Athletes

- Miscellaneous Causes of SCD
  - Cocaine/Other Rx, Steroids, “Huffing”
  - Atherosclerotic coronary artery disease
    - 2-3% of SCD in athletes < 40 years
    - Is this on the rise?
  - Anorexia nervosa/bulemia
    - electrolyte imbalances, conduction abnormalities
SCD in Young Athletes

How do we cost-effectively reduce the incidence of sudden death in our athletes?
Primary Prevention

- Do not let anyone participate in sports
Primary Prevention

• Pre-participation Screening
  – Padua, Italy: Nationwide systematic athletic screening implemented in 1982 (ages 12 – 35 years)
  – There was a significant reduction of SCD in this population (0.56 vs. 0.21)
  – The causes of SCD differ in Italy (ARVD, HCM)
  – Stringent disqualification
  – The Lausanne Recommendations (International Olympic Committee)
  – European Society of Cardiology Consensus Statement
What is the US Consensus?

- **1996**, the American Heart Association consensus panel recommended pre-participation cardiovascular screening for young, competitive athletes based on “ethical, legal and medical grounds”.

- **The Bethesda Conference**  
  "JACC 2005"
Screening athletes

- AHA recommends “a complete and targeted personal and family history and physical examination (including brachial artery blood pressure measurement) performed by a qualified examiner and include 12 key AHA-recommended elements as well as parental verification of medical history for high school and middle school athletes”
Screening Athletes

- The 12 Element AHA Recommendations for Preparticipation CV Screening of Competitive Athletes (2006-7)
- Medical History
- Personal History
  - Exertional chest pain/Discomfort
  - Unexplained Syncope/Near syncope
  - Prior recognition of heart murmur
  - Elevated systemic blood pressure
Screening Athletes

- **Is there Family History?**
  - **Premature death** (sudden, and unexpected, or otherwise) before age 50 years due to heart disease, in > 1 relative
  - **Disability from heart disease** in a close relative < 50 years of age
  - Specific knowledge of **certain cardiac conditions** in family members: hypertrophic or dilated cardiomyopathy, long QT syndrome or other ion channelopathies, Marfan syndrome, or clinically important arrhythmias
Screening Athletes

- **Physical Exam**
  - Heart murmur
  - Femoral pulses to exclude aortic coarctation
  - Physical stigmata of Marfan syndrome
  - Brachial artery blood pressure (sitting position)
Screening Athletes

- **NBA** requires echo and ECG on all players on an annual basis.
- **NFL** teams generally perform ECG’s and obtain echos if clinically indicated.
- **US Olympic Committee** administers preparticipation H & P and further noninvasive testing is performed only when warranted.
Screening Athletes

• International Olympic Committee and European Society of Cardiology recommend ECG’s in all young competitive athletes

• AHA panel does not believe it to be either prudent or practical to recommend the routine use of tests such as 12-lead ECG or echocardiography in the context of mass, universal screening
Screening Athletes

- More than 5 million competitive athletes in high school
- Greater than 500,000 collegiate athletes
- Greater than 5,000 professional athletes
- Population available for screening may be as large as 10 million
Screening Athletes

- **ECG** abnormal in > 90% HCM
- Detects ARVC, ion channelopathies
- Misses catecholaminergic polymorphic ventricular tachycardia
- **ECG** has low specificity as a screening test in athletic populations
SCD in Young Athletes

- Concerns re: widespread use of noninvasive testing
  - False-positive test results
    - unnecessary anxiety
    - unjustified exclusion from sports
    - unjustified exclusion from life insurance coverage
Screening Athletes

Screening for SCD

8,000,000 competitive sports participants in the U.S. (high school & college)

200,000 estimated to be screened to detect 1000 at risk and 1 who would die
SCD in Young Athletes

- Cost of Non-Invasive Screening
  - 6,000,000 high school athletes
    - Screening exam $100
    - ECG $70
    - Echocardiogram $1500
    - Total cost / participant $1670

Total US health care cost $10,020,000,000
Screening athletes

- **Cost of Non-Invasive Screening**
  - 10 million high school athletes
    - Screening exam $25
    - ECG $50
    - Echocardiogram $400

Total US health care cost $2 billion to prevent each preventable death.

(Per suspected athlete $330,000)
Normal Conduction Pathway in the Heart and the ECG

- Sinoatrial (SA) Node
- Atrioventricular (AV) Node
- Left Bundle Branches
- Right Bundle Branch
- Purkinje Fibers

P = Atrial Depolarization
QRS = Ventricular Depolarization
T = Ventricular Repolarization
Normal Sinus Rhythm

Sinoatrial Node
Ventricular Tachycardia

12:57  29MAR96  PADDLES X1.0  HR = 214
Ventricular Fibrillation

Diagram of ventricular fibrillation showing rapid, chaotic electrical activity in the heart, resulting in ineffective pumping of blood.

Waveform indicating the irregular electrical activity typical of ventricular fibrillation.
Defibrillation: The Only Effective Treatment for Ventricular Fibrillation
LIFEPAK® 500
Automated External Defibrillators
How to Defibrillate

- Verify the victim is unconscious, not breathing, without a pulse or signs of circulation
- Turn on AED and attach electrodes
- ANALYZE heart rhythm
- Follow the voice prompts and screen messages
Defibrillation Electrode Placement

Anterior-lateral placement
Defibrillation Electrode Placement

- Correct electrode position optimizes the amount of current flowing through the ventricles
Sudden Cardiac Death

• A public health crisis:
Sudden Cardiac Death

Why is early defibrillation so important?

Chance of survival from SCA diminishes 7-10% with every minute after collapse.
Averting SCD in Young Athletes

- “Play Ball!” “Just DO it!”
- Screen appropriately with expert providers
- Sometimes old-fashioned H&P are the best
- Keep AED handy, in working condition, and educate users
• USA in Beijing
March 2008!
ECG Findings

Distribution of 3 ECG categories with respect to sporting disciplines among 1005 athletes. ECGs that were distinctly abnormal (black bars), mildly abnormal (gray bars), and normal or with minor alterations (white bars) are depicted as proportions of all the athletes participating in each sporting discipline. Only sports with 12 participants are shown. X-C indicates cross-country.