Sudden Death in Athletes
Finding the Needle in the Haystack

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Outline

• Epidemiology
  – Causes of sudden death – what are we looking for?
  – Young vs Old
  – Race and Sex?
• Finding the needle
  – Pre-participation physical exam
  – The ECG Debate
• Preventing deaths – helping the rest of the haystack
  – Other non-cardiac concerns
  – AED use

Epidemiology

What sport causes the most serious injuries?
Rates of direct fatalities and injuries (per 100,000)

<table>
<thead>
<tr>
<th>Activity</th>
<th>High School</th>
<th>College</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ice Hockey</td>
<td>3.11</td>
<td>11.55</td>
</tr>
<tr>
<td>Gymnastics</td>
<td>2.46</td>
<td>17.39</td>
</tr>
<tr>
<td>Football</td>
<td>1.76</td>
<td>6.96</td>
</tr>
<tr>
<td>Lacrosse</td>
<td>0.23</td>
<td>3.30</td>
</tr>
<tr>
<td>Baseball</td>
<td>0.45</td>
<td>1.25</td>
</tr>
<tr>
<td>Track</td>
<td>0.31</td>
<td>0.84</td>
</tr>
<tr>
<td>Basketball</td>
<td>0.05</td>
<td>0.81</td>
</tr>
</tbody>
</table>

Cantu RC, Mueller FO. Phys Sportsmed, 1999

Sudden Death

• Defined as unexpected death occurring as a result of natural causes in which loss of all functions occurred instantaneously or within six hours of the onset of symptoms or collapse

Maron et al., J Am Coll Cardiol, 1986.

Most common causes of Sudden Death

Non-traumatic causes

In individuals older than 35 years old is

Unsuspected coronary artery disease / Myocardial infarction

Most common causes of Sudden Death

Sport-related sudden deaths < 35 yrs old

Cardiac causes

• Hypertrophic Cardiomyopathy (HCM)
• Arrhythmogenic right ventricular cardiomyopathy
• Anomalous origin of a coronary artery
• Dilated cardiomyopathy (viral)
• Marfan’s syndrome

• Commotio Cordis
• Doping
Most common causes of Sudden Death

Sport-related sudden deaths < 35 yrs old

Noncardiovascular deaths

- Exertional hyperthermia
  - 13/30 (43%) of noncardiovascular deaths
- Sickle Cell
- Hyponatremia
- Stroke (Ruptured Aneurysm)
- Bronchial asthma


Epidemiology

- High school or college athletes 1/200,000 – 1/300,000
- 1/70,000 over 3 year high school career

- Exercise related deaths for joggers and marathon racers 1/15000 – 1/50000


What is Normal?
The Athletic Heart

<table>
<thead>
<tr>
<th>Pattern of LVH</th>
<th>Hypertrophic Cardiomyopathy</th>
<th>Athlete’s Heart</th>
</tr>
</thead>
<tbody>
<tr>
<td>Left Ventricle cavity dimension</td>
<td>&lt; 45 mm</td>
<td>&gt; 55 mm</td>
</tr>
<tr>
<td>Left Atrium</td>
<td>Dilated</td>
<td>Non dilated</td>
</tr>
<tr>
<td>LV diastolic filling</td>
<td>Abnormal</td>
<td>Normal</td>
</tr>
<tr>
<td>Gender</td>
<td>Male and female</td>
<td>Male</td>
</tr>
<tr>
<td>Effect of detraining</td>
<td>No reduction of LVH</td>
<td>Regression of LVH</td>
</tr>
<tr>
<td>Family history</td>
<td>Positive/Negative</td>
<td>Negative</td>
</tr>
</tbody>
</table>

Maron et al., Circulation, 1995.

Race and Sudden Death

- More than 50% of SCD in high school and college student-athletes occur in blacks (usually due to HCM)
- Between 2003 and 2007, 300 nationally ranked black male athletes (mean age 20.5 years) underwent 12-lead electrocardiogram and 2-dimensional echocardiography
- Black athletes exhibited greater LV wall thickness and cavity size compared with sedentary black and white individuals

Basavarajaiah et al., J Am Coll Cardiol, 2008
**Race**

- Black athletes had greater LV wall thickness compared with white athletes ($11.3 \pm 1.6$ mm vs. $10 \pm 1.5$ mm; $p < 0.001$)
- In absolute terms, 54 black athletes (18%) had LV wall thickness $>12$ mm compared with 12 white athletes (4%), and 3% of black athletes exhibited LV wall thickness $>15$ mm compared with none of the white athletes
- More false positives in blacks vs risk factor?

*Basavarajaiah et al., J Am Coll Cardiol, 2008*

**Sex and Sudden Death**

- Sudden death in Females:Males occurs in a ratio of 1:9
- Rare in young females
- Lower participation?
- Different sports? No football?

**Arrhythmia**

- Ventricular fibrillation is usually the underlying fatal rhythm in athletes
- Can we save them?

**Cardiac Causes of Sudden Death**

Maron et al., Circulation, 2007
Hypertrophic cardiomyopathy

- Estimated prevalence is 1:500
- Prevalence is higher among blacks (0.24 percent) than among whites (0.10 percent) (Maron, 1995)
- Phenotypic expression of HCM may not be evident or complete until adolescence
- DNA testing for one of the 10 most common HCM-causing mutant genes available

Hypertrophic Cardiomyopathy (HCM)

- High QRS voltages and diffuse negative T waves

Hypertrophic cardiomyopathy

- Screening with history is unreliable
- Patient typically does not complain of syncope or have a FH of sudden death Maron BJ, et al., Circulation, 1995.
- Electrocardiogram is abnormal in more than 90% LaCorte MA, et al., Clin Cardiol, 1989.

Hypertrophic Cardiomyopathy (HCM)

- Septum usually < 13 mm
Hypertrophic Cardiomyopathy (HCM)

- Fibrotic and disorganized, scarred myocardium consequent to episodes of myocardial ischemia

Arrhythmogenic Right Ventricular Cardiomyopathy (ARVC)

- ARVC is a genetic condition causing fibrosis and fatty infiltration of the myocardium
- Progressive wall thinning and dilation of the right ventricle
- Associated with ventricular tachyarrhythmias, particularly in relation to psychophysical stress
- May be completely asymptomatic or may refer palpitations, syncope/presyncope at rest and/or during effort

Cardiac physical exam may be negative
ECG is often abnormal
- QRS complex widening (“epsilon” wave), due to right ventricular activation delay
- Repolarization abnormalities (negative T waves in right precordial leads)
- Ventricular premature beats
ECHO and MRI are used to make diagnosis
Anomalous Coronary Artery

- Young athletes with CCAA have a 70-fold increase in relative risk of dying during effort than at rest (Corrado D, JACC, 2003)
- Most common anomaly linked with sudden death is a left main artery arising from the right sinus of Valsalva
- Abnormal course to the usual position is between pulmonary artery and aortic root


Anomalous Coronary Artery

- Physical exam usually normal
- Rest and stress ECG usually normal
- Only about 1/3 of subjects had symptoms (chest pain, palpitations and/or syncope/presyncope)
- If symptoms are present, usually during or immediately after a strenuous effort
- ECHO, in experienced laboratories, can be useful as first diagnostic approach
- MRI and angio-coronary computed tomography (angio-CT) scans best to assess coronary arteries anatomy
**Dilated Cardiomyopathy / Myocarditis**

- Inflammatory condition of the myocardium, usually due to a viral infection
- Can be asymptomatic but may show subtle symptoms (exercise intolerance, palpitations, tachycardia at rest and excessive heart rate increase on exercise, etc.), which appear days or weeks after a viral illness

**Marfan’s Syndrome**

- Genetic (Fibrillin)
- Tall (above 97th percentile)
- Scoliosis (60% of Marfan’s patients)
- Pectus carinatum / excavatum
- Arm span to height ratio than 1.05
- Long fingers and extremities
- Joint hypermobility

**Dilated Cardiomyopathy / Myocarditis**

- ECG may be abnormal and dysrhythmias are frequently present when investigated with stress ECG and Holter monitoring
- Malignant ventricular arrhythmias most likely responsible sudden death in presence of either active or healing myocarditis
- Athlete can safely return only after full recovery

**Marfan’s Syndrome**

- Ocular system
  - Lens dislocation
- Cardiovascular
  - Aortic root dilation, MVP with regurg (50-80%), Aortic regurg
- Pulmonary, skin and dura
- Spontaneous pneumothorax, dural ectasia

Wolf-Parkinson’s White Syndrome

• Disease of the cardiac conduction system, characterized by the congenital presence of one (or more) additional electrical pathways
• Prevalence ranges from 0.15% to 0.2% of the general population, but the risk of ESD regards only the 0.1% of these subjects
• Abnormal pathways typically result in atrial impulses bypassing the atrio-ventricular node and can reach the ventricles more rapidly

In cases of atrial tachyarrhythmias (atrial fibrillation) a desynchronization of ventricular activity may occur deteriorating in a ventricular fibrillation (VF)
• Invasive or transoesophageal electrophysiological study is mandatory in symptomatic subjects to assess risk for sudden death

Channelopathies

• Rare, malignant arrhythmias
• Long and short QT syndromes, Brugada syndrome, polymorphic cathecolaminergic ventricular tachycardia, etc.
• An accurate analysis of rest and stress-ECG needed
• If present, withdrawal of these subjects from competitive sports is recommended

Commotio Cordis

• An electrophysiological event caused by blunt precordial chest impact
• Can occur in individuals usually free from heart disease
• Chest impact is delivered within a narrow, electrically vulnerable period of the cardiac cycle, it may start a VF
Baseball

Commotio Cordis
- 158 reported cases, 95% male, 87% white
- 10-14 year old is peak incidence
- 10% survival w/o AED, 50% with AED

Solutions?
- Chest protectors
- Larger balls
- AED on field?

Performance Enhancing Drugs

Stimulants (epinephrine, ephedrine, cocaine etc.)
- Sympathetic drugs all have a pro-arrhythmic effect, particularly in presence of dehydration, abnormal electrolytes or in association with other heart abnormalities

Erythropoietin
- Related deaths mainly due to blood hyper-viscosity and increased thrombogenesis

Anabolic steroids
- May cause cardiac hypertrophy, myocardial fibrosis, and accelerated atherosclerosis

The Pre-participation Physical Exam

False Sense of Security

- Ability to detect serious problems by PPE is limited
- Athlete, parents, coaches and medical staff should avoid false expectations that PPE rules out possibility of injury or sudden death during sports
- No evidence that injuries are reduced by PPE
- Risk Management
History is Key

- Around 30% of athletes will report an injury needing MD treatment in past
- 70% of orthopaedic or medical problems resulting in disqualification are identified by history alone

AMA, Arch Pediatr Adolesc Med, 1994

Results of PPE studies

- Referrals for further evaluation or treatment = 2.1% to 11.9%
- Disqualifications = 0.2% to 1.9%


Disqualification

- Musculoskeletal condition was the most common reason (43.4%), second was cardiac problems (0.37%)
- N= 2,739 high school athletes


Case 1

Who? 16 year old basketball player
When? Comes in for PPE and try-outs start next week
What? 2/6 Systolic Crescendo/decrescendo
Murmur heard L sternal border, decreases with Valsalva; Player is asymptomatic and has played basketball all summer
Case 1

- Will you sign the paper to clear him to play?
- Do you want an EKG?
- Do you want an ECHO?

Case 2

- Will you sign the paper to clear him to play?
- Do you want an EKG?
- Do you want an ECHO?

Medical history

Maron et al., Circulation, 2007

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

With parental verification of responses*
Medical History

Maron et al., Circulation, 2007

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 cardiac 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
Precordial auscultation in both supine/sitting and standing positions to identify heart murmurs consistent with left ventricular outflow tract obstruction
10. Femoral pulses to exclude aortic coarctation
11. Physical stigmata of Marfan syndrome
12. Brachial artery blood pressure (sitting position)

H C M

- Hypertrophic cardiomyopathy is characterized by:
  - A harsh systolic ejection murmur
  - Decreases with squatting
  - Increases in intensity upon standing or during a Valsalva maneuver

H C M

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Heart

- Auscultate supine

Heart

- Auscultate supine
- Auscultate standing
- Increase murmur
- Rule out HCM

“I was told I have a Heart Murmur”

- Loud murmurs (grade 3 or more)
- Systolic: holosystolic, late systolic, systolic ejection with or without or click
- Any diastolic
- Continuous murmurs
  Warrant cardiac investigation
- If the murmur is innocent, full participation is permitted

“My heart races or skips beats with exercise”

Racing heart
- PSVT
- WPW
- Atrial fibrillation
- Usually athlete is simply out of shape

Syncope
- Long QT Syndrome
- Arrhythmia

Chest Pain
- HCM
Cardiac Screening Effectiveness

- CV history identified 2% and exam identified 3.2% of abnormalities (n=5165)
- B.P. measurement (abn = 0.3%) (5 disqualified for severe HTN)
- History and physical exam
  - 3% sensitivity, specificity 97.8%, FP rate 2.2%
- Electrocardiograms (abn = 15.7%)
  - 60-70% sensitivity, spec 97.4%, FP rate 2.6%
- Echo and Stress test used if abnormalities

US studies

- Cost of identifying a significant problem $4,537 (n=763)
- 15/16 identified by history; 2/16 disqualified
  Risser etal, J School Health, 1985
- Cardiovascular history and physical exam $84,000/yr
- ECG $44,000/yr
- Ultrasound $200,000/yr

EKG vs No EKG?

- N= 12,550 athletes (mean 9 yr follow up)
- Of the 81 athletes with abnormal ECGs, 5 (6%) ultimately proved to have cardiomyopathies; one died suddenly at the age of 24 years (2 arrests)
- 3 developed HCM; one death
- No deaths in controls
- No routine screening EKG’s recommended by the AHA

Big in Japan

- Standard CV screening with history and ECG (Gr. 1, 7, 10)
- N = 69,033 Gr. 7; 37,807 6-yr follow up
- 1876 sent for secondary screening (2.7%)
- 9 students “high-risk”; 3 sudden deaths
- Estimate $8,800 per year of life saved
Common European Proposal

- Based on 25-year Italian experience with competitive athletes
- Italian screening with 3 x greater cost-effectiveness
- Suggest routine PPE with systematic screening by 12-lead ECG

Consensus of the Study Group on Sports Cardiology of the Working Group of Cardiac Rehabilitation and Exercise Physiology and the Working Group of Myocardial and Pericardial Diseases of the European Society of Cardiology

Corrado et al., Eur Heart Journal, 2005

Other Cardiac Examinations

- Stress ECG can identify a large number of subjects with potentially lethal diseases
- Further instrumental investigations
- ECHO
- Holter
- MRI
- Angio-CT

Picking the Right Activity

- PPE, Physician and Sports Medicine, McGraw Hill, 2005 (new edition soon to be published)

Use Bethesda Guidelines

- Young athletes with the unequivocal diagnosis of HCM are discouraged from competitive athletic participation, **with the exception of low-intensity sports (1A Classification Sports)**
Use the Bethesda Guidelines

Recommendations:

1. Detection of **coronary anomalies** of wrong sinus origin in which a coronary artery passes between great arteries should result in exclusion from all participation in competitive sports.

2. **Participation in all sports three months after successful operation** would be permitted for an athlete without ischemia, ventricular or tachyarrhythmia, or dysfunction during maximal exercise testing.

**Use the Bethesda Guidelines**

1. Athletes with episodes of **atrial flutter/fibrillation** and syncope or near syncope whose maximal ventricular rate at rest (without therapy) ... exceeding 240 beats/min should be considered for catheter ablation therapy...

2. Athletes with no structural heart disease who have had **successful catheter or surgical ablation** of the accessory pathway, are asymptomatic, and have normal AV conduction and **no inducible arrhythmia by follow-up electrophysiologic study** can participate in all competitive sports in several days. Those without an electrophysiologic study and no spontaneous recurrence of tachycardia for two to four weeks after ablation can participate in all competitive sports.

**Medicolegal**

- Must practice within your scope, at the same level of standard of care of your peers
- Don't be negligent
- Document, document, document
- Disqualify athlete if medically necessary
- Medical waiver and informed consent if athlete wishes to compete against medical advice (but NOT perfect)
Treating the rest of the Haystack

Sports is a Risk Taking Activity

Sports Medicine is Managing Risk

Heat Can Kill
"21 died from 95 to 2001”
National Center for Catastrophic Sport Injury, 2003

- EHS death rate football: 1/350,000/yr
- 5 heat-related deaths reported in summer 2006
- Soccer heat illness was 0.6 cases/1000 player-hours under “normal” conditions compared to a rate of 2.8/1000 player-hours during “hot” years

Who’s at Risk?
Young Athletes
- Reduced heat tolerance
- Take longer to acclimatize
- Less sweating
- Lower voluntary fluid intake
Obese athletes
- Have poorer thermoregulation
- Need to acclimatize to hot environment
Specific Sports

- Outside
- Long exercise duration
- Risk of dehydration
  - X-country
  - Long Track events
  - Marathons
  - Tennis
  - Soccer
  - Football

Too Much Water is DANGEROUS

Hyponatremia

- Can look like dehydration
- Athlete may be confused, obtunded
- Lowering sodium can lead to seizures, even death

Hyponatremia

Risk factors:
- Racing time > 4 hours
- Female sex
- Low Body Mass Index < 20
- Excessive drinking > 3 liters
- Postrace weight > prerace weight

Other mechanisms?
- Hormonal, excessive salt loss

Sudden Cardiac Arrests

- Multiple events at high schools over the last five years
- Some saved using an Automated External Defibrillator (AED).
- Incidence of pediatric sudden cardiac arrest varies (2.6 to 19.7 annual cases per 100,000)
- Estimating the number of pediatric SCA events is difficult.
Save A Life

Automated External Defibrillator
- There are only two rhythms that a defibrillator will shock: Ventricular Tachycardia and Ventricular Fibrillation
- 40% of cardiac arrest victims have this on first rhythm analysis
- The longer it takes to get the AED and get it connected to the patient, the less likely there will be a rhythm to shock
- In one study, untrained 6th graders operated AEDs almost as quickly as trained paramedics

2004 AAP and AHA Guidelines

- Establish an effective communication system
- Develop an Emergency Action Plan and practice yearly
- Identify staff and students with life threatening conditions
- Train staff in CPR and AED use
- Implement a lay rescuer AED program

Good Samaritan Law

- Any citizen “acting in good faith” can administer CPR or use an AED in an emergency.
- If your school has an AED, someone at the school MUST be trained in its use.

PlaySafe Sports Physicals

- May 2, 2009 for high school students in SF
- Complete pre-participation physical exam consisting of a thorough medical, orthopaedic, and cardiac screening
- A cardiac exam consisting of an EKG screening will be given to each participant
- An echocardiogram (ultrasound) is available on-site for participants that require additional screening
- Program is supported by UCSF Orthopaedics and Cardiology, National Football Foundation and College Hall of Fame, and CIF: San Francisco Section
EKG Screening Debate

Solutions

• Increase awareness about symptoms and signs of cardiovascular disease
• Offer EKG as a screening tool
• Need more research before adopting EKG as standard of care

• GOAL should be IMPROVE the PPE

“You see what you look for and diagnose what you know”

Take Home points:
1. Improve your PPE – encourage proper timing
2. Practice standard of care
   – Ask the right questions
   – Check for murmurs
3. Consider an EKG – the debate continues
4. Be prepared – AED available?
Can the Athlete Play Safely?

- Make a working diagnosis
- Is there potential for worsening injury?
- Is there potential for a new secondary injury
- MD or trainer decides: CAN THE ATHLETE PLAY SAFELY?
- If there’s a medical problem and you can’t clear them, put your foot down

Can the Athlete Play Effectively? Pain-Free?

- Athlete, coaches and medical staff decide can the athlete play effectively
- Athlete needs to indicate can he/she play relatively pain free