Prevention of Sudden Cardiac Death In Competitive Young Athletes: Is ECG the Answer?

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Objectives

- To review the epidemiology of cardiac sudden death in athletes.
- Understand the cardiac causes of cardiac sudden death in athletes.
- Identify athletes at increased risk for cardiac sudden death.
- Understand the limitations in prevention of cardiac sudden death.
Sudden Cardiac Death

- True incidence of SCD in athletes is not known.
- Frequency of SCD in US < 100 deaths per year (Maron, 2009).
- Estimated incidence of SCD:
  - High school athletes: 1 in 100,000 – 200,000
  - College athletes: 1 in 65,000 - 69,000
  - General population: 55 in 100,000
Sudden Cardiac Death

- Maron et al 2009
  - 27 year registry of SCD
  - 1866 athletes identified
  - 56% due to CV causes
    - 36% of CV due to HCM
    - 17% of CV due to coronary anomalies
  - 3% commotio cordis
CV SCD Among US Athletes <40y
1980 -2006

Data from Maron et al Circ 2009

n=1049
Non-CV SCD in US Athletes <40y 1980-2006

Data from Maron et al Circ 2009

n=1866
## Estimated Incidence of SCD in Young Athletes

<table>
<thead>
<tr>
<th>Location</th>
<th>Incidence/100,000 person-yrs</th>
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<tbody>
<tr>
<td>Italy-Veneto region (Corrado et al 2006)</td>
<td>0.87</td>
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<tr>
<td>Minnesota (Maron et al 2008)</td>
<td>0.93</td>
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<tr>
<td>US (Maron et al 2009)</td>
<td>0.6</td>
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</table>
SCD in US and Italy

From Link and Estes Prog in CV Diseases 2008
SCD in NCAA Athletes

- NCAA database used to identify SCD over 5 years (no autopsy confirmation)
- 273 deaths over 1.9m athlete-yrs
- 68% non-medical (accidents 51%)
- 29% (n=80) medical
  - CV=45 (56% of medical, 16% of total)
- SCD incidence 1:43,770/yr

Harmon et al Circ 2011
Deaths have occurred in several well known collegiate, Olympic, and professional athletes resulting in a great deal of publicity and increased public concern.

Thomas Herrion SF 49ers
Age 23 HCM
Reggie Lewis  27 Boston Celtics
HCM
Hypertrophic Cardiomyopathy

- Epidemiology
  - Prevalence = 1 in 500 (0.2%)
  - Accounts for up to 36% of exercise related sudden death in individuals 35 yrs or younger
Hypertrophic Cardiomyopathy

- Inherited as autosomal dominant trait with variable penetrance

- Disease causing mutations have been identified in 10 genes encoding sarcomere and regulatory proteins
  - Beta myosin heavy chain
  - Cardiac troponin T
  - Myosin-binding protein C

- Overall incidence of sudden death in patients with HCM is ~ 1% per year (prior estimates 3-6%/yr)
  - Disease subsets - 10-20% of HCM cases - may be at higher risk ~5%/yr
Hank Gathers age 23 LMU
HCM
Gathers Collapses, Then Dies
Hypertrophic Cardiomyopathy

- Distinguishing features

  - Histologically:
    - Myocardial disarray
    - Abnormal intramural coronary arteries
    - Myocardial scarring (likely due to ischemic events)
  - Thickened left ventricular wall (>13mm) in a non-dilated LV
  - Asymmetric thickening of LV
    - Ventricular septum to free wall ration >1.3
  - Thickening of the mitral valve, atrial dilatation, LV outflow tract obstruction
Hypertrophic Cardiomyopathy

- Patient identification
  - History:
    - Family history of sudden death (or HCM), syncope, presyncope, dyspnea, exertional angina, fatigue
    - Symptoms do not correlate with disease expression
Hypertrophic Cardiomyopathy

- ECG
  - 75-95% will have abnormal 12 lead ECG
  - No characteristic pattern
Hypertrophic Cardiomyopathy

- Echocardiography
  - Diagnosis is best made with echo even in the absence of symptoms
  - Hypertrophied but non-dilated LV
  - Findings:
    - LV thickness > 15mm in an adult (13 - >30mm)
    - Septum : free wall ratio of >1.3
    - Septum : LVESD > 0.48
  
- Mutant HCM gene can be present without LVH
Anomalous Coronary Artery
Marfan’s Syndrome

Flo Hyman 31y
Aortic Dissection
Ronny Turiaf enlarged aortic root
Primary Electrical Abnormalities

- Structurally normal heart
  - Long QT
  - Brugada
  - Short QT
  - Catecholaminergic polymorphic VT
  - WPW
ARVD

- Replacement of RV myocardium with fatty and fibrous tissue
- Creates milieu for malignant arrhythmias
- Most common cause of SCD in NE Italy (Corrado et al)
- Tw inversions and epsilon waves in R precordial leads
  - VTs and LBBB
Commotio Cordis

The syndrome of relatively minor blunt chest wall impact producing cardiac arrest in the absence of structural cardiac disease and no acute cardiac injury identifiable at autopsy.

Maron 1997, Link 1997
George Boiardi was a healthy college student. He was a senior history major at Cornell University and a co-captain of the lacrosse team. However, on March 17, he was pronounced dead at Cayuga Medical Center in New York. Boiardi was hit in the chest by a lacrosse ball during a game against Binghamton. With less than three minutes left in the game and Cornell leading, Binghamton player Nate Kerstein shoot the ball towards the goal when it hit Boiardi in the chest, causing him to collapse to the turf. Doctors pronounced him dead by the end of the day.

The official cause of Boiardi's death is thought to be commotio cordis.
Screening Measures

- Athletes < 35 yrs
  - History: can be helpful, but also can be unreliable
  - Physical exam: sensitivity is poor
  - ECG is sensitive, but not specific (except in cases of long QT syndrome, WPW, or sinus node disturbances)
  - ECG: Potential for false positives
    - Leads to additional testing
    - Temporary restriction
    - Possible erroneous exclusion
# Causes of Sudden Cardiac Death

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<thead>
<tr>
<th>Condition</th>
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<th>Physical Exam</th>
<th>ECG</th>
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<tr>
<td>HCM</td>
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<td>Anomalous Coronary Arteries</td>
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<tr>
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Screening for SCD with ECG

Identifiable with ECG
- ARVD
- HCM
- Long QT
- WPW

Not Identifiable with ECG
- Anomalous Coronary Arteries
- Marfan’s
- Commotio Cordis
- Premature CAD
ECG Screening

- Pelliccia et al 2000
  - 1050 athletes screened with hx, exam, ECG, and echo
  - 40% had abnormal ECG
  - 14% distinctly abnormal; 26% mildly abnormal
    - 10% with distinctly abnormal ECG had CV abnormality on echo
    - 5% with mildly abnormal ECG had CV abnormality
    - 4% with normal ECG had CV abnormality

Pelliccia et al Circulation 2000;102:278-284
ECG Screening

- 53 athletes had CV abnormality on echo
  - 27 had abnormal ECG
  - 26 had normal ECG

ECG
- sens = 51%  spec = 61%
- + PV = 61%  - PV = 96%

Pelliccia et al Circulation 2000;102:278-284
ECG Screening

- Pellicia et al 2007 Eur Heart J
  - 32,652 subjects 8-78y (median 17y)
  - 11.8% with abnormal ECG
  - 4.8% distinctly abnormal ECG
ECG Screening

- Baggish et al 2010 Ann Int Med
  - 510 athletes
  - ECG and echo
  - 11 had abnormal echo
    - H&P detected 5/11
    - ECG detected an additional 5

- H&P 45% sens. 94% specific
- ECG + H&P sens 91% spec 83%
- ECG false pos of 17%
Screening for SCD

- Corrado et al 2006
- Screening of athletes with Hx, PE and ECG
- SCD between 1979-2004 recorded for region
- 55 cases of SCD among athletes
- Incidence of 1.9/100,000 person-years

Screening for SCD

- Corrado et al 2006
  - Incidence of SCD among athletes declined from 4.0 prior to screening to 0.87 /100,000 person-yrs at end of study period

- For non-athletes SCD incidence was unchanged (0.77 to 0.81)
• Annual incidence of SCD in athletes decreased by 89%

• 2% disqualified due to CV reasons

Screening for SCD

- Corrado et al 2006
  - Causes of CV disqualification
    - Sig increase in DQ b/c of ARVD
    - No increase in DQ b/c of HCM
  - Most of reduction in SCD due to decrease in deaths from cardiomyopathies
    - ARVD had 84% decline 0.9 to 0.15
    - No sig change in incidence of HCM deaths
Screening for SCD (Corrado et al 2006)

- Limitations:
  - Population demographics
  - Retrospective
  - Uncontrolled
  - Observational
  - ECG
  - Extraordinarily high annual mortality rate in early 1980s
Screening Measures

- AHA estimates cost of screening in US = $2 billion/yr
- Cost of preventing one death = $3.4 million

Screening Measures

- Estimates in UK:
  - 1,520,021 would be screened
  - 140,361 referred
  - W/o screening, estimated 196 SCDs/yr, 64 in athletes
  - 40 SCDs prevented with screening
  - 24 SCDs in athletes despite screening
  - 791 athletes would be DQed for every life saved

Elston J and Stein K. BJSM 2009 epub
Current Screening Recommendations

- Current screening procedures either lack the sensitivity or specificity to detect disease, or are not cost effective.

- “It is unrealistic to expect that standard large-scale athletic screening examinations can exclude all clinically relevant diseases.”

Sun sets on Smith’s UW career

By Jonathan Linder BadgerNation.com
Date: Aug 27, 2004

Badger tailback practiced once this fall; was asymptomatic until fall camp physical caught a heart murmur
Summary

- Exercise-related sudden cardiac death is rare.
- In young athletes in the US, it is usually a consequence of a HCM.
- The use of ECG to prevent SCD from HCM has not been proven.
- Successful SCD screening strategies remain elusive and controversial.
THE DEATH OF REGGIE LEWIS: SO MANY QUESTIONS
Thank you!