Sudden Cardiac Death in the Young and Implications for the Preparticipation Physical Exam

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Children’s Mercy Hospitals and Clinics

No Disclosures
SCDY and the PPE

• Objectives
  – Review epidemiology and causes of SCDY
  – Discuss the current PPE cardiac evaluation
  – Review recent medical literature, including controversy surrounding the role of the PPE in preventing SCDY
  – Discuss rationale/aims for the NIH/CDC prospective SCDY registry
Sudden Cardiac Death in the Young

- Sudden cardiac death in the young: rare but catastrophic events
- Estimated incidence varies widely: 0.6–6.2 deaths per 100,000 persons
- Diagnoses related to SCDY include:
  - Hypertrophic cardiomyopathy (HCM) - Genetic
  - Coronary artery anomalies
  - Cardiac Arrhythmias - Genetic
  - Myocarditis
  - Marfan Syndrome - Genetic

12 young sudden deaths a week
and every loss is heartbreak

Young at risk of sudden death
Figure 2. Percent distribution of all deaths to teenagers 12–19 years, by cause of death: United States, 1999–2006

- Unintentional injury (48%)
- Homicide (13%)
- Suicide (11%)
- Malignant neoplasms (6%)
- Heart disease (3%)
- Congenital anomalies (2%)
- Other causes (residual) (17%)

- Percent distribution of unintentional injury deaths by detailed mechanism of injury:
  - Motor vehicle traffic accident (73%)
  - Unintentional poisoning (7%)
  - Unintentional drowning (5%)
  - Other land transport accident (3%)
  - Unintentional discharge of firearm (2%)
  - Other unintentional deaths (10%)

Sudden Cardiac Death in the Young

- National/international discussion regarding most effective screening for conditions related to SCDY
  - History and physical alone?
  - EKG?
  - Echo?
  - Genetic Testing?
  - Athletes vs. non-athletes?
Causes of SCDY

- **Cardiomyopathy**
  - Hypertrophic cardiomyopathy
  - Dilated cardiomyopathy, myocarditis
  - ARVC

- **Arrhythmias**
  - Ventricular dysrhythmias: Long QT, Brugada, ARVC
  - Bradycardia: sick sinus syndrome, complete AV block
  - SVT: WPW, AV node re-entry, atrial tachycardia

- **Structural heart disease**
  - LV outflow obstruction: aortic valve stenosis, HCM
  - Coronary anomalies
  - Marfan: aortic root dilation
Hypertrophic Cardiomyopathy
Familial Syndromes Associated with Ventricular Arrhythmia

- **Autosomal dominant inheritance**
  - Romano-Ward Long QT syndrome
  - Brugada syndrome
  - Arrhythmogenic Right Ventricular Cardiomyopathy

- **Autosomal recessive inheritance**
  - Jervell and Lange-Nielsen syndrome
    (congenital deafness)
Familial Syndromes Associated with Ventricular Arrhythmia

• The Long QT syndromes (LQTS) characterized by:
  – Syncope
  – Seizures
  – Sudden death

  Polymorphic ventricular tachycardia, “torsades de pointes”
Wolff-Parkinson-White
Wolff-Parkinson-White

Rapid conduction of atrial dysrhythmia
Single coronary artery (RCA) with LCA coursing between great vessels

Marfan Syndrome: aortic dissection
## Preparticipation Physical Evaluation

### History Form

(Note: This form is to be filled out by the patient and parent prior to seeing the physician. The physician should keep this form in the chart.

<table>
<thead>
<tr>
<th>Date of Exam</th>
<th>Name</th>
<th>Sex</th>
<th>Age</th>
<th>Grade</th>
<th>School</th>
<th>Sport(s)</th>
<th>Date of birth</th>
</tr>
</thead>
</table>

#### Medications and Allergies

Please list all of the prescription and over the counter medications and supplements (herbal and nutritional) that you are currently taking.

<table>
<thead>
<tr>
<th>Do you have any allergies?</th>
<th>Yes</th>
<th>No</th>
<th>If yes, please identify specific allergies:</th>
</tr>
</thead>
</table>

#### General Questions

1. Have you ever been diagnosed with or treated for a heart problem?
2. Do you have any heart disease or heart rhythm problems?
3. Have you ever had a heart attack?
4. Have you ever been told you have high blood pressure?
5. Do you have any eye disease?
6. Have you ever had a stroke?
7. Have you ever had a seizure or convulsion?
8. Do you have any family history of heart or chest pain?
9. Have you ever had a stroke?
10. Do you have any family history of cancer?

#### Medical Questions

13. Do you have diabetes?
14. Do you have high cholesterol?
15. Do you have any other medical conditions?
16. Do you have any other health problems?
17. Do you have any other conditions that need to be treated by a doctor?
18. Do you have any other conditions that need to be managed by a nurse or other health professional?
19. Do you have any other conditions that need to be monitored by a doctor or other health professional?

#### Heart Health Questions About Your Family

1. Has anyone in your family had a heart attack or stroke?
2. Has anyone in your family been diagnosed with or treated for high blood pressure?
3. Has anyone in your family been diagnosed with or treated for diabetes?
4. Has anyone in your family been diagnosed with or treated for cancer?

#### Bone and Joint Questions

1. Have you ever had a bone fracture?
2. Have you ever had an injury to a bone?
3. Have you ever had any other injuries?
4. Have you ever had any surgery?

#### Personal Statements

I hereby state that, to the best of my knowledge, my answers to the above questions are complete and correct.

Signature of patient or parent/guardian

Use of this form is authorized.

Children’s Mercy
Hospitals & Clinics
www.childrensmercy.org
HEART HEALTH QUESTIONS ABOUT YOU

5. Have you ever passed out or nearly passed out DURING or AFTER exercise?
6. Have you ever had discomfort, pain, tightness, or pressure in your chest during exercise?
7. Does your heart ever race or skip beats (irregular beats) during exercise?
8. 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: ______________________
9. Has a doctor ever ordered a test for your heart? (For example, ECG/EKG, echocardiogram)
10. Do you get lightheaded or feel more short of breath than expected during exercise?
11. Have you ever had an unexplained seizure?
12. Do you get more tired or short of breath more quickly than your friends during exercise?

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13. 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)?

14. 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?

15. Does anyone in your family have a heart problem, pacemaker, or implanted defibrillator?

16. Has anyone in your family had unexplained fainting, unexplained seizures, or near drowning?
The PPE Cardiac Physical Exam

- Cardiac Exam
  - Blood pressure
  - Marfan syndrome features
  - Heart murmurs (auscultation standing, supine, +/- Valsalva)
  - Location of point of maximal impulse (PMI)
  - Pulses: simultaneous femoral and radial pulses
Syncope (Fainting) in Children

- Common
- Dramatic events
- Potential for injury
- RARELY, self-limited expression of a potentially lethal condition

The Common Faint

• Inciting factors
  – Noxious stimulus (blood draw)
  – Illness
  – Prolonged standing
  – Warm environment
  – Recent exercise

• Associated symptoms/signs/events
  – Prodrome: dizziness, visual changes, pallor
  – Seizure
  – Nausea/vomiting
  – Injury
PATH TO DESTRUCTION

MUNICIPAL BONDS
TREASURIES
BANKRUPTCY

LIVE
FOX NEWS
channel

ADAHN ATTRIBUTES GLOBAL ECONOMIC CRISIS TO “THE GREAT...”
Common Faint - Vasodepressor Mechanism

BP (mm Hg)  HR (b/min.)

140  70
120  60
100  50
80   40
60   30
40   20
20   10

ONSET
STOP
TILT TEST

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Syncope in Children

• Exercise or Stress-related Syncope:

RED Flag Event
Syncope in Children

- **The Electrocardiogram**
  - Hypertrophic Cardiomyopathy
  - Long QT; Brugada syndrome
  - Wolff-Parkinson-White

- **Consensus Statements**
Syncope in Children

• Cardiac Workup beyond ECG indicated:
  – Faint with exercise
  – Syncope without prodrome, while supine, preceded by chest pain or palpitations
  – Event triggers: loud noise, fright, extreme emotional stress
  – Known or suspected heart disease
  – Family history: unexplained or sudden death, known familial heart disease
Reducing SCDY

- Is the current PPE enough?
- Does mass EKG screening make sense?
- What about non-athletes?
- What should we do for asymptomatic athletes?
- Do we exclude athletic participation for asymptomatic heart disease?
- What are the physical, social and mental health costs of exclusion?
Trends in Sudden Cardiovascular Death in Young Competitive Athletes After Implementation of a Preparticipation Screening Program
Corrado et al. JAMA 2006; 296(13): 1593-1601

Table 1. Number and Annual Incidence Rates of Total and Cause-Specific Sudden Cardiovascular Death in Screened Athletes and Unscreened Nonathletes in Relation to 3 Screening Periods*

<table>
<thead>
<tr>
<th></th>
<th></th>
<th></th>
<th></th>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>No. of Events</td>
<td>Incidence Rate (95% CI)</td>
<td>No. of Events</td>
<td>Incidence Rate (95% CI)</td>
<td>No. of Events</td>
</tr>
<tr>
<td>Total sudden deaths in athletes</td>
<td>14</td>
<td>4.19 (1.78-7.59)</td>
<td>29</td>
<td>2.35 (1.94-2.75)</td>
<td>12</td>
</tr>
<tr>
<td>Cardiomyopathies</td>
<td>5</td>
<td>1.50 (0.21-2.78)</td>
<td>7</td>
<td>0.57 (0.26-0.87)</td>
<td>2</td>
</tr>
<tr>
<td>Coronary artery disease</td>
<td>3</td>
<td>0.90 (0.3-1.2)</td>
<td>5</td>
<td>0.41 (0.09-0.72)</td>
<td>3</td>
</tr>
<tr>
<td>Cardiac conduction disease</td>
<td>1</td>
<td>0.30 (0-1.56)</td>
<td>2</td>
<td>0.16 (0-0.40)</td>
<td>1</td>
</tr>
<tr>
<td>Myocarditis</td>
<td>1</td>
<td>0.30 (0-1.56)</td>
<td>4</td>
<td>0.32 (0.02-0.63)</td>
<td>2</td>
</tr>
<tr>
<td>Congenital coronary anomalies</td>
<td>1</td>
<td>0.30 (0-1.56)</td>
<td>4</td>
<td>0.32 (0.02-0.63)</td>
<td>2</td>
</tr>
<tr>
<td>Mitral valve prolapse</td>
<td>1</td>
<td>0.30 (0-1.56)</td>
<td>4</td>
<td>0.32 (0.02-0.63)</td>
<td>1</td>
</tr>
<tr>
<td>Other†</td>
<td>2</td>
<td>0.60 (0-1.87)</td>
<td>3</td>
<td>0.24 (0-0.52)</td>
<td>1</td>
</tr>
<tr>
<td>Total sudden death in nonathletes</td>
<td>29</td>
<td>0.77 (0.26-1.26)</td>
<td>110</td>
<td>0.79 (0.69-0.88)</td>
<td>126</td>
</tr>
<tr>
<td>Cardiomyopathies</td>
<td>8</td>
<td>0.21 (0.10-0.33)</td>
<td>35</td>
<td>0.25 (0.17-0.33)</td>
<td>40</td>
</tr>
<tr>
<td>Coronary artery disease</td>
<td>7</td>
<td>0.19 (0.07-0.30)</td>
<td>23</td>
<td>0.17 (0.12-0.22)</td>
<td>25</td>
</tr>
<tr>
<td>Cardiac conduction disease</td>
<td>3</td>
<td>0.08 (0-0.28)</td>
<td>8</td>
<td>0.06 (0-0.10)</td>
<td>12</td>
</tr>
<tr>
<td>Myocarditis</td>
<td>4</td>
<td>0.10 (0-0.34)</td>
<td>15</td>
<td>0.11 (0-0.16)</td>
<td>20</td>
</tr>
<tr>
<td>Congenital coronary anomalies</td>
<td>2</td>
<td>0.05 (0-0.17)</td>
<td>5</td>
<td>0.04 (0-0.06)</td>
<td>7</td>
</tr>
<tr>
<td>Mitral valve prolapse</td>
<td>2</td>
<td>0.05 (0-0.17)</td>
<td>9</td>
<td>0.06 (0-0.11)</td>
<td>8</td>
</tr>
<tr>
<td>Other†</td>
<td>3</td>
<td>0.08 (0-0.28)</td>
<td>15</td>
<td>0.11 (0-0.15)</td>
<td>14</td>
</tr>
</tbody>
</table>

Abbreviations: CI, confidence interval; RR, relative risk.
*Incidence rates are shown as events per year per 100,000 athletes aged 12 to 35 years. Number of events represent the actual number of events.
‡Includes myocardial bridge, aortic stenosis, aortic rupture, and pulmonary thromboembolism.
Trends in Sudden Cardiovascular Death in Young Competitive Athletes After Implementation of a Preparticipation Screening Program
Corrado et al. *JAMA* 2006; 296(13): 1593-1601

**Study Objective:** Determine the incidence of SCD in a population of high school athletes undergoing “screening” with standardized sports preparticipation history and physical evaluation (PPE)

**Study Design:** Retrospective, observational

**Population:** “unduplicated” athletes aged 12-19

**Time Frame:** 1993-2012, deaths by review of catastrophic insurance records

**Incidence of death determined:** \( \frac{\text{# deaths}}{\text{# unduplicated athlete-years}} \)

**Results:** 1,666,509 unduplicated athlete years

4 deaths during the study period: 2 cross-country, 1 basketball, 1 wrestling

0.24 deaths/100,000 athlete years

**Conclusion:** Low incidence of SCD, ECG screening not warranted as part of PPE
Maron et al. *Am J Card* 2009; 104: 276-280
Incidence of Sudden Cardiac Death in National Collegiate Athletic Association Athletes

Table 1. Incidence of SCD in NCAA Athletes According to Sex, Ethnicity, and Division, 2004–2008

<table>
<thead>
<tr>
<th></th>
<th>No. of Athlete-Years</th>
<th>No. of Deaths</th>
<th>Death Rate (per Year)</th>
</tr>
</thead>
<tbody>
<tr>
<td>NCAA athletes</td>
<td>1,969,663</td>
<td>45</td>
<td>1:43770</td>
</tr>
<tr>
<td>Sex</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Male</td>
<td>1,126,557</td>
<td>34</td>
<td>1:33134</td>
</tr>
<tr>
<td>Female</td>
<td>843,106</td>
<td>11</td>
<td>1:76646</td>
</tr>
<tr>
<td>Ethnicity</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Black</td>
<td>300,835</td>
<td>17</td>
<td>1:17696</td>
</tr>
<tr>
<td>White</td>
<td>1,583,635</td>
<td>27</td>
<td>1:58653</td>
</tr>
<tr>
<td>By division</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Division I</td>
<td>788,023</td>
<td>27</td>
<td>1:29186</td>
</tr>
<tr>
<td>Division II</td>
<td>424,572</td>
<td>10</td>
<td>1:42457</td>
</tr>
<tr>
<td>Division III</td>
<td>760,258</td>
<td>8</td>
<td>1:95032</td>
</tr>
</tbody>
</table>

SCD indicates sudden cardiac death; NCAA, National Collegiate Athletic Association.

*Circulation. 2011;123:1594-1600.*
## Incidence of Sudden Cardiac Death in National Collegiate Athletic Association Athletes

### Table 2. Incidence of NCAA SCD by Sport, 2004–2008

<table>
<thead>
<tr>
<th>Sport</th>
<th>Number of Deaths</th>
<th>Overall Incidence*</th>
<th>Incidence in Males</th>
<th>Incidence in Females</th>
<th>Incidence in African Americans</th>
<th>Incidence in Caucasians</th>
</tr>
</thead>
<tbody>
<tr>
<td>Basketball</td>
<td>14</td>
<td>1:11,394</td>
<td>1:6,993</td>
<td>1:37,799</td>
<td>1:5,743</td>
<td>1:21,824</td>
</tr>
<tr>
<td>Division I</td>
<td>9</td>
<td>1:5,451</td>
<td>1:3,126</td>
<td>1:23,901</td>
<td>1:5,284</td>
<td>1:6,135</td>
</tr>
<tr>
<td>Division II</td>
<td>3</td>
<td>1:12,631</td>
<td>1:11,330</td>
<td>1:15,232</td>
<td>1:9,503</td>
<td>1:20,822</td>
</tr>
<tr>
<td>Division III</td>
<td>2</td>
<td>1:24,681</td>
<td>1:13,646</td>
<td>†</td>
<td>1:6,952</td>
<td>†</td>
</tr>
<tr>
<td>Swimming</td>
<td>4</td>
<td>1:21,293</td>
<td>1:34,552</td>
<td>1:16,457</td>
<td>†</td>
<td>1:20,981</td>
</tr>
<tr>
<td>Lacrosse</td>
<td>3</td>
<td>1:23,357</td>
<td>1:19,770</td>
<td>1:30,531</td>
<td>†</td>
<td>1:23,357</td>
</tr>
<tr>
<td>Football</td>
<td>8</td>
<td>1:38,497</td>
<td>1:38,497</td>
<td>†</td>
<td>1:59,814</td>
<td>1:14,401</td>
</tr>
<tr>
<td>Cross-country</td>
<td>3</td>
<td>1:41,695</td>
<td>1:59,484</td>
<td>1:32,801</td>
<td>1:12,043</td>
<td>1:51,033</td>
</tr>
</tbody>
</table>

NCAA indicates National Collegiate Athletic Association; SCD, sudden cardiac death.

*SCD incidence is expressed as number of athletes per year.

†No deaths for incidence calculation.
## Sudden Cardiac Death in the Young

<table>
<thead>
<tr>
<th></th>
<th>Cardiac Deaths</th>
<th>Incidence (per 100,000 AY)</th>
<th>Relative Risk</th>
</tr>
</thead>
<tbody>
<tr>
<td>MSHSL 1993-2012</td>
<td>4</td>
<td>0.24</td>
<td>Reference</td>
</tr>
<tr>
<td>MSHSL 2003-2012</td>
<td>1</td>
<td>0.11</td>
<td>0.45</td>
</tr>
<tr>
<td>Italy 1979-81*</td>
<td>14</td>
<td>4.19 (1.78, 7.59)</td>
<td>17.5</td>
</tr>
<tr>
<td>Italy 1982-1992</td>
<td>29</td>
<td>2.35 (1.94, 2.75)</td>
<td>9.79</td>
</tr>
<tr>
<td>Italy 1993-2004</td>
<td>12</td>
<td>0.87 (0.46, 1.28)</td>
<td>3.6</td>
</tr>
<tr>
<td>Division 1 NCAA</td>
<td>27</td>
<td>3.45</td>
<td>14.3</td>
</tr>
<tr>
<td>Division 2 NCAA</td>
<td>10</td>
<td>2.38</td>
<td>9.9</td>
</tr>
<tr>
<td>Division 3 NCAA</td>
<td>8</td>
<td>1.05</td>
<td>4.4</td>
</tr>
</tbody>
</table>

* Before pre-participation screening

Corrado et al. *JAMA* 2006; 296(13): 1593-1601
Harmon et al. *Circulation* 2011; 123 (15): 1594-1600
<table>
<thead>
<tr>
<th>Age of Population Under Study</th>
<th>Identification of Sudden Cardiac Death</th>
<th>Which deaths included</th>
</tr>
</thead>
<tbody>
<tr>
<td>Minnesota Study</td>
<td>12-19</td>
<td>Review of catastrophic insurance records</td>
</tr>
<tr>
<td>Italian Study</td>
<td>12-35</td>
<td>Registry on Juvenile Sudden Death of the Veneto Region of Italy, review of regional newspapers</td>
</tr>
<tr>
<td>NCAA Study</td>
<td>17-24</td>
<td>NCAA database &amp; Parent Heart Watch database as well as NCAA catastrophic insurance claims</td>
</tr>
</tbody>
</table>
Sudden Cardiac Death in the Young

- No current evidence in US population that screening reduces incidence of SCDY
- What’s needed:
  - Prospective epidemiology and etiology of SCDY
  - Performance of screening methodology in a target population
  - Optimal management of asymptomatic heart disease uncovered by screening
  - Impact of ECG screening on the individual, family, community and society

NIH and CDC launch registry for sudden death in the young

“This registry will collect comprehensive, population-based information on sudden unexpected death in youths up to age 24 in the United States. It is a critical first step toward figuring out how to best prevent these tragedies.”

October 24, 2013, 9:00 AM EDT
Summary Points

• During the PPE, personal and family heart health history is key to determine need for in-depth cardiac evaluation

• Kids who faint should have an EKG

• Fainting or dizziness during exercise is a RED flag symptom

• Higher risk populations exist: in-depth screening (Division I basketball, professional)

• NIH/CDC registry should help us shape data-driven screening recommendations for the future