Sudden Cardiac Death in Young Athletes

Ivan Anderson, MD
RIHNVH Cardiology
Outline

• Background
• Specific Causes
Outline

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• Specific Causes
Background

- Rare
  - Incidence is ~ 1 per 100,000 person-years
  - One study (cited below) in a population of 1.23 million filtered down to 361 cases over 30 years
- Represents 75% of all fatalities during sports
- Survival < 15% by many studies

Circulation. 2012;126:1363-1372
Causes of death in the US population aged 1 to 21 years.
Outline

• Background
• Specific Causes
Outline

• Background
• Specific Causes
Causes of Sudden Death

1. HCM
2. Anomalous Coronary Artery
3. Myocarditis
4. ARVC

Other congenital HD (2%)
Ion channelopathies (3%)
Aortic rupture (2%)
Sarcoidosis (1%)
Dilated C-M (2%)
AS (3%)
CAD (3%)
Tunneled LAD (3%)
MVP (4%)

Indeterminate LVH - possible HCM (8%)

Other (3%)
Normal heart (3%)

Circulation. 2007;115:1643-1655
Causes of Sudden Death in Athletes

1. Hypertrophic cardiomyopathy
2. Anomalous coronary artery
3. Myocarditis
4. Arrhythmogenic right ventricular cardiomyopathy
Causes of Sudden Death in Athletes

1. Hypertrophic cardiomyopathy
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Hypertrophic Cardiomyopathy
Normal Heart

RV VS LV
Disarrayed Myocytes in Hypertrophic Cardiomyopathy

Normal Myocardium

Myocyte disarray and fibrosis with hypertrophic cardiomyopathy
LA = left atrium, LV = left ventricle, RV = right ventricle, IVS = intraventricular septum, AoV = aortic valve, MV = mitral valve
Athlete’s Heart versus HCM

- Symmetric hypertrophy
- Hypertrophy is rarely greater than 17 mm.
- LV cavity dimension is increased, whereas it is decreased in HCM.
- Diastolic function is normal (Ea >7 cm/sec).
- Tissue Doppler velocities and strain values are normal.
Causes of Death in Hypertrophic Cardiomyopathy
Ventricular Tachycardia
Markers for Sudden Death in Hypertrophic Cardiomyopathy

2° prevention
- Cardiac arrest. Sustained VT

1° prevention
- Familial sudden death
- Unexplained syncope
- Multiple-repetitive NSVT (Holter)
- Abnormal exercise BP response
- Massive LVH

Potential arbitrators
- End-stage phase
- LV apical aneurysm
- Marked LV outflow obstruction (rest)
- Extensive delayed enhancement
- Alcohol septal ablation (?)
- Modifiable
  - Intense competitive sports
  - CAD

% PATIENTS WITH SD

<table>
<thead>
<tr>
<th>MAX. LV WALL THICKNESS (MM)</th>
<th>0</th>
<th>2</th>
<th>4</th>
<th>6</th>
<th>8</th>
<th>10</th>
<th>12</th>
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ICD
Hank Gathers
Causes of Sudden Death in Athletes

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Causes of Sudden Death in Athletes

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R = Right, L = Left,
A = Anterior, P = Posterior

- Aorta
- Left coronary artery
- Right coronary artery
- Pulmonary artery

Normal anatomy
Anomalous Coronary Artery
“Pistol” Pete Maravich
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Myocarditis
<table>
<thead>
<tr>
<th>Virus Type</th>
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<tbody>
<tr>
<td>Enterovirus</td>
<td>Varicella</td>
</tr>
<tr>
<td>Coxsackie A</td>
<td>Mumps</td>
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<tr>
<td>Coxsackie B</td>
<td>Measles</td>
</tr>
<tr>
<td>Echovirus</td>
<td>Rabies</td>
</tr>
<tr>
<td>Poliovirus</td>
<td>Hepatitis B, C</td>
</tr>
<tr>
<td>Adenovirus</td>
<td>Rubella</td>
</tr>
<tr>
<td>Parvovirus B19</td>
<td>Rubeola</td>
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<tr>
<td>Cytomegalovirus</td>
<td>Respiratory syncytial virus</td>
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<tr>
<td>Herpesvirus</td>
<td>Human immunodeficiency virus</td>
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<tr>
<td>Influenza A and H1N1</td>
<td>Epstein-Barr virus</td>
</tr>
<tr>
<td>Human herpesvirus 6</td>
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</table>
Diagnosis: Dallas Criteria

1. Lymphocyte infiltration
2. Myocyte necrosis
Reggie Lewis
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Arrhythmogenic Right Ventricular Cardiomyopathy (ARVC)
The Desmosome and It’s Function (Disrupted in ARVC)

Circulation. 2010;121:1533-1541
Fibrofatty Myocardial Replacement in ARVC
ECG Findings in ARVC

Epsilon wave
Antonio Puerta
Conclusion

• Sudden cardiac death in young athletes is a rare, but tragic phenomenon that results from a number of pathologies, most commonly hypertrophic cardiomyopathy.
Questions/Comments?
Having dumped the bag of ashes on the table, Stew hid behind the door and waited for the X-ray technician’s reaction.
<table>
<thead>
<tr>
<th>Family History</th>
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<tbody>
<tr>
<td>1 Premature sudden cardiac death</td>
</tr>
<tr>
<td>2 Heart disease in surviving relatives &lt; 50 yr</td>
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<table>
<thead>
<tr>
<th>Personal History</th>
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<tbody>
<tr>
<td>3 Heart murmur</td>
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<tr>
<td>4 Systemic hypertension</td>
</tr>
<tr>
<td>5 Fatigue</td>
</tr>
<tr>
<td>6 Syncope, near-syncope</td>
</tr>
<tr>
<td>7 Excessive, unexplained exertional dyspnea</td>
</tr>
<tr>
<td>8 Exertional chest pain</td>
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<thead>
<tr>
<th>Physical Examination</th>
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<tbody>
<tr>
<td>9 Heart murmur (supine/standing)</td>
</tr>
<tr>
<td>10 Femoral arterial pulses (to exclude coarctation of aorta)</td>
</tr>
<tr>
<td>11 Stigmata of Marfan syndrome</td>
</tr>
<tr>
<td>12 Brachial blood pressure measurement (sitting)</td>
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</table>
Sodium Channel and Heart Conditions

- Atrial fibrillation (AF)
- Brugada syndrome (BrS)
- BrS or LQTS
- Cardiac conduction defect (CCD)
- Dilated cardiomyopathy (DCM)
- Drug-induced torsades de pointes (drug-TdP)
- Long QT syndrome (LQTS)
- Mixed phenotype (BrS with SSS and/or CCD)
- Rare and common missense variants in health
- Sick sinus syndrome (SSS)
- Sudden infant death syndrome (SIDS)
Long QT Syndrome

- **KCNQ1 (LQT1)**
  - Swimming
  - Exertion/emotion
  - 35%

- **KCNH2 (LQT2)**
  - Auditory triggers
  - Postpartum period
  - 30%

- **SCN5A (LQT3)**
  - Sleep
  - Rest
  - 10%
Brugada Syndrome