Sudden Cardiac Death in the Pediatric Population

Maggie Gray, MD
Department of Pediatric Cardiology
Palmetto Health/University of South Carolina School of Medicine
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Objectives

• Describe the incidence of sudden cardiac death in the pediatric population
• Review the most common cardiac etiologies for sudden cardiac death
• Discuss the risk of sudden cardiac death in the young athlete
• Discuss the risk of sudden cardiac death in patients receiving stimulant medications
• Describe evidence-based primary prevention modalities
• Identify the role of the pediatrician in implementing secondary prevention measures
Incidence and Etiology
Ronald Rouse, 18, dies during game

Associated Press
Oct 6, 2012

Lewis Simpkins
2001-2016

Heart condition, heat combined to kill River Bluff football player, coroner says

Autopsy: Adams died of cardiac arrest

Jan 18, 2015

Surviving sudden cardiac arrest: One teen athlete's story
Background

• Abrupt and unexpected cardiac arrest
• Within 1 hour from onset of symptoms
• Devastating for family and community

Prodrome → Onset → Arrest → Death or Resuscitation

Incidence

• 0.5 – 20 per 100,000 pt years\(^1\)
• Up to 2x more frequent in athletes\(^2\)
• 2-3x more frequent in males vs females\(^3\)
• Most frequent in African American males
• Most frequent during basketball, swimming, football, cross country/track\(^4\)

Etiology

- Hypertrophic cardiomyopathy¹-³
- Anomalous coronary artery
- Myocarditis
- Arrhythmogenic right ventricular cardiomyopathy
- Channelopathy
  - Long QT syndrome
  - Brugada syndrome
- Catecholaminergic polymorphic ventricular tachycardia
- Aortic dissection
- Aortic stenosis

SCD ≤2 years
- CHD
- Myocarditis
- Arrhythmia
- MVP

SCD 3-13 years
- CHD
- LQTS
- MVP
- HCM
- Arrhythmia

SCD 14-24 years
- CHD
- LQTS
- DCM
- HCM
- Myocarditis
- Arrhythmia

SCD Athletes
- HCM
- Coronary Anomaly
- ARVC
- Other
Hypertrophic Cardiomyopathy

- 0.2-0.5% of general population\textsuperscript{1,2}
- AD inheritance
- Pathophysiology
  - Thickened, nondilated LV
  - Disorganized myocytes $\rightarrow$ arrhythmia and diastolic dysfunction
  - 80% with abnormal coronary architecture $\rightarrow$ ischemia and scarring $\rightarrow$ more arrhythmia and diastolic dysfunction
  - Dynamic LVOTO
- High Risk
  - Prior arrest or VT/VF
  - Family hx of SCD due to HCM
  - Syncope
  - LV wall $\geq$ 30 mm

Hypertrophic Cardiomyopathy

• Clinical features
  • Exam
    • Prominent apical impulse
    • Murmur – LLSB/apex medium pitch systolic murmur
  • ECG
    • Abnormal in up to 90%
    • LVH, TWI, LAE, Q waves, LAD
  • Echo
    • LV wall ≥ 15 mm anywhere
    • Systolic anterior motion of mitral valve
    • LVOTO

• Management
  • Beta blockers, myectomy, ICD
  • Sports restriction (class IA)

Pathologic LVH vs Physiologic LVH

“Gray zone” of LV wall thickness

HCM

Athlete’s heart

+ Focal LVH pattern
+ LV cavity <45 mm
+ LV cavity >55 mm
+ LA enlargement
+ Bizarre ECG patterns
+ Abnormal LV filling
+ Female gender
+ Family history of HCM
+ Thickness with deconditioning
- VO₂ >110 percent
+ Late gadolinium enhancement
+ Pathogenic sarcomere mutation

Hugh D. Allen, R.E.S., Daniel J. Penny, Timothy F. Feltes, Frank Cetta, Moss and Adams' Heart Disease in Infants, Children, and Adolescents.
Coronary Artery Anomalies

• Abnormal origin of the coronary artery\(^1,2\) (0.2%)
  • MC is circumflex from right sinus
    • No clinical significance
  • LMCA from right sinus of Valsalva (1-3%)
    • Multiple possible courses
    • Interarterial associated with SCD
  • RCA from left sinus of Valsalva (30%)
    • Risk of SCD not well established
• Single coronary artery origin (5-20%)
  • Risk of SCD not well established

Coronary Artery Abnormalities

• Clinical features\(^1\)
  - Exam
    - Usually normal
  - ECG
    - Usually normal
  - Echo
    - Show coronary arteries by 2D and color
    - May require CTA confirmation

• Management\(^2\)
  - Surgical intervention
  - Sports restriction
    - L from R – only class IA
    - R from L – sports ok if asymptomatic and EST negative
    - Return 3 months after surgery if asymptomatic and EST negative

Myocarditis

- Incidence between 4-5%
- Signs/symptoms
  - Nonspecific
  - Viral prodrome
  - Malaise, fatigue, pallor, diaphoresis, dyspnea
- Chest x-ray
  - Cardiomegaly
  - Pulmonary vascular congestion
- ECG
  - Sinus tachycardia
  - T wave inversion
  - Q waves
  - PVCs, VT, SVT, AV block

Myocarditis

• Echo
  • Dilated LV
  • Decreased function
  • Pericardial effusion

• Biopsy (Dallas criteria)
  • Inflammatory infiltrate of the myocardium
  • Necrosis/degeneration of myocytes

• Labs
  • +viral pcr
  • Elevated BNP and cardiac enzymes
Myocarditis

• Management
  • Supportive care
    • Medication
    • Mechanical ventilation
    • ECMO
  • IVIG, steroids debated
  • Heart failure management
  • Transplantation

• Sports Restriction
  • 3-6 months
  • No competitive sports if symptomatic

Arrhythmogenic Right Ventricular Cardiomyopathy

- Incidence 1 in 5000\(^1,2\)
- AD inheritance
- Fibrofatty infiltration of the right ventricle
  - Thinning and dysfunction
  - Arrhythmia nidus
- Palpitations, syncope most common presenting symptoms
- ECG
  - T wave inversion in V1-V3
  - Epsilon wave
- Echo
  - RV dysfunction
  - RV dilation and thinning

Arrhythmogenic Right Ventricular Cardiomyopathy

- Management
  - ICD
  - Antiarrhythmics
- Sports restriction
  - Consider class IA participation

Long QT Syndrome

- Incidence 1 in 5000
- AD with variable penetrance
  - Mutation in sodium or potassium channels
  - Leads to delayed repolarization
- Associated with deafness (Jervell Lange-Nielsen syndrome)
- Grey area with QTc
- Genetic testing important

Table 1: Suggested Bazett-Corrected QTc Values for Diagnosing QT Prolongation

<table>
<thead>
<tr>
<th>Rating</th>
<th>1-15 yrs</th>
<th>Adult Male</th>
<th>Adult Female</th>
</tr>
</thead>
<tbody>
<tr>
<td>Normal</td>
<td>&lt;440</td>
<td>&lt;430</td>
<td>&lt;450</td>
</tr>
<tr>
<td>Borderline</td>
<td>440-460</td>
<td>430-450</td>
<td>450-470</td>
</tr>
<tr>
<td>Prolonged</td>
<td>&gt;460</td>
<td>&gt;450</td>
<td>&gt;470</td>
</tr>
</tbody>
</table>

Table 2: Diagnostic Criteria for LQTS

<table>
<thead>
<tr>
<th>Electrocardiographic‡</th>
<th>Finding</th>
<th>Score</th>
</tr>
</thead>
<tbody>
<tr>
<td>Corrected QT interval, ms</td>
<td>≥480</td>
<td>3</td>
</tr>
<tr>
<td></td>
<td>460-470</td>
<td>2</td>
</tr>
<tr>
<td></td>
<td>450 (in males)</td>
<td>2</td>
</tr>
<tr>
<td></td>
<td>Torsades de pointes‡</td>
<td>1</td>
</tr>
<tr>
<td></td>
<td>T-wave alternans</td>
<td>1</td>
</tr>
<tr>
<td></td>
<td>Notched T-wave in 3 leads</td>
<td>1</td>
</tr>
<tr>
<td></td>
<td>Low heart rate for age‡</td>
<td>0.5</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Clinical history</th>
<th>Finding</th>
<th>Score</th>
</tr>
</thead>
<tbody>
<tr>
<td>Syncope‡</td>
<td>With stress</td>
<td>2</td>
</tr>
<tr>
<td></td>
<td>Without stress</td>
<td>1</td>
</tr>
<tr>
<td>Congenital deafness</td>
<td></td>
<td>0.5</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Family history</th>
<th>Finding</th>
<th>Score</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Family members with definite LQTS</td>
<td>1</td>
</tr>
<tr>
<td></td>
<td>Unexplained SCD in immediate family members &lt;30 yrs old</td>
<td>0.5</td>
</tr>
</tbody>
</table>

Long QT Syndrome

• Management¹
  • Antiarrhythmic (beta blocker**)  
  • ICD  
  • Avoid QT prolonging medications (sads.org)

• Sports restriction²
  • No competitive sports  
  • Gene +, pheno – ok after evaluation and avoidance of triggers (AED)  
  • Consider return if asymptomatic after treatment  
  • Participation ok with ICD if asymptomatic

Brugada Syndrome

• First described in 1992\textsuperscript{1}
• 5 out of 10,000 (kids and adults)
• Characteristic ECG findings
  • Coved ST elevation
  • +/- T wave inversion
  • Right precordial leads
• AD inheritance (SCN5A mutation)
• Management\textsuperscript{2}
  • ICD
  • Sports Restriction

Primary Prevention
Early identification of children at risk for SCD

Apply appropriate sports limitations

Pursue available medical therapy

Genetic testing for family members

All while limiting adverse effects of screening and treatment

Current Practices

• AHA/AAP recommend\(^1\):
  • Comprehensive PPE prior to participating in competitive sports
  • Not universal ECG screening
  • Cardiology referral for any red flags\(^3\)

• Other recommendations\(^2\)
  • ESC PPE + ECG
  • NCAA PPE + ECG
  • NBA PPE, ECG, echo
  • NFL PPE, EKG +/- echo

• SC State Law mandates PPE prior to sports enrollment
  • Can be done by physician, NP, or PA
  • Type of form used not specified
  • Does not YET include ECG screening

14 Point Screening Checklist

• Personal History
  • Exertional chest pain/tightness
  • Palpitations
  • Syncope or dizziness*
  • Exertional fatigue/dyspnea*
  • Prior history of a heart murmur
  • Prior history of ↑BP
  • Prior restriction from sports
  • Prior cardiac testing

14 Point Screening Checklist

• Family History
  • Death prior to age 50 due to cardiovascular causes
  • Disability from heart disease prior to age 50
  • HCM, DCM, Long QT syndrome (or other channelopathy), Marfan syndrome

14 Point Screening Checklist

• Physical Exam
  • Heart murmur
  • Femoral pulses
  • Stigmata of Marfan Syndrome
  • Brachial artery pressure (seated)

<table>
<thead>
<tr>
<th>HCM Murmur</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Louder</td>
<td>Softer</td>
</tr>
<tr>
<td>Standing</td>
<td>Supine</td>
</tr>
<tr>
<td>Valsalva</td>
<td>Squatting</td>
</tr>
<tr>
<td>Nitroglycerin</td>
<td>Handgrip</td>
</tr>
</tbody>
</table>

## AAP PPE Form

### Heart Health Questions About You

<table>
<thead>
<tr>
<th>Question</th>
<th>Yes</th>
<th>No</th>
</tr>
</thead>
<tbody>
<tr>
<td>Have you ever passed out or nearly passed out DURINGS or AFTER exercise?</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Have you ever had discomfort, pain, tightness, or pressure in your chest during exercise?</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Does your heart ever race or skip beats (irregular beats) during exercise?</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Has a doctor ever told you that you have any heart problems? If so, check all that apply:</td>
<td></td>
<td></td>
</tr>
<tr>
<td>High blood pressure</td>
<td>A heart murmur</td>
<td></td>
</tr>
<tr>
<td>High cholesterol</td>
<td>A heart infection</td>
<td></td>
</tr>
<tr>
<td>Kawasaki disease</td>
<td>Other:</td>
<td></td>
</tr>
<tr>
<td>Has a doctor ever ordered a test for your heart? (For example, ECG/EKG, echocardiogram)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Do you get lightheaded or feel more short of breath than expected during exercise?</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Have you ever had an unexplained seizure?</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Do you get more tired or short of breath more quickly than your friends during exercise?</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

### Heart Health Questions About Your Family

<table>
<thead>
<tr>
<th>Question</th>
<th>Yes</th>
<th>No</th>
</tr>
</thead>
<tbody>
<tr>
<td>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)?</td>
<td></td>
<td></td>
</tr>
<tr>
<td>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?</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Does anyone in your family have a heart problem, pacemaker, or implanted defibrillator?</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Has anyone in your family had unexplained fainting, unexplained seizures, or near drowning?</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

### Examination

<table>
<thead>
<tr>
<th>Measurement</th>
<th>Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Height</td>
<td></td>
</tr>
<tr>
<td>Weight</td>
<td></td>
</tr>
<tr>
<td>BP</td>
<td></td>
</tr>
<tr>
<td>Pulse</td>
<td></td>
</tr>
<tr>
<td>Vision</td>
<td>R 20/</td>
</tr>
</tbody>
</table>

### Medical

- **Appearance**
  - Marfan stigmata (kyphoscoliosis, high-arched palate, pectus excavatum, arachnodactyly, arm span > height, hyperflexity, myopia, MVP, aortic insufficiency)
- **Eyes/ears/nose/throat**
  - Pupils equal
  - Hearing
- **Lymph nodes**
- **Heart**
  - Murmurs (auscultation standing, supine, +/- Valsalva)
- **Pulses**
  - Simultaneous femoral and radial pulses
- **Lungs**
- **Abdomen**
- **Genitourinary (males only)**
- **Skin**
  - HSW, lesions suggestive of MRSA, linea corporis
- **Neurologic**
USA
PPE only

Italy
PPE + ECG

Israel
PPE + ECG

Japan
PPE + ECG for 1st, 7th, 10th graders

Denmark
No universal screening

Maron, B.J., et al., Eligibility and Disqualification Recommendations for Competitive Athletes With Cardiovascular Abnormalities: Preamble, Principles, and General Considerations: A Scientific Statement From the American Heart Association and American College of Cardiology. Circulation.
Universal ECG?

• Estimated at $70-204,000 per quality year life added$^{1,2}$
  • Could avert up to 130 SCDs per million children
  • $900,000 per averted SCD

• Positive predictive value impacted by incidence$^3$

• Pilot studies in PA, IL, TX, NV, NC$^4$
  • 2-7% required additional testing
  • 1-3% with “potentially serious conditions”

• PPE alone has not reduced mortality

Universal ECG

• ECG abnormalities frequent among athletes
• Common training related changes
  • Increased QRS voltage
  • Early repolarization in V3-V6
  • Increased vagal tone
  • Axis shift to -30° to 115°
  • First degree AV block
  • Incomplete right bundle branch block

Present in 50% of nonathletes and almost 90% of athletes.

Universal ECG?

• Concerning findings
  • Q waves
  • QRS >120 ms
  • RVH
  • RAA or LAA
  • T wave inversion
  • ST depression
  • ≥2 premature ventricular contractions
  • QTc ≥440 ms in men and ≥460 ms in women
  • QTc <380 ms
  • Pre-excitation (PR <120 ms)
  • Type 1 Brugada (RBBB morphology with 2 mm ST elevation)

Present in 20% of nonathletes and 30% of athletes.

Evaluation of a High Risk Patient

- Referral to pediatric cardiology
- Treatment may include\textsuperscript{1,2}:
  - Sports restriction (2015 AHA/ACC guidelines)
  - Medication administration
  - Medication avoidance
  - ICD implantation
  - Emergency preparedness
- Genetic Screening for family members
- Management of emotional distress

Genetic Screening

• 49% of SCD due to potentially heritable condition\(^1\)
• Allows further diagnosis when heart is structurally normal at autopsy\(^2\)
• SCD is 1\(^{st}\) manifestation in up to 50%
• Molecular autopsy and screening of first degree family members should be standard of care\(^3\)
• Allows for better primary prevention

3. Tester, D.J. and M.J. Ackerman, *The molecular autopsy: should the evaluation continue after the funeral?* Pediatr Cardiol.
ADHD and SCD

• Stimulant medications\(^1\)
  • 70% efficacy
  • Generally accepted as safe\(^2\)
  • Mild increase in HR and BP (1-5bpm and 3-4 mmHG)
  • No significant change in QTc
• “Use caution in children with known structural heart disease, cardiomyopathy, or arrhythmia”
• AAP recommendations\(^3,4\)
  • Thorough patient and family history prior to Rx
  • Monitor heart rate and BP response
  • Increase research into risk factors for SCD

Pediatric patient under consideration for or currently being treated with stimulant medication

Known cardiac disease?

No

Patient history, family history or physical exam suggestive of cardiac disease?

No

Treatment with stimulants does not require additional cardiac testing

Yes

Further evaluation – if indicated, obtain input from a pediatric cardiologist

No

After initiating treatment, does history or exam change to suggest possible cardiac disease?

Yes

Figure 1. Cardiac evaluation of children and adolescents receiving or being considered for stimulant medications (reproduced with permission. Copyright © 2008 American Academy of Pediatrics)
ADHD and SCD

- No increased risk of SCD compared to general population\(^1,2\)
- Universal ECG not recommended
- Established increased risk of poor outcomes if ADHD not adequately treated\(^3,4\)
  - Injuries
  - Academic underachievement
  - Peer rejection
  - Substance abuse
  - Automobile accidents
  - Antisocial behavior

3. Berger, S., *Cardiac evaluation of patients receiving pharmacotherapy for attention deficit hyperactivity disorder*. UpToDate.
Secondary Prevention
Widespread CPR and AED training

Access to AED

Prompt recognition of signs/symptoms

Engage emergency response plan

Berger, S., A Reflection on Pediatric Sudden Cardiac Arrest. American College of Cardiology.
Emergency Response

• Survival from pediatric OHCA is abysmal (<10%)\(^1\)

• Survival increased when\(^2\):
  • Witnessed arrest
  • Initial rhythm VT/VF
  • Bystander CPR initiated
  • Public location

• Significant reduction in mortality after compression only CPR introduced

Emergency Response

- 83% of US high schools have a plan\(^1\)
- 40% practice and review annually
- SC bill passed in 2016 mandates CPR/AED training for high school students ("Ronald Rouse Law")
  - Requires training in CPR (including compression only) and AED use
  - Begins 2017-2018 school year
- SC bill passed in 2008 requiring an AED on school grounds
  - Does not provide funding
  - Does not require at sporting events
  - Does not mandate emergency response plan

National Registry

• No national mandatory registry
• Better understanding of SCD events may improve prevention
• Attempts to quantify and study SCD rely on piecemeal data sets
• National Center for the Review and Prevention of Child Deaths\(^1\)
  • Reviews deaths age 0-21 years
  • 35 states involved (SC included)
  • State specific review teams
  • Traditionally aimed at injuries, abuse, neglect, homicide

In Summary – The AAP Stance

• Standardize the PPE
  • Perform thorough CV family history
• Refer to cardiology for any red flags
• Autopsy (including genetics) for SCD patients
• Comprehensive cardiology evaluation (including genetics) for SCD survivors
• Recognize signs and symptoms of SCD
• Promote education for CPR and AED
• Promote school emergency response plan
• Support nationwide registry
• Encourage EBM review of screening modalities
Works Cited


35. Berger, S., Cardiac evaluation of patients receiving pharmacotherapy for attention deficit hyperactivity disorder. UpToDate, 2016.

