Syncope and Sudden Death: Back to Titanic

Ric Samson, MD
Childrens Heart Center Nevada
The Titanic
Syncope vs. Sudden Death
The Dilemma
Syncope vs. Sudden Death

syncope

sudden death
Definitions

• **syncope**: a brief sudden loss of consciousness and muscle tone secondary to ischemia, or inadequate substrate delivery to the brain

• **sudden death**: an unexpected death from natural causes which can occur instantaneously or within 24 hours of the patient’s initial symptoms
Incidence Syncope

- Adults
  - 1 - 6% of hospital admissions
  - 3% of emergency room visits
- Children
  - 15% experience a syncopal episode during childhood
Incidence
Sudden Death

• Adults – seemingly healthy
  – competitive athletes: 4 per million per year
  – recreational athletes <30: 10-25 per million per year
  – US Air Force recruits - 1 per 3 million hours of exercise (1 per 735,000 per year)

• Children
  – Mayo clinic: ages 1-22 in Olmstead County, MN:
    » 1.3 per 100,000 per year
  – Gajewski, Annals Ped Cardiol 2010
    » 0.6-6.2 per 100,000

• 30% of all non-trauma deaths in people < 35
• <20% have cardiac symptoms prior to death
Headlines - Sudden Death

Hank Gathers

Wes Leonard: Michigan High School Basketball Star Dies After Game-Winning Shot
Patient Presentation #1

- 12 year old boy presents with transient loss of consciousness following blood draw
- episode preceded by nausea, feeling of warmth
- no previous cardiac symptoms
- past medical history unremarkable
- family history negative for cardiac disease
- physical exam normal
Autonomic Syncope
Vasovagal

• stimulus induced
  – blood, pain, objectionable sights
  – nausea, anxiety, flushing, warmth

• others:
  – tussive
  – swallowing
  – defecation
  – breath holding spells
  – carotid hypersensitivity
Patient Presentation #2

• 16 yo female presents with syncopal episode while singing in the church choir
• 3-4 similar episodes in the past year
• typically occur in an upright position, e.g., in choir or while standing in line
• no palpitations or chest pain
• not associated with exercise
• physical exam normal
Neurocardiogenic syncope

- associated with upright posture
- exaggerated manifestation of a normal physiologic reflex (Bezold-Jarisch reflex)

\[\text{decreased venous return} \rightarrow \text{decreased CO}\]

\[\text{adrenergic stimulation} \rightarrow \text{stimulation of cardiac mechanoreceptors}\]

\[\text{catecholamine release} \rightarrow \text{brainstem and vagal stimulation}\]

\[\text{adrenergic withdrawal} \rightarrow \text{vasodepressor response}\]

\[\text{cardioinhibitory response} \rightarrow \text{SYNCOPE}\]
Pathophysiology of neurocardiogenic syncope

- Supine to upright
  - Venous pooling in legs > decreased venous return > decreased cardiac output > lower BP > Compensatory release of intrinsic catecholamines
  - Catechols > increased vasoconstriction, HR, contractility
  - Increased contractility sensed by cardiac β mechano-receptors elicits paradoxical reflex
  - Unmyelinated C fibers within myocardium
    - WHOA! Heart is beating too hard > Brainstem vagal stimulus
    - Bradycardia (cardioinhibitory response)
    - Vasodilation (vasodepressor response)
  - SYNCOPE
  - Return to supine increases venous return, reverses process
Neurocardiogenic Syncope

• not life-threatening
• diagnosis of exclusion; must rule out life-threatening conditions first
• confirmed by tilt-test
• neurocardiogenic syncope may be associated with the catecholamine release from physical exertion
• vasodepressor syncope can be seen after exertion due to continued vasodilation and decreased cardiac output
• cardioinhibitory response can be associated with profound asystole
Neurocardiogenic Syncope
Tilt Table Test

- Tilt table test -
  - high sensitivity
  - low specificity - many false positives
    » as high as 30% positive in asymptomatic people
  - Confirms a diagnosis of exclusion
  - Can provoke profound asystole
Cardioinhibitory Syncope
Neurocardiogenic syncope: Treatment

• Increased fluid and salt intake
• $\beta$ blockers to block increase in contractility
  – Atenolol, metoprolol, propranolol
• Mineralocorticoids to cause fluid retention
  – Fludrocortisone
• Vasoconstrictors to block vasodilation
  – Midodrine, pseudoephedrine
• Anti-cholinergics to block vagal response
  – Atropine, glycopyrronium (Robinul), disopyramide
Patient Presentation #3

• 16 year old female - previously healthy competitive swimmer

• while swimming in event, became syncopal, sinking to bottom of pool

• required CPR, brought to ER

• PMH negative

• FH history negative for syncope, sudden death

• Physical exam normal
Causes of Sudden Cardiac Death in Children

<table>
<thead>
<tr>
<th>Causes</th>
<th>Relative incidence (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hypertrophic cardiomyopathy</td>
<td>36</td>
</tr>
<tr>
<td>Increased cardiac mass</td>
<td>10</td>
</tr>
<tr>
<td>Coronary artery anomalies</td>
<td>24</td>
</tr>
<tr>
<td>Marfan’s syndrome</td>
<td>6</td>
</tr>
<tr>
<td>Congenital heart disease</td>
<td>5</td>
</tr>
<tr>
<td>Myocarditis</td>
<td>3</td>
</tr>
<tr>
<td>Dilated cardiomyopathy</td>
<td>3</td>
</tr>
<tr>
<td>Arrhythmogenic right ventricular dysplasia</td>
<td>3</td>
</tr>
<tr>
<td>Ischemic heart disease</td>
<td>2</td>
</tr>
<tr>
<td>Commotio cordis</td>
<td>&lt;1</td>
</tr>
</tbody>
</table>

Table 1: Causes of sudden death [8]
Coronary Artery Anomalies

- Most often an autopsy diagnosis
- Not always easily identified by echo
- Confirmed by cardiac CT and/or catheterization
Marfan’s Syndrome

- characteristic body habitus
- skeletal, cardiac, ocular involvement
- progressive aortic root dilation and risk of dissection
- autosomal dominant: + fam history
- intrinsic defect in connective tissue
- genetic defect traced to the fibrillin gene located on chromosome 15

Flo Hyman
Other causes

• Myocarditis – inflammation of myocardium related to a viral infection
  – Often reversible if given IVIG
• Dilated cardiomyopathy – enlarged heart with poor ventricular function
  – Some forms associated with genetic mutations
• Commotio cordis – impact striking chest over precordium at a precise instant during cardiac cycle (vulnerable period) and inducing VF
  – “very bad luck”
Syncope/Sudden Death

CHD Causes

• “non-life threatening” arrhythmias can become significant in patients with CHD
  – SVT in D-TGA (Mustard/Senning)
  – SVT in single ventricle (Fontan)
  – Bradycardia in CHD patients
    » complete heart block
    » sick sinus syndrome
Syncope/Sudden Death: CHD Causes

• obstruction to flow
  – aortic stenosis, pulmonary stenosis
    » loud murmur
  – pulmonary hypertension
    » loud P2 component of S2

• Tetralogy of Fallot
  » hypercyanotic spells
  » s/p repair - ventricular tachycardia

• Primary arrhythmias
Primary arrhythmias (<1%)

- Intrinsic abnormalities of cardiac conduction
  - Arrhythmogenic right ventricular dysplasia
  - Long QT syndrome
  - Andersen–Tawil syndrome
  - Short QT syndrome
  - Brugada syndrome
  - Catecholaminergic polymorphic ventricular tachycardia
  - Wolff–Parkinson–White syndrome

- 12-lead ECG is usually diagnostic in these conditions
  - CPVT is the exception; requires exercise testing to elicit
ECG “Markers of Sudden Death”

- **Hypertrophic cardiomyopathy**
  - LVH, deep Q waves in III, V6

- **Long QT syndrome – prolonged QTc interval**
  - Bazett’s formula: \( QTc = \frac{QT}{\sqrt{\text{preceding R-R interval}}} \)

- **Short QT syndrome – QTc < 350 ms**

- **Wolff-Parkinson-White syndrome**
  - Delta wave, “ventricular pre-excitation

- **Brugada syndrome**
  - RsR’ in V1, V2 with ST segment elevation

- **Arrhythmogenic right ventricular dysplasia/cardiomyopathy**
  - Epsilon wave
Short QT Syndrome

- QTc interval <350 ms
- Intrinsic abnormality of potassium channels
- Very rare, very lethal condition
Brugada syndrome

RsR’ in V1 and V2 with ST segment elevation
WPW - ECG

“Short PR interval with Bundle Branch Block”
Wolff-Parkinson-White Syndrome
Delta Wave

SA node
AV node
bypass tract
Wolff-Parkinson-White Syndrome

- < 1% risk of sudden death
- Of those, 20% present with sudden death as first manifestation
WPW: A-fib leading to V-fib
Arrhythmogenic right ventricular cardiomyopathy

Epsilon wave – slurred terminal portion of QRS
Long QT Syndrome

- Torsade de Pointes ventricular tachycardia
- corrected QT interval > 0.45

\[
\text{QT}_c = \frac{\text{QT interval (sec)}}{\sqrt{\text{preceding R-R interval (sec)}}}
\]
Catecholaminergic polymorphic ventricular tachycardia

- Presents with syncope associated with exercise or emotional stress
- Thus far, isolated to genetic mutations encoding for proteins which handle calcium within cardiac cells
- PE, echo, ECG and MRI are normal
- Diagnosed by exercise treadmill testing showing PVCs of at least 2 morphologies
- Genetic testing confirms about 50% of cases
# Syncope Evaluation:

history, physical exam, ECG, CXR, and labs

---

<table>
<thead>
<tr>
<th>Causes</th>
<th>Relative incidence (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hypertrophic cardiomyopathy</td>
<td>36</td>
</tr>
<tr>
<td>Increased cardiac mass</td>
<td>10</td>
</tr>
<tr>
<td>Coronary artery anomalies</td>
<td>24</td>
</tr>
<tr>
<td>Marfan’s syndrome</td>
<td>6</td>
</tr>
<tr>
<td>Congenital heart disease</td>
<td>5</td>
</tr>
<tr>
<td>Myocarditis</td>
<td>3</td>
</tr>
<tr>
<td>Dilated cardiomyopathy</td>
<td>3</td>
</tr>
<tr>
<td>Arrhythmogenic right ventricular dysplasia</td>
<td>3</td>
</tr>
<tr>
<td>Ischemic heart disease</td>
<td>2</td>
</tr>
<tr>
<td>Commotio cordis</td>
<td>&lt;1</td>
</tr>
</tbody>
</table>

---

*Catecholaminergic Polymorphic VT*
### Syncope vs. Sudden Death Evaluation - History

- **stimulus, event or body position related**
- **exertion related**
- **associated symptoms**
  - nausea, flushing, diaphoresis
  - dyspnea, palpitations, chest pain with exercise
- **recurrent or infrequent**
  - multiple episodes less likely to be serious
- **associated with injuries**
- **hydration status/environmental**
Syncope vs. Sudden Death
Evaluation - Physical Exam

• Vital Signs
  – orthostatic BPs

• Cardiac Exam
  – S2 physiologically split, P2 not loud
  – murmur
    » aortic stenosis - loud SEM USB
    » hypertrophic cardiomyopathy - SEM louder when upright

• Neurologic Exam
  – rule out seizures
Sudden Death vs. Syncope
Diagnostic Tests

- ECG - highly sensitive, moderately specific
- 24-hour Holter, event recorder - document heart rhythm at the time of symptoms
- Echo - rule out CHD, cardiomyopathy
- Exercise stress test - rarely diagnostic, but reassuring, a must for exertion-related syncope
- Tilt test – to confirm neurocardiogenic syncope
- Cath, EP study - when structural, rhythm abnormalities suspected