Syncope (and Pre-Syncope) of Potential Cardiac Origin

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“Passing Out”
Hippocrates

“Sudden Prostration” “Falling into a Swoon”
Galen

“One in five children experience at least one episode before age 15 years”

“Nearly one percent of all Peds ER visits”

Categorical Causes of Syncope (& Pre-Syncope)

- Neurologic
- Cardiac
- Traumatic
- Autonomic
- Psychological
- Endocriological
- Pulmonary
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96% of all cases are due to traumatic causes.

50% of all sudden deaths are due to cardiac causes.

The Journey

- Cardiac Origins of Syncope
  - Structural
  - Electrical
  - Hemodynamic
  - Autonomic Correlates
  - A Diagnostic Approach

Cardiac Origins of Syncope

- Structural
  - Mitral Stenosis/Insufficiency
  - Hypertrophic Cardiomyopathy
  - Dilated Cardiomyopathy
  - Restrictive Cardiomyopathy
  - Severe Aortic Stenosis
  - Aortic Valve Stenosis
  - Supravalvular Aortic Stenosis
  - Coronary Artery Disease
  - Arteriovenous Fistula
  - Intracardiac Tumor
  - Pericardial Effusion

- Electrical
  - Bradycardia
  - High Degree AV Block
  - Sick Sinus Syndrome
  - Block ADT Tachycardias
  - Atrial Flutter
  - Atrial Tachycardia
  - Concealed Pathways (SVT/VT)
  - Manifest Pathways (WPW)
  - Ion Channelopathies (LQTS…)
  - Unique Cardiomyopathy (ARVC)
  - Idiopathic (VT/VT)

- Hemodynamic
  - Pulmonary Hypertension
  - Idiopathic
  - Hereditary Hemorrhagic Telangiectasia
  - Eisenmenger complex
  - Anemia

Mechanism of Syncope

- Limitation of cardiac output
- Fixed due to structural limitations
- Dynamic with cardiac movement
- Impaired by arrhythmias
- Stopped due to arrest

Contributing Factors

- Increased demands/requirements
- Emotional state
- Potentially other factors
Structural Heart Disease

Hypertrophic Cardiomyopathy
- Multiple genetic causes and variable penetrance makes disease expression very variable.
- Both "obstructive" and "non-obstructive" forms.
- Loss of intracavitary volume may be significant in some forms.

Aortic Stenosis
- Infrequent as an "unknown cause" of syncope.
- Gradients may change quickly leading to events.
- Dysrhythmias may be precipitated in the vulnerable heart.

Supravalvar Aortic Stenosis
- Often occurs in syndromic cases (William syndrome).
- Gradient may belies risk.
- Frequently co-morbid coronary artery involvement.

Coronary Artery Anomalies
- Variable manifestations arise from coronary morphogenesis.
- Syncope may be the first presentation (usually exertion-dependent).

Electrical Cardiac Causes of Syncope

High-Grade Atrioventricular Block
- May vary from asymptomatic to highly symptomatic.
- Frequently confused with neurologic disease when intermittent.
- Resting ECG may be normal for intermittent cases.

Index of Suspicion is Essential!
Electrical Cardiac Disease

Paroxysmal Supraventricular Tachycardia

- Syncope rarely occurs in isolated SVT
- Palpitations are frequently reported at onset
- Classic prodromal symptoms rarely observed

Wolff-Parkinson-White Syndrome

- Frequently clinically indistinguishable from other forms of SVT
- Syncope is a potential harbinger of an increased risk of sudden death (> 0.1%/year)
- Prompt referral of any syncopal patients with WPW

Ion Channelopathies

Family of disorders characterized by abnormalities of the cardiac action potential, leading to electrical instability

- Short QT Syndrome
- Long QT Syndrome
- Brugada Syndrome
Electrical Cardiac Disease

Ion Channelopathies
- Heterogeneity makes diagnosis a challenge
- History Must Guide Work-Up!
- Genetics should be reserved for treatment guidance and family screening

Heterogeneity makes diagnosis a challenge! History Must Guide Work-Up! Genetics should be reserved for treatment guidance and family screening.

Compromise of cellular communication and early cell death can lead to both myopathic and arrhythmic states.

Electrical Cardiac Disease
Common Threads for Electrical Diseases
- Provocations may vary significantly from none to extreme activity to emotive states
- Absence of an “autonomic prodrome” is the key
- Complete exclusion of dysrhythmias is not possible unless the rhythm is captured

Electrical Cardiac Disease
Interface of Cardiomyopathies & Channelopathies

Hemodynamic Causes of Syncope

Pulmonary Artery Hypertension
- Multiple potential causes with most frequent being idiopathic in children.
- Syncope is a late manifestation; may reflect hypertensive crisis
- Delayed diagnosis is the rule — but physical examination holds the key (RV heave, telangiectasia, etc.).
Dysautonomia

The Most Common Cause of Syncope

- Significant confusion in literature about nature/type
- Impedes evaluation & study
- It is not all “dehydration…”

Autonomic Correlates

What is the Diagnosis?

Dysautonomia

A Framework for Diagnosis and Management

Persistent
- Orthostatic Hypotension
- Blood Pressure Problem
- Postural Orthostatic Tachycardia Syndrome
- Heart Rate Problem
- Mixed Form
  - BP & HR Problem

Episodic
- Vasovagal Syncope
- Blood Pressure Problem
- Cardioinhibitory Syncope
- Heart Rate Problem
- Mixed Form
  - BP & HR Problem

A Diagnostic Approach

It all begins with the history of the precipitating event:
- Accurate timeline of events
- Accurate description of symptoms
- Review of similar events (near misses?)
- Review of potential exacerbating/mitigating factors
  - May include past history, medications, family history, etc.

The physical examination serves to rule in disease:
- A head-to-toe evaluation
- Focus on cardiac, neurologic and dermatologic findings

A Diagnostic Approach

An electrocardiogram (E.C.G) is advocated for all first time syncopal events:
- Many electrical (and some structural) causes may be occult
- Significant implications for family screening
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An electrocardiogram (ECG) is advocated for all first time syncopal events:

- Many electrical (and some structural) causes may be occult
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Echocardiography may be reserved for high-risk cases:
- History not compatible (or partially compatible) with dysautonomia
- Abnormal physical exam findings
- Planned high-risk future activities (e.g., competitive contact athletics)

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Take Home Messages

- The potential cardiac causes of syncope are multiple though collectively quite rare.
- Look for patients for whom an autonomic cause "doesn't seem to fit."
- Evaluation for potential cardiac causes requires a thorough history, a complete physical examination and (for first time syncopal episodes) a resting electrocardiogram.

Parting Thought

"We shall not cease from exploration and the end of all our exploring will be to arrive where we started and know the place for the first time."

T.S. Eliot, Four Quartets