Beware the Red Flags in the Management of Syncope

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I am going to discuss Red Flags in clinical scenarios of children and adolescents after an initial presentation of syncope that presented to me.
Is it this.......... Or that..........
Initial Presentation of Syncope – My View

No Worry  Mild Worry  Moderate Worry  Severe Worry
Case #1

February 2007

• 14 year old male presents with syncope
• Playing basketball game
• towards end of game – dizzy; weak
• sat down on half-court line slumped unconscious
• pale, no tonic-clonic movements
• admitted to ER; received IV hydration – discharged
• finished out basketball season – no problems
December 2008

- varsity basketball – running sprints - collapsed
no left ventricular hypertrophy
no wpw
no qt prolongation
no brugada
• no symptoms
• maximum heart rate 208 bpm
• maximum blood pressure 166/96 mmHg
Anomalous Left Coronary Artery From Right Facing Sinus
Normal Left Coronary Artery Origin

- RVOT to PA
- LAD
- AO
- CX
Syncope with Exertion
DO NOT STOP UNITL YOU FIND A CAUSE
Case #2

- 4 year-old girl who presents for a heart murmur
- On the clinic note indicated that she has had two syncopal events?
- The most recent episode occurred when she was standing at the window, she saw dad pulling into the garage and fainted. Mom thought she just tripped.
Episode #2:

– standing in the kitchen, eating a banana when the phone rang and she fell forward.

– Mom thought she was “just playing” as she was back to normal a few seconds later.

• Examination: Normal

• Family History: Negative
Long QT Type II

QTc 560
Syncope with Exertion
Syncope with Sudden Noise
Case #3

- 7 y.o. boy new-onset seizure while running at recess
- Admitted neurology, diagnosed with a seizure disorder
- No EKG – no cardiology consult is obtained
- Family history is “negative for sudden death”
- Discharged the following day on a seizure medication
- 6 months later drowns in the swimming pool.
- The 5 y.o. brother is referred for evaluation
- EKG on arrival of the 5 year-old brother: ... a miss
EKG: Long QT & T wave alternans
• when I met with the family – I had asked the parents if any close relatives had died under 30 y of age. The answer was “No”
• when I asked the dad how many brothers and sisters he has he told me: 2 brothers and 1 sister. He had a brother that died before he was born
• how did the brother die?
  – (swimming age 5, but “no one talks about it”)
• the dad’s EKG was also c/w LQT
Long QT Syndrome

- Inherited disease (most autosomal dominant)
- Prolongation of cardiac repolarization
- "Longer time for the heart to relax"

**Genetic Basis**
- Cardiac ion (potassium, sodium, calcium) channel disease
- LQT 1, 2, 3 account > 85-90% all LQT cases
Channelopathies

- Diseases that affect cardiac ion channels
- LQTS prototype channelopathy
Cardiac Action Potential

Sustained Na into cell

Inability to expel K+ from cell
abnormal potassium channel

Sodium ($\text{Na}^+$) goes in

Potassium ($\text{K}^+$) cannot leave

Cell has more (+) charge = cell excited and cannot relax
Clinical Manifestations of LQT

- None
- Fainting
- Palpitations
- Sudden cardiac death
Clues to LQT Syndrome

• **Personal or Family History of:**
  – Stress induced fainting / palpitations
  – Stress induced seizures (1/3 of LQT misdiagnosed as epilepsy)
  – Unexplained drowning / Near drowning
  – Unexplained motor vehicle accidents

• **Family History of:**
  – Unexplained Sudden Death <40 y/o
  – SIDS
Occurrence of Gene-Specific Triggers

- LQT 1
- LQT 2
- LQT 3

Percent:

- EXERCISE
- EMOTION
- REST

Circ 2001;103:89-95
how good you are on obtaining a family history is only as good as the effort you put in
QTc 640 ms

patient clearly has LQT – why gene test?
This individual is positive for the deleterious mutation SCN5A Thr 220 Ile.
This individual is positive for the possible deleterious mutation KCNQ1 Arg 507 Trp.
Careful in the Waiting Room
Syncope with Exertion
Syncope with Sudden Noise
Syncope with a Concerning Family History
Case #4

• 13 year old boy presents to the hospital with a syncopal event while playing soccer on a typical Arizona summer day
• He was dizzy running down field and fainted.
• Fell hit head, small subdural hematoma
• Admitted to Neurosurgery Service
Left temporo-parietal hemorrhage
an astute medical student....

- Met family and found out that this is the 3rd syncopal event that occurred while playing soccer but the other two episodes it was not hot outside.
- **Time for a Cardiology Consultation**
- According to the resident exam normal, ECHO normal, no other concerning elements
- **Resident Request:** “You just need to clear (is he OK) him before anesthesia takes him down for another MRI”
A few more questions

• Was he well when he had these passing out episodes? – “Well he had a fever all three times”

• Let’s go through the family history one more time, let’s go through each of your siblings

• Uncovered that dad’s brother died suddenly at age 15 in the midst of a fever- it was attributed to a viral infection of the heart.
Brugada Syndrome

• Inheritable electrical disease
• Characterized by a typical ECG pattern
• Structurally normal heart
• Presents with syncope, sudden death, seizures due to polymorphic ventricular tachycardia
• Adolescents and young adults
Syncope with Exertion
Syncope with Sudden Noise
Syncope with a Concerning Family History
Syncope With a Fever
9 year old: 6 month history of exercise intolerance, chest pain, and a recent syncopal episode playing soccer

Moved from midfield to striker to goalie, so he can stand there.
12 Lead Electrocardiogram
42 segments of coronary narrowing
Started beta-blockers
Placed dual chamber ICD
Referred for transplant evaluation
Hypertrophic Cardiomyopathy

- Uncommon (0.1-0.2% general population)
- Most common cause of sudden death in young
- Mutations in genes encoding sarcomere
- Sudden death may be 1st symptom
- Variably hypertrophied (mild to massive)
- Autosomal dominant
- Variable penetrance
Hypertrophic Cardiomyopathy

Myocardial Disarray

Normal Muscle Structure
Symptoms of Hypertrophic Cardiomyopathy

- Tremendous variability in symptoms
- No symptoms
- Palpitations
- Dizziness
- Passing out
- Exercise intolerance
- Sudden cardiac death
Hypertrophic Cardiomyopathy Treatment

- Beta blockers and Calcium channel blockers to reduce HR and contractility
- Avoid dehydration
- AICD if septal thickness significant or symptomatic
- Muscle resection for relief of LV obstruction
- SCD prevention by diagnosing before disease manifests (genetic testing)
- Exercise restrictions is very effective
Syncope with **Exertion**
Syncope with **Sudden Noise**
Syncope with a **Concerning Family History**
Syncope with a **Fever**
Syncope with **Chest Pain**
Adolescent with a 1 month history of palpitations (appointment scheduled with cardiology next week)

Syncope During Dodge Ball, Connected to AED
Syncope with **Exertion**
Syncope with **Sudden Noise**
Syncope with a **Concerning Family History**
Syncope with a **Fever**
Syncope with **Chest Pain**
Syncope with **Palpitations**
7-year-old boy brought to the cardiology office after a vague syncopal episode while playing soccer.
No previous syncopal events while running.
Baseline EKG – normal sinus rhythm
Echo: Normal
No family history of syncope or SCD
Peak Exercise – “bidirectional couplets”

Started on beta-blockers - repeat EST unchanged

Added calcium-channel blockers - repeat EST unchanged
3 weeks later while running to 2nd grade class, collapses in front of school nurse’s office—(school has an AED)

Catecholaminergic Polymorphic VT
Catecholaminergic Polymorphic VT

- Polymorphic (bidirectional) VT in children
- Structurally normal heart
- Uncontrolled release of calcium sets up life-threatening arrhythmias
- 1/3 family history of syncope or sudden death
- Sudden death during adolescence
- Mutations RyR2 – cardiac ryanodine receptor
Natural History of CPVT

- 67% of patients have exercise/emotional fainting
- 33% sudden cardiac death
- 80% of patients will have symptoms < 40 years
- One of the most severe arrhythmia disorders
Treatment of CPVT

- Beta-blockers are the mainstay of treatment
- If fainting persists or there are continued ventricular arrhythmias despite beta-blockers an ICD is warranted
Syncope with **Exertion**
Syncope with **Sudden Noise**
Family Histories Often Provide A Clue
Syncope with a **Fever**
Syncope with **Chest Pain**
Syncope with **Palpitations**
Even Being Aware of Red Flags: Secondary Precautions are Important
Know Where the AED Is Located!

Where Did You Say the AED was??
Syncope with Exertion
Syncope with Sudden Noise
Family Histories Often Provide A Clue
Syncope with a Fever
Syncope with Chest Pain
Syncope with Palpitations
Even Being Aware of Red Flags: Secondary Precautions are Important

Syncope with NO SYMPTOMS
Goals of Assessment of Syncope

1. Evaluate for underlying heart diseases that alter
   – ion channel function
   – cardiac myocyte function
   – intracellular matrix
2. Determine safety of returning to sports
3. Who else in family needs to be tested
4. Reassurance
5. Therapy if needed
Prove Vasovagal Syncope Not...

• Vasovagal syncope is
  – transient loss of consciousness
  – inability to maintain postural tone
  – spontaneous recovery
  – absence of...

• seizure
• arrhythmia
• heart disease
Vasovagal Syncope
Detailed History of Event

- Predisposing situations
- Prodromal symptoms
- Physical signs
- Recovery time and symptoms
Predisposing Situations

• Prolonged standing, sitting, position change
• Medical, painful, “yucky”
Prodromal Symptoms

- Warmth
- Nausea
- Clouded thinking
- Vision/hearing changes
- Tunnel Like Vision
- Dizziness

Syncope without prodrome or with injury is worrisome
Prodrome

- May be falsely reassuring
- $> \frac{1}{2}$ patients with channelopathy reported a prodrome before a syncopal event

MacKormik Heart Lung Circulation 2011
Physical Signs

• Pallor
• Diaphoresis
• Hypotension
• Relative bradycardia
• Myotonic movements may be interpreted as seizure
Recovery Time and Symptoms After

- Unconsciousness for < 2 minutes, full recovery sluggish
- Tired for minutes-hours after
- Headache

Careful and focused history makes the diagnosis
**Personal/Family History**

<table>
<thead>
<tr>
<th>Personal History</th>
<th>Family History</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Previous syncope/dizziness</td>
<td>• Sudden death</td>
</tr>
<tr>
<td>• Palpitations</td>
<td>• Known heart disease</td>
</tr>
<tr>
<td>• Chest pain</td>
<td>• SIDS</td>
</tr>
<tr>
<td>• Drugs including prescribed, illicit, supplements and</td>
<td>• Drowning</td>
</tr>
<tr>
<td>“sports enhancing”</td>
<td>• Recurrent fetal loss</td>
</tr>
</tbody>
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Testing?

• History/Physical
• ECG

• History/Physical
• ECG
• Echo
• Exercise test
• Tilt
• Holter
• EP Study
• Loop recorder
• MRI
• Genetic testing
16-year-old boy faints at the end of the high school state cross country finals.

Elite athlete – headed for scholarship to an Ivy League

No chest pain and no palpitations

“I was getting a little dizzy before the finish line went black”

Self-recovered

Cool outside

Pushing himself super hard
I Saw This A Lot!!!!!!

In Arizona, we have 2 types of weather...

HOT.

a little less hot.
Why do endurance athletes faint?

- Abnormal BP response - **depressed carotid baroreceptor sensitivity**
- Increased stroke volume, more compliant, distensible ventricles - chronic volume overload
  - delivery large volume of blood to muscles
- Disadvantage during orthostasis
- Results in large decrease in SV when filling pressure reduced

Whyte Br J Sports Medicine 2004
My Approach to the Exercise Syncope (syncope before the finish line) with No Easy Answer

- Assuming normal exam and family history
- ECG - Normal
- ECHO – Normal
- Stress Test – Normal
- Holter – No concerning PVC burden, normal
- CT scan or MRI (coronaries unless echo 100%)
- Implantable loop recorder (if truly exertional)
- Resume competition (must have AED available, my advocacy)
- Treatment: Vasovagal approach
Management of Vasovagal Syncope

- Be Aware of Situations
- Hydration, salt, support stockings
- No other therapy unless syncope persists, danger of injury or drowning
  - pharmacotherapy
    - midodrine
    - florinef
  - 2017: beta-blockers
- Lower body (+) pressure maneuvers after exercise may limit/eliminate events
Thank You

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