History, not Hysteria: A Rational Approach to Syncope in Children

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Disclosures: none

Evaluation of Syncope in Children

• Objectives
  – To review the epidemiology and physiology of syncope in children.
  – To discuss appropriate clinical assessment and treatment of the fainting child.
  – To highlight and distinguish the presentation of syncope associated with structural cardiac disease and cardiac arrhythmias.

Evaluation of Syncope in Children

• Common
  – Dramatic, disruptive events
  – Potential for injury
  – RARELY, self-limited expression of a potentially lethal condition.

Evaluation of Syncope in Children

• Syncope: sudden, transient loss of consciousness and postural tone
• Presyncope: sensory and/or postural impairment without complete loss of consciousness

Transient impairment cerebral perfusion

Evaluation of Syncope in Children

- Inciting factors
  - Noxious stimulus
  - Acute illness
  - Orthostatic challenge
  - Warm environment
  - Drugs
  - Recent exercise

- Associated symptoms/signs/events
  - Prodrome: dizziness, flushed, clammy
  - Tonic-clonic movements
  - Nausea/vomiting
  - Pallor
  - Injury

- Syncope during exercise:
  Red Flag Event

- Neurally-mediated Syncope
  - Neurogenic syncope
  - Neurocardiogenic syncope
  - Vasovagal syncope
  - Vasodepressor syncope
  - Reflex syncope
  - “Common faint”
Evaluation of Syncope in Children

- Differential Diagnosis
  - Seizures
  - Migraine
  - Hypoglycemia
  - Breath-holding spells
  - Hyperventilation
  - Psychogenic/Pseudo-syncope
  - Cardiac syncope

Cardiac Syncope

- Structural heart disease
  - Obstruction: hypertrophic cardiomyopathy, aortic valve stenosis
  - Coronary anomalies
- Cardiomyopathy
  - Dilated cardiomyopathy, myocarditis
  - ARVC
- Pulmonary Vascular
  - Pulmonary hypertension
  - Pulmonary embolus
- Arrhythmias
  - Bradycardia: sick sinus syndrome, complete AV block
  - SVT: WPW, AV node re-entry, atrial tachycardia
  - Ventricular dysrhythmias: ion channelopathies, ARVC

Evaluation of Syncope in Children

- Goals of evaluation
  - Accurate characterization of event
  - Appropriate diagnostic testing
  - Logical treatment
  - Follow-up to assess effectiveness
Evaluation of Syncope in Children

- History is the KEY to diagnosis
  - History: child
  - History: eyewitness account
  - History: what, where, when, who, how
- Family History
  - Syncope
  - Sudden, unexpected deaths in young people
  - Seizures
- Physical Exam
  - Evidence of structural heart disease
  - Orthostatic BP changes

Evaluation of Syncope in Children

- The Electrocardiogram
  - Hypertrophic Cardiomyopathy
  - Cardiac Ion Channelopathy
  - Wolff-Parkinson-White
- Consensus
  - ESC Guidelines for the Diagnosis and Management of Syncope. EHJ 2009;30; 2631-2671.
  - Syncope: Therapeutic Approaches. JACC 2009;53;1741-1751.

Evaluation of Syncope in Children

- Cardiac Workup beyond ECG indicated:
  - Faint with exercise
  - Syncope without prodrome, while supine, or preceded by chest pain or palpitations
  - Event triggers: loud noise, fright, extreme emotional stress
  - Known or suspected heart disease
  - Family history: unexplained or sudden death, known familial heart disease

Wolff-Parkinson-White

- Rapid conduction of atrial dysrhythmia

Familial Syndromes Associated with Ventricular Arrhythmia

- Autosomal dominant inheritance
  - Romano-Ward Long QT syndrome
  - Brugada syndrome
  - Arrhythmogenic Right Ventricular Cardiomyopathy
- Autosomal recessive inheritance
  - Jervell and Lange-Nielsen syndrome (congenital deafness)
Familial Syndromes Associated with Ventricular Arrhythmia

- The Long QT syndromes (LQTS) characterized by:
  - Syncope
  - Seizures
  - Sudden death
  Polymorphous ventricular tachycardia, “torsades de pointes”

Romano-Ward Long QT Syndrome

- ECG Findings in LQTS
  - Prolonged QT, (QT/RR) > 450 msec
  - Abnormal T waves: biphasic, notched, T wave alternans
  - Sinus bradycardia: 20% - 30% of patients
  - Rarely, atrioventricular block
- Impaired Cardiac Na⁺ or K⁺ Channel Function
  - Prolonged ventricular repolarization prolongs period of vulnerability

Genetic Diagnosis of LQTS

  - RFLP/Southern Blot
  - 250 DNA probes
  - H-ras 1: K+ channel protein
- Single gene defect?
  - Genetic homogeneity?

EcG phenotypes in LQTS

Genetic Diagnosis of LQTS

- LQT1-KCNQ1 (KvLQT1) 11p15.5
- LQT2-HERG; LQT3-SCN5A; LQT4-AnkyrinB; LQT5-MinK; LQT6-MirP1; LQT7-KCNJ2; LQT8-CACNA1C; LQT9-Caveolin3; LQT10-SCN4B

KvLQT1
Brugada Syndrome
SCN5A

Evaluation of Syncope in Children

- Diagnostic evaluation
  - Electrocardiogram: inexpensive, occasionally useful
  - Event recorder: low-yield; frequent episodes with palpations
  - 24-hour Holter monitor: low-yield; daily episodes
  - Echocardiography: expensive, low-yield

- Routine Tilt-table testing
  - Poor specificity in kids: 34% false positive
    - Lewis, JACC 1997;30:1657
    - Non-predictive of outcome
      - Levine, Pediatr Cardiol 1999;20:331
      - Costly: $1065/pt for neurogenic syncope
        - Knilans, J Pediatr 2005;146:355-8
  - When?
    - Multiply recurrent or atypical events: mechanism of reflex
    - In combination with exercise if syncope with exertion
    - Psychogenic/Pseudosyncope

- Role of Echocardiography:
  - Only if know/suspect structural heart disease or Syncope with exercise

- Ritter (Pediatrics 2000;105;5;e58):
  - 480 patients (458 non-cardiac, 14 Long QT)
  - 322 echocardiograms ($387,000)
  - 2 patients with cardiomyopathy (both with abnormal EKG)
Syncope Treatment

North Otago Times, Volume XVIII, Issue 712, 15 March 1872

Syncope in Children: Treatment

• Non-pharmacologic
  – Avoidance: prodrome recognition
  – Behavioral modification: aggressive hydration and added salt
• Pharmacologic
  – Florinef: 0.1 mg daily
  – Beta-blockers: effective though side effects not trivial, Atenolol


Aborting Syncope: Counter-maneuvers

Syncope in Children: Treatment

• Pharmacologic
  – Alpha-agonists: pseudoephedrine, mitodrine; multiple daily dosages
  – Serotonin agonists: sertraline
  – Disopyramide: vagolytic, effective for refractory syncope, high side-effect profile

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