Adolescent Syncope

Brett Goudie, M.D.
Pediatric Cardiology
14 yo boy presents to clinic following syncopal episode. Standing in church, felt hot and lightheaded before passing out. Witnessed event, parents described brief seizure like activity while he was down. No loss of bowel or bladder control, no CPR, no resp support. Brief LOC, woke up 5 sec later, was soon back to being alert and oriented. He noted visual changes and hearing became distant prior to the episode.

- Cause?
- Further testing?
- Referral?
- Recommendations?
Syncope

From the Greek word **synkoptein**, which means “to cut short”

Syncope – “temporary loss of consciousness and postural tone resulting from an abrupt, transient, and diffuse cerebral dysfunction (hypoperfusion), and followed by spontaneous recovery” – cerebral blood flow ↓ 30-50%

Syncopized????

Pre-syncopize?

Pre-syncope – prodromal dizziness and lightheadedness that precedes LOC – aka near-syncope

Sadly I have no financial disclosures related to this topic
• 17 yo girl (no PMHx) presents after syncope with exercise. She was running the 100 m dash, and passed out suddenly while running. No preceding symptoms. Woke up 4-5 sec later, was confused, had trouble recognizing her track coach or track team.
  • Same questions
  • Cause?
  • Further testing?
  • Referral?
  • Recommendations?
• Common - 15-25% of children experience at least one episode (peak ages 15-19) (♀ > ♂)
• Expensive clinical problem / testing / Up to 3 % of ER visits / treatable
• Most pediatric syncopal episodes are totally benign
• 80-90% is vasodepressor or cardioinhibitory → vasovagal
• Only 1% of time due to arrhythmia or structural cardiac disease – hence there is a risk of sudden death
• ANGST!!!!!
Vasovagal Syncope Terminology

- Vasodepressor (Vaso)
- Cardioinhibitory (Vagal)
- Neurocardiogenic
- POTS – postural orthostatic tachycardia syndrome
- Orthostatic Hypotension or Intolerance
- Situational
- Reflex
- Neurally mediated
- Dysautonomia
Bryan Cannon, Philip Wackel
Pediatrics in Review Apr 2016, 37 (4) 159-168
“Fainting Goat”
Myotonia Congenita
Presyncope

- Lightheaded, dizziness
- Visual changes - blurry vision, tunnel vision, spots, double vision
- Altered hearing – distant voices
- Altered mentation, confusion, panic, h/a
- Weakness, poor balance
- Warmth / rush, or cold and clammy
- Sweating, nausea
- Pallor or gray
Syncope, at the Carotid Sinus

- Upright posture, 25% of blood redistributed to lower extremities
- Preload diminished, leads to diminished stroke volume by as much as 40%, CO = HR * SV
- Baroreceptors activated (Carotid sinus)
- Increased sympathetic activity increases HR, contractility, and SVR (systemic vascular resistance)
- Failure of this response leads to vasodepressor syncope

https://medicine.uiowa.edu/iowaprotocols/carotid-body-and-carotid-sinus-general-information
Bezold-Jarisch Reflex

Tilt

↓ Venous Return

↓ BP

↑ Catecholamines

Reflex

↓ Venous Return

↓ BP

↑ Carotid Sinus Baroreceptors

Ventricles

↓ Reflux

Vagal Afferent

Brain Stem

↑ Vagal Efferent

↓ HR

↓ HR

Vasodilation

Sympathetic Withdrawal

↓ BP

Increase HR, contractility, and BP

18 year old with syncope after exercise

Recovery 0:14  HR=189 bpm  BP=115 systolic
18 year old with syncope after exercise

Recovery 0:30  HR=178 bpm  BP=60 systolic
Albert von Bezold (1836-1868)

- "Bezold-Jarisch reflex" is a triad of responses (bradycardia, hypotension, and apnea) from paradoxical stimulation of vagal mediated ventricular C fiber receptors.
- Reflex is involved with the “Cardioinhibitory” or “Neurocardiogenic” Syncope
- Bezold - intravenous injection of veratrum alkaloids, +/- vagal nerve ligation.
- Bezold - Munich, then in Univ of Berlin under du Bois-Reymond (nerve action potential)
- Adolf Jarisch Jr (1891–1965) replicated study in cats with reversible cooling of vagal nerve.
Veratrum viride or album

- Potent alkaloids, antagonism of adrenergic receptors
- Highly toxic – nausea / vomiting, cold sweat and vertigo, slowing of respiration, bradycardia, hypotension, death
- 1950-1960s extract known as alkavervir – antihypertensive
- Some Native American nations – historical use for elections of new leader
Bezold-Jarisch Reflex

Tilt

↓ Venous Return

↓ BP

Carotid Sinus Baroreceptors

↑ Catecholamines

↓ Reflex

Ventricles

↓ Vagal Afferent

Brain Stem

↓ Vagal Efferent

Brain Stem

↓ HR

↓ BP

Sympathetic Withdrawal

Vasodilation

Increase HR, contractility, and BP

Oh God, my mind is going a mile an hour.
• What are the circumstances leading to events? Sick with URI or other illness pre or post? Preceding mono or viral illness weeks or months prior? Growth spurt?
• Abrupt onset or prodrome? Exercise? Post trauma?
• Are there palpitations, visual changes, chest pain, shortness of breath, dizziness vs vertigo?
• LOC details – duration?
• Injury? Seizure? Loss of bowel/bladder control? CPR?
• Eat or drink? Is there a history of caffeine use? Is his/her urine concentrated or clear? 2nd daily void
Syncope

History – Red Flags

- Syncope after surprise, loud noises, anger
- No warning or while supine
- Prodrome of palpitations/tachycardia/severe chest pain
- Syncope with exercise ***
- Prolonged down time, pulseless, CPR, residual neuro issue
- Other red flags
  - Abnormal exam or EKG
  - F Hx of sudden death / SIDS
  - Hx of CHD
- Adult Syncope – more likely to be cardiac
Syncope

History – Green Flags

- Benign Prodrome
- Brief Episode
- Single Episode
- Only while Upright
- Change of position
- Poor hydration
- Lots of dizziness with standing

OR

- Presyncope only – no flaccid LOC, full memory
Family History / Physical Exam / EKG

- Arrhythmias
- Sudden death
- SIDS
- Cardiomyopathy
- Pacemakers
- Defibrillators / ICD
- Deafness
- Congenital Heart Disease
- Fainters
• Heart rate and blood pressure, both supine and standing, then standing again in a few minutes
• Are the first and second heart sounds normal?
• Is there a murmur? Does it get louder with standing?
• Any ectopy noted?
• Is there a click, gallop, rub, or filling rumble?
• Leg plethora, edema, swelling
Cardiac Syncope

Structural Heart Disease
- Severe aortic stenosis
- Hypertrophic cardiomyopathy
- Severe pulmonic stenosis
- Coronary artery anomalies
- Primary pulmonary hypertension

Myocardial Dysfunction
- Dilated cardiomyopathy
- Myocarditis
- Bradyarrhythmias
  - Atrioventricular block
  - Sinus node dysfunction

Arrhythmia
- Tachyarrhythmias
  - Supraventricular tachycardia
  - Ventricular tachycardia
  - Long QT syndrome
  - Catecholaminergic polymorphic ventricular tachycardia
  - Wolff-Parkinson-White

Bryan Cannon, Philip Wackel
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Psychogenic Syncope

- Pseudoseizures or behavioral spells
- No physiologic changes prior to syncope
- Certain times, certain places, daily
  - > age 10. LOC > 10 min
- Emotionally charged
- Common after abuse (emotional or sexual)
- 15 yo girl “passing out” many times per day.
  - Fluid, salt, cut caffeine
  - Florinef, mitodrine.
- Put on the brakes BP high.
- Episode during stress test, actually passed out with no BP change, no EKG change, nothing. No EKG change ?????
Fainting Lark
Everybody thinks that I am crazy and that tells me that I am the sanest person I know.
History / Physical Exam / EKG

- Is the rhythm SINUS?
- Is the rate appropriate?
- Is there AV block?
- Is there evidence of hypertrophy or strain?
- Is there pre-excitation (short PR or delta)?
- Is the QTc normal?
- Any PVCs?
EKG post syncope
Diagnostic tests

• Orthostatic HR and BP
• ECG
• Head Up Tilt Test - usually not necessary
• Exercise stress test – syncope with exercise
• 24 hour Holter / 30 day event/loop monitor / Reveal monitor
• Echocardiogram with emphasis on chamber size, obstructive lesions, coronary arteries
• Electrophysiology study
• Labs – U/A (spec grav), CBC, glucose, Lytes, TSH, pregnancy, drugs
• Brain imaging – only for focal neuro signs
• EEG – can be abnormal after benign syncope
The Tilt Lab
Neurally-Mediated Syncope
Tilt Table Response

Arrhythmias Causing Syncope

• Bradycardia
  • Sinus Node Dysfunction
  • AV Block

• Tachycardias
  • Wolff-Parkinson-White Syndrome
  • Long QT Syndrome
  • Brugada Syndrome
  • Arrhythmogenic RV Dysplasia (ARVD)
  • Catecholaminergic Polymorphic VT (CPVT)
Sinus Node Dysfunction

- Inappropriately slow (or fast) heart rate
- SA node origin
- Frequently seen in post-op hearts
- <3 years old <100
- 3-9 years old <60
- 9-16 years old <50
- >16 years old <40
Complete AV Block
Wolff-Parkinson-White Syndrome

Sinus Rhythm

Orthodromic Atrioventricular Reentrant Tachycardia
Not-so-subtle WPW
Atrial Fibrillation in WPW
The Long QT Syndrome

- LQTS is present in 1:5,000 persons (over 50,000 people) in the USA
- Estimated to cause as many as 3000 deaths (mostly in children and young adults) in the USA each year.
- It is present in all races and ethnic groups
- In as many as 30-40%, sudden death is the first event (8-40% of asymptomatic patients have SCD as first event)
- 57% of patients who die will die by age 20
- Events rates are 5%/yr. for syncope and 0.9-2.5%/yr. for cardiac death
Corrected QT Interval (QTc)

- Include U wave if > 50% of T wave height
- Upper limits of normal 450

<table>
<thead>
<tr>
<th>PPV 100%</th>
<th>Male</th>
<th>&gt;470</th>
</tr>
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<tbody>
<tr>
<td></td>
<td>Female</td>
<td>&gt;480</td>
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<tr>
<td>PPV 93%</td>
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<td>&gt;460</td>
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<td>NPV 100%</td>
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<td>&lt;400</td>
</tr>
<tr>
<td></td>
<td>Female</td>
<td>&lt;420</td>
</tr>
</tbody>
</table>
Torsades de Pointes
ECG Phenotype of LQT3, 2, and 1

LQT3
Chromosome 3

LQT2
Chromosome 7

LQT1
Chromosome 11

II

aVF

V5
Varieties of LQTS

- LQT 1-Tend to have events at times of exertion or excitement.
- LQT 2-Tend to have events when startled or with swimming.
- LQT 3-Tend to have events when asleep.
- LQT 1 and 2 account for 85% of total. LQT 3 accounts for about 10%.
TdP following a sudden arousal (alarm clock) in a patient with a HERG defect.

Wilde et al. JACC. 1999;33:327-32
Treatment of LQTS

• Avoid all strenuous exercise, esp. swimming

• Avoid startling (alarm clocks etc.)

• Beta blocker therapy (Propranolol, Nadolol) w/ or w/o bradycardia pacing

• ICD (QTc>550, noncompliant, FHx SCD)

• Left Stellate ganglionectomy
### Drugs – QTDRUGS.ORG

#### Updated April 10, 2021
Please check our website for the most current information.

<table>
<thead>
<tr>
<th>Drug (Brand Names)</th>
<th>Drug Class (Clinical Usage)</th>
<th>QT</th>
<th>TP</th>
<th>Comments</th>
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<tbody>
<tr>
<td>Amiodarone (Cordarone)</td>
<td>Antiarrhythmic (heart rhythm)</td>
<td>QT</td>
<td>Tdp</td>
<td>F-M, Tdp Cases in Lit</td>
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<td>Artesic (Ioxides) (Theroux)</td>
<td>Anticancer (leukemia)</td>
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<td>Tdp Cases in Lit</td>
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<td>Bupropi (Zyban)</td>
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<td>Chlorpromazine (Thorazine)</td>
<td>Antipsychotic/Antiemetic (schizophrenia/nausea)</td>
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<td>Tdp</td>
<td>Tdp Cases in Lit</td>
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<td>Clopamide (Propulsor)</td>
<td>GI stimulant (stimulates GI motility)</td>
<td>QT</td>
<td>Tdp</td>
<td>Tdp Cases in Lit</td>
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<tr>
<td>Clonazepam (Biaxin)</td>
<td>Antibiotic (bacterial infection)</td>
<td>QT</td>
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<td>Tdp Cases in Lit</td>
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<td>Desipramine (Norpramin)</td>
<td>Antidepressant (depression, others)</td>
<td>QT</td>
<td>Tdp</td>
<td>Tdp Cases in Lit</td>
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<td>Disopyramide (Norpace)</td>
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<td>Dofetilide (Tikosyn)</td>
<td>Antiarhythmic (heart rhythm)</td>
<td>QT</td>
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<td>Doxepin (Sinequan) (Zonalon)</td>
<td>Antidepressant (depression, pain, other)</td>
<td>QT</td>
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<td>Tdp Cases in Lit</td>
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<td>Droperidol (Inapsine)</td>
<td>Sedative/Hypnotic (anesthesia adjunct)</td>
<td>QT</td>
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<td>Tdp Cases in Lit</td>
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<td>Erythromycin (E.E.S., Erythrocin)</td>
<td>Antibiotic (stimulant infection)</td>
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<td>Flecainide (Tambocor)</td>
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<td>Fluoxetine (Prozac, Sarafem)</td>
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<td>Foscarnet (Foscavir)</td>
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<td>Fosphenytoin (Levetiracetam)</td>
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<td>Gattifloxacin (Tequin)</td>
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<td>Haloperidol (Haldol)</td>
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<td>Ibutilide (Corvert)</td>
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<td>Tdp Cases in Lit</td>
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<td>Imipramine (Tofranil)</td>
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<td>Indapamide (Lozol)</td>
<td>Diuretic (stimulates urie &amp; salt loss)</td>
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<td>Tdp</td>
<td>Tdp Cases in Lit, TQT in Animals</td>
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<td>Iratidine (Dynacor)</td>
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<td>Levofloxacin (Levaquin)</td>
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<td>Levomethadyl (Orafun)</td>
<td>Opiate agonist (narcotic dependence)</td>
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<td>Mesoridazine (Sensid)</td>
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<td>Moxifloxacin (HCTZ) (Uniret)</td>
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<td>Moxifloxacin (Avisox)</td>
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<td>Naranil (Amegam)</td>
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<td>Nicardipine (Cardene)</td>
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<td>Ocotrode (Sandostatin)</td>
<td>Endocrine (acromegaly/carcinoid diarrhea)</td>
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<td>Paroxetine (Paxil)</td>
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<td>Pentamidine (Pentam, Nustro, Pentil)</td>
<td>Anti-infective (Pneumocystis pneumonia)</td>
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<td>Pimozide (Orap)</td>
<td>Antipsychotic (Tourette's tic)</td>
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<td>F-M, Tdp Cases in Lit</td>
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<td>Probufol (Lubope)</td>
<td>Antilipemic (lowers cholesterol)</td>
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<td>F-M</td>
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<td>Procanamide (Procan, Proventil, Procan)</td>
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<td>Quinidine (Cardioquin)</td>
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<td>Salmeterol (Serevent)</td>
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<td>Sotalol (Betapace)</td>
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<td>Sulfamethoxazole - trimethoprim (Bactrim, Septra)</td>
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<td>Sumatriptan (Imitrex)</td>
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<td>Tacrolimus (Prograf)</td>
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<td>Tamoxifen (Nolvadex)</td>
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<td>Theophylline (Mellaril)</td>
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<td>Tizanidine (Zanaflex)</td>
<td>Muscle relaxant</td>
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<td>Venlafaxine (Effexor)</td>
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<td>Zonisamide (Geodone)</td>
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<td>Zomig (Zomig)</td>
<td>Migraine treatment</td>
<td>QT</td>
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</tr>
</tbody>
</table>

### Drugs to avoid (partial list)

**Antiiarrhythmics**
- procainamide
- quinidine
- disopyramide
- sotalol
- amiodarone
- ibutilide
- dofetilide

**Psychotropics**
- haloperidol (Haldol®)
- risperidone (Risperdal®)
- droperidol (Inapsine®)
- thiothixene (Navane®)
- doxepin (Sinequan®)
- phenothiazines
- tricyclics
- tetracyclines (Zonlalon®)
- fluvoxamine (Luvox®)

**Other**
- epinephrine (adrenaline)
- diuretics (water pills)
- probucol
- cisapride (Propulsid®)
- ketanserin
- bepridil (Vasco®)
- pimozide (Orap®)

### Antimicrobial & Antifungals (some)
- sulfamethoxazole - trimethoprim (Bactrim, Septra®)
- penicillin
- amantadine (Symmetrel®)
- erythromycin
- chloroquine
- halofantrine
- ketoconazole
- itraconazole
- clindamycin (Cleocin®)**
- clarithromycin (Biaxin®)**
Brugada Syndrome

- Leading cause of death in young Asian males
- Arrhythmias tend to occur during febrile periods (exercise) and during sleep
- Defect is in cardiac sodium channel
Brugada Syndrome

Wilde A, et al., 2002
Catecholaminergic Polymorphic Ventricular Tachycardia (CPVT)

- aka Syncopal paroxysmal tachycardia, malignant paroxysmal VT, multifocal ventricular premature beats, paroxysmal VF, bidirectional tachycardia, Familial Polymorphic VT
- Defect identified in the ryanodine receptor (RyR2) and Calsequestrin genes
- Rarely symptomatic before 3 years. Mean age of presentation 8 yrs for RYR2 defect, later for others.
- Male predominance in RYR2 variety
CPVT – increased ectopy with exercise
Bidirectional VT in CPVT

Priori S, et al, 2002
History Findings Suspicious Arrhythmias

- Syncope preceded by palpitations
- Syncope with exertion or excitement
- Abrupt syncope without a prodrome
- Syncope while swimming
- Family history of syncope, seizures, SIDS, sudden death, or deafness
Syncope and Structural Heart Disease

• Hypertrophic Cardiomyopathy
• Coronary artery anomalies
  • Abnormal take-off
  • Single coronary
• Marfan syndrome
• Arrhythmogenic RV Dysplasia
• AS, PS, TOF, pulmonary hypertension, MVP (0.5% risk of sudden death annually)
Hypertrophic Cardiomyopathy

• Leading cause of sudden death in healthy young athletes
• Typically die of arrhythmias (V fib)
• Prevalence as high as 1/500
• At least 50% are inherited
• Family history is prognostic
Hypertrophic Cardiomyopathy
Single Coronary Arteries

From Perloff J. The Clinical Recognition of Congenital Heart Disease, 1994
Sudden Death in Young Athletes - Coronary Artery Anomalies

• Study of 27 patients (Basso C, et al., 2000)
  • 23 had anomalous LCA from R cusp
  • 4 had anomalous RCA from L cusp

• 41% HS students, 44% JHS, college 2%

• Sports included
  • Basketball 8
  • Soccer 7
  • Football 3
  • Distance running 3
  • Track 2
  • Hockey, rugby, softball, swimming 1 each
Coronary Artery Anomalies

• 16 died during training, 11 during competition
• 10/12 patients with clinical history available
  • 5 chest pain (3 with exertion)
  • 4 syncope (3 with exertion)
  • 2 palpitations unrelated to exercise
  • 1 palpitations and presyncope with exercise
• ECG normal 9/9
• EST normal 6/6
• Echo normal in 2/2
ALCAPA – anomalous LCA from PA
Arrhythmogenic RV Dysplasia

• Second leading cause of sports related death in parts of Europe
• Occurs in 1 in 5000 individuals
• Defect in cardiac ryanodine receptor (calcium release protein)
• Arrhythmias occur during times of stress
• Associated with fibrosis and fatty infiltrates of the RV free wall, best detected by MRI
• ECG may show classic “epsilon” waves
Arrhythmogenic RV Dysplasia

Gear K et al., 2003
Arrhythmogenic RV Dysplasia

Soler, Rafaela, et al., 2003
Epsilon Wave of ARVD

Alter P, et al., 2004
Marfan Syndrome

• Tall thin with disproportionately long arms
• Long lower body
• Arachnodactyly
• Hypermobility (thumb sign, wrist sign)
• Scoliosis
• Pectus deformity
• Lens dislocation
• Aortic root dilatation

• In one study (Fornes P, et al, 2003) 3/11 sports related SCD were due to ruptured aortic aneurysm
Marfan Syndrome

Braverman AC, 1998
Cardiac Syncope is 1% of all Syncope

- History
- Family History
- Exam
- EKG
Syncope Treatment - acute

- Lay down
- Legs up
- Give them air
- Get them a drink

- Presyncope – same instructions, or while standing initiate counterpulsation.
Neurocardiogenic Syncope
Nonpharmacologic Interventions

• Hydration
  • 64 fluid oz of fluid/day (4-6 ½L waters/day)
  • 32 oz by noon
  • Urine should be clear, void every 2-3 hrs,
  • Avoid diuretics (caffeine and alcohol)
• Increase salt intake (if not hypertensive)
• Support hose, shorter showers
• Tilt training – leg strengthening, leg flex
• Time – Viral illness preceding, Growth spurt
• Regular exercise program with cool down
“How many times must I remind you? Eight glasses a day!”
Why don't you explain this to me like I am an eight-year-old?
Pharmacologic Interventions

• Florinef acetate 0.1 mg po QD
  • Avoid if hypertensive, or glaucoma
  • Recent study suggests no more effective than placebo, and placebo very effective.
    • (Salim M, et al. 2005)

• Beta blocker therapy (Atenolol or Nadolol) 1 mg/kg div. b.i.d. or q.d.
  • Lack of controlled studies

• Midodrine (ProAmatine) – 2.5-10mg TID
  • Shown to be effective in acute setting
    • (Kaufmann H, et al, 2002)
  • Side effects often intolerable – HTN, blurry vision, H/A
How to Approach the Patient With Syncope

- History, History, History
- Family history
- Physical exam including orthostatic BP and auscultation in prone and upright position
- All patients deserve an ECG
- The remainder of tests (echo, tilt, EST, EP study, cardiac MRI) should be confirmatory, not exploratory
When to Worry...

- Syncope during exertion / excitement / swimming
- Family history of sudden death
- Abrupt syncope without a prodrome
- Sustained seizure activity
- Apnea, CPR, chest compressions
- Prior cardiac surgery or congenital heart disease
When to Be Suspicious of psych issue...

- Frequent absence from school
- Selective loss of neurological function
- Risky social or sexual behavior
  - SEXUAL ABUSE
- Multiple episodes daily at same time
- Vague changing character of symptoms
Who to refer to the Cardiologist

- Syncope with exertion, swimming, palpitations, or chest pain
- Abnormal ECG
- Abnormal cardiac exam
- Family history of sudden cardiac death
- Congenital heart disease or prior heart surgery
- Anyone who’s syncope is not obviously neurally mediated, orthostatic, CNS, or psychogenic
What is the most clinically beneficial long term treatment for benign vasovagal syncope in a 16 yo adolescent?

A) Florinef 0.1 mg daily
B) Salt tabs or increased dietary salt
C) Caffeine Avoidance
D) 2-3 L of daily oral clear fluid intake
Question 2

What patient worries you the most, with the most concerning syncopal presentation?

A) 14 yo passed out while standing in church
B) 10 yo with 5 second period of LOC, passed out after blood draw
C) 17 yo collapses while running 100 m dash
D) 8 yo girl faints while having her hair brushed
E) 9 yo who passes out after stubbing his toe and cries, while down has brief clonic posturing
Question 3

What is the most useful test in patient with single episode of benign syncope by history, FHx, and exam?

A) No testing needed
B) EKG
C) ECHO
D) Tilt Test
E) Stress Testing
Bonus question – who can summarize Bezold-Jarisch reflex?
Breath Holding Spells

• Pallid
  • Usually provoked by pain
  • Child becomes pale and limp
  • Caused by reflex asystole

• Cyanotic
  • Usually provoked by anger
  • Infant holds breath and becomes cyanotic

• In both cases, the child may have tonic or tonic-clonic activity
Pallid Breath Holding Spell
18 month old
Hyperventilation Syndrome

• Typically provoked by anxiety
• Subjective SOB (inability to fill lungs)
• Paresthesias of hands and lips
• Visual changes-tunneled vision
• Lightheadedness
• Chest pain (tightness) very common
• Treatment: Breath into a paper bag!