“When It All Goes to POTS”
Assessment and Treatment of Pediatric Syncope

Camden Hebson, M.D.
Sibley Heart Center, Children’s Healthcare of Atlanta
Emory University School of Medicine
## Camden Hebson, M.D.
### Personal/Professional Financial Relationships with Industry

<table>
<thead>
<tr>
<th>External Industry Relationships *</th>
<th>Company Name</th>
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<td>Industry funds to Emory for my research</td>
<td>None</td>
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<tr>
<td>Other</td>
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</tbody>
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## Today’s AM Schedule

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Outline

• Defining the problem(s)
• Mechanism of neurocardiogenic syncope and POTS
• Assessment pearls
• Treatment
Defining the Problem(s)

- Reflex syncope
- Neurocardiogenic syncope
- Vasovagal syncope
- Orthostatic intolerance
- Orthostasis
- Syncope
- Presyncope
Defining the Problem(s)

Syncope
- sudden, brief loss of consciousness and postural tone with spontaneous recovery
- 15% of children prior to end of adolescence*

Presyncope = Near syncope

Defining the Problem(s)

Orthostasis (Orthostatic intolerance)
-symptoms (dizzy, LH, vision change, syncope, heart racing) brought on by standing / position change

Orthostatic hypotension
-drop in BP with standing, associated with symptoms
-typically > 20-30 mmHg

Orthostatic (postural) tachycardia
-as above, but increase in HR by > 20-30 bpm
Tilt Table Record from a Patient with Orthostatic Intolerance

From Stewart, J Peds 2013
Defining the Problem(s)

Vasovagal syncope = Neurocardiogenic syncope = Reflex syncope
-syncope, mediated by a reflexive drop in BP and HR, brought on by position change or long-standing
-can also be brought on by emotional startle, neck manipulation (hair braiding), etc.
Postural Orthostatic Tachycardia Syndrome (POTS)

- Exaggerated increase in HR with position change/prolonged standing
- Associated with significant symptoms – dizziness, LH, vision change, palpitations, fatigue
- Recurrent
- Decreased quality of life
Quality of Life in Patients With Postural Tachycardia Syndrome

Lisa M. Benrud-Larson, PhD; Melanie S. Dewar, BS; Paola Sandroni, MD, PhD; Teresa A. Rummans, MD; Jennifer A. Haythornthwaite, PhD; and Phillip A. Low, MD

- N = 94
- Mean age = 34
- Physical functioning, bodily pain, general health, vitality, and social functioning were similarly poor to other chronic disabling conditions.
Healthy Person after shower

Dysautonomia Patient after shower

- exhaustion
- brain fog
- loss of vision
- tachycardia
- pre-syncope
- leg swelling
- blood pooling

red patches of skin
Vasovagal Syncope

POTS

Orthostasis
Outline

• Defining the problem(s)
• Mechanism of neurocardiogenic syncope and POTS
• Assessment pearls
• Treatment
“You were dehydrated”
How I Explain It To Patients

Sudden or Prolonged Standing

Gravity
↑ Pooling in the legs
↓ Blood return to the heart

Hair braiding
Emotional startle

Dizziness
Vision changes
Heart racing

Syncope

↓ Perfusion to the brain
Adrenaline “surge”

Brain reflexes
Reflex ↓ HR/BP

Hair braiding
Emotional startle

POTS
(Extreme, recurrent)

Supine
- restore brain perfusion
- regain consciousness

Brain reflexes
Reflex ↓ HR/BP

Dizziness
Vision changes
Heart racing

High metabolism = increased needs for fluids/salt/energy compared to adults or children – you need a lot!!!! If inadequate = symptoms
# Left Ventricular Geometry and Function Preceding Neurally Mediated Syncope


| TABLE 2. Echocardiographic Findings at Baseline and After 5 Minutes of Upright Tilt |
|---------------------------------|-----------------|-----------------|-----------------|-----------------|
|                                 | Supine          |                 |                 | Upright         |
|                                 | Negative Tilt   | Positive Tilt   | P               | Negative Tilt   | Positive Tilt   | P               |
| EDVI, mL/m²                    | 63±12           | 63±14           | NS              | 60±14           | 48±13           | 0.05            |
| ESV, mL/m²                     | 26±8            | 28±11           | NS              | 24±7            | 23±11           | 0.70            |
| EF, %                          | 68±18           | 63±18           | NS              | 66±22           | 44±13           | 0.01            |
| MWS                            | 0.31±0.06       | 0.30±0.08       | NS              | 0.32±0.06       | 0.28±0.09       | 0.35            |
| CI, mL/min per m²              | 2174±395        | 2724±900        | NS              | 2364±683        | 1964±483        | 0.14            |
| EF                             | 0.58±0.09       | 0.57±0.11       | NS              | 0.59±0.09       | 0.54±0.14       | 0.36            |
| mESS, kdynes/cm²               | 94±36           | 101±33          | NS              | 85±30           | 74±26           | 0.37            |
| cESS, kdynes/cm²               | 177±60          | 190±53          | NS              | 163±50          | 142±44          | 0.33            |
| SV, mL                         | 1697±388        | 1596±484        | NS              | 1619±456        | 1993±716        | 0.20            |
| Stress (cESS)-corrected FS, %  | 118±13          | 122±18          | NS              | 117±23          | 94±19           | 0.03            |
| Stress (cESS)-corrected MWS, % | 110±14          | 107±15          | NS              | 106±28          | 84±17           | 0.03            |

EDVI indicates end-diastolic volume indexed to body surface area; ESVI, end-systolic volume indexed to body surface area; SV, stroke volume; MWS, midwall shortening; CI, cardiac index; EF, ejection fraction; and SVR, systemic vascular resistance.
### Decreased Stroke Volume

#### Table 2: Stroke Volume at Baseline and After 5 minutes of Upright Tilt, as Assessed by Echocardiogram

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<td>Supine</td>
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<td>Stroke Volume (mL)</td>
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<td>63 ± 18</td>
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<td>66 ± 22</td>
<td>44 ± 13</td>
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Liu, et al Circulation 2000
### Decreased Stroke Volume

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<td>Negative Tilt</td>
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Liu, et al Circulation 2000

- Exaggerated venous pooling
- Inadequate CV response
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• Defining the problem(s)
• Mechanism of neurocardiogenic syncope
• Assessment pearls
• Treatment
Cardiac Syncope

Red flags
• With exercise
• Preceding palpitations
• Preceded by loud noise
• While swimming, driving
• No prodrome
• Abnormal cardiac exam
• Associated cardiac sx
• Significant family history

Cardiac causes
• Cardiomyopathy
  – HCM, DCM
• Rhythm disturbance
  – LQTS, CPVT, CHB
• Structural heart disease
  – AS, coronary anomaly
• Acquired heart disease
  – Myocarditis
• Pulmonary hypertension
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Assessment of the Syncopal Patient

• History
  – Frequency of syncope **AND** dizziness
    • Position at time of syncope
    • Prodromal symptoms
  – Fluid intake (AM routine, breakfast)
  – Salt intake
  – Preceding event (illness, surgery, concussion, growth spurt, vaccines???)
  – Neuropsychiatric assessment
  – FH and Meds
Assessment of the Syncopeal Patient

Example of Blood Pooling and Heart Rate Before and After Standing

<table>
<thead>
<tr>
<th>Sitting</th>
<th>After standing for 10 mins</th>
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</thead>
<tbody>
<tr>
<td>Pulse: 80</td>
<td>Pulse: 120</td>
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<tr>
<td>Blood Pressure: 100/72</td>
<td>Blood Pressure: 99/76</td>
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To Tilt or Not to Tilt?

- Sensitivity and specificity less than perfect
- Reproducibility of a positive test over a period of 3-7 days is ~66%
- Most useful if psychiatric cause is being considered
Outline

- Defining the problem(s)
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Treatment of Pediatric Syncope/Near Syncope

- Fluid and salt: Focus on dizziness
- Non-pharmacologic measures
- Exacerbating factors
- Meds
Fluids

• Hypovolemia makes symptoms worse
  – Decreased volume return to the heart
  – Decreased cerebral perfusion

• 2L or 64-72 fluid oz/day
  – Lionshare in the AM
  – Keep at bedside
  – Track intake at first – prove it!

• Water or Gatorade/Powerade

• Non-caffeinated

• EBM
  – Improves symptoms, improves low BP, decreases dizziness

2. Low, PA. Curr Opin Neuro, 1994
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Salt

- Salt expands blood volume
- Sodium
- 3-5 g Na/day
  – In those with recurrent dizziness
- Will not cause weight gain
- HTN?
- EBM – symptom improvement*

*Raj SR. Circulation, 2013

↑LE venous pooling
↓Ventricular filling
↓Stroke volume

Children’s Healthcare of Atlanta | Emory University
# High Salt Diet

(3000-5000 mg Na/day)

<table>
<thead>
<tr>
<th>Food Item</th>
<th>Mg of Sodium (Google)</th>
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</thead>
<tbody>
<tr>
<td>Bacon (3 slices)</td>
<td>390</td>
</tr>
<tr>
<td>Table salt (1 tsp)</td>
<td>2300</td>
</tr>
<tr>
<td>Pretzel snack</td>
<td>1400</td>
</tr>
<tr>
<td>Salted nuts (1/2 cup)</td>
<td>420</td>
</tr>
<tr>
<td>Hamburger</td>
<td>690</td>
</tr>
<tr>
<td>Enchilada</td>
<td>1300</td>
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<tr>
<td>Dill pickle</td>
<td>1430</td>
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<tr>
<td>Soy sauce (1 tbsp.)</td>
<td>870</td>
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<tr>
<td>Beef Jerky</td>
<td>420 mg</td>
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Bloody Mary Mix
~ 1500 mg Na/serving
Salt Tablets

• “Old athlete trick”
• 180 mg Na per tablet
• 2-3 tablets 1 hour prior to sports (+hydration)
• Over the counter
Focus on the dizziness

• Anticipate what makes you dizzy
  – AM routine, quick standing, periods of standing
  – How to circumvent?
• “The more you are dizzy, the more you are dizzy” \(\rightarrow\) recurrent “adrenaline surges”
• Less dizziness \(\rightarrow\) less likely dizzy with same situation 2 weeks from now. . . then less fatigue, brain fog, headaches with time
  – Lag time to improvement (can be 1-2 months)
  – Need day after day of symptom control
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Treatment of Pediatric Syncope/Near Syncope

Meds

Exacerbating factors

Non-pharmacologic measures

Fluid and salt Focus on dizziness
Counter pressure maneuvers

• Decrease symptoms by decreasing amount of LE venous pooling
  – Standing with legs crossed, sitting knee to chest, tightening leg and pelvic muscles during standing
  – “Muscle pump” and venoconstriction

• EBM
  – Tensing the leg muscles while standing → improved NIRS and MCA blood velocity (echo)*

*van Lieshout. Stroke, 2001
Compression hose

- Goal: decrease LE venous pooling
- Strength: 20-40 mmHg
- Brightlifedirect.com
- $30-50.00 per
- Best if waist high, at least knee high
- Also abdominal binders
Elevating the head of the bed
Elevating the head of the bed

- Raise HOB ~ 10-15°
- Not just pillows
  - Wedge
  - Bed risers
- Nocturnal antidiuretic effect → increased plasma volume
  - Decreased orthostasis with tilt testing*
- “Orthostatic training”

Exercise/Physical Therapy

- Avoid deconditioning
- Strong leg muscles
- Swimming, recumbent stationary bikes
- Expert recommendations*
  - 3 times a week, 20 minutes/session
- CHOA
  - Cheryl Miller, PT
  - Modified Dallas Protocol for POTS

*Grubb BP. J Cardiovasc Electrophysiol, 2006
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- Fluid and salt focus on dizziness
- Non-pharmacologic measures
- Exacerbating factors
- Meds
Medications to Avoid*

- Vasodilators
  - ACEi, CCB, beta-blockers (while still dizzy)
- TCA
- Diuretics
- Opiates
- ETOH
- Phenothiazines

*Grubb & Karas.  Pacing and Clin Electrophysiol, 1999
Iron Deficiency Anemia

- Increased incidence in adolescents with POTS$^1$
- Correcting anemia can improve orthostatic tolerance$^2$
- Labs: cbc, ferritin, +/- iron studies
- Ferrous sulfate TID
- Hormonal therapy for menorrhagia

2. Low PA. Curr Opin Neurol, 1994
Vitamin D deficiency

- Associated with orthostatic hypotension in the elderly*
- Association in children/teens has not been well-defined
- **Goal:** 25-Vitamin D > 30-40 ng/mL
- **Dose:** Vitamin D3 4000 IU daily

*Annweiler C. J Intern Med, 2014
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<td>Fitzherbert, Eugene</td>
<td>Headaches</td>
</tr>
<tr>
<td>8:30</td>
<td>Hamada, Hiro</td>
<td>Syncope after getting out of bed</td>
</tr>
<tr>
<td>8:45</td>
<td>Dunbroch, Merida</td>
<td>Sports clearance</td>
</tr>
<tr>
<td>9:00</td>
<td>McStuffins, Dottie</td>
<td>Syncope while running. Shortness of breath</td>
</tr>
<tr>
<td>9:15</td>
<td>La Bouff, Charlotte</td>
<td>Well-check</td>
</tr>
<tr>
<td>9:30</td>
<td>Von Schweetz, Vanellope</td>
<td>Diabetes</td>
</tr>
<tr>
<td>9:45</td>
<td>Calisto, Miles</td>
<td>Syncope, dizzy all the time, missing school, headaches, fatigue</td>
</tr>
<tr>
<td>10:00</td>
<td>Kion the Lion</td>
<td>Hoarse</td>
</tr>
<tr>
<td>10:30</td>
<td>Apatosaurus, Arlo</td>
<td>Well-check</td>
</tr>
<tr>
<td>10:45</td>
<td>Turtle, Crush</td>
<td>Shell issues</td>
</tr>
</tbody>
</table>
Treatment of Pediatric Syncope/Near Syncope

- Fluid and salt focus on dizziness
- Non-pharmacologic measures
- Exacerbating factors
- Meds
Fludrocortisone (Florinef)

- Mineralocorticoid steroid / aldosterone analogue
- Acts in the kidney to conserve Na and H2O, thus increasing plasma volume
  - Potassium excretion
- **Dose**: 0.1 mg qday to bid
- **SE**: abdominal bloating, low K
- Evidence
Fludrocortisone for the Prevention of Vasovagal Syncope
A Randomized, Placebo-Controlled Trial

Robert Sheldon, MD, PhD,\textsuperscript{a} Satish R. Raj, MD, MSci,\textsuperscript{b} M. Sarah Rose, PhD,\textsuperscript{a} Carlos A. Morillo, MD,\textsuperscript{c} Andrew D. Krahm, MD,\textsuperscript{d} Eduardo Medina, MD,\textsuperscript{e} Mario Talajic, MD,\textsuperscript{f} Teresa Kus, MD, PhD,\textsuperscript{g} Colette M. Seifer, MD,\textsuperscript{h} Malgorzata Lelonek, MD, PhD,\textsuperscript{i} Thomas Klingebhein, MD,\textsuperscript{j} Ratika Parkash, MD,\textsuperscript{k} Debbie Ritchie, MN,\textsuperscript{a} Maureen McRae, RN,\textsuperscript{a} for the POST 2 Investigators

- \( N = 210 \)
- Mean age = 30 years
- 1-year treatment period

\( p = 0.019 \)
Midodrine

- Alpha-1 agonist → vasoconstrictor
- Decreases blood pooling in the legs and abdomen
- **Dose:** 5-10 mg TID
- **SE:** pins/needles, abdominal cramping, itching, urinary urgency
- Avoid concomitant use of stimulants
And Beyond . . .

• SSRIs
  – Inhibit synaptic reuptake of serotonin (vasoconstrictor)
  – POTS + anxiety/depression
  – Venlaxafine: NE and serotonin reuptake inhibition

• Beta blockers
  – Don’t use too early (dizzy, low BP)
  – Can be very effective if BP is reasonable/dizziness controlled and tachycardia is main complaint (POTS)
The Deep End

- Mestinon (pyridostigmine bromide)
- Ivabradine
- Stimulants (brain fog, fatigue)
- Octreotide
- Wellbutrin
- ddAVP (dizziness)
- Celexa (cardioinhibitory syncope)
- Licorice root
Prognosis

• Teens, metabolism, and orthostasis
• Symptoms resolve in many with time
• Improvement in symptoms in almost all with adherence to therapy
• Be positive!
# Today’s AM Schedule

<table>
<thead>
<tr>
<th>Time</th>
<th>Patient</th>
<th>Complaint</th>
</tr>
</thead>
<tbody>
<tr>
<td>8:00</td>
<td>Otterton, Emmitt</td>
<td>Turned savage</td>
</tr>
<tr>
<td>8:15</td>
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<td>Kion the Lion</td>
<td>ANGRY RE: WAIT TIME</td>
</tr>
<tr>
<td>10:30</td>
<td>Apatosaurus, Arlo</td>
<td>ANGRY RE: WAIT TIME</td>
</tr>
<tr>
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<td>Turtle, Crush</td>
<td>ANGRY RE: WAIT TIME</td>
</tr>
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</table>
Sibley Syncope Clinic

- 2016 initiative
- Specifically designed for challenging pts
- **Longer time slots**
  - Initial visit: 60 min
  - Follow-up: 40 min
- Time to hear family’s concerns
- Core group
The most important thing for you to do to treat dizziness, fatigue, headaches and/or passing out is to concentrate on NEVER getting dizzy. When you get dizzy, your body has to compensate in order to provide adequate blood flow to your brain. It is this compensation that leads to the fatigue and headaches. Once you get control of the dizziness, it may be as long as 2 months before your energy level begins to come back.

Here are some ways to stop getting dizzy:

- Increase fluid intake to 64-72 fluid oz/day. At least. Drink a large share of this in the morning. Drink immediately when getting out of bed (keep a thermos on the nightstand). If you cannot sit up without getting dizzy, drink cold water from a thermos cup using a straw while you are lying down. This will constrict the blood flow to your stomach and allow more blood to be available for your brain.

- Drinking fluids until the urine is clear and colorless is one way to know that hydration is assured. If your urine is yellow, you are not drinking enough. If you need an excuse for bathroom breaks at school, please call us.

- Avoid rapidly changing position and instead do this gradually. Decreasing the number of dizzy episodes tends to lead to even less dizziness with time.

- Increase salt in your diet, especially at breakfast. A diet that leads to at least 3 g of sodium intake per day is a good place to start. You can continue to add more and more salt to your diet until you stop getting dizzy. Salt does not have calories, and will NOT make you gain weight. You can also take Thermotabs, which are salt tablets available over the counter as a salt source. Some patients take as many as twenty a day, but start with one each meal. You will know you have done enough when you can change position without getting dizzy.

- Avoid caffeine as much as possible. Caffeine makes you urinate, and this can worsen your dizziness.

- Counter-pressure maneuvers have been shown to help lessen dizziness. These include making arm muscles (bicep contractions), leg crossing, and leg crossing with lower body muscle tensing. These were discussed today.

- Elevating the head of the bed and sleeping at an elevation of ~ 10 degrees can help to retrain mechanism involved and decrease symptoms.
Summary

Focus on Dizziness

Vasovagal Syncope

POTS Orthostasis

How I Explain It To Patients

- Gravity
  - Pooling in the legs
  - Blood return to the heart
- Hair braiding
  - Emotional startle
- Brain reflexes
  - Reflex HR/BP
- Supine
  - Restore brain perfusion
  - Regain consciousness

Dizziness
- Vision changes
- Heart racing

POTS (Extreme, recurrent)

Syncope

Sibley Syncope Clinic

- 2016 initiative
- Specifically designed for challenging pts
- Longer time slots
  - Initial visit: 60 min
  - Follow-up: 40 min
- Time to hear family’s concerns
- Core group of cardiologists

Chin up!
Thank you!

Mike McConnell

Heather Phelps
Treatment of Pediatric Syncope/Near Syncope

- Fluid and salt
- Non-pharmacologic measures
- Exacerbating factors
- Meds

Children’s Healthcare of Atlanta | Emory University
• Simplified mechanism
• “Dehydrate” and AM passing out or routine dizziness
• Urine color and daily dizziness as checks
• QOL
Standing (head above heart)

↑ LE venous pooling
↓ Ventricular filling
↓ Stroke volume

Vasoconstriction
Mild ↑ heart rate

No/minimal symptoms

Blood evenly distributed in veins

Blood pools in leg veins
Standing (head above heart)

↑ LE venous pooling
↓ Ventricular filling
↓ Stroke volume

Inadequate

↑↑ Heart rate
↓ Cerebral perfusion

Prodrome (Dizzy, vision change, etc)

POTS (Extreme, recurrent)

SNS hyperactivity
LE excess venous pooling
Standing (head above heart)

↑LE venous pooling
↓Ventricular filling
↓Stroke volume

Inadequate

Heart rate
↓Cerebral perfusion

Prodrome
(Dizzy, vision change, etc)

SNS hyperactivity
LE excess venous pooling

Reflex

Carotid sinus
LV C-fibers

POTS
(Extreme, recurrent)
Children's Healthcare of Atlanta | Emory University

Standing (head above heart)

↑LE venous pooling
↓Ventricular filling
↓Stroke volume

Inadequate

SNS hyperactivity
LE excess venous pooling

↑↑Heart rate
↓Cerebral perfusion

Prodrome (Dizzy, vision change, etc)

Brainstem reflexes
- Vagal response
- Sympathetic withdrawl

Supine
- restoration of cerebral perfusion
- restore consciousness

Carotid sinus LV C-fibers

Vasodilation/decrease BP
+/− Decrease HR
↓↓↓Cerebral perfusion
Prodrome (dizzy)

POTS
(Extreme, recurrent)

Syncope

Reflex
Children’s Healthcare of Atlanta | Emory University

**Brainstem reflexes**
- Vagal response
- Sympathetic withdrawal

**Hair braiding**
- Micturition
- Emotional startle

**Carotid sinus LV C-fibers**
- Vasodilation/decrease BP
- +/- Decrease HR
- ↓↓↓Cerebral perfusion
- Prodrome (dizzy)

**Syncope**
- Supine
  - restoration of cerebral perfusion
  - restore consciousness
Hill’s Criteria of Causation

- Temporal relationship
- Dose-response
- Consistency
- Plausibility
- Strength
- Consideration of alternative explanations