Diagnosis and Management of Dysautonomia in the Pediatric Population

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Disclosures
Relevant Disclosures: None.
Irrelevant Disclosures: Too Many to List....

On Discovery...

“What is wanted is not the will to believe, but the will to find out, which is the exact opposite.”

Bertrand Russell
1872-1970

My Goals for Giving This Talk

• Pass on personal knowledge/anecdotes
• Stimulate interest in research in the area
• Convince you I am not totally crazy
  ... Only Partially

Considerations

• Dysautonomia is both acknowledged and controversial
• Dysautonomia has been poorly studied
• Dysautonomia is one of the more common conditions you will confront in the late child and adolescent age group

Where We Are Going

• Explore the Normal Physiology of the Autonomic Nervous System & Possible Pathophysiology
• Describe Age-Specific Autonomic Conditions
  – Clinical Presentation
  – Diagnostic Criteria/Evaluation
  – Treatment Options
• Prognostic Thoughts
Autonomic Nervous Physiology

- A part of the “primitive” central/peripheral nervous system
- Responsible for regulating subconscious homeostatic processes within the body and making them response to both internal and external stimuli

Primary Effects

Secondary Effects
- Cerebral Autoregulation
- Digestive Control
- Thermal Autoregulation

Tertiary Effects
- Fatigue/Sleep
- Decreased Sense of Well Being

Evidence in Favor of Developmental Maturation

- Gestational age correlates with HR variability
  - Longin E, et al 2006
- HR variability correlates with sleep parameters and gestational age
  - Clairambault J, et al 1992

Maturation is a process of increased parasympathetic control
- Eyre EL, et al 2014

Disorders in other organ systems (such as OSA) can affect sleep and HR variability through enhanced sympathetic activity

Framework for Autonomic (Cardiovascular) Disorders

<table>
<thead>
<tr>
<th></th>
<th>Persistent Dysautonomia</th>
<th>Episodic Dysautonomia</th>
</tr>
</thead>
<tbody>
<tr>
<td>Blood Pressure</td>
<td>Orthostatic Hypotension</td>
<td>Vasovagal Syncope</td>
</tr>
<tr>
<td>Heart Rate</td>
<td>Postural Orthostatic Tachycardia Syndrome</td>
<td>Cardioinhibitory Syncope*</td>
</tr>
<tr>
<td>Both</td>
<td>Mixed</td>
<td>Mixed</td>
</tr>
</tbody>
</table>

* Known as “Pallid Infantile Syncope” in the newborn
Episodic Dysautonomia

Seizures and Syncope

Pallid Infantile Findings

Infantile Correlates

Pallid versus Breatholding Syncope

Toddler/School-Age Presentations

<table>
<thead>
<tr>
<th></th>
<th>Pallid Syncope</th>
<th>Cyanotic Syncope</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Age of Onset</strong></td>
<td>2-12 mo</td>
<td>6-48 mo</td>
</tr>
<tr>
<td><strong>Appearance</strong></td>
<td>White</td>
<td>Blue</td>
</tr>
<tr>
<td><strong>Precipitated by Crying</strong></td>
<td>Occasional</td>
<td>Frequent</td>
</tr>
<tr>
<td><strong>Tonic-Clonic Activity</strong></td>
<td>Frequent</td>
<td>Common</td>
</tr>
<tr>
<td><strong>Mechanism</strong></td>
<td>Asystole</td>
<td>Hypoxia/Bradycardia</td>
</tr>
<tr>
<td><strong>Treatment</strong></td>
<td>Pacing</td>
<td>Reassurance</td>
</tr>
<tr>
<td></td>
<td>? Reflux Treatment</td>
<td>? Iron</td>
</tr>
<tr>
<td></td>
<td>? Stimulants</td>
<td>? Stimulants</td>
</tr>
<tr>
<td><strong>Prognosis</strong></td>
<td>Resolution by 6 yrs age</td>
<td>Resolution by 4-6 yrs age</td>
</tr>
</tbody>
</table>
Toddler/School-Age Presentations

- Episodic Dysautonomia
  - Syncope/Convulsions
  - Pre-syncope (Pallor, Inattention)
  - Activity Avoidance
  - School Performance Issues
- Persistent Dysautonomia (?)

Pediatric/Adolescent Triggers

- Persistent Dysautonomia (?)

Adolescent Presentations

Persistent Dysautonomia

<table>
<thead>
<tr>
<th>Onset Characteristic</th>
<th>&quot;Infectious Form&quot;</th>
<th>&quot;Developmental Form&quot;</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age of Onset</td>
<td>12-21 years</td>
<td>4-16 years</td>
</tr>
<tr>
<td>Initial Symptoms</td>
<td>Fatigue, Pre-Syncope</td>
<td>Non-specific complaints &quot;Stomach Aches&quot;</td>
</tr>
</tbody>
</table>

Associated Disorders/Syndromes

- Connective Tissue Disorders
  - Ehler-Danlos syndrome
  - Loey-Dietz syndrome
  - Marfan syndrome
- Mitochondrial Disorders
  - Riley-Day syndrome
- Medications
  - High-Dose Beta-Blockade
- Co-Morbid Disease
  - Diabetes Mellitus

Diagnostic Approach to Dysautonomia

- Historical Considerations
  - The cornerstone of the evaluation
  - Review of all prescribed and alternative medications
  - Head-to-toe Review of Systems

Diagnostic Approach to Dysautonomia

<table>
<thead>
<tr>
<th>System</th>
<th>Symptoms</th>
</tr>
</thead>
<tbody>
<tr>
<td>Neurologic</td>
<td>Headaches, Fatigue, Sleep disturbance, Loss of consciousness, Pain</td>
</tr>
<tr>
<td>Ophthalmologic</td>
<td>Vision changes, &quot;bags&quot;</td>
</tr>
<tr>
<td>Cardiovascular</td>
<td>Chest pain, palpitations, near syncope, syncope</td>
</tr>
<tr>
<td>Gastrointestinal</td>
<td>Nausea, emesis, constipation, diarrhea, abdominal pain</td>
</tr>
<tr>
<td>Dermatologic</td>
<td>Venous stasis, Thermal dysregulation</td>
</tr>
</tbody>
</table>
**Physical Examination**

**Diagnostic Approach to Dysautonomia**

- History & Physical Examination
  - Electrocardiogram
  - Selected Bloodwork (CBC, Electrolytes, TFTs)
  - Echocardiogram
  - Tilt Table Testing
  - Ambulatory Monitoring

**Examples of Significant Findings**

**Tilt Table Testing**

**Examples of Tilt Table Findings**

**Examples of Tilt Table Findings**
Examples of Tilt Table Findings

Examples of Tilt Table Findings

Treatment Options

- Stimulus Avoidance
- Physical Conditioning
- “Hyperhydration”
- Pharmacologic Treatment
- Device Therapy
- Psychological Counseling

Treatment Approaches

Physical Conditioning

Cardiac Origins of the Postural Orthostatic Tachycardia Syndrome

Fu, et al. JACC 2010;55(25):2858
Treatment Approaches

“Hyperhydration”

- “Dehydrated” versus “Underhydrated”
- Primacy of salt
  - Minimum goal 3-6 g
- Water that follows
  - 8-10 8 ounce glasses of non-caffeinated beverages

Treatment Approaches

Pharmacological Options

Psychological Counseling

Mayo Clinic Publication, 2014

Prognostic Thoughts

- Any pathophysiologic condition that is difficult to “objectify” is difficult to treat
- Being a “cheerleader” is also more difficult than providing a discrete therapy
- The line between “trying to help” and “doing harm” can be very grey…
Where We’ve Been

- Explore the Normal Physiology of the Autonomic Nervous System & Possible Pathophysiology
- Describe Age-Specific Autonomic Conditions
  - Clinical Presentation
  - Diagnostic Criteria/Evaluation
  - Treatment Options
- Prognostic Thoughts

The 10%ers...

“The New England Journal of Medicine reports that 9 out of 10 doctors agree that 1 out of 10 doctors is an idiot.”

Jay Leno

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