Co-Managing The Adolescent with Orthostatic Intolerance

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Background

• Postural orthostatic tachycardia syndrome (POTS) is a multi-system dysfunction disorder that primarily affects those organ systems with significant autonomic input.

• Organ systems affected include the central nervous, cardiovascular, gastrointestinal, cutaneous and muscular-skeletal systems.

• Patient symptoms may be limited to just a single organ system, but more commonly are complex with a wide distribution throughout multiple body regions.
Background

• POTS causes significant disability, limiting physical activity (sports participation) and school attendance in adolescent patients, or the ability to work a full day when employed in adults patients.

• POTS is not a rare disease and is estimated to affect between 500,000 to 1 million Americans.

• Age at presentation 14-45 years.

• Female preponderance – 4-5/1.
Background

- POTS patients may present to the cardiologist, neurologist or gastroenterologist. The mode of presentation depends on the predominant symptom complex.

- While seemingly different because of the primary mode of clinical presentation, these patients ultimately share many common features and overlap of symptoms.
Terminology

- Grinch syndrome
- Chronic orthostatic intolerance
- Orthostatic tachycardia
- Sympathotonic orthostatic hypotension
- Hyperdynamic beta adrenergic state
- Idiopathic hypovolemia
- Mitral valve prolapse syndrome
- Neurocirculatory asthenia
- Irritable heart
- Soldier's heart
- Effort syndrome
Clinical Presentation

• Dizziness, lightheadedness, weakness, blurred vision and fatigue upon standing.
• Orthostatic symptoms of palpitations, tremulousness and anxiety.
• Gastrointestinal symptoms – nausea, abdominal cramps, early satiety, bloating, constipation or diarrhea.
• Peripheral extremities – acro-cyanosis and edema when upright.
Clinical Presentation

• Intractable headaches, that are medication resistant.
• Abnormal sleep – insomnia, and frequent awakening during sleep.
• Mood disorder – panic disorder, anxiety and depression.
• Exercise intolerance.
• Cognitive dysfunction – mental “fog”, ADD
• Syncope is relatively unusual.
  – Occurs in up to 40%
• Symptoms may appear abruptly after a viral infection, surgery or trauma (head concussion). Others experience a more insidious onset.
Clinical Presentation

• Severity of symptoms quite variable – mild to profoundly incapacitating.
• We follow many children unable to attend school on a regular basis (Home-Hospital Study Program).
• Symptoms can be worsened by menstrual cycle and relative dehydration.
• The course of the disorder may be self-limited or follow a relapsing remitting course over several years.
  – Majority of children improve by young adult life
Diagnosis

• Tilt Table Testing or Bedside Standing Test
  
  – Exaggerated increase in HR usually without associated hypotension
  
  – Greater than 40 bpm or an increase to ≥130 bpm for ages 13 years and younger and to ≥ 120 bpm for ages 14 years and older within the first 10 minutes of being upright
  
  – Development of typical clinical symptoms when upright
  
  – Resolution of tachycardia and clinical symptoms when placed supine
Secondary POTS

- Anemia – Iron deficiency
- Vitamin B12 deficiency
- Bedrest - Deconditioning
- Side effect of Medications – diuretics, vasodilators
- Dehydration / Moderate weight loss
- Endocrinologic Abnormalities
  – Hyperthyroidism
  – Adrenal insufficiency
  – Hypoparathyroidism
  – Hypoaldosteronism
- Diabetes Mellitus
- Connective Tissue Disease - Sjögren's syndrome, systemic lupus erythematosus
- Mitochondrial Disease
Associated Disorders

- Chronic fatigue syndrome
- Mitral valve prolapse syndrome
- Mast cell activation abnormalities – episodes of flushing associated with increases in urinary methylhistamine, and elevated plasma histamine and tryptase levels
- Ehlers-Danlos syndrome
- Joint Hypermobility Disorder
Multi-Disciplinary Approach

• **CADE Clinic – Clinic for Autonomic Disorders Evaluation**
  – Cardiology
  – Pain Medicine
  – Psychology
  – Sleep Medicine
  – Diabetes
  – Other Subspecialty Components Available
    • Gastroenterology
    • Neurology
    • Physical Therapy – Cardiac Rehabilitation

• **Primary HealthCare Provider**
  – Continue your involvement with the medical – psychosocial care of your patient
  – Leverage your long-term relationship with the patient and family to help in the “road to recovery”
  – Help your patient to identify needed resources in the local community
  – Help patient and family overcome high level of non-compliance and inertia to enact change
Non-Pharmacologic Treatment

- **Non-pharmacologic Rx** maybe as useful or more so than pharmacologic Rx

- Avoid exacerbating factors:
  - Medications - Vasodilators or Sympathomimetic drugs
  - Dehydration
  - Menstrual cycle (modify menstrual cycle w Long acting BCPs)
  - Sudden changes in posture – arise slowly and in stages
    - Supine, seated and then standing
  - Inactivity and/or prolonged recumbency
  - Avoid prolonged standing and walking in hot weather
  - High environmental temperatures (showers)
  - Large meals
Non-Pharmacologic Treatment

• Lower extremity support stockings - + 25 mm Hg, custom fit, and ankle to waist height

• Raise head of bed 10-30 degrees

• Exercise – aerobic exercise with lower extremity muscle strengthening regimen.
  o See CNHS recommend muscle strengthening and aerobic exercise program
Non-Pharmacologic Therapy

• Sleep hygiene
  • Possible sleep study if no improvement
• Cognitive behavioral therapy for pain and symptom management
  • Coping skills - distraction and positive self-talk
  • Stress management
  • Wellness / Mindfulness instruction
  • Activity pacing
  • Help family identify local resources
• Operant learning strategies
  • POTS is a “Familial Disorder”, not just the teenager!
  • Elimination of pain behaviors and parent behaviors that may serve as secondary reinforcement for pain behavior
• Resume academic activity – going back to school, may need to be gradual and incremental.
  • Try to avoid the patient initiating the cycle of not going to school
Non-Pharmacologic Therapy

- Identification and treatment of co-morbid psychiatric illnesses - depression and anxiety disorders
- Training in relaxation strategies is a core treatment element in many pain management programs
Non-Pharmacologic Therapy

Mindfulness-Based Stress Reduction (in-person) group (n=10)
6 weekly sessions, conducted by Vicki Freedenberg, RN and Robin Fabian

- Intervention consisted of meditation, yoga, group support, group discussions about stressors
  - 66% (4/6) of those not in school pre-intervention went back to school at least part-time
  - 60% said meditation helped decrease their stress level
  - 60% liked yoga
  - 90% said meeting others with POTS helped them deal with their issues better, and didn’t feel “alone” anymore
  - 10% felt physically worse after yoga
  - 10% found nothing helpful about the program
Non-Pharmacologic Therapy

Pain Medicine – Teen Treatment and Education Series

• GOALS of Program:
  • Meet other teens with POTS
  • Discuss challenges related to Sx’s of POTS
  • Develop strategies for conquering limitations imposed by POTS Sx’s
  • Make positive decisions to begin living a normal life
  • For parents to meet other parents and discuss common issues surrounding the management of POTS
Treatment of Low Plasma volume

- High fluid intake (avoid caffeine)
  - 64-80 oz fluid intake, without caffeine
- High salt diet or Salt supplementation
  - Salt Tablets, ThermoTabs or Sodium Chloride Salt Stick capsules
  - 1-2 gms NaCl po bid
  - 24 hour urine Sodium > 170 mmole/day
- Fludrocortisone (0.1-0.4 mg/day)
  - Monitor for development of edema, hypertension, hypokalemia, mood changes and hirsutism
  - Encourage diet with high potassium intake
  - Monitor BP every other day for 2-4 weeks
- Acute ingestion of 16 oz cold water in less than 5 minutes
- Intravenous fluids (500-2000 ml, acutely and daily), if necessary
Pharmacologic Treatment

• Alpha adrenoreceptor agonists
  – Midodrine (2.5 -10 mg 3-4X/day q 4 hours)
  – Pseudoephedrine 30-60 mg every 4-6 hours
  – Methylphenidate 5-30 mg po bid q 4 hours
  – Monitor BP

• Acetylcholinesterase inhibitor
  – Pyridostigmine 30-60 mg tid

• Beta-Blockers
  – Propranolol 10 - 30 mg 3x/day, Propranolol LA 60 mg/day
  – Low dose better than high dose
Pharmacologic Treatment

- Selective Serotonin Reuptake Inhibitors
  - Prozac - fluoxetine
  - Effexor - venlafaxine
- Erythropoietin
- Non-steroidal Inflammatory Drugs
- DDAVP
- Octreotide - SC/IV
- Clonidine – may help with sleep dysfunction
- IVIG (Maybe helpful in those with autonomic / neuronal antibodies or diffuse autonomic dysfunction)
Treatments Rarely Used

- Sinus node ablation for persistent sinus tachycardia
  - May result in need for pacemaker implantation
- Phenobarbital
- Metyldopa