

# 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  $\geq 130$  bpm for ages 13 years and younger and to  $\geq 120$  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 B<sub>12</sub> 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.
  - 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
- Methyldopa