Co-Managing POTS Patients: Interdisciplinary Specialist and Primary Care Treatment

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Disclosures

• Nothing to Disclose.
POTENTIAL POSTURAL ORTHOSTATIC TACHYCARDIA SYNDROME

- It is not a disease
- It has no specific diagnostic test
- It has no specific treatment
Postural Orthostatic Tachycardia Syndrome (POTS)

POTS is

• A real set of symptoms which have the potential to profoundly impact functioning and quality of life
• The symptoms come from a variety of causes which might help direct treatment
• Treatment is directed at improving as many symptoms resulting in as few side effects as possible
• The most important therapies are non pharmacologic
• THE END...
POTS Definition

• “...sustained heart rate increment of ≥30 beats/min within 10 min of standing or head-uptilt in the absence of orthostatic hypotension.
• The standing heart rate for all subjects is often ≥120 beats/min.
• These criteria may not be applicable for individuals with low resting heart rates.
• For individuals aged 12-19 years, the required increment is at least 40 beats/min
• The orthostatic tachycardia may be accompanied by symptoms of cerebral hypoperfusion and autonomic overactivity that are relieved by recumbency.”

- 2011 Consensus Statement
# POTS: Office Screen

**Table 1.**

**Postural Tachycardia Syndrome Screening Test**

<table>
<thead>
<tr>
<th>Protocol</th>
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<tbody>
<tr>
<td>Record baseline heart rate and blood pressure while patient is recumbent ≥10 minutes</td>
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<td>Ask patient to stand in place calmly for 10 minutes</td>
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<td>Record heart rate and blood pressure every 2 minutes or monitor continuously</td>
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<td>Encourage reporting of symptoms</td>
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<td>Recommend sitting if fainting seems imminent</td>
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Consider the diagnosis of POTS when all of the following are present:

- Sustained rise in heart rate of 40 bpm or absolute heart rate of 120 bpm
- Orthostatic symptoms correspond with a rise in heart rate and resolve with recumbency
- Blood pressure does not drop ≥20 mm Hg systolic or ≥10 mm Hg diastolic
- Patient has longstanding, day-to-day symptoms
- Medicines that can alter hemodynamics were held at least five half-lives prior to testing
- No signs of anemia, acute dehydration, or hyperventilation (while standing)

Source:
POTS

- POTS is a symptom complex rather than a disease entity itself, with underlying heterogeneous pathophysiologies such as:
  - Neuropathic
  - Mast Cell activation
  - Autoimmune/Immunologic
  - Mitochondrial disease
  - Post-traumatic (concussion)

- Maybe it’s “all of the above”
  - In the future, it could be broken down into individual diagnoses, each with diagnostic criteria and treatment
Conceptualizing POTS

- Genetic
  - TPSAB1, TRAP

- Post-infectious
  - Anti-NMDA, FIRES, Rheumatic fever

- Metabolic / Mitochondrial

- Concussion

- Functional brain disorder
  - Autism
  - Seizures (Rolandic epilepsy)
  - Anxiety / Depression
What makes POTS POTS?

- **Orthostatic Intolerance**
  - HR, BP
- Dizziness / Syncope
- Fatigue
- “Brain Fog” and cognitive difficulties
  - Executive function problems
  - Memory problems
- Migraines / headaches

- Sweating
- Venous Pooling / Mottling
- Tremors
- Stomach pain / Nausea
- Physical Deconditioning
- Confused with worsening OI and fatigue
- Anxiety / Depression
- Somatic hypervigilance
Autonomic Nervous System

- Heartbeat
- Blood flow
- Breathing
- Skin temperature
- Digestion

- Autonomic dysfunction can cause blood to pool in the body and circulation to slow down:
  - Blood pooling in limbs upon standing causes tachycardia to try to get the blood back to the heart (dizziness and tachycardia)
  - After eating, not enough blood flow to digestive organs to stimulate digestion (nausea)
  - Too much blood flow to intestines when they haven’t eaten (stomach cramping)
Autonomic Nervous System

- ENS (Enteric NS)
  - ACh, SAS, 5HT, DA, VIP
- PNS (Parasympathetic Cholinergic NS)
  - ACh
- SNS (Sympathetic Noradrenergic Syst)
  - Purines, NE
- SAS (Sympathetic Adrenergic Syst)
  - Epi
- SCS (Sympathetic Cholinergic Syst)
  - ACh
Structural vs. Functional Problems

**Structural**: a well-defined change in ANS structure produces disease - examples:
- Multiple system atrophy
- Diabetic autonomic neuropathy
- Baroreflex failure due to neck radiation

Hardware -> Solution is to fix hardware, replace parts (mostly we don’t have them – factory is closed)

**Functional**: a change in ANS function is involved in disease production, but is (1) less well-defined; (2) a link in a pathogenic chain, not primary:
- Postural tachycardia syndrome
- Irritable bowel syndrome
- Syncope

Software -> Solution is to reprogram neural networks

“Functional” NOT “non-organic”
POTS Co-Morbidities

- Chronic fatigue
- Headache / Migraines
- Hypermobility (including Ehlers-Danlos)
- MALS
- Chronic vomiting or nausea
- Constipation or Irritable Bowel Syndrome
- Fibromyalgia
- Pelvic pain and interstitial cystitis
- Depression, Anxiety, PTSD, ADHD
- High achievement
POTS: Headaches

• Significant overlap with patients with Migraines, New Daily Persistent Headache, and Chronic Daily Headache
  • Headache type did not reliably predict POTS diagnosis in a group of adolescent patients (Heyer, 2013)

• May also mimic symptoms of a low-pressure headache/intracranial hypotension
  • Post-LP
  • CSF leak is rare in adolescents
  • Imaging with CSF leak: pachymeningeal enhancement, brain sag, subdural fluid collections
  • Low CSF pressures

• Treatment approach is similar
  • Lifestyle modifications: sleep, nutrition, hydration, exercise
  • Behavioral and coping strategies
  • Medications when indicated (abortives/preventives)
  • **Counseling on Medication Overuse—high risk population**
POTS Mimics…or are they? It’s complicated!

- Prolonged bedrest
- Hypovolemia/Dehydration
- Anemia
- Syncope
- Medications
- Anxiety/Hyperventilation
- Thyroid disease
POTS: Treatment

- Knee high stockings
  - Ankle/Waist 25mm/Hg
  - Other compression garments
- Abdominal binder
- Spansks compression garments
- 2-3 liters water daily, IV saline (non-caffeinated beverages)
- 3-5 teaspoons salt daily, salt tablets
- Exercise
- CBT

- Pyridostigmine
- Midodrine, Clonidine
- Propranolol
- Fludrocortisone
- DDAVP
- SSRIs
- Methylphenidate
- Other: IVIG
  - Case-based
POTS: Pharmacotherapy

- Alpha-adrenergic agonist (i.e. Midodrine)
- Beta blockers (i.e. Propranolol)
  - may increase fatigue/exercise intolerance
- Cholinesterase inhibitors
- Fludrocortisone
  - may increase headache/migraine symptoms and vertigo
- Amitriptyline (to treat depression, insomnia, and/or headache)
  - may exacerbate tachycardia
- Amphetamines (to treat attention, fatigue)
  - may exacerbate tachycardia
Integrative Approach

- Physicians (general)
  - Specialists
- Nurses
- Psychology
  - Psychiatry
- Physical Therapy

- Parents
- School
- Community
Treatment: PT for POTS

• Fatigue and exercise intolerance
• Physical activity beneficial
• Physical therapist or personal trainer

• Levine Protocol: Exercise guidelines
  • Endurance training- recumbent or semi recumbent position (recumbent bike, rowing machine, aquatic therapy), gradually work up to upright position (Goal 4-5 x/week)
  • Lower extremity strength training (1-2 x/week) 

*Fu and Levine, 2015*
Rehabilitation Approach to POTS

- **Goal is to focus on INCREASING FUNCTIONING**
  - Results in a decrease in POTS-related symptoms

- Increase activity level
  - Daily schedule/routine
  - Attend school daily
  - Gradually increase exercise
    - May require Physical Therapy referral & treatment
    - Goal: reconditioning
  - Avoid laying down during waking hours

- In general, recovery is faster for patients with less functional disability
Lifestyle Recommendations

• Behavioral modifications across several key areas can greatly reduce POTS symptoms & associated functional disability
  • Sleep Hygiene (see next slides)
  • Increase hydration
    • Exact recommendations based on weight
    • Typically 80-100oz water/day
    • Avoid caffeine
    • Increased salt in diet or add salt tabs
    • Eating: keep a regular schedule, eat frequent snacks

• Note: Making changes to routines/behaviors is difficult –
  • Some patients will require CBT support to make changes
Cognitive Behavioral Therapy (CBT): Overview

- Co-occurring anxiety/depression – exacerbate POTS symptoms
- Psychoeducation about the relationship among thoughts, emotions, and behaviors
  - Impact of stress on pain; impact of pain on stress
  - Prevention of depression and avoiding anxiety cycles
- Understanding physiological responses, identify emotions, and individual stress responses
- Thoughts/behaviors/somatic precursors of pain exacerbations
  - Identification of “Negative/Unhelpful Thoughts”
  - Thought stopping and Challenging Negative thoughts
    - “I will never make it through the school day with this dizziness”
Cognitive Behavioral Therapy (CBT): Components

- CBT for POTS may include:
  - Behavioral Activation
  - Relaxation training
    - Diaphragmatic breathing, guided imagery, progressive muscle relaxation
    - Active Distraction
  - Identifying/challenging negative/anxious thoughts
  - Biofeedback
  - Mindfulness
  - Acceptance and Commitment Therapy
  - Problem solving implementing lifestyle strategies
  - Stress management
  - Activity Pacing
  - Create school reintegration plans, coordinate with school
Common Patient Characteristics

• Anxiety
  • Pre-existing, related to POTS symptoms, related to functional impairment

• Depression
  • Pre-existing, frustration/irritability related to symptoms (dizziness, fatigue) and functional limitations
  • Loss of identity/social network if not attending school or stopped sports/activities

• Dysregulated sleep
  • Shifted weekday/weekend schedule, frequent naps, sleeping too much or too little, difficulty falling asleep
  • Spending long periods of time in bed awake
Lifestyle Recommendations: Sleep Habits

• **Sleep Hygiene**
  • Behaviors that help to improve sleep quality
  • Examples:
    • Bed reserved only for sleep
    • Maintain consistent sleep/wake schedule (vary no more than 2 hours)
    • Avoid screens 1 hour prior to bed
    • Avoid naps
    • Avoid time laying down during awake hours
    • Increase daytime activity/exercise
    • Avoid excessive caffeine or caffeine late in the day
    • Set schedule for gradually shifting sleep patterns
      • Shifting wake up time 15-30 minutes earlier each morning
      • Shifting bed time by 15-30 minutes earlier each evening
Difficulty with Sleep Changes

• If making change is difficult, refer to CBT psychologist to:
  • Identify barriers to changing sleep habits
  • Problem solve new strategies
  • Behavioral Therapy for Insomnia if needed

• Consider Sleep Study referral for:
  • Patients who implement sleep hygiene changes and continue to have difficulty with limited or excessive sleep AND endorse any of the following:
    • Family history of sleep disorders
    • Restless sleep
    • Snoring
Strategies for Working with Parents

• The symptoms are real
• POTS is not life threatening
• Do not ask about pain/symptoms
  • Focusing on pain can increase pain perception
    • No “status checks” for patients with chronic pain
  • Parents to help child focus on functioning and coping strategies
    • “How can I help you feel more comfortable?”
  • Distraction: walk, talk to a friend, coloring books, play with a pet, bake
  • Limit worry/ “venting” time
    • Catastrophic thinking, worst-case scenario, focus on symptoms
    • Negative/worry thoughts lead to increased arousal & increased focus on symptoms
Strategies for Working with Parents

• Identify potential secondary gains
  • A day with no activity or missed school should not be reinforced with pleasurable activities/rewards.
  • Missed school = no TV/video games
  • Continue to require homework completion

• Importance of attending school and normal responsibilities (chores)

• Moderation and activity pacing
  • Encourage child to take breaks both at school & during activities

• School: Request 504 Plan Meeting (NOT homebound)
  • Accommodations: take short breaks, access to water/snacks, etc.
Missing School

• Pain is not a reason to miss school
• Orthostatic dizziness is not a reason to miss school
  • Frequent LOC *may be* a reason to miss school
• Reasons to miss school:
  • Contagious
  • Danger to self or others
  • Rest and/or recovery are a necessary part of the therapy
• ?? Possible Reason to Miss: Safety/liability inhibits good judgment sound practice
School Recommendations

• **Daily school attendance is important**
• POTS Sx are not reasons to miss school
• Frequent LOC *may be* a reason to miss school

• Golden Rules to miss school:
  • Communicative disease/Contagious
  • **Fever of 102 or higher**
  • Danger to self or others (instead should be seen in ER)
  • Profuse bleeding or other broken bone
  • Hospitalized
  • Death of a family member/friend
School Recommendations

• Schedule a meeting with appropriate teachers and advisor/counselor.
• Explain POTS
• Work out a plan that is best for the patient and which takes into account how the patient is feeling and how the school can accommodate their needs.
• Some people may need to have some partial home schooling (attend school part-time), then gradually increase school time, depending on how they are doing.
  • If possible, do any “homebound” schooling/teaching outside of the house.
School Recommendations: 504 Plan

- Flash Pass
- Access to hydration & nutrition
- Extra time for assignments, exams, & projects
  - ONLY while catching up on missed work/after missing several consecutive days/weeks (limited time)
- Modify schedule and start classes later to increase time spent in school
- More travel time between classes
- Reduce class load (if you can)
- Find your best time of the day and try to modify your schedule to get the most of it

Note: 504 Plan can be beneficial; can also be counter-productive
School Recommendations

- Communicate, Communicate, Communicate
  - Ideal if patient can advocate without their parents!

- Patient recommendation: Speak assertively so people understand your feelings and what you need.

- Schedule a meeting with appropriate teachers and advisor/counselor

- Be prepared to show documentation from doctors
School Recommendations

• Homebound/Home & Hospital is generally NOT recommended

• Leads to decreased functioning
  • May require BRIEF partial Homebound if patient has been out of school for an extended period of time
    • Attend school daily for shortened time, while following gradual return to school plan
    • Ideally, do “home tutoring” outside of the house
    • Gradually increase time at school
Referrals to Behavioral Medicine

• **Framing it is Key**
  • Focused on teaching skills:strategies that can reduce the symptoms
  • Skills are relevant to help everyone deal with stress and anxiety/frustration that are inevitable

• **Tips for referring patients to a psychologist for CBT**
  • Contact insurance company for list of providers
  • “Cognitive Behavioral Therapy”
  • Psychologist with training in Behavioral Medicine or pediatric psychology
Referrals to Behavioral Medicine

• For more complex patients or patients with more severe functional disability, refer to Children’s National for interdisciplinary team evaluation:

  • Chronic Pain Clinic – 202-476-6765
Resources for Parents/Youth

• Dysautonomia International Website
  • http://www.dysautonomiainternational.org/
  • They also have a yearly conference for patients and families

• Dysautonomia Youth Network America
  • http://www.DynaKids.com

• Mayo Clinic – Teen Dysautonomia Handout
  • https://med.uth.edu/pediatrics/files/2014/02/MAYO_Clinic_Teens_Dysautonomia_copy.pdf
Resources for Pediatricians

Take Home Messages

• Increase activity, minimize time laying down

• Attend school daily

• DO NOT SIGN THE SCHOOL FORMS without consulting with co-treating providers
QUESTIONS?

Thank you for your attention today!