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

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Disclosures

- Nothing to Disclose.

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POSTURAL
ORTHOSTATIC
TACHYCARDIA
SYNDROME

- It is not a disease
- It has no specific diagnostic test
- It has no specific treatment

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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…

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POTS Definition

- "…sustained heart rate increment of $\geq$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 $\geq$120 beats/min.
- These criteria may not be applicable for individuals with low resting heart rates.
- For individals 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[a]

Standing-test protocol to screen for POTS in adolescent patients

- Record baseline heart rate and blood pressure while patient is recumbent $\geq$10 minutes
- Ask patient to stand in place calmly for 10 minutes
- Record heart rate and blood pressure every 2 minutes or monitor continuously
- Encourage reporting of symptoms
- Recommend sitting if fainting seems imminent

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 $\geq$20 mm Hg systolic or $\geq$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)

Abbreviation: bpm, beats per minute; POTS, postural tachycardia syndrome.
[a]Consider subspecialty referral when criteria are met.

Source:
Heyer GL. Postural Tachycardia Syndrome: Diagnosis and Management in Adolescents and Young Adults. Pediatr Ann. 2017 Apr 1;46(4):e145-e154. doi: 10.3928/19382359-20170322-01.

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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*

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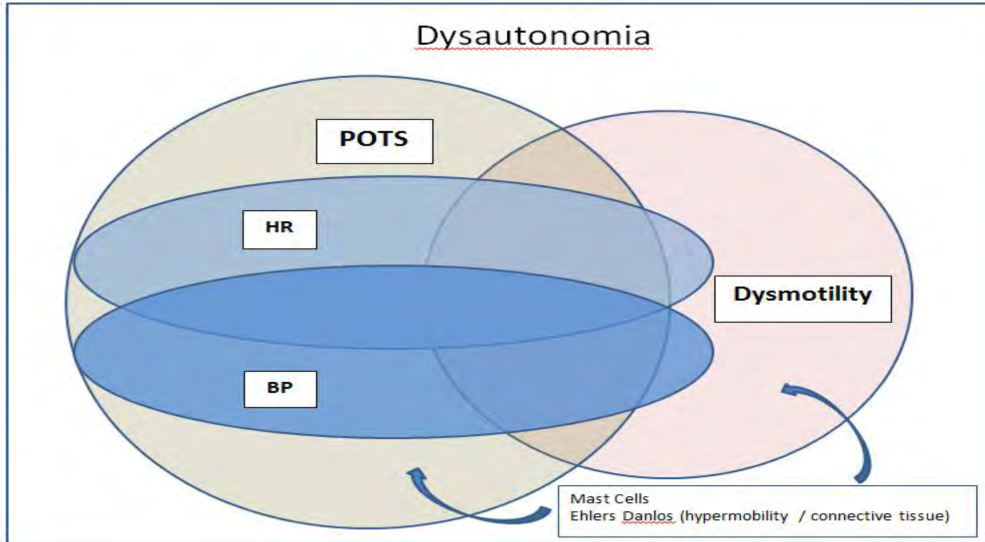
Conceptualizing POTS



- 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

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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)

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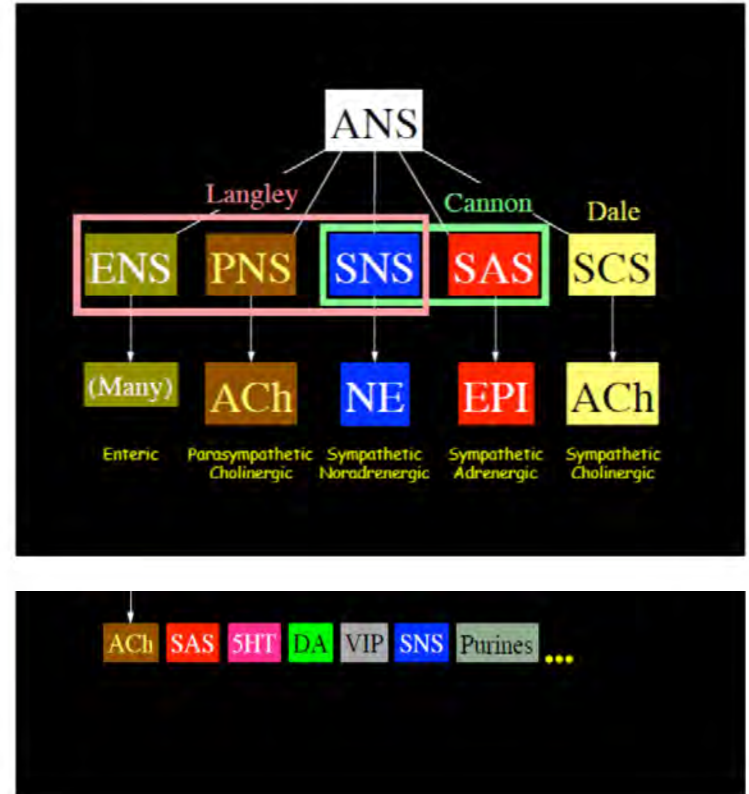
Autonomic Nervous System

- ENS (Enteric NS)
  - ACh, SAS, 5HT, DA, VIP
- PNS (Parasympathetic Cholinergic NS)
  - ACh
- SNS (Sympathethic Noradrenergic Syst)
  - Purines, NE
- SAS (Sympathetic Adrenergic Syst)
  - Epi
- SCS (Sympathetic Cholingeric 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"

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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*

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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**

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POTS Mimics…or are they? It's complicated!

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

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POTS: Treatment

- Knee high stockings
  - *Ankle/Waist 25mm/Hg*
  - *Other compression garments*
- Abdominal binder
- Spanks 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

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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

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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

- **<u>Goal is to focus on INCREASING FUNCTIONING</u>**
  - 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

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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

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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”

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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

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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

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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

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Difficulty with Sleep Changes

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

- <u>Consider Sleep Study referral for</u>:
  - 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

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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 <u>focus on functioning</u> and <u>coping strategies</u>
    - “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

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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.

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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

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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

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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.

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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

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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

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School Recommendations

- **Homebound/Home & Hospital is generally NOT recommended**

- Leads to decreased functioning
  - May require BRIEF <u>partial</u> 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

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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

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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

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Resources for Pediatricians

- Kizilbash SJ, et al. Adolescent fatigue, POTS, and recovery: A guide for clinicians. Current Problems in Pediatric and Adolescent Health Care. 2014;44:108.

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Take Home Messages

- Increase activity, minimize time laying down
- Attend school daily
- DO NOT SIGN THE SCHOOL FORMS with out consulting with co-treating providers

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QUESTIONS?

Thank you for your attention today!