Postural Orthostatic Tachycardia Syndrome
POTS

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Learning Objectives

After completion of this course, learners will be able to:

1. Identify and describe the orthostatic changes that are characteristic of Dysautonomia.
2. Speak to the wide variety of symptoms and concurrent conditions that Dysautonomia patients present with.
3. Have a general understanding of the testing for POTS diagnosis.
4. Understand the evidence based approach used to manage POTS in the rehabilitation setting and disciplines involved in the collaborative care treatment model for patients with Dysautonomia.
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• Multidisciplinary Team Approach
• Nurse Coordinator
• Sleep psychologist
• Rheumatologist
• Internal medicine
• Gastrointestinal physicians
• Genetics counseling.
• Speech
• Occupational therapy
• Physical therapy
What is Dysautonomia?
What is POTS?

Dysautonomia symptoms vary widely and range from mild to severe – chest pain, palpitations, blurred vision, sleep disorders, mood swings, headache, nausea, anxiety, panic attacks and memory problems.

POTS is a form of dysautonomia by excessive heart rate and orthostatic intolerance.
Postural Tachycardia Syndrome – Common Criteria

- Orthostatic tachycardia > 30 bpm
  - >40 bpm required if <18 years
- No consistent orthostatic hypotension
  - ΔBP > 20/10 mmHg
- Symptoms of sympathetic activation
  - Worse upright; better recumbent
- Chronic symptoms > 6 months

Testing

- Q-Sweat test
- Heart Rate Deep Breathing
- Valsalva
- Tilt table
  - Real time BP on finger
  - HR monitoring
  - Supine measurements
  - Tilt table to 70°
Testing

• Other possible testing:
  – Blood work up
  – Thyroid function testing
  – Echocardiogram

Prevalence of POTS

• Young adults and children, usually age 13-50 years
• 500,000 to 3,000,000 Americans
• Female to male ratio of 4-5:1 for unknown reasons
• Due to the debilitating nature 25% if POTS patients are unable to work
Diagnosis of POTS

“Is like diagnosing a fever while there are criteria for each a fever and POTS, and each can benefit from direct treatment (such as acetaminophen for a fever), there are multiple underlying disorders that can lead to POTS”

~Raj 2018

WHY do they have POTS?

“… ‘final common pathway’ of hundreds of genetic and acquired autonomic and cardiovascular entities”

~David Robertson
Causes of POTS symptom Onset

- Acute stressors such as pregnancy, surgery, viral infection, vaccinations, traumatic events
- Genetic
- Inflammatory and autoimmune disorders—Sjogren’s, Lupus, mixed connective tissue disorders

Borris et al. study 2018

- 2.8 trauma or surgery
- 11.6 by Concussion
- 23.9% of cases were triggered by infection.
- Among the infection triggers
  - Most common were Epstein–barre virus and mononucleosis (18.6%)
  - Upper respiratory infections (18%)
  - Gastroenteritis (11.4%)
5 subtypes of POTS

1. Neuropathic
2. Hypovolemic
3. Primary hyperadrenergic
4. Joint hypermobility-related
5. Immune related
Five Subgroups of POTS

- **Neuropathic**
  - SFN
  - SFN small fiber neuropathy, NET norepinephrine transporter, hEDS Ehlers-Danlos syndrome, hypermobility type

- **Primary Hyperadrenergic**
  - NET deficiency

- **Immune-Related**
  - Mast Cell Activation Syndrome
  - Autoimmune involved

- **Joint-Hypermobility Related**
  - hEDS


POTS Subgroup: Neuropathic

- Sympathetic denervation in the legs activates the sympathetic system causing tachycardia(Sheldon and Grubb 2015, Jacobs 2000)

- Within the neuropathic POTS 50% of patients have distal sudomotor neuropathy, a form of small fiber neuropathy(Sheldon 2015, Peltier 2010, Singer 2004)
POTS Subgroup: Hypovolemic

- Unclear if this is a part of neuropathic POTS
- Many patients in this group have reduced total blood volume, plasma volume and red blood cell volume (Fu 2010, Raj 2005, Fouad 1998)
- In 70% of POT patients blood volume is reduced, but not all patients may have hypovolemia some have a dramatic reduction in blood volume on standing. (Raj 2005)

POTS Subgroup: Primary Hyperadrenergic

- “Central activation of the sympathetic nervous system has reported to be an underlying problem due to persistent increase in sympathetic activity with elevated levels of plasma norepinephrine at resting supine and greater increase in standing (Garland 2015, Raj 2013, Furlan 1998)
- 50% of POTS patients have this subtype (Sheldon 2015)
- This group experiences primarily sympathetic symptoms tachycardia, palpitations, tremor and anxiety. (Sheldon 2015, Garland 2007)
POTS Subgroup Joint Hypomobility – Related

- Associated with Ehlers-Danlos syndrome which may be an underlying mechanism of POTS
- EDS is an inherited connective tissue disorder in which patients have delicate connective tissue, hyperextensible skin, and hypermobility of joints. (Moon 2016)
- 80% of EDS (the most common type of hypermobility) have POTS without orthostatic hypotension.
- 18% of all POTS meet the criteria for EDS

POTS Subgroup: Immune-related

- Immune related pathways may be involved especially if POTS occurs with mast cell activation syndrome (MCAS).
- Patients have skin flushing associated with tachycardia
- These patients have hypertension with the orthostatic tachycardia upon standing (Rah 2013)
MISCONCEPTIONS ABOUT POTS:

“IT IS A FAINTING DISORDER”
MISCONCEPTIONS ABOUT POTS:

“IT’S ALL IN YOUR HEAD”
POTS: “It is not all in your head!”

- Approximately 60% of the patient’s we have seen in clinic have stated that they were told their symptoms were “all in their head” prior to being diagnosed with POTS.
- After diagnosis 73% of POTS patients encounter physicians who had never heard of POTS (Steles et al., 2017)
- An average diagnosis delay of 4 years and seeing an average of 7 doctors before being diagnosed with POTS with 23% seeing more the 10 physicians (Steles et al., 2017)
- Approximately 50% of patient’s had to traveled 100 miles from home to receive POTS related specialty care and 21% have traveled over 500 miles

Is the HR Increase in POTS due to Blood Pooling in Legs or Anxiety?

![Graph showing HR and MAP responses to LBNP and MAST inflation.](image-url)
Anxiety (ASI) Scores

CAARS DSM-IV Inattention Scores
MISCONCEPTION ABOUT POTS: “IT IS A PSYCHIATRIC DISORDER”

Is POTS…a Psychiatric Disorder?

• Patients with POTS did not have an increased prevalence of major depression or anxiety disorders, including panic disorder, compared to the general population.
Non-orthostatic symptoms of POTS Sleep problems and Chronic Fatigue

Sleep Problems Correlate with Poor HRQL

Physical

Mental

$R^2=0.62$

$R^2=0.60$

Modified from K Bagai et al., J Clin Sleep Med 2011

Symptoms
Non-Orthostatic Symptoms of POTS

- Chronic fatigue
- Day timer sleepiness
- Migraine Headaches
- Hypermobility

Symptoms related to Autonomic Dysfunction

- Gastrointestinal (abdominal pain, nausea irritable bowel syndrome)
- Bladder symptoms
- In addition to initial symptoms 66% of patient’s report 10 more symptoms and 50% experience at least 14 or more and 30% experience up to 26 symptoms.

~Borris 2018
Clinical presentation with POTS:

- 50% of patients report discoloration, swelling or edema occurs in the legs (Raj 2006, Garland 2015, Freeman 2002)
- Venous pooling may be due to lack of vasoconstriction (Stewart 2002)

Physical Deconditioning

It is unclear whether deconditioning is a cause or effect of POTS
Non-Pharmacological Treatment Options

Diet

• Diet/Nutrition
  – Whole Food Plant Based Diet
  – Anti-inflammatory Diet Dr. Andrew Weil

Compression garments

• Sleep
• Exercise
The Blind Men & The Elephant

- It was six men of Hindustan
  To learning much inclined,
  Who went to see the Elephant
  (Though all of them were blind),
  That each by observation
  Might satisfy his mind

- They conclude that the elephant is like a
class, snake, spear, tree, fan or rope,
dependent upon where they touch.

Ancient Hindu Parable retold by John Godfrey Saxe (1816–1887)
Multiple Disciplinary Rehabilitation Approach

- Speech
- Occupational therapy
- Physical therapy
- Integrative Health

Physical Therapy: Subjective

- Focus on current activity limitations and work from biggest to smallest complaint
  - Ask about sleep
  - Digestion issues
  - Level of fatigue
  - Current water intake
  - Do they add salt to their diet
  - Current activity level/ exercises/activities
Physical Therapy: Objective

- Posture
- Gait
- Range of motion
- Flexibility - isolate out muscle length vs joint hypermobility
- Strength
  - assess the core activation
  - For extremities use functional testing in neutral
Meet the patient where they are the day they come in for the evaluation

- Continuum of activity level from one patient to another

- Assess what their energy level is and prioritize Instructions for the day and tests performance.

- Have pt. use a heart rate monitor with a chest strap throughout the session.

- Find a motivating factor to make the improvement in their current level of activity tolerance and level of fatigue meaningful to them.

Empirical Evidence

- Dr. Benjamin Levine Study
  - Peak heart rate is the same but peak stroke volume and cardiac output are greater after training
  - Heart rate recovery from peak exercise is significantly faster after training, indicating an improvement in autonomic circulatory control
  - Patients with POTS have no intrinsic abnormality of heart rate regulation during exercise
  - The tachycardia in POTS is due to a reduced stroke volume.
  - Cardiac remodeling and blood volume expansion associated with exercise training
Levine exercise protocol

- 8 months progressive aerobic exercise protocol
  - 3 sessions of aerobic exercise per week
  - 2 sessions of strengthening exercises per week
- Start recumbent position and progress to upright
Exercise in POTS: Cardiovascular

Starting in a recumbent position

- Must be regular
  - Every other day or 4-5 days a week
- 30 minute sessions working quickly up to 45-60 minute sessions
- NO UPRIGHT EXERCISES
  - Rowing
  - Recumbent cycling
  - Swimming once vitals are stable and always with a partner
- Takes 6-8 weeks to start noticing benefits
Exercise in POTS: Strengthening

- Progress leg and core strengthening adding the arms if there is not a significant increase in heart rate if doing so
- Start in supine progress to upright low resistance
- Modified Pilates, Core First. Sahrmann based exercises, modified yoga if appropriate
- Carefully consider exercises to address individuals with hypermobility

Abdominals
Work Out Fluid Intake

- Patient should drink at least 16 oz (1/2 a liter) 30 minutes prior to, during and following exercise
- If patient is increasing their salt intake a salty snack 1 hour before working out
- Daily fluid intake goal is set by physician
  - Ranging from 2-4 liters

Counter Measures

To Increase Blood Flow Back to Heart

- Toe Raises
- Leg Crossing
  In standing or sitting
- Squat
- Wall Sit
  45° knee bend
- Half Kneeling
- Foot on step
- Jendrassik maneuver
International POTS registry: Evaluating the efficiency of an exercise training intervention in a community setting

- George Bivens et al 2016 working with Dr. Levine
- Reproduction of the original Levine study in the community.

**Results:**
- Of the patients that completed the program 71% no longer qualified for POTS
- Reported improved quality of life (SF36)
- A small group of patients was followed for 6-12 months and the effects persisted
- Reasons for patient drop out not being able to afford a gym, exercises were too hard other medical reason

**Outcome Measures**

- **Brief Fatigue Inventory**
  - consists of 9 items that look at fatigue in the past and then rating it from 0 no fatigue to 10 completely interferes with work/activities
  - Score
- **Timed Up and Go**
  - 3 trials with heart rate recorded before and after each trial; and perceived exertion for each trial is recorded.
- **10 Minute Stand**
  - Used by neurologists
  - 5 minutes supine with heart rate measured at 2, 5, 7, and 10 minutes post standing
Borg Perceived Exertion Scale

• Rating of Perceived Exertion (PRE)
  – A subjective scale of cardiovascular work out intensity on a scale of 6-20:
  – (used primarily for when a patient is on beta blockers)
  – 6 very, very, very easy
  – 11 fairly easy
  – 13 somewhat hard
  – 15 hard
  – 17 very hard
  – 19 very, very hard

Sample Patient Home Instruction

• Cardiovascular Program
  – Begin recumbent exercise during your cardio workout, 3 days/week for 20 minutes. Perform a 5 minute warm-up (RPE 11 or less), 10 minutes base pace exercise (RPE no higher then 13) and 5 minute cool down (PRE 11 or less).

• Strength program
  – Continue with current strengthening exercises focused on core 2 x a week, but not on days you perform cardio

Activity Instructions While Participating in Rehabilitation

• Decrease activity level at home
  – Spread out/ modify projects and chores throughout the week (laundry, house cleaning, cooking etc.)
  – Hold off on additional leisure activities unless discussed with your physical therapist (hiking or participating in group exercise classes).

• Exercising at the appropriate exertion level for the training zone is a must
  – No activities should exceed a 15 on RPE scale.


Long term Outcome and Maintenance

• Typically patients notice an increase in fatigue the first month
• It typically takes 2-3 months of consistent participation for patients to start noticing a difference.
• Important to realize that they will continue to exercise the rest of their lives
• Participating in their exercise program on a regular basis is essential to making improvements.
• Frequent set backs
  – Catching a cold allergies, medical procedures
• Some patients do return to running and playing sports
• Physical therapy for POTS is a marathon, not a sprint it takes months to years to improve their level of function.

Helpful Resources for Patients

POTS Center: http://myheart.net/pots-syndrome/
National Institute of Neurological Disorders and Stroke:
Dysautonomia International:
http://www.dysautonomiainternational.org •
Dysautonomia Youth Network of America, Inc.:

Common Comorbidities

• Ehlers-Danlos Syndrome
• Mast Cell activation
Ehlers-Danlos Syndrome

- Hypermobility: Generalized joint laxity with associated musculoskeletal complaints in absence of any systemic disease
- Inherited form of generalized connective tissue disorder

EDS relationship to POTS

- Orthostatic intolerance is significant in EDS
- Prevalence 18% in POTS population compare to .02% in the general population
Types of Pain

Nocioceptive: Mechanical inflammatory
Neuropathic: peripheral, central
Central sensitization: Neuroplasticity
Different patients have different responses to the same level of pain or stimuli
Each type of pain is addressed differently

Physical therapy general approach to each type of pain

- **Mechanical***- muscle setting, strength balance proprioceptive retraining
- **Inflammatory**: pool proprioceptive input mindful movement
- **Nerve**- Nerve flossing cardio mindful movement, posture training
- **Central Sensitization** Cardiovascular exercise, Medication, Breathing mindful movement
Mast Cell Activation Disorder

• “Mast cells are hardwired to recognize and react to with a defined set of chemicals and physical responses in order to contain usual suspects pathogens and harmful substances”

• Mast cells are found throughout the body and play a role in allergic and anaphylactic reactions and other inflammatory disorders in the skin respiratory tract, joints gastrointestinal tract, nervous system and bladder

  • Information from slides Anne Maitland’s slides at 2017 EDS Global Learning Conference

Mast Cell Activation

Dr. Anne Maitland’s presentation at EDS conference
Symptoms of Mast Cell Activation Disorder

- Hives and red rash
- Brain fog
- Digestive issues
- Bladder irritability
- Swelling in limbs supraclavicular region

Mast Cell Triggers

Management of Mast Cell Activation

- **Identify triggers:** alcohol, heat, medication, food sensitivity, excessive exercise
- **Manage physical and emotional stress**
- **Exercise regularly**

Measure of Hypermobility

- **Beighton Scale**
- **Revised Diagnostic Criteria for EDS**

Cirque du soeil.com
Exercise Intervention for EDS

- Focus on large muscle groups
- Decreased emphasis on Stretching
- Less emphasis on passive based exercises
- Bergin at a lower level of intensity
- Progress slowly monitor symptoms
- Focus on stability and control
- Avoid end range motion

Patient’s perspective of POTS

9 COMMON POTS SYMPTOMS!

- Fatigue!
- Dizziness/lightheadedness
- Wacky heart rate and blood pressure
- GI issues/discomfort
- Palpitations
- Blood pooling
- Temperature intolerance
- Brain fog
- Anxiety

Postural Orthostatic Tachycardia Syndrome
Awareness Week

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You know you have POTS when...

...you make a cup of tea and your heart thinks you’ve run a marathon.

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• POTS Patient story
What POTS Patients want Physician and Therapists to Know

• It is very frustrating having to travel great distances and seeing 7-10 doctors before getting a diagnosis.
• Having to go through numerous tests and different treatments before receiving a diagnosis
• Long periods of missing school or work
• Symptoms of shortness of breath, racing heart, lightheadedness, unusual tingling and numbness can be frustrating and frightening

Tips for Discussing Exercise with your POTS Patients

• Education and explanation of the benefits of regular exercise to increase fitness level, blood volume, cardiac remodeling and normalize sympathetic activity
• Acknowledge that exercising with an orthostatic disorder may be difficult especially the first month.
• Don’t blame the patient for exercise intolerance but work with them to help them identify how they are feeling that day and to chose the appropriate level of activity
• Be a cheerleader for them encourage the m to stick with their program
Wholistic approach: Using PT tools to think outside the box

- Seated Exercise ball routine
- Seated yoga routine if appropriate
- Seated Tai Chi routine
- Mindfulness

Tai Chi Quigong Seated

- Major goal is to improve an individual’s ability to become more aware or attentive to the present moment.
- Tai Chi translated means extreme ultimate recognizing that everyone is born with the Supreme Potential to deal with the ultimate energy of the Universe that presents both difficult challenges and wonderous opportunities.
Tai Chi Video

Benefits of seated Tai Chi

- Engagement of core muscles for postural stability
- Engagement of the large muscles of the legs to promote blood flow back toward the heart
- Focus on breathing
- Physical and emotional sense of well being
- Pain reduction
- Mental distraction
Diaphragmatic Breathing

Seated instructions:
1. As you breathe in through your nose, don’t let your chest rise, but let your stomach expand instead.
2. As you breathe out, slowly and evenly allow your stomach to move or pull back in.
3. During this breathing process, your hand on your stomach should move, but your hand on your chest should remain almost still.

Instructions Lying Down: Some may find it easier to lie down:
1. Lie on your back on a flat surface or in bed, with your knees bent and your head supported. You may use a pillow under your knees to support your legs if desired. Place one hand on your upper chest and the other just below your rib cage. This will allow you to feel your diaphragm move as you breathe.
2. Breathe in slowly through your nose so that your stomach moves out against your hand. The hand on your chest should remain as still as possible.
3. Let your stomach fall inward as you exhale. The hand on your upper should remain.

Mindfulness/Medication

- Medication Video
- (Dan Harris medication 101)
Mindfulness Apps

- The Mindfulness App.
- Headspace.
- Calm.
- MINDBODY.
- Buddhify.
- Insight Timer.
- Smiling Mind.
- Meditation Timer Pro.
- Stop breathe, & Think

Mindfulness Video
What We have learned from our patients

Electrolytes to add to water
- Liquid I.V. hydration multiplier
- Banana bag
- Coconut water

Precautions

• Symptoms can change rapidly it is important to use a heart rate monitor
• Ehlers Dalos Syndrome/Hypermobility
• Mitochondrial syndrome
• Minimize use of arms and arms above shoulder height due to often increasing heart rates
Hints to help with follow through

• Have them get a heart rate monitor and check into gyms in their area before the next session
• Use examples for fight or flight that is why they are so fatigued or “gum” analogy for why the heart rate goes so high when they stand up
• Use teach back method
• Reassure them they will get better and make improvements

Patient Education

• Important that they get a heart rate monitor
• Look into a gym with seated equipment and possibly a pool
• Log their exercises
• On bad days modify their exercise or perform core exercises at home
• Take water/drink with electrolytes with them to their workout
• Cooling towel if they get too warm
• Plan ahead in the warmer months to be able to stay cool
Cooling Vests

A cooling vest is a piece of specially made clothing designed to lower or stabilize body temperature and make exposure to warm climates or environments more bearable. There are many different types of cooling vests and they vary in price from $25 to $800 and more.

There are five common types of cooling vests including:

- **Ice pack Cooling Vests**
- **Water Activated Evaporative Cooling Vests**
- **Phase Change Cooling Vests**
- **Air Cooling Vest (fan system)**
- **Circulating cooling vests**

Cooling Towels

- **Microfiber** – Microfiber towels are made with specialized fabrics that retain water more than a standard cotton towel. Microfiber cooling towels are incredibly soft to the touch and provide prolonged cooling. They are durable, machine washable, and do not dry stiff like PVA towels and similar moisture wicking towels. Microfiber towels are easy to reactivate, by re-soaking in water, wringing, and then a quick snap in the air.
- **Polyvinyl Acetate**– Polyvinyl Acetate or PVA cooling towels retain the most amount of water compared to other towels. However, they also drip more than microfiber towels, yet less than cotton towels. PVA towels work well at cooling you down, but please know they tend to retain smells and as noted above, dry stiff. As long as you keep them wet, they work well.
- **Hybrid** – There are also mesh microfiber hybrid towels. These types of towels also absorb water, retain it, and create a cooling effect. The mesh-microfiber towel tends to be more lightweight than other cooling towel and subsequently are less bulky. These are perfect for outdoor activities and are very soft to the touch whether they are wet or dry.

Top rated towels:
- Riptgear
- Alfamo Sport
- Chill-Its 6602

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POTS Take Home Messages

1. POTS is not a diagnosis but a grouping of symptoms that can be divided into five subgroups with similarities and over many variable symptoms.
2. There are some common misconceptions about what POTS
3. It takes a multidisciplinary team approach to effectively assist patient’s in managing their rehabilitation process.
4. Important to meet the patient where every they are on the spectrum of activity level
5. Find out what is most meaningful to them to address first
6. A collaborative team approach and working toward a self-directed home exercises program
7. Be creative think outside of the box take your PT skills and mold them to the specific needs of the patient to be successful
8. Provide education to assist the patient better understand their symptoms and more importantly that exercise will help them feel better.
9. Encourage patients to seek out counseling as appropriate to help them learn to live with fact that their lives might be different then what they initially had in mind for their future. We can assist with reframing that their life will be different then is was before.
10. We as individuals can often be the initiator but it takes a group effort to change things.

Cognitive Behavioral Therapy

THOUGHTS
What we think affects how we feel and act

EMOTIONS
What we feel affects how we think and act

BEHAVIORS
What we do affects how we think and feel

CHANGING PERCEPTIONS

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"Remember to look up at the stars and not down at your feet. Try to make sense of what you see and wonder about what makes the Universe exist. Be curious. And however difficult life may seem, there is always something you can do and succeed at. It matters that you don’t just give up."
- Professor Stephen Hawking
Thank You

"I fight for my health everyday in ways most people don’t understand. I’m not lazy. I’m a warrior."
• U-Tube video: Suggestions of how to live a better life
References

The whole issue of Am J Med Genet C Semin Med Genet. 2017;175(1) is devoted to EDS, including hEDS and HSD. These articles, including many cited below, are available at https://www.ehlers-danlos.com/2017-eds-international-classification/.


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