Acknowledgements

Naomi Bauer, PT, DPT, CCS
Tiffany Champion, PT, DPT
Cynthia Harrell, PT
Camille Frazier-Mills, MD
Director, Duke Syncope and Dysautonomia Clinic

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Objectives

- Discuss the epidemiology, pathophysiology, diagnosis, and medical management of dysautonomia.
- Identify appropriate cardiovascular and orthopedic physical therapy evaluation and examination techniques for patients with dysautonomia.
- Implement a physical therapy plan of care with appropriate goals for patients with dysautonomia.
- Identify potential barriers to patient compliance with physical therapy treatment.

What is Dysautonomia?

- Umbrella term used to describe medical conditions resulting in a malfunctioning autonomic nervous system
- Can affect many parts of the body
  - Neurocardiogenic/Vasovagal Syncope
  - Inappropriate Sinus Tachycardia
  - Diabetic Autonomic Neuropathy
  - Autonomic Dysreflexia
- Can be associated with/caused by autoimmune diseases

What is POTS?

- A form of dysautonomia characterized primarily by orthostatic intolerance resulting in excessive increases in heart rate.
- Often idiopathic
- Can occur in conjunction with many other conditions
Diagnostic Criteria

- Symptoms for ≥6 months
  - AND
- Heart rate increase within 10 minutes of standing
  - ≥30 beats per minute for adults
  - ≥40 beats per minute for adolescents
  - OR
  - >120 beats per minute
- No orthostatic hypotension (drop in blood pressure of 20mmHg systolic or 10 mmHg diastolic with 5 minutes of standing)
- Symptoms exacerbated with standing and relieve when recumbent
- No other overt causes of orthostatic symptoms or tachycardia can be identified
- If POTS is suspected, a tilt table test is the gold standard for confirming the clinical diagnosis

Common Signs and Symptoms of POTS

- Palpitations
- Fatigue
- Exercise intolerance
- Nausea, GI issues
- Brain fog
- Headache
- Tremulousness
- Light-headedness
- Dizziness
- Sleep disturbances
- Poor thermoregulation
- Syncope
- Near syncope

Epidemiology and Demographics of POTS

- Not rare, just rarely diagnosed
- First described in 1860’s
- Has been known as Civil War Syndrome, Soldier’s Heart, Irritable Heart Syndrome, and Effort Syndrome among others
- First named as “POTS” in 1993 by the Mayo Clinic
- Prevalence is estimated at about 0.2% of the general population
  - Approx. 650,000 people in the U.S.A.
  - Some reports say up to 3,000,000 people in the U.S.A.
- Predominantly affects women
- Age of onset is most commonly between the ages of 15 and 25
- Difficult to track patients due to no specific ICD-10 code

Physiology of POTS

- Two main types of POTS: Neuropathic and Hyperadrenergic
  - Neuropathic
    - More common and commonly referred to as just “POTS”
    - Hypotensive BP
    - Decreased vasoconstriction in standing
  - Hyperadrenergic
    - Less common
    - Increased sympathetic activity
    - Hypertensive BP
Causes of POTS and Comorbidities

Causes
- Viral Infection
  - Onset of symptoms following acute viral infection
  - 1/3rd of cases, best prognosis
- Genetic
  - Runs in families
- Inflammatory and autoimmune disorders
  - Sjogrens, Lupus, mixed connective tissue disorders
- Idiopathic

Common Comorbidities
- Mast cell disorders
- Ehlers-Danlos Syndrome
- Celiac Disease
- Other digestive disorders
- Migranes

Treatment for POTS

- Medical/Pharmacological management
- Non-pharmacological management/lifestyle modification
  - Dietary modification
  - Exercise

Medical Management of POTS

Three-pronged approach
- Heart Rate Control
  - Beta blockers
  - Ivabradine
- Peripheral Vasoconstriction
  - Midodrine
  - Droxidopa
  - Pyridostigmine
- Volume Expansion
  - Fludrocortisone
  - Desmopressin

Non-Pharmacological Treatment Options

- Diet
  - Limit caffeine
  - Non-hyperadrenergic (Hypotension)
    - 4-6 grams of salt per day
    - 2-4 liters of fluid (half water, half electrolyte replacement)
  - Hyperadrenergic (Hypertension)
    - NO extra salt
    - 2-4 liters of water per day
  - Exercise
    - Start slowly, increase gradually

- Compression garments
  - Socks
  - Leggings
  - Abdominal (e.g. Spanx)
- Sleep
  - Avoid over-sleeping
  - < 12 hours per day
  - Incline head of bed a few inches
- IV Fluids
  - For volume bolus when drinking is not enough

Prognosis for POTS

- No cure
- ~50% of patients with post-viral POTS make full recovery
- Secondary POTS symptoms may subside if underlying cause is treated
- Can be effectively managed with medication and lifestyle modification
- ~25% of patients experience severe and ongoing symptoms and disability
- Little longitudinal research to determine long term effects of treatment

Helpful Resources for Patients

- POTS Center: [http://myheart.net/pots-syndrome/](http://myheart.net/pots-syndrome/)
- Dysautonomia International: [http://www.dysautonomiainternational.org](http://www.dysautonomiainternational.org)
- Dysautonomia Youth Network of America, Inc.: [http://www.dynainc.org](http://www.dynainc.org)
Exercise Intolerance

- Study by Raj and Levine showed that on echocardiogram, POTS patients found to have smaller hearts (Raj and Levine, 2013).
- Another study found POTS patients have increased heart rate and reduced stroke volume at any absolute workload compared with healthy sedentary individuals, but there is no difference in the heart rate responses between groups at relative workload (Takeshita et al., 2010).
- Tachycardia may be related to decreased stroke volume rather than intrinsic abnormality of HR regulation (Fu et al., 2010, Masuki et al., 2007b, Shibata et al., 2012).
- CO = HR x SV → Decreased stroke volume results from smaller heart and lower blood volume.

Effects of Cardiovascular Training

- Endurance training:
  - expands blood volume and plasma volume (Saltin et al., 1968)
  - increases cardiac size and mass (Dorfman et al., 2007)
  - improves orthostatic tolerance (Saltin et al., 1968).
- Study done by Dr. Benjamin Levine et al. confirmed these effects of exercise training in POTS patients.

Physical Therapy Management

- General PT evaluation: subjective, systems screen, strength testing, orthostatics, submaximal exercise testing.
- Treatment includes:
  - Progressive cardiovascular exercise
  - Strength training
  - Static standing tolerance
- Plan of care frequency usually 2-3 sessions per week until stable
- Decreasing frequency as patient’s symptoms decrease and independence increases
- Encouraged to get equipment for home or join gym.

Levine Exercise Protocol

- 8 month protocol for progression of aerobic exercise
  - 3-4 sessions of aerobic activity per week
  - 6 weeks of strength training per week
- Each exercise session includes a 5, progressing to 10, minute warm-up and cooldown
- Progressively increases “base pace” time to reach 30 minutes of moderate intensity exercise
- Starts in recumbent position and progresses to upright position
- Incorporates higher heart rate zones and interval training for intensity later in the program.

Levine Protocol

- Developed by Benjamin Levine, MD, et al. in Dallas, Texas
- Study done on patients with POTS found that many patients can be “cured” with exercise (no longer met POTS diagnostic criteria)
  - Un-medicated patients, tested positive on tilt table
  - Participated in 3 months of exercise training
  - 8-12 grams of salt daily
  - 2-4 liters of water daily
- Other institutions have developed their own protocols
  - Children’s Hospital of Philadelphia (CHOP)
  - Mayo

Measure of Exercise Intensity

- Measured using the Borg 6-20 RPE Scale
- Warm-up/recovery/cool-down should be 10-12
- Base pace should be 13-14
- HR range not useful in patients on beta-blockers
- Maximal steady state, race pace, and intervals used late in training.
If the patient becomes symptomatic…

- Decrease exercise intensity
- Ensure they are hydrating
- If necessary, have them lie supine to recover – “The floor is their friend”

After-Care

- Inform patient that they will likely be very fatigued and symptomatic following initial therapy sessions
- Instruct on adequate hydration
- Encourage patients to go home and rest
  - Don’t try to clean the house
  - Don’t try to go shopping
- NO NAPPING!

PT: Strength Training

- Progressive UE, LE, and core strength training
- Initiate in supine at low resistance
- Progress to upright position
- Focus on core stability
- Pilates and modified yoga may be used
- Strengthening specifically focused on LE and thighs has been shown to significantly improve POTS symptoms (Raj, 2016)
- Address hypermobility in patients with EDS

PT: Postural Training

- Initiate static standing against wall as long as is tolerated without symptoms
  - Cannot move or fidget with LEs
  - Progress by 30 seconds when previous level no longer causing symptoms, usually 1 week
- Goal is 10 minutes standing without symptom exacerbation

PT: Fluid Intake

- Patients should drink at least 1/2 liter (16 ounces) of fluid 30 minutes prior to, during, and after exercise.
- This is in addition to their normal fluid intake for the day.
- The patient will typically have a daily fluid intake goal set by their MD
  - Generally 2-4 liters per day

Outcome Measures

- Fatigue Severity Scale (FSS)
  - 9-item questionnaire
  - Measures severity and effect of fatigue on daily activities and lifestyle
- Fatigue Impact Scale (FIS) (McDonald 2014)
- Orthostatic Grading Scale (OGS)
  - 5-item questionnaire
  - Assess frequency and severity of orthostatic symptoms
- Orthostatic Intolerance Questionnaire, Orthostatic Hypotension Questionnaire
Outcome Measures

• Ferrans and Powers Quality of Life Index Cardiac Version IV
  – Assesses quality of life with a scaled score of 0-30
  – SF-36 (George 2016, Moon 2016)
  – QOL of patients with POTS has been found to be comparable to a
    patient with COPD or CHF

• Center for Epidemiological Studies Depression Scale
  – Assesses symptoms of depression
  – Patients are at high risk of depression and typically have high scores
    pre-rehab (Bruce 2016)
  – Rehab improves scores and decreases risk of depression (Bruce 2016)
  – Beck Depression Inventory (Moon 2016)

Long Term Outcomes and Maintenance

• Typically increased fatigue for first month
• Patients generally require 3-4 months of exercise compliance before they notice improvements
• Many patients recover to lead relatively normal lifestyles
• Some return to jogging and playing sports
• Must continue to exercise for the rest of their life to maintain benefits
• Frequent setbacks
  – Catching cold, allergies, medical procedures

PT for POTS is a marathon, not a sprint. It may take months to years for patients to return to their PLOF!

History

• 1901 recognized by a dermatologist Edvard Ehlers
• 1908 Henri-Alexandre Danlos skin extensibility and fragility were the cardinal features of the syndrome

Hypermobility Syndrome

• Kirk Ansell and Bywaters in 1967 coined the term hypermobility syndrome
• Defined as: Generalized joint laxity with associated musculoskeletal complaints in the absence of any systemic disease
• Inherited form of generalized connective tissue disorder (Grahame et al 1999)

Relationship with POTS

• Orthostatic intolerance is significantly more prevalent in EDS (74-78% than controls 10-34%) (Gazit 2003 and Wandele 2014)
• Prevalence of 18% in POTS patients compared to general population of 0.02% (Wallman et al 2014)
Categories of EDS

- Classical Type (Type 1 and 2)
- Hypermobility Type (Type 3)
- Vascular Type (Type 4)
- Kyphoscoliosis Type (Type 6)
- Arthrochalasia Type (Type 7A and 7B)
- Dermatosparaxis Type (Type 7C)

Theories of pain in hypermobility

- Impairment of joint proprioception, balance, coordination (Mallik 1994 Hall 1995)
- Result of joint micro trauma from overuse and misuse of tissues that have an inherent weakness in their collagen structure (Kirk et al 1967, Russek 2000)
- Pain due to sensory nerve endings which are overstretched by stretch, but which are poorly supported by collagen fibrils and hence are overstretched as the lax capsule is stretched (Child 1986)

Theories of pain in hypermobility

- May begin as a localized joint pain but frequently develops into chronic pain with alterations in central nervous system processing, also evidence that dysfunction in autonomic nervous system may play a part (Gazit et al 2003)
- Kinesiphobia leading to disuse and deconditioning

Examination of the Hypermobile patient
Subjective examination

Previous history
- Pain as a child
- Growing pains
- Gymnastics or ballet when younger
- More flexible when younger
- Better/worse in pregnancy
- Any dislocation/subluxations
- History of fractures (complete or stress)

Present history
- Family history of increased flexibility
- Dislike of sustained postures such as standing/sitting
- Dislike too much activity
- Bruise easily
- Uncomfortable all of the time
- Short term relief with stretching and massage

Measurement of joint hypermobility

- Beighton Scale
- Revised Diagnostic Criteria for Ehlers-Danlos Hypermobility Type

Beighton Scale

Revised Diagnostic Criteria for Ehlers-Danlos Hypermobility Type (Grahame et al 2000)

Major Criteria
• Beighton score of ≥ 4/9
• Pain for longer than 3 months in ≥ 4 joints

Minor Criteria
• Beighton score of 1-3
• Pain for longer than 3 months in 1-3 joints or back pain for more than 3 months
• Dislocation/subluxation in more than one joint, or in one joint on more than one occasion
• Three or more locations of soft tissue pain
• Marfanoid habitus
• Abnormal skin extensibility
• Eye drooping
• Varicose veins or hernia or uterine/rectal prolapse

Treatment
Joint Stability

Control or neural and feedback subsystem

Passive musculoskeletal subsystem

Active musculoskeletal subsystem

(Panjabi 2003)

Neutral zone

Norma l

Hypermobility Syndrome

(Modified from Panjabi 2003)

Treatment

• Focus on large muscle groups
• Decrease emphasis on stretching
• Less emphasis on passive based treatments

Case Study

DukeHealth

Treatment

• Insufficient studies regarding long term physical therapy treatments
• 63.4% of patients in physical therapy reported a positive outcome (Rombart 2011)
• Current evidence is to treat impairments of increased extensibility and decreased strength
• Keep in mind positional tolerance of patient with POTS when performing strengthening exercise
Case Study

- 31-year-old female diagnosed with POTS 3 years prior
- Had symptoms for the year previous to diagnosis
  - Sxs started during a hike in Greece after she became dehydrated
  - Was worked up for cardiac, GI, psychiatric complaints
  - POTS diagnosis came because her therapist had been treating another patient diagnosed with POTS and so was familiar with the presentation
- Previously led an active lifestyle: running, yoga, business owner
- Now unable to exercise except for some UE and core strengthening in supine
- Endorses chronic fatigue
- Unable to stand for longer than 2 minutes

Case Study

- Lives alone in a 2-story apartment
- Does not venture into the community
- Relies on friends and family to complete any grocery shopping and walk her dog
- Goals
  - Reduce fatigue
  - Return to exercise
  - Improve QOL (go out to eat with friends, go to movies, walk dog)

Case Study

- PMH significant for
  - Asthma
  - Fibromyalgia
  - Chronic fatigue
  - Migraines
  - IBS
  - Gluten intolerance
- Medications
  - Atenolol, started 6 weeks prior
  - Escitalopram
  - Levalbuterol
  - Midodrine
  - Sumatriptan

Case Study - Patient Presentation

- Well appearing
- Reluctant to sit or stand
- Strength WNL throughout
- Beighton score of 8
- Emotional lability during evaluation, crying at times, fearful of starting extensive exercise protocol
  - Due to emotional state, patient unable to complete exercise test at evaluation, spent most of evaluation lying on table crying

Case Study - Exercise Progression

- Symptomatic sitting in waiting room
  - Unable to tolerate sitting upright
  - Arranged for patient to proceed directly to hi/lo table in treatment area
- Started on NuStep level 1, 13 minutes, 11 watts
- Able to tolerate 2-3 minutes before needing to rest in supine
- Initial focus of exercise prescription was to increase duration of activity, decrease number of rest breaks

Case Study – Exercise Progression

- 2 weeks: introduced core and LE strengthening in supine using body weight and resistance bands (EDS assumed)
- 1 month: patient tolerating 20 minutes on NuStep level 1, but still requiring 4 supine rests; introduced UE strengthening in supine using resistance bands
- 2 months: patient tolerating 30 minutes on NuStep level 6 but still requiring 4 supine rests
  - Starting to engage in community activities: limited grocery shopping, cooking dinner, socializing with friends at home
  - Completing strength training at home independently
  - Increasing understanding of symptoms and how far she is able to push with exercise
Case Study – Exercise Progression

- 3 months: fluctuating symptoms, but able to complete part of her exercise on the recumbent bike
  - Officially diagnosed with EDS
  - Progression of strength exercises to sitting, following precautions for EDS
  - Reviewed joint protection strategies
- 4 months: symptoms improving, exercise tolerance still fluctuating due to heat, but completing recumbent bike 30-45 minutes at home 1-2 days/week
  - Patient starting to drive self to therapy sessions
  - Progression of some strength exercises to standing
- 5 months: beginning ambulation for exercise

Case Study - Summary

- Very symptomatic with very poor activity tolerance
- Initial exercise progression was to increase duration of activity in recumbent position while respecting symptoms
- As patient was able to tolerate more activity, exercise progressed to interval training on bikes with more upright positioning then to standing exercise
  - Continued to use a less upright position for warm-up and cooldown when initially working on tolerance to exercise position

Takeaways

- These patients will likely be very symptomatic for the first month
- Most patients will take 3-4 months to begin to see improvements
- It can take months to years of targeted exercise to reach a point of feeling “normal” again
- It is almost never a smooth progression with exercise training
- These patients must be diligent with exercise for the rest of their lives to maintain benefits
- Exercise is like a prescription drug for these patients
- There is no one exercise plan that works for everyone
- Hydration is very important for ALL POTS patients
- Comorbidities can further complicate treatment
  - EDS, migraines, fibromyalgia, anxiety, depression

Case Study - Summary

- Did not follow a specific protocol closely
- Reported return to most daily activities and starting to “feel like a normal person” 11 months after initiation of PT
- Due to diagnosis of EDS, required frequent musculoskeletal treatments to deal with hip, back, and shoulder issues
  - Joint protection and appropriate strengthening
  - Manual therapy
  - Modalities for pain, cold pack to initially reduce symptom exacerbation during summer heat

References

References

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24. Questions

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