Cardiac and Orthopedic Treatment for Patients with Dysautonomia

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Acknowledgements

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What is Dysautonomia?

- Umbrella term used to describe medical conditions resulting in a malfunctioning autonomic nervous system
- Can affect many parts of the body
- Dysautonomia is very common
  - 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
Confirming The Diagnosis: Tilt Table Testing

• If POTS is suspected, a tilt table test is the gold standard for confirming the clinical diagnosis.

• Procedure:
  – Patient is connected to vital sign monitoring equipment and vitals are taken at regular intervals.
  – Patient rests supine for 20 minutes.
  – Table tilted to 70º.
  – Blood pressure and heart rate are monitored continuously and recorded at minutes 2, 5, and 10.
  – Patient is kept in upright position for 40 minutes, or until they faint.

• The test is considered positive if heart rate parameters are met.
Common Signs and Symptoms of POTS

- Palpitations
- Fatigue
- Exercise Intolerance
- Nausea
- Diminished concentration
- Headache
- Tremulousness
- Lightheadedness
- 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
Distribution of People with POTS in the U.S.
Distribution of Physicians Who Treat POTS in the U.S.
Physiology of POTS

- Two main types of POTS: Neuropathic and Hyperadrenergic
  - Neuropathic
    - More common and commonly referred to as just “POTS”
    - Hypotensive BP
  - Hyperadrenergic
    - Less common
    - Hypertensive BP
- Low blood volume
- Small hearts
  - Grinch Syndrome
# 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**
  - Sjogren's, Lupus, mixed connective tissue disorders
- **Idiopathic**

## Common Comorbidities
- Mast cell disorders
- Ehlers-Danlos Syndrome
- Celiac Disease
- Other digestive disorders
- Migraines
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 - BP Low**
    • 4-6 grams of salt per day
    • 2-4 liters of fluid (half water, half electrolyte replacement)
  – **Hyperadrenergic - BP High**
    • 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/
- Dysautonomia International: http://www.dysautonomiainternational.org
- Dysautonomia Youth Network of America, Inc.: 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 (Shibata et al., 2012)
- 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 (Dorfman et al., 2007)

• Study done by Dr. Benjamin Levine et al. confirmed these effects of exercise training in POTS patients
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
Levine Exercise Protocol

- 8 month protocol for progression of aerobic exercise
  - 3 sessions of aerobic activity per week
  - 2 sessions of strength training per week
- Progressively increases “base pace” time to reach 45 minutes of moderate intensity exercise
- Start in recumbent position and progress to upright position
- Incorporates interval training using maximum steady state (MSS) for intensity
  - MSS calculated based on heart rate
Physical Therapy Management

- General PT evaluation: subjective, systems screen, strength testing, orthostatics, exercise testing
- Recumbent bike exercise test at evaluation
  - Resistance and watts goal increased each minute
  - Test stopped when pt becomes symptomatic or can no longer maintain goal watts
  - Monitored on ECG
- Progressive cardiovascular exercise
- Strength training
- Static standing tolerance
PT: Cardiovascular Exercise

- Initiate in recumbent position – typically bike
- Alternating warm-up/recovery/cool-down pace (RPE < 12) with base pace (RPE 13-15)
- Goal of 30+ minutes continuous base pace
- Progress to upright exercise
- Vital signs monitored at regular intervals
- Come to PT 2-3 times per week until stable (1-3 months typically)
  - Transition from individual sessions to participating in POTs group 1-2 times per week
  - Encouraged to get equipment for home, or join gym
- Swimming is a great option once pt’s vitals are stable*

Criteria for discharge from therapy:
- Adherence to exercise regimen
- Transition to HEP when pt’s vitals are stable and appropriate
POTS Group

- Group was implemented due to our overwhelming caseload of POTS patients
- Offered 3 times per week (Mon, Wed and Saturday)
- Education sessions are offered every Monday on topics such as: Energy conservation and energy expenditure, joint protection, Nutrition, Relaxation and meditation, core strengthening class, patient support group, etc.
- Group is for patients who are relatively stable  
  – those who are very symptomatic continue with 1-on-1
- Patients continue to progress with exercise regimen, vitals are checked less often then during individual sessions.
- Started 1/6/16
- Patients come 2 days per week to start, then decrease in frequency based on progress.
- Currently cardiovascular exercise only with plans to add LE strengthening and core strengthening on a regular basis
- Allows patients to meet and talk to other patients with POTS
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
- Extremely important to stick to the above exertion levels during each training zone.
- HR range not very useful in patients on beta-blockers
- Maximal Steady State may be used late in training

<table>
<thead>
<tr>
<th>Perceived Exertion</th>
<th>Description</th>
<th>Training Zone</th>
</tr>
</thead>
<tbody>
<tr>
<td>6</td>
<td>Rest</td>
<td>Recovery</td>
</tr>
<tr>
<td>7</td>
<td>Extremely light</td>
<td>Warmup, Coldown, Recovery</td>
</tr>
<tr>
<td>8</td>
<td>Very light</td>
<td>Warmup, Coldown, Recovery</td>
</tr>
<tr>
<td>9</td>
<td>Easy walking at comfortable pace</td>
<td>Warmup, Coldown, Recovery</td>
</tr>
<tr>
<td>10</td>
<td>Can easily talk; could continue activity &gt; 1 hour</td>
<td>Warmup, Coldown, Recovery</td>
</tr>
<tr>
<td>11</td>
<td>Light</td>
<td>Base Pace</td>
</tr>
<tr>
<td>12</td>
<td>Somewhat hard</td>
<td>Base Pace</td>
</tr>
<tr>
<td>13</td>
<td>Hard</td>
<td>Base Pace</td>
</tr>
<tr>
<td>14</td>
<td>Very hard</td>
<td>Maximal Steady State (MSS)</td>
</tr>
<tr>
<td>16</td>
<td>Very very hard, fatigued, can continue &lt; 10 minutes</td>
<td>MSS, Race Pace</td>
</tr>
<tr>
<td>17</td>
<td>Vigorous breathing, can't talk, can continue &lt; 5 minutes</td>
<td>MSS, Race Pace</td>
</tr>
<tr>
<td>18</td>
<td>Extremely hard</td>
<td>MSS, Interval</td>
</tr>
<tr>
<td>19</td>
<td>Counting down to quit time (&lt; 2 minutes)</td>
<td>MSS, Interval</td>
</tr>
<tr>
<td>20</td>
<td>Maximal</td>
<td>Interval</td>
</tr>
</tbody>
</table>
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”
Aerobic Exercise After Visit Instructions

- 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 postural muscles
- Pilates and modified yoga may be used
- Strengthening specifically focused on LE and thighs has been shown to significantly tachycardia and improve POTS symptoms (Raj 2016)
- Helps address hypermobility in pts who also have EDS.
PT: Postural Training

- Initiate with 1 minute static standing against wall, or as long as is tolerated
  - Cannot move or fidget with LEs
  - Can perform minimal movement with UEs such as texting
  - Perform with supervision or in front of a bed
- Progress by 30 seconds when previous level no longer causing symptoms
- Goal is 10 minutes static standing without symptom exacerbation
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
  - Other Measures:
    - Fatigue Impact Scale (McDonald 2014)

- **Self-report Orthostatic Grading Scale**
  - 5-item questionnaire
  - Assess frequency and severity of orthostatic symptoms
  - Other measures:
    - Orthostatic Intolerance Questionnaire
    - Orthostatic Hypotension Questionnaire

- **Ferrans and Powers Quality of Life Index Cardiac Version IV**
  - Assesses quality of life with a scaled score of 0-30
  - Higher score = better QoL
  - Other Measures:
    - SF-36 (George 2016, Moon 2016)

- **Center for Epidemiological Studies Depression Scale**
  - Assesses symptoms of Depression
  - Higher score = more at risk for depression
  - POTS patients are at a high risk of depression and typically have very high scores pre-rehab (Bruce 2016)
  - Rehab improves scores and decreases risk of depression (Bruce 2016)
  - Other measure:
    - Beck depression inventory-II (Moon 2016)
Outcome Measures

• Research shows depression and decreased QOL are huge issues in POTS patients

• Many POTS patients are young and this age alone increases risk for depression

• Referral to psychiatrist/psychologist may be necessary temporarily

• An exercise program may lead to improvements in depression and QOL.

• Some individuals may not recognize the improvements they make with rehab and may have unchanged or worsening scores post-rehab
Sample Patient HEP Instructions

Cardio exercise program:
• Begin using the bike during your cardio workouts, 3 days/week for 20 minutes. Perform a 5 minute warm-up (RPE 11 or less), 10 min base exercise (RPE no higher than 13) and 5 min cooldown (RPE 11 or less).

Strength program:
• Continue with current pilates exercises that you have been doing at home. Perform 2 days/week but not on days you are performing cardio.
Sample Patient Instructions

Postural training:

- Begin static standing against a wall for 3 minutes, three times a day. If you start to experience any symptoms (dizziness, lightheadedness, nausea, chest pain, headache, blood pooling), you can stop before 3 minutes. Expect some days to be better than others. Continue at least 1 week. If you see improvement after 1 week, increase the time by 30 seconds. If not, maintain time at 3 minutes until you do see improvement at which point you may increase the time by 30 seconds.
- Remaining upright as much as possible during day.
Activity Instructions

• Decreasing activity level at home
  – Spreading out/modifying chores throughout the week
    • laundry, house cleaning, cooking, etc
  – Refraining from additional leisure activities (unless cleared by PT)
    • Hiking with friends
    • Jogging
    • Participating in group exercise classes
• Exercising at the appropriate exertion level for the training zones is a must.
• No activity should exceed a 15 on RPE scale.
Long Term Outcomes and Maintenance

- Typically increased fatigue for first month
- Patients generally require 2-3 months of PT 2-3 times per week 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 – Exercise should be treated like a drug
- Adherence to exercise regimen is crucial to making improvement
- 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)
• Arthrochalasemia 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 overstimulated by stretch, but which are poorly supported by collagen fibrils and hence are overstimulated 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
Key point

Joint Hypermobility + Symptoms = Joint Hypermobility Syndrome
Examination of the Hypermobile patient
Aches

Numbness oo00

Pins/Needles 0000

Burning xxxx

Stabbing 1/1

Vision Issues

Facial Pain

Vision Difficulty

Nausea

Tightness

Tight

Shoulders

Elbow Wrist Fingers

Unstable

Headaches

Shoulders

Unstable

Unstable

Unstable

Hips unstable

Knees, lock, click
Unstable pain

Ankles unstable pain

Toes Ditto
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
<table>
<thead>
<tr>
<th>CLINICAL MANEUVER</th>
<th>UNABLE TO PERFORM (0 POINTS)</th>
<th>ABLE TO PERFORM (1 POINT)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Apposition of thumb to forearm</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Right</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>Left</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>Extension of fifth finger beyond 90 degrees</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Right</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>Left</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>Extension of elbow beyond 10 degrees</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Right</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>Left</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>Extension of knee beyond 10 degrees</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Right</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>Left</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>Forward flexion of trunk, legs straight, palms touching floor</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td><strong>Total Beighton Score</strong> (sum of points for each maneuver)</td>
<td>0 to 9 points</td>
<td></td>
</tr>
</tbody>
</table>
Revised Diagnostic Criteria for Ehlers-Danlos Hypermobility Type

**Major Criteria**
- Beighton score of $\geq 4/9$
- Pain for longer than 3 months in $\geq 4$ 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
- Grahame et al 2000
Treatment
Joint Stability

Control or neural and feedback subsystem

- Passive musculoskeletal subsystem
- Active musculoskeletal subsystem
Neutral zone

Normal

Hypermobility Syndrome
Interventions

• Focus on large muscle groups
• Decrease emphasis on stretching
• Less emphasis on passive based treatments
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
- Must keep in mind positional tolerance of patient with POTS when performing strengthening exercise
Selected References


22. Dysautonomia International. www.dysautonomiainternational.org
