

JUL 2019

Volume 3

ISSN 2575-7644 (Print)
ISSN 2575-7652 (Online)

JHBC



Journal of the Heartbeat Clinic



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Journal of the Heartbeat Clinic



Dr. Amer Suleman

Dr. Amer Suleman completed his studies at the Ochsner Clinic in New Orleans, the Mid America Heart Institute, and the Krannert Institute of Cardiology at Indiana University. He also held resident and clinical instructor positions at the State University of New York and the Mayo Hospital (in Pakistan). His Fellowship in Cardiology was completed at Alton Ochsner Medical Center.

In addition to being certified in internal medicine, clinical cardiac electrophysiology, cardiovascular medicine, pacemakers and defibrillators, and echocardiography, Dr. Suleman is a specialist in clinical hypertension. He is Testamur of NASPeXAM in cardiac pacemaker and defibrillator management, and has worked across the state of Texas as a consultant in cardiac electrophysiology and cardiovascular medicine. Over the years, he has participated in numerous clinical trials and investigations.

Dr. Suleman has received several awards and grants, and is a regular contributor to medical and scientific communities as both a lecturer and writer. Currently, he serves as CME Editor of Cardiology for www.eMedicine.com, the largest source of medical literature on the Internet. Dr. Suleman is an Adjunct professor at UT Dallas and Chief of Cardiology at Heartbeat Clinic. He is Editor-In-Chief of JHBCC and has a passion contributing research to the field.



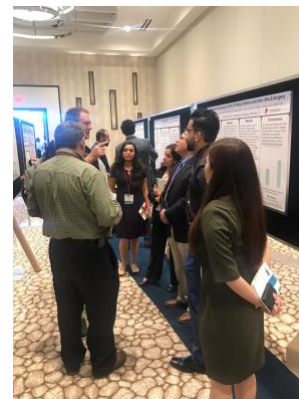
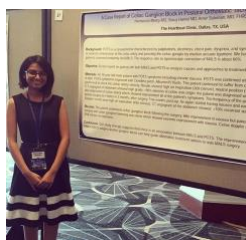
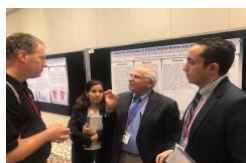
Dr. Sami B. Alam

Salam@sunbeamresearch.com

Dr. Sami Bin Alam is the Editor of JHBCC. He was born in New York and has lived in North Carolina, Ohio and now Texas. He obtained his Medical Education from Dow International Medical College in Karachi, Pakistan. He has completed the USMLE Steps (Step 1 | Step 2 CS, CK | ECFMG certified). He joined The Heartbeat Clinic in 2017 as the Research Director and since been published 30+ times. He aims to continue pursuing his career in medicine by securing a Family Medicine residency and supplementing that with a Board Certification in Lifestyle or Integrative Medicine. Other medical interests beyond the clinic are within the realms of Community Medicine, Global Health, Medical Journalism, Exercise, Nutrition, Stress Management, and Social Connection.

Journal Designed By: Jeanana Khan

Dr. Amer Suleman and the Heartbeat Clinic Team @ Medical Conferences





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Patient's Perspective

A Day in the Life with POTS

When you wake up in the morning, what are some of the first challenges you face?

Taking a shower is the toughest part of the morning. I can't stand up while in the shower at all. My symptoms are worse in the morning just after I wake up, which doesn't help much either.

What are your daily symptoms?

Common everyday symptoms for me are tachycardia, fatigue, dizziness, blurred vision, shortness of breath, near faints, nausea, heat intolerance, and hypotension. Throughout the day I have periods of time where walking and standing causes so much fatigue, shortness of breath, and hypotension that I have to stop what I'm doing to sit or lay down for a while and recover. This takes out a big chunk of time.

What are some things you do in order to recover from these symptoms throughout the day?

I take my medication daily, but other than that I drink a lot of water. I also take electrolyte tablets to adjust my electrolyte levels. Each time I do get out of breath or feel as if my blood pressure is dropping, I have to stop and lay down somewhere until the symptoms pass.

How has POTS changed your life?

I used to be very active. I used to go to my university, then to work, and then have other evening activities and hobbies. I can't do that anymore; life's discolored. My free time goes by in front of the TV instead because of how POTS affected my body.



Abdominal Binder with Insulin Therapy for better Glycemic Control in a Patient with POTS and DM-Type 1

Wael Almardini*, MD, Hemanshi Mistry, MD, Sabih Alam, MD, Sami Bin Alam, MD, Nida Noor, MD, Mohamad Asaad Nasri, MD, Zan Rehman, Merin Rajumon, Luay Ben Gaied, Belaal Sheikh, MD, Amer Suleman, MD

Postural Orthostatic Tachycardia Syndrome (POTS) is a heterogeneous group of disorders and a form of autonomic dysfunction that could be associated with other comorbidities. We hereby present a case of POTS and Diabetes Mellitus type 1 treated with subcutaneous insulin injections, and abdominal binder for better glycemic control.

History: An 18 year-old female with Diabetes Mellitus type 1 and Postural Orthostatic Tachycardia Syndrome (POTS) presented with longstanding symptoms of dizziness, lightheadedness, palpitations and syncope. A detailed workup also revealed a moderate form of hypermobility. Patient complained of having difficulty in controlling her blood sugar levels with the use of subcutaneous insulin Lispro injections after meals, and subcutaneous insulin glargine injections before going to bed. She used continuous glucose monitoring to check random blood glucose levels that ranged between 300–500 mg/dL. A therapeutic trial of using an abdominal binder with insulin injections for 1 week was suggested to see if this could help in achieving better glycemic control.

Theoretically, the abdominal binder should help with improving circulation and decreasing the effect of blood pooling associated with POTS, and possibly improving absorption of insulin to the circulatory system. During the therapeutic trial, the patient used an elastic abdominal binder during the day, Lispro insulin was injected after meals, and Glargine insulin was injected before bed time. On day 4 post treatment, normalization in blood glucose levels were appreciated using a continuous glucose monitoring device. The patient continued using the abdominal binder with her insulin therapy, and her average random blood glucose level was 170 mg/DL post treatment.

Conclusion: In patients with POTS and DM type 1. The use of an abdominal binder along with insulin injections may help in achieving better glycemic control. This could be explained by increasing circulation in POTS patients, which may facilitate the absorption of insulin subcutaneously injected in the abdominal area. Therefore, we suggest that patients with POTS and signs of blood pooling may benefit from the use of abdominal binder in combination with the insulin therapy to achieve a tighter blood glucose control. Further studies should be conducted in order to better understand this implementation.



Incidence of May-Thurner Syndrome in Postural Orthostatic Tachycardia Syndrome (POTS) Patients

Ameen Khan*, Merin Rajumon, Zan Rehman, Sami B. Alam, MD, Hemanshi Mistry, MBBS, Wael Almardini, MD, Nida Noor, MD, Luay Ben Gaied, Sabih Alam, MD, Mohamad Asaad Nasri, MD, Belaal Sheikh, MD, Amer Suleman, MD

Background: Patients with Autonomic Dysfunction, such as POTS, often have co-existing vascular disorders. May-Thurner Syndrome refers to a condition where the left iliac vein is compressed by the right iliac artery, which increases the risk of deep vein thrombosis (DVT) in the left extremity. Often, the syndrome acts as a permissive lesion and becomes symptomatic when something else happens such as, following trauma or a change in functional status. An incidental compression of 50% or more in the Left Iliac Vein is seen in about a quarter of the General Population.

Objective: We aim to study the Prevalence of Prevalence of May-Thurner Syndrome in POTS patients.

Method: We did a retrospective study of 876 patients randomly selected from our pool of POTS patients. Their electronic medical records were reviewed and the CT Angiogram Abdomen & Pelvis were evaluated for their diagnosis. Patients were diagnosed based on a compression of 50% or more of the Left Iliac Vein.

Results: 209 (23.74%) out of 876 POTS patients (age = 32.94 ± 12.21) were tested for May-Thurner Syndrome. 33 of 209 (15.79%) tested Positive whereas 176 of 209 Patients (84.21%) tested Negative for May-Thurner. Out of the 33 Patients that tested Positive for May-Thurner; 2 (6.06%) were Male (Age 31.5 ± 4.95) whereas 31 (93.94%) were Female (Ages 30.97 ± 11.11). 2 out of 33 (6.06%) had Leg Edema and 0 out of 33 (0.00%) had a DVT.

Conclusion: Our study reports show that out of 209 POTS patients that had CT Abdomen & Pelvis for May-Thurner testing, approximately 15.79% tested positive. Therefore, we do not see an increased incidence of May-Thurner Syndrome in POTS patients compared to the general population. Further studies must be conducted in order to better understand this relationship.



Compression and Velocity of the Left Renal Vein as a Measure of Severity of Nutcracker Syndrome in Postural Orthostatic Tachycardia Syndrome (POTS) Patients

Zan Rehman*, Merin Rajumon, Belaal Sheikh, MD, Sami B. Alam, MD, Wael Almardini, MD, Hemanshi Mistry, MBBS, Ameen Khan, Nida Noor, MD, Luay Ben Gaied, Sabih Alam, MD, Mohamad Asaad Nasri, MD, Amer Suleman, MD

Background: Previously, we have seen a relationship between Nutcracker Syndrome and POTS. Nutcracker syndrome is the compression of left renal vein between Abdominal Aorta (AA) and Superior Mesenteric Artery (SMA) causing symptoms such as flank pain, lower abdominal pain, hematuria, pelvic congestion, varicocele and varicose veins due to the disturbance of blood flow from the Left Renal Vein (LRV) into the Inferior Vena Cava.

Objective: We aim to study the Compression and Velocity of the Left Renal Vein in POTS patients as a possible indicator of the severity of Nutcracker Syndrome based on prevalence of symptoms.

Method: We did a retrospective study of 876 POTS patients referred to our clinic. Electronic medical records were reviewed and the CT angiograms of Abdomen (w/ & w/o contrast) evaluated for Nutcracker Syndrome and its respective symptoms. Renal Dopplers were assessed and Left Renal Vein velocities were collected. The findings from both tests were compared to each other.

Results: 208 (23.74%) out of 876 POTS patients were tested for Nutcracker Syndrome. 24 of 208 (11.54%) Patients were diagnosed with Nutcracker syndrome and had data on their Systolic Velocity. Out of the 24 Patients that had Nutcracker syndrome; 1 (4.17%) was Male (Age 37) whereas 23 (95.83%) were Female (Ages 28.96 ± 10.59). Our findings showed that 5 out of 24 patients suffered from Lower Abdominal Pain (20.83%); 6 out of 24 patients suffered from Leg Pain [(25%) 3 in only the Left Leg and 3 in Both Legs]; 6 out of 24 patients showed Pelvic Congestion (25%); 4 out of 24 patients had Flank Pain [(16.67%) 3–Left Flank and 1–Right flank]; 3 out of 24 patients had Varicose Veins (12.50%); and 2 out of 24 patients suffered from Hematuria (8.33%). 7 out of 24 (29.17%) patients had Mild Compression; 17 out of 24 (70.83%) patients had 50% or More Compression. Mean Systolic LRV Velocity = (31.33); No correlation was observed between Degree of Compression and LRV/RRV Velocity Ratio ($R^2=0.053$); No correlation was observed between Degree of Compression and LRV Systolic Velocity ($R^2=0.021$) and a Pearson Correlation Coefficient (R Value = -0.145) was obtained for the same.

Conclusion: Our study showed that there is no correlation between the compression and velocity of the Left Renal Vein and symptoms of Nutcracker Syndrome. This is a preliminary study and further studies need to be conducted for a better understanding.



Silicone Breast Implant Rupture as a Cause of Postural Orthostatic Tachycardia Syndrome (POTS)

Wael Almardini*, MD, Sami B. Alam, MD, Mohamad Asaad Nasri, MD, Hemanshi Mistry, MBBS, Nida Noor, MD, Sabih Alam, MD, Zan Rehman, Merin Rajumon, Luay Ben Gaied, Belaal Sheikh, MD, Amer Suleman, MD

Postural Orthostatic Tachycardia Syndrome (POTS) is a heterogenous group of disorders that has been associated with multiple etiologies. We hereby present a case of POTS most probably caused by rupture of breast implants.

History: A 41-year-old female presented with an acute onset of symptoms of dizziness, lightheadedness, and syncope after her colonoscopy. A detailed workup revealed joint hypermobility, evidence of exaggerated heart rate response to tilt test that was consistent with postural tachycardia, normal heart rate with deep breathing, normal Valsalva maneuver and no evidence of small fiber neuropathy in Q-SWEAT. A complete diagnostic workup including abdominal ultrasound and referral to an immunologist was done. No definite cause of her symptoms was established and she was stabilized on Florinef and IV fluids. The patient was barely able to get back to work.

During one of her incidental self-exams, she found an asymmetric breast. She consulted the plastic surgeon who had performed her breast implant surgery, and was told follow up after an MRI was done. An MRI scan of the breasts showed rupture of the right breast implant. The area on the right side was found to have gross ecchymosis with inflammation of the right breast. Surgical removal of both breast implants was done and resulted in almost immediate improvement in the patient's symptoms of orthostatic intolerance. A repeat tilt that was done post breast implant removal showed exaggerated heart rate response to tilt that was less than the original tilt table test.

Conclusion: We conclude that in this particular case for a patient with joint hypermobility, breast implant rupture was associated with acute onset of orthostatic symptoms that coincided with a colonoscopy. There was a strong relationship with resolution of the patient's symptoms and removal of the breast implants. It has resulted in significant improvement in the quality of life and the patient is able to return back to work and back to her baseline. We still suspect that because of her joint hypermobility, she may still be prone to developing postural tachycardia from another cause. The patient has been taken off her medications.

We recommend that in appropriate subjects, breast implant rupture should be considered as a potential cause of postural orthostatic tachycardia syndrome, especially in the group of patients with joint hypermobility. MRI of the breast or self-examinations may aid in the diagnosis.



Case Report of Successful May-Thurner Syndrome (MTS) Surgery in Postural Orthostatic Tachycardia Syndrome (POTS)

Sabih Alam, MD, Hemanshi Mistry, MBBS, Wael Almardini, MD, Sami B. Alam, MD, Nida Noor, MD, Mohamad Asaad Nasri, MD, Zan Rehman, Merin Rajumon, Ameen Khan, Luay Ben Gaied, Amer Suleman, MD, FHRS, FAIS, FSCAI*

Background: POTS is a condition characterized by an abnormal increase in heart rate upon changes in posture & presents with an array of symptoms like headache, dizziness, syncope, palpitations, nausea, abdominal discomfort, fatigue, and blood pooling of hands and feet. MTS is a compression of the left common iliac vein (LCIV) by the right common iliac artery (RCIA) and may present with DVT, leg swelling, varicose veins. Theoretically, treating MTS will improve venous flow back to the heart, which will reduce the POTS symptoms. Previously, a case report was presented of a patient who reported no improvement of their POTS symptoms after MTS surgery without a hemodynamic study prior to the procedure.

Method: A case of a 38-year-old female with POTS and MTS symptoms with failure of medical management is presented. Imaging studies such as Mesenteric & Renal Doppler, and Magnetic Resonance Venogram–abdomen & pelvis (MRV–AP) were performed prior to the surgery. Patient proceeded with MTS surgery done through stenting of LCIV and RCIA known as “kissing stents”.

Results: Celiac, and Renal Doppler were normal. MRV–AP showed narrowing of the proximal LCIV distal to the bifurcation of the Inferior Vena Cava as it coursed between the lumbar vertebral body and the RCIA. Significant short-term improvement of the patient’s symptoms after MTS surgery were noted. She reported a significant decrease in her fluctuating heart rate, she was able to stop taking the prescribed Ivabradine; significant decrease in symptoms of dizziness, chest pressure, and palpitations from postural/positional changes. A significant decrease in swelling & discoloration of her lower extremities and slight–moderate increase in her energy levels were also noted.

Conclusion: MTS surgery using two stents, with prior hemodynamic study, was found to be beneficial in relieving the POTS symptoms in this patient. We are currently studying the long term benefits and management of MTS surgery in POTS patients, while using hemodynamic studies to guide us.



A Comparison Study of Heart Rate with Deep Breathing Variability (HRVdb) in Female patients with intermediate Tilt Table Test results V/S Postural Orthostatic Tachycardia Syndrome Patients

Hemanshi Mistry*, MBBS, Belaal Sheikh, MD, Sabih Alam, MD, Wael Almardini, MD, Mohamad Asaad Nasri, MD, Sami B. Alam, MD, Nida Noor, MD, Zan Rehman, Merin Rajumon, Luay Ben Gaied, Ameen Khan, Amer Suleman, MD, FHRS, FAIS, FSCAI

Background: Often, patients with Orthostatic Intolerance (OI) do not meet criteria for Postural Orthostatic Tachycardia Syndrome (POTS) or Orthostatic Hypotension. There was one previous study that showed relative inability to increase HR upon posture changes. This was attributed to enhanced vagal tone. That study used frequency domain analysis for HRVdb. HRVdb is a non-specific but reproducible marker to check for parasympathetic cardiac function.

Objective: We aim to study whether time-domain analysis for HRVdb also shows enhanced cardiovagal tone in Female patients who have intermediate Tilt Test response, compared to female POTS patients.

Method: A retrospective study was performed by randomly selecting 41 OI Female patients using the intermediate tilt table test criteria (HR increase between 15–30 bpm within 10 min of tilt) as our case group. We selected our control group as 50 POTS Female patients (HR increase 30 bpm or exceeds 120 bpm within 10 min of tilt) minimizing the confounders like age and gender. We utilized time-domain analysis for HRVdb results and performed a Chi-Square test to see if there are any significant differences between case and control group.

Results: Our study identified a total of 41 Female OI patients (Age= 36.5 years + 10.4); Their Tilt Table Test results had: Mean HR change= 24.225 bpm + 4.393, SEM= 0.694, and HRVdb mean = 19.014 + 8.633, SEM= 1.365. The study had a total of 50 Female POTS patients (Age= 31.372 years + 11.549); Tilt Table Test results had: Mean HR change = 45.725 bpm + 13.523, SEM=1.893 and HRVdb mean = 21.675 + 8.175, SEM= 1.144. Comparing HRVdb among these two groups showed Chi-square statistic of 1.7144 and p-value of 0.190412.

Conclusion: Our study showed that there was no statistically significant difference in the case group V/S control group and did not show there was enhanced vagal tone in patients with OI symptoms (Non-POTS patients). This is a preliminary study and further studies need to be conducted for better understanding.



Prevalence and Treatment of Small Intestinal Bacterial Overgrowth (SIBO) in Patients with Postural Orthostatic Tachycardia Syndrome (POTS)

Zan Rehman*, Merin Rajumon, Sami B. Alam, MD, Waiel Almardini, MD, Hemanshi Mistry, MBBS, Ameen Khan, Nida Noor, MD, Luay Ben Gaied, Sabih Alam, MD, Belaal Sheikh, MD, Mohamad Asaad Nasri, MD, Amer Suleman, MD

Background: POTS is a form of orthostatic intolerance characterized by a myriad of symptoms including syncope, dizziness, poor concentration, exaggerated heart rate response (palpitations) upon postural changes, fatigue, and nausea. Patients with Autonomic Dysfunction, such as POTS, often present with GI motility issues. SIBO refers to a condition in which large amounts of bacteria are present in the small intestine. The two processes that most commonly predispose to bacterial overgrowth are diminished gastric acid secretion and small intestine dysmotility.

Objective: We aim to study the Prevalence of Small Intestinal Bacterial Overgrowth in POTS patients. We also aim to analyze the improvement of POTS symptoms by treating SIBO in POTS patients.

Method: We did a retrospective study of 876 patients randomly selected from our pool of POTS patients. Their electronic medical records were reviewed and the SIBO tests were evaluated to determine their diagnosis.

Results: 67 (7.65%) out of 876 POTS patients (age = 32.94 ± 12.21) had SIBO testing for clinically unexplained or unresolved Nausea, Vomiting, and Bloating. 39 of 67 Patients (58.21%) tested Positive whereas 28 of 67 Patients (41.79%) tested Negative for SIBO. Out of the 39 Patients that tested Positive for SIBO; 6 (15.38%) were Male (Age 28 ± 9.34) whereas 33 (84.62%) were Female (Ages 26.69 ± 10.08). 14 of 39 SIBO (+) Patients, were prescribed Antibiotics (Rifaximin or Metronidazole) to alleviate associated symptoms out of which 4 patients were available for follow-up results. 2 Patients treated with Rifaximin (200 mg BID) reported improvement in abdominal pain, sleep, dizziness and chest pain. The other 2 patients treated with Metronidazole denied improvement in GI symptoms and/or had no change in symptoms.

Conclusion: Our study reports show that out of 67 POTS patients that had SIBO testing, approximately 58.21% tested positive and those that were treated with Rifaximin reported an improvement in their POTS and GI symptoms. It has been clinically observed, that POTS patients respond to lower doses of medication than the general population. Our study shows that SIBO is a pathology that can be associated with POTS and must be acknowledged for more comprehensive treatment. Further studies must be conducted in order to better understand this relationship.



Sleep Indices and Patterns in Postural Orthostatic Tachycardia Syndrome (POTS)

Chandralekha Ashangari, Amer Suleman, MD. (American Society of SLEEP 2015 Annual Meeting; Seattle, WA)
Journal of Sleep and Sleep Disorders Research. ISSN: 0161-8105, Volume 38,2015.

Objectives: The aim of this study is to assess Sleep Indices and Patterns in Postural Orthostatic Tachycardia Syndrome (POTS)

Background: POTS affects primarily young women. POTS patients have a wide variety of symptoms. Poor sleep is one of the symptom found in the POTS patients.

Methods: 51 patients with POTS were referred to our clinic from January to November. Sleep study is done in all the patients. During the sleep study, surface electrodes are put on the patients face and scalp and electrical signals are recorded by the measuring equipment. These signals, which are generated by brain and muscle activity, are then recorded digitally. Belts will be placed around the chest and abdomen to measure the breathing. A bandage-like oximeter probe will be put on the patient's finger to measure the amount of oxygen in the blood. Epworth Sleepiness Scale, N, N2, N3, REM stages, Apnea/Hypopnea Index (AHI), SpO2 Sleep HR bpm, PLMS index and Total sleep efficiency (TSE) are recorded .

Results: Out of 51 patients, 88% are females (n=45; age 29.20 ± 9.26), 12% are males (n=6; age 28.50 ± 13.64) Epworth scale (10.21 ± 5.19); N1 ($8.94\% \pm 5.68\%$); N2 ($56.74\% \pm 10.69\%$); N3 $19.63\% \pm 9.718\%$; REM (15.60 ± 6.34) , AHI (1.59 ± 3.18), Spo2 (Spo2 = 86.66 ± 14.06) , Sleep HR bpm (72.57 ± 7.94 bpm), PLMS index (3.32 ± 7.53) , TSE% ($69.14\% \pm 15.50\%$), Out of 51 patients, 21 patients had >10 Epworth scale; 24 patients had N1 <8% and 27 patients had N1 >8%; 4 patients had N2 <45% and 47 patients had N2 >45%; 23 patients had N3 <22% and 28 patients had N3 >22%; 7 patients had REM >23% and 44 patients had REM <23%; 6 patients had AHI >5 and 45 patients had AHI <5; 45 patients had < 5 PLMS index and 6 patients had > 5 PLMS index; 6 patients had >85% TSE and 45 had <85% TSE, Snoring observed in 48 patients (11 patients had Severe snoring BMI 30.11 ± 5.20 ; 18 patients had moderate snoring BMI 27.46 ± 6.22 ; 19 Patients had mild snoring 20.37 ± 4.62)

Conclusion: Patients with POTS had increased N1, N2, N3, daytime sleepiness, Sleep apnea, Periodic limb movements, Snoring, decreased REM sleep stages and decreased Total sleep efficiency. 2. Sleep heart rate is normal in all the patients.



Exercise Capacity in Postural Orthostatic Tachycardia Syndrome (POTS) Patients through Six Minute Walk test and its Correlation with Delta Heart Rate from Tilt Table Test

Chandralekha Ashangari, Amer Suleman, MD. (American Autonomic Society 25th Annual Conference, November 2015 Puerto Rico) (Late Breaking Abstract).

Objectives: The purpose of this research is to understand the relation of delta heart rate from tilt table test and number of laps covered in 6 Minute walk test.

Background: The Postural Orthostatic Tachycardia Syndrome (POTS) affects primarily young women. The purpose of the six minute walk test is to assess exercise tolerance in chronic respiratory disease and heart failure. We used this test as a performance based measure of functional exercise capacity in POTS.

Methods: Six minute walk test was done in 50 patients at Heartbeat Clinic and delta heart rate was recorded from tilt table test in Autonomic lab. Comparisons were analyzed with Correlation coefficient between number of laps (1 Lap=51 Meters) covered in Six Minutes with delta heart rate in Postural Orthostatic Tachycardia Syndrome.

Results: Out of 50 patients 46 were females and 4 were males. There is a moderate correlation between the delta heart rate recorded from tilt table test and number of laps covered in Six Minute walk test with correlation coefficient of 0.32.

Patients with Borg scale more than 6 covered less than or equal to 9 laps and those with less than 6 Borg scale covered more than 10 laps with a strong correlation coefficient of -0.60 .

Conclusion: There is a significant correlation between Borg scale and number of laps covered in a six minute walk test ,There is a moderate correlation between delta heart rate recorded from tilt table and number of laps covered in a six minute walk test.



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A Comprehensive Echocardiogram Study in Postural Orthostatic Tachycardia Syndrome (POTS)

Chandralekha Ashangari, Amer Suleman. (American Heart Association-BCVS 2015 Conference, New Orleans, LA) Circulation Journal. ISSN 1524-4539 Sep/Oct issue supplement.

Objectives: The aim of this study is to determine the echocardiographic findings in Postural Orthostatic Tachycardia Syndrome (POTS) patients.

Background: The Postural Orthostatic Tachycardia Syndrome (POTS) affects primarily young women. POTS is a form of dysautonomia that is estimated to impact between 1,000,000 and 3,000,000 Americans, and millions more around the world.

Methods: 320 POTS patients were referred to our clinic, 73 POTS patients were randomly selected and underwent an echocardiogram. Echocardiogram results were reviewed comprehensively in all the patients and we performed cumulative distribution of comprehensive echocardiogram results in the top 10 percentile to see the abnormalities.

Results: Out of 73 patients, 96% are females (n=70; age 29.20 ± 9.26), 4% are males (n=3; age 28.50 ± 13.64). Our findings are as follows (mean \pm standard deviation) RVDD (2.37 cm \pm 0.30 cm), LVED (4.34 cm \pm 0.35 cm), IVS (0.82 cm \pm 0.12 cm), LVEF (63.7% \pm 2.61%), LVES (2.93 cm \pm 0.25 cm), LVPW (0.83 cm \pm 0.13 cm), LVOT Diam (1.82 cm \pm 0.20 cm), AO Root (2.70 cm \pm 0.31 cm), LA Diam (3.13 cm \pm 0.36 cm), AV Velocity (1.12 m/sec \pm 0.13 m/sec), LVOT Vel (0.83 m/sec \pm 0.31 m/sec), Mean Gradient (2.81 mmHg \pm 0.68 mmHg), Peak Gradient (4.91 mmHg \pm 1.40 mmHg), AVA (2.65 cm² \pm 0.44 cm²), E Velocity (0.77 m/sec \pm 0.13 m/sec), A Velocity (0.54 m/sec \pm 0.10 m/sec), E/A Ratio (1.45 \pm 0.36), Mean Gradient (0.83 mmHg \pm 0.29 mmHg), Peak Gradient (2.21 mmHg \pm 0.82 mmHg), PHT (48.25 m/sec \pm 11.13 m/sec), MVDT (151.40 m/sec \pm 31.44 m/sec), MVA (4.85 cm² \pm 1.12 cm²), Mitral Regurgitation (Mild) in 82 % patients TV Velocity (0.61 m/sec \pm 0.13 m/sec), RAP (est) (10 mmHg \pm 0 mmHg), TR Velocity (20.78 m/sec \pm 5.62 m/sec), RSVP (est) (29.04 \pm 4.54), TR Mild in 86 % Patients, PV Vel (0.72 m/sec \pm 0.16 m/sec). Top 10 percentile parameters are follows RVDD 2.75 cm, LVED 4.79 cm, IVS 0.97 cm, LVEF 67.04 %, LVES 3.25 cm, LVPW 0.97 cm, LVOT Diam 2.08 cm, Aortic Root 3.10 cm, LA Diam 3.59 cm AV Velocity 1.29 m/sec, LVOT Vel 1.23 m/sec, Mean Gradient 3.68 mmHg, Peak Gradient 6.70 mmHg, AVA 3.21 cm², E Velocity 0.94 m/sec, A Velocity 0.67 m/sec, E/A Ratio 1.91, Mean Gradient 1.20 mmHg, Peak Gradient 3.26 mmHg, PHT 62.51 m/sec, MVDT 191.69 m/sec, MVA 6.29 cm² TV Velocity 0.78 m/sec, RAP (est) 10mmHg, TR Velocity 27.98 m/sec, RSVP (est) 34.81, PV Vel 0.93 m/sec.

Conclusion: 1) POTS patients have normal echocardiographic parameters however even the top 10 percentile of these patients had smaller LVEDD. Similarly the internal dimensions of cardiac chambers does appear to be smaller though still within accepted normal parameters. 2) Further studies should be taken to better de-fine this subgroup.



A Comprehensive Echocardiogram Study in Ehlers-Danlos Syndrome

Chandralekha Ashangari, Amer Suleman. (American Society of Echocardiography 2015 Annual meeting Boston MA) Journal of American Society of Echocardiography (JASE) June 2015. Volume 28, Issue 6, B135–B159.

Background: Ehlers–Danlos syndrome (EDS) is a group of inherited disorders marked by extremely loose joints, hyperelastic skin that bruises easily, and easily damaged blood vessels. The apparent high prevalence of cardiovascular abnormalities in hospitalized patients with Ehlers–Danlos syndrome necessitates a careful cardiovascular evaluation. The aim of this study is to determine the echocardiographic findings in Ehlers–Danlos syndrome (EDS) patients

Methods: 139 EDS patients were selected randomly from our clinic and underwent echocardiogram. Echo–cardiogram results were reviewed comprehensively in all the patients and performed cumulative distribution of results in top 10 percentile to see the abnormalities.

Results: Out of 139 patients (n=139), 96% are females (n=133; age 31.88 ± 11.36), 4% are males (n=6; age 23.83 ± 9.19). Our findings are as follows (mean \pm standard deviation) RVDD ($2.27 \text{ cm} \pm 0.20 \text{ cm}$), LVED ($4.45 \text{ cm} \pm 0.43 \text{ cm}$), IVS ($0.79 \text{ cm} \pm 0.11 \text{ cm}$), LVEF ($61.9\% \pm 2.48\%$), LVES ($2.93 \text{ cm} \pm 0.25 \text{ cm}$), LVPW ($0.82 \text{ cm} \pm 0.12 \text{ cm}$), LVOT Diam ($1.82 \text{ cm} \pm 0.21 \text{ cm}$), AO Root ($2.70 \text{ cm} \pm 0.33 \text{ cm}$), LA Diam ($3.17 \text{ cm} \pm 0.33 \text{ cm}$), AV Velocity ($1.11 \text{ m/sec} \pm 0.14 \text{ m/sec}$), LVOT Vel ($0.82 \text{ m/sec} \pm 0.31 \text{ m/sec}$), Mean Gradient ($2.92 \text{ mmHg} \pm 0.68 \text{ mmHg}$), Peek Gradient ($4.81 \text{ mmHg} \pm 1.40 \text{ mmHg}$), AVA ($2.85 \text{ cm}^2 \pm 0.34 \text{ cm}^2$), E Velocity ($0.77 \text{ m/sec} \pm 0.17 \text{ m/sec}$), A Velocity ($0.64 \text{ m/sec} \pm 0.10 \text{ m/sec}$), E/A Ratio (1.42 ± 0.34), Mean Gradient ($0.81 \text{ mmHg} \pm 0.27 \text{ mmHg}$), Peak Gradient ($2.22 \text{ mmHg} \pm 0.81 \text{ mmHg}$), PHT ($48.21 \text{ m/sec} \pm 11.16 \text{ m/sec}$), MVDT ($157.40 \text{ m/sec} \pm 30.33 \text{ m/sec}$), MVA ($4.72 \text{ cm}^2 \pm 1.16 \text{ cm}^2$), Mitral Regurgitation (Mild) in 87 % patients TV Velocity ($0.69 \text{ m/sec} \pm 0.16 \text{ m/sec}$), RAP (est.) ($10 \text{ mmHg} \pm 0 \text{ mmHg}$), TR Velocity ($20.88 \text{ m/sec} \pm 6.62 \text{ m/sec}$), RSVP (est.) (29.08 ± 6.54), TR Mild in 83 % Patients. PV Vel ($0.92 \text{ m/sec} \pm 0.23 \text{ m/sec}$) Top 10 percentile parameters are follows RVDD 2.53 cm, LVED 5.00 cm, IVS 0.93 cm, LVEF 65.08 %, LVES 3.25 cm, LVPW 0.96 cm, LVOT Diam 2.09 cm, Aortic Root 3.12 cm, LA Diam 3.59 cm, AV Velocity 1.29 m/sec, LVOT Vel 1.22 m/sec, Mean Gradient 3.79 mmHg, Peek Gradient 6.60 mmHG, AVA 3.29 cm², E Velocity 0.99 m/sec, A Velocity 0.77 m/sec, E/A Ratio 1.86, Mean Gradient 1.16 mmHg, Peak Gradient 3.25 mmHg, PHT 62.51 m/sec, MVDT 207.80 m/sec, MVA 6.21 cm², TV Velocity 0.90 m/sec, RAP (est) 10mmHg, TR Velocity 29.36 m/sec, RSVP (est.) 37.46, PV Vel 1.21 m/sec.

Conclusion: Our study attempts to define echo characteristics of a large cohort of patients with Ehlers Danlos Syndrome. In this subgroup no ventricular or atrial enlargement is seen. There is no increased prevalence of mitral valve prolapse seen. The increased LVOT and aortic velocities may be related with Hyperadrenergic state that happens in these patients. Diastolic parameters are preserved.



Headache Symptoms in Postural Orthostatic Tachycardia Syndrome (POTS)

Chandralekha Ashangari, Amer Suleman. (57th Annual Scientific Meeting of American Headache Society Washington DC) *The Journal of Head and Face Pain*. Volume 55, Issue Supplement S3.P127–190.

Objectives: The aim of this study is to determine the Headache Symptoms in Postural Orthostatic Tachycardia Syndrome (POTS) patients.

Background: The Postural Orthostatic Tachycardia Syndrome (POTS) affects primarily young women. POTS is a form of dysautonomia that is estimated to impact between 1,000,000 and 3,000,000 Americans, and millions more around the world. Symptoms of orthostatic intolerance and often severe headaches may be of such intensity that the patient may be functionally disabled, the etiology is unclear but appears to reflect a transient period of autonomic imbalance that occurs in rapidly growing adolescents.

Methods: 146 female Postural Orthostatic Tachycardia Syndrome patients' proceedings were reviewed comprehensively from Electronic Medical records and performed Data analysis (Patients age mean \pm standard deviation and percentage calculation).

Results: Out of 51 patients, 96% are females ($n=45$; age 29.20 ± 9.26), 4% are males ($n=6$; age 28.50 ± 13.64); Out of 51 patients 43 patients (84%) had headache symptoms in which 22 patients (43%) had Tension-Type Headache, 9 patients (18%) had chronic headaches, 7 patients (14%) had Migraine headache, 5 patients (10%) had Sinus headaches.

Conclusion: Patients with Postural Orthostatic Tachycardia Syndrome (POTS) have a very high prevalence of Headache symptoms.



Headache Symptoms in Ehler-Danlos Syndrome (EDS)

Chandralekha Ashangari, Amer Suleman, MD. (57th Annual Scientific Meeting of American Headache Society Washington DC) *The Journal of Head and Face Pain*. Volume 55, Issue Supplement S3.P127–190.

Objectives: The aim of this study is to determine the Headache Symptoms in Ehlers–Danlos Syndrome (EDS) patients.

Background: Ehlers–Danlos syndrome (EDS) is a heterogeneous group of heritable connective tissue disorders, characterized by articular (joint) hypermobility, skin extensibility and tissue fragility. Chronic recurrent headaches may constitute the neurologic presentation of EDS in the absence of structural, congenital or acquired CNS lesions that correlate with their symptoms. Individuals with EDS may be prone to migraine due to an inherent disorder of cerebrovascular reactivity or cortical excitability.

Methods: 139 patients referred to our clinic from January to December with EDS. Reviewed the medical records of 139 EDS patients for headache symptoms.

Results: Out of 139 patients (n=139), 96% are females (n=133; age 31.88 ± 11.36), 4% are males (n=6; age 23.83 ± 9.19). Out of 139 patients, 97 patients (69.7%) had headache symptoms in which 45 patients (32%) had Tension–Type Headache, 25 patients (18%) had Migraine headache, 13 patients (9%) had chronic headaches, 11 patients (8%) had combination of Migraine and Tension–Type Headache, 3 patients (2%) had Sinus headaches.

Conclusion: Patients with Ehlers Danlos Syndrome (EDS) have a very high prevalence of Headache symptoms.



Median Arcuate Ligament Syndrome in Postural Orthostatic Tachycardia Syndrome (POTS)

Chandralekha Ashangari, Amer Suleman, MD, Tam H.Le. *Proceedings of International Society for Autonomic Neuroscience (ISAN) 2015 Annual conference In conjunction with Federation of European Autonomic Societies (EFAS), American Autonomic Society (AAS), Japan Society of Neurovegetative Research (JSNR), Stresa, Italy. The Journal of Autonomic Neuroscience: Basic and Clinical. ISSN: 1566-0702.*

Background: POTS is a form of orthostatic intolerance that is associated with the presence of excessive tachycardia and symptoms like fatigue, headaches, lightheadedness, heart palpitations, exercise intolerance, nausea, diminished concentration, tremulousness (shaking), syncope (fainting), coldness or pain in the extremities, chest pain and shortness of breath upon standing. Median arcuate ligament syndrome (MALS, also known as celiac artery compression syndrome) is a condition characterized by abdominal pain, delayed gastric emptying, nausea, weight loss and other symptoms of autonomic dysfunction attributed to compression of the celiac artery and possibly the celiac ganglia by the median arcuate ligament. The researchers suggest that MALS should be considered in POTS patients who have persistent gastrointestinal symptoms.

Objectives: The purpose of this research is to study the celiac artery velocity in in Postural Orthostatic Tachycardia Syndrome (POTS) patients.

Methods: 93 patients are referred to our clinic with POTS. Celiac Doppler is conducted in POTS patients with family history of MALS and persistent gastric symptoms. Peak systolic velocity (PSV), End Diastolic velocity (EDV) is calculated in all the patients. PSV greater than 200 cm/s, EDV greater than 55 cm/s are suggestive of celiac artery stenosis associated with MALS.

Results: Out of 93 patients, 95% patients are Females (n=88, age 28.88 ± 9.36), 5% patients are Males (n=5, age 25.83 ± 6.19). PSV (cm/sec) 223.84 ± 109.52 , EDV (cm/sec) 71.07 ± 54.46 , PSV (neutral) 179.77 ± 83.01 , PSV Expiration 218.58 ± 109.79 . PSV (cm/sec) > 200 in 51 patients (55%), EDV (cm/sec) > 55 in 50 patients (54%), PSV Expiration (cm/sec) > 200 in 52 patients (56%). PSV (cm/sec) > 300 in 18 Patients.

Conclusion: Our research study demonstrated that higher percentages of POTS patients (>50%) are found to have MALS. MALS was found to be more common in females POTS Patients. There appears to be tremendous improvement in symptoms of two POTS patients after surgery for MALS.



Ehlers–Danlos syndrome (EDS) and its Associated Dermatologic Symptoms

Chandralekha Ashangari, Amer Suleman, MD. (American Academy of Dermatology 2015 Summer Academy Meeting - New York, NY)

Objectives: The aim of this study is to determine the associated Dermatologic Symptoms in Ehlers–Danlos syndrome (EDS) patients.

Background: Ehlers–Danlos syndrome (EDS) is a group of inherited disorders characterized by an increased elasticity of the skin, hyperextensibility of the joints, and increased fragility of the skin and blood vessels

Methods: 203 patients randomly referred to our clinic from January 2014 to December 2014 with EDS were included in the study, reviewed the medical records of 203 patients and dermatologic symptoms.

Results: Out of 203 patients, 193 patients are female (95 %; n=193 age 30.88 ± 10.36), 5 are males (5% ;n=5; age 22.83 ± 8.19), Our findings are as follows 101 patients (50%) had loose skin, Fragile skin, Poor wound Healing, Bruising, 33 patients(16%) had Discoloration of legs and arms and multiple concussions, 30 patients (15%) had Eczema, Fungemia, Keratosis pilaris, Dark skin pigmentation, 15 patients (7%) had Folliculitis, Lupus, Nevus, 15 patients (7%) had Skin Rash, Nevus, Rosacea, Sjogren syndrome, 5 Patients (2%) had Translucent skin, 2 Patients (1 %) had Skin cancer.

Conclusion: Our Study indicates that Patients in Ehlers–Danlos syndrome (EDS) has strong association with Dermatologic Symptoms (92% EDS patients have Dermatologic Symptoms).



Detecting Resting Adrenal Hyperactivity by Analyzing PR Intervals in patients with Postural Orthostatic Tachycardia Syndrome

Ramtej Atluri*, MBBS, Vishak Kumar, MD, Sri Mandava, MBBS, Yashi Bu, PhD, Sami bin Alam, MD, Telle Ang, Medical Student, Arathi Kumar, MBBS, Amroo Dada, MPH, Belaal Sheikh, MD, Amer Suleman, MD, FHRs, FSCAI (ENDO Annual Meeting 2019 - New Orleans, LA)

Introduction: Postural Orthostatic Tachycardia Syndrome (POTS) is a form of dysautonomia which primarily affects young women (15–45 years of age). These patients frequently experience symptoms of orthostatic intolerance in response to postural changes. Autonomic reflexes are often spared in many of these patients and the hallmark of this condition is an exaggerated heart rate increase in response to changes in posture. The symptoms of POTS are often due to cerebral hypoperfusion and include generalized weakness, dizziness, lightheadedness, darkening of the visual fields and in some, palpitations and loss of consciousness.

Diagnostic criteria for POTS are a heart rate increase of greater than 30 beats per minute or an increase to 120 beats per minute or greater within the first 10 minutes of a tilt table testing.

The aim of this study is to determine if patients with severe POTS have resting Adrenal Hyperactivity by utilizing PR-Intervals.

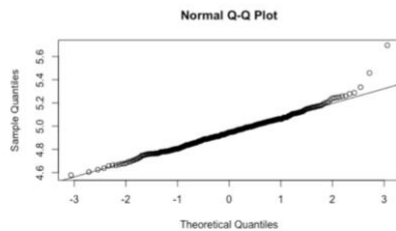
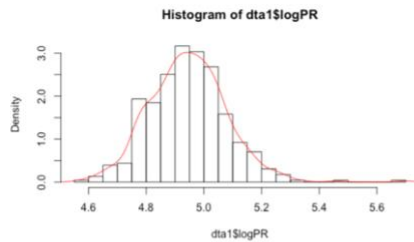
PR interval is determined by the AV node conduction delay which is normally 120–200 ms. Pacemaker action potential is determined by the conduction of Na^+ , Ca^{2+} , and K^+ ions into the pacemaker cells. The Funny current (iF channel or Heterocyclic nucleotide channel) transports Na^+ into the pacemaker cells. The T-type and L-type Ca^{2+} channels transports Ca^{2+} into the pacemaker cells and the K^+ leak channels allow for K^+ to re-enter the pacemaker cells during repolarization. Increased sympathetic activation results in increased cAMP, activation of the iF channel, and increased Na^+ conduction through the iF channel, thus increasing the slope of the pacemaker action potential and increasing the conduction through the AV node. Increased conduction through the AV node results in a shorter PR interval and a faster heart rate.



Methods: A team of researchers randomly selected patients with POTS from our clinic. This group consisted of 455 patients and these patients' tilt table results as well as their resting ECG's were reviewed from their electronic medical records. Patients were then categorized based on the degree of change in heart rate during the tilt table test. *The categories were as follows: Orthostatic intolerance (<30 beats per minute), mild POTS (30–50 beats per minute), and severe POTS (>50 beats per minute).* There were a few patients in the data set that did not have tilt table results so they were removed as we could not categorize these patients. We then noted the resting PR intervals of these patients from their ECG's prior to the day of the tilt, on the day of the tilt, and after the day of the tilt. ECG's on the day of the tilt were taken in the morning prior to the procedure. The mean of these values for each patient were then used. We then converted the data into log scale and then tested for Normality and for Homogeneity of Variance. The log transformed resting PR intervals have normality based on histogram, QQ plot and the Anderson–Darling normality test results. Levene's test for Homogeneity of Variance was then used to determine if there was equal variance across the three categories. With a significance level of 0.05, Levene's test has a P-value slightly higher than 0.05 (0.081) and accepts null hypothesis of homogeneity. With the normality and homogeneity assumptions being verified, it is safe to conduct Analysis of Variance (ANOVA) using the data with log transformed resting PR intervals to determine if there was any statistically significant difference between the various categories. After using ANOVA to analyze the data set, a second team of researchers, blinded to the first team, collected the same data set of patients with POTS from our clinic. This was done in order to minimize observer bias. This team of researchers further filtered out 8 patients whose data did not meet the criteria required to categorize the patients, thus the second data set consisted of 447 patients. Their tilt table results and resting PR intervals were taken from their electronic medical records and the patients were then categorized in the same manner as the first data set. We converted the second data set into log scale and then tested for Normality and for Homogeneity of Variance as well. The log-transformed second data set also had normality based on histogram, QQ plot and the Anderson–Darling normality test. Levene's test for Homogeneity of Variance was then used to determine if there was equal variance across the three categories in the second data set. With a significance level of 0.05, Levene's test has a P-value higher than 0.05 (0.4044) and accepts null hypothesis of homogeneity. Studentized Breusch–Pagan test was also used to determine if there was equal variance across the categories and with a P-value of 0.1063 (at a significance level of 0.05) the null hypothesis of homogeneity was accepted. Once normality and homogeneity assumptions were verified for the second data set, ANOVA was conducted to determine if there was any statistically significant difference between the various categories.



Group 1 (n=455):



Shapiro-Wilk normality test	Anderson-Darling normality test
data: dt.trans W = 0.98063, p-value = 9.2e-06	data: dt.trans A = 0.6264, p-value = 0.1022

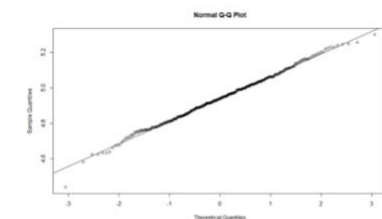
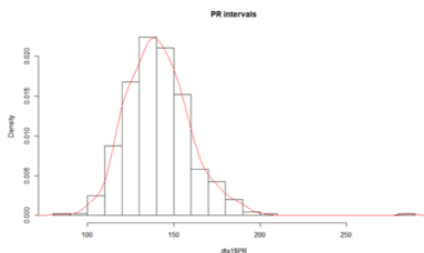
Homogeneity of variance assumption

- Using the log transformed PR data, the homogeneity assumption requires they have equal variance across three POTS groups. Levene test is a test for homogeneity of variance, with null hypothesis of equal variance.

Levene's Test for Homogeneity of Variance (center = median)		
group	Df	F value Pr(>F)
2	2	2.5222 0.08141
452		
Signif. codes: 0 '***' 0.001 '**' 0.01 '*' 0.05 '.' 0.1 ' ' 1		

With a significance level of 0.05, Levene test has a p value slightly higher than 0.05, and accepts null hypothesis of homogeneity.

Group 2 (n=447):



Shapiro-Wilk normality test	Anderson-Darling normality test
data: dt.logPR1 W = 0.99664, p-value = 0.4806	data: dt.logPR1 A = 0.21786, p-value = 0.8401

```
> leveneTest(y = dt.logPR1, group = dt.POT1)
Levene's Test for Homogeneity of Variance (center = median)
Df F value Pr(>F)
group 2 0.9071 0.4044
443
```

Equal variance (homogeneity) assumption

Anderson-Darling normality test	studentized Breusch-Pagan test
data: dt.logPR1 A = 0.21786, p-value = 0.8401	data: dt.logPR1 ~ dt.POT1 BP = 4.4832, df = 2, p-value = 0.1063

In AD test, with pval=0.84, it fails to reject null hypothesis. Namely, AD test supports null hypothesis that the data is homogeneous.

The studentized Breusch-Pagan has p-value = 0.1063. When using a significance level of 0.05, the test also fails to reject null hypothesis, which suggesting data is homogeneous.

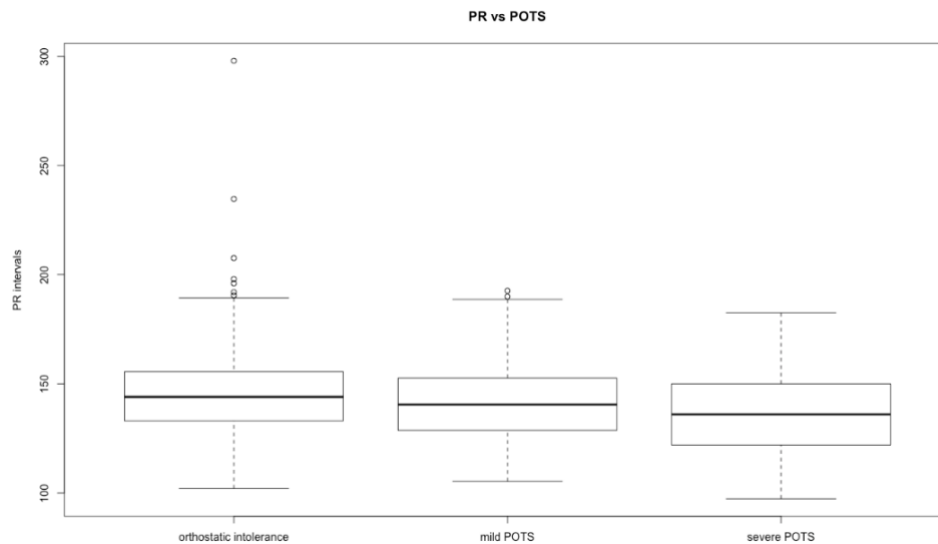


Results: In the group of 455 patients (n=455), 29% (n=133) are categorized as having orthostatic intolerance, 52% (n= 237) are categorized as mild POTS, and 18.6% (n=85) are categorized as severe POTS. Using a significance level of 0.05, the P-value was lower than 0.05 (0.00116), thus ANOVA fails to accept the null hypothesis that the means of the PR-intervals across the various categories are equal. In the second group of 447 patients (n=447), 28.6% (n= 128) are categorized as having orthostatic intolerance, 51.6% (n=231) are categorized as mild POTS, and 19.6% (n=88) are categorized as severe POTS. Once again using a significance level of 0.05, the P-value was lower than 0.05 (0.00787), thus ANOVA fails to accept the null hypothesis that the means of the PR-intervals across the various categories are equal. That is to say, statistically the resting PR intervals are not equal across the different POTS categories in both groups with a significance level of 0.05.

Group 1 (n=455):

Average PR intervals:

- Orthostatic intolerance: 146.88 ms
- Mild POTS: 140.74 ms
- Severe POTS: 137.09 ms



ANOVA

With the normality and homogeneity assumptions being verified, it is safe to conduct ANOVA using the data with log transformed PR intervals.

	Df	Sum Sq	Mean Sq	F value	Pr(>F)
POTS	2	0.243	0.12160	6.86	0.00116 **
Residuals	452	8.012	0.01773		

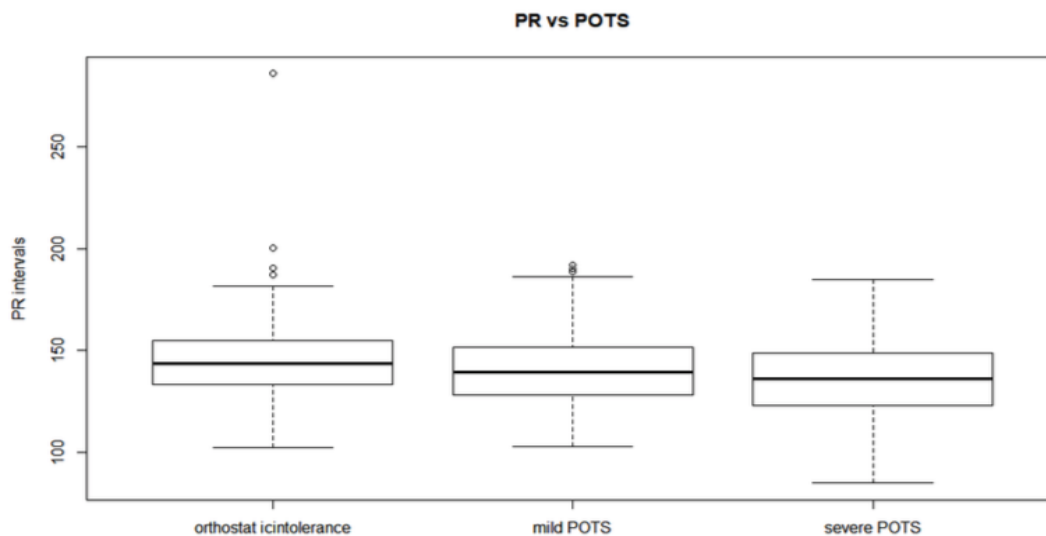
Signif. codes: 0 '***' 0.001 '**' 0.01 '*' 0.05 '.' 0.1 ' ' 1					

Using a significance level of 0.05, the ANOVA fails to accept null hypothesis that group means are equal. That is to say, statistically it can be claimed that PR intervals are not equal across different POTS groups with a significance level of 0.05

**Group 2 (n=447):**

Average PR intervals:

- Orthostatic intolerance: 145.59 ms
- Mild POTS: 140.42 ms
- Severe POTS: 137.0 ms

**ANOVA test**

```
> summary(model.1m)
              Df Sum Sq Mean Sq F value    Pr(>F)
dt.POT1         2   0.155   0.07766    4.898 0.00787 **
Residuals      443   7.024   0.01586

---
Signif. codes:
  0 '***' 0.001 '**' 0.01 '*' 0.05 '.' 0.1 ' ' 1
```

ANOVA null hypothesis is that PR intervals averages are equal for different POTS levels. With a significance level of 0.05, the result p-value = 0.00787 will reject null hypothesis. Thus, it can be claimed that PR intervals are not equal across different POTS groups, with a significance level of 0.05.

Conclusion: There is a statistically significant increase in resting Adrenal Hyperactivity in patients with severe POTS (>50 beats per min) as opposed to patients with orthostatic intolerance (<30 beats per min); the severity of which may have long-term prognostic significance. Future studies will need to be conducted to further investigate the long-term sequelae of the varying degrees of this condition as well as the efficacy of various treatment modalities among the different categories.



Renin Angiotensin Aldosterone System Inactivation in Postural Orthostatic Tachycardia Syndrome

Vishak Kumar, MD, Sabahat Usmani, MD, Sami B. Alam, MD, Muhammad Asad Fraz, MD, Dutt Patel, MBBS, Hemanshi Mistry, MD, Nida Noor, MD, Wael Almardini, MD, Mariam Fayyaz, Thomas Burphy, Amer Suleman, MD, FHRS, FAIS, FSCAI. (ENDO Annual Meeting 2019 – New Orleans, LA)

Abstract: Postural Orthostatic Tachycardia Syndrome (POTS) is a clinical syndrome characterized by the presence of tachycardia in the absence of orthostatic hypotension; symptoms of orthostatic intolerance (presyncope and syncope) are present secondary to autonomic dysfunction (1). It is thought that one of the implications for POTS involves plasma volume disturbances leading to blood pooling. The Renin Angiotensin Aldosterone System (RAAS) is one system that assists in plasma volume regulation (2). In this study we hypothesized that these disturbances are brought about by an inactivation of RAAS. This was done by retrospectively reviewing the medical records of POTS patients. In these patients, diagnosis of POTS was established by an increase in Heart Rate of over 30 bpm within 10 minutes of tilt. Stroke Volume was used as a measure of volume disturbances; those **POTS patients whose stroke volumes had not increased by the End of the Tilt as compared to their 6 minute supine baseline were considered to have an inactivation of RAAS.** Our **total sample of POTS patients** numbered at **447**, out of which **417** patients had lower stroke volumes at the end of their tilt table tests when compared to their baseline; this makes for **92.87% of our POTS Patient Sample.** The average Stroke Volume at the **6 minute mark** was calculated to be **74.67**; the average stroke volume at the **end of the tilt table test (30 minutes)** calculated was calculated to be **46.33**; . The results seen in this study show that **more than 90% of our POTS Patient Sample** there is **no recovery of the stroke volume** by the **end of the tilt table test**, this volume disturbance can be the result of decreased RAAS activation. **This observation is supported by the difference found between average stroke volumes seen at 6 minute baseline and at 30 minutes of tilt.** In normal individuals, we should see an increase in stroke volume at the end of the tilt as RAAS is activated. However, as patients with POTS are known to have lower plasma volumes (2), compounded with the inactivation of RAAS; the resulting hypovolemia will amplify the symptoms of POTS, increasing their severity. These patients should respond well to fludrocortisone, as there will be a correction of hemodynamic impairments (RAAS inactivation) leading to symptomatic relief (3). Further studies and analyses are needed to look into the levels of Renin and Aldosterone at Baseline & at Tilt, as well as the effects of fludrocortisone on such patients with RAAS inactivation.

References: (1) Satish R. R., Circulation. 2013. June 11; 127(23): 2336–2342. (2) Satish R. R. et al., Circulation. 2005. March 2; 111(13): 1574–1582. (3) Freitas. J et al., Clin Auton Res. 2000. October; 10(5): 293–303.



Migraine Treatments Utilized by Postural Orthostatic Tachycardia Syndrome Patients

Tabbassum Fayyaz, BS, Ramtej Atluri, MBBS, Sri Mandava, MD, Sabahat Usmani, MD, Sami B. Alam, MD, Amroo Dada, MPH, Nida Noor, MD, Sabih Alam, MD, Mohamad Nasri, MD, Wael Almardini, MD, Hemanshi Mistry, MD, Amer Suleman, MD, FHRS, FSCAI (American Headache Society – 61st Annual Meeting)

Background: Postural Orthostatic Tachycardia Syndrome is a form of dysautonomia associated with a wide array of symptoms, like dizziness, syncope, anxiety, tremors, palpitations and headaches. POTS and migraines are often comorbid conditions. However, the prevalence and clinical features of headache and how it relates to clinical signs and symptoms in POTS patients have not been widely reported.

Migraine headaches are a debilitating neurological disorder with incapacitating symptoms including throbbing headache confined to one half or the whole head, photosensitivity, nausea, vomiting, sensitivity to sound and odors, paresthesias and visual disturbances.

The aim of this study is to examine the variety of treatment modalities sought by POTS patients suffering from migraines.

Methods: A complete list of POTS patients was generated from the electronic medical records and patients with a diagnosis of migraines were selected. The final list of 316 candidates was prepared and their drug history chart was reviewed retrospectively. The treatment modalities were organized into the following categories: Anticonvulsants, Antidepressants, Triptans, Narcotics, Analgesics, OTC pain relievers (including Excedrin Migraine), Botox and Nerve Block.

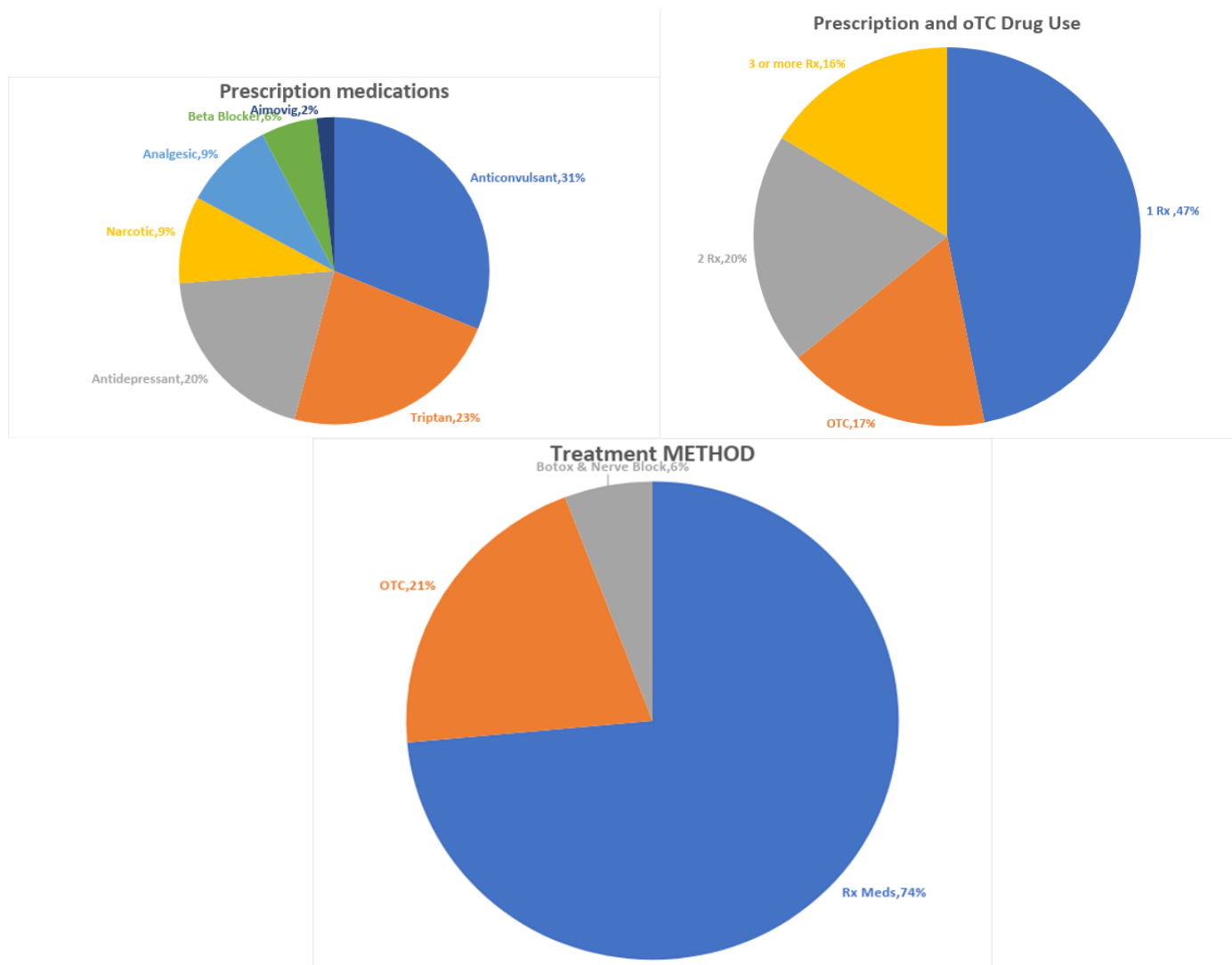
Results: At our clinic, 1142 POTS patients were selected, out of which 316 patients or 27% suffered from migraines. 93.3% were Female (n=295) and 6.7% were Male (n=21). 81.6% (n=258) of those patients reported using medical treatment; 46.9% (n=121) used one prescription medication, 19.8% (n=51) used two prescription medications, 16.3% (n=42) used three or more prescription medications and 17.0% (n=44) used only OTC pain relievers. The following are the major prescription drugs used along with the most common drug in each category: Anticonvulsant 31% (Topiramate 46.2% n=48, migraine prophylaxis), Triptan 23% (Sumatriptan 48.7% n=37, abortive treatment), Antidepressant 20% (Amitriptyline 37.5% n=24, migraine prophylaxis), Narcotic 9% (Hydrocodone 51.6%, n=16), Analgesic 9% (Cafegot 33.1%, n=10), Beta Blocker 6% (Propranolol 84.2%, n=16), and Erenumab 2% (CGRP-R blocker, n=6). In regard to treatment modalities, 73% (n=331) used prescription drugs, 21% OTC (n=93) and 6% Botox and nerve blocks (n=26).



Conclusion: The results of our study suggest that the prevalence of Migraines in POTS patients is 27%. About 47% of patients in our study sought at least one prescription medication, either for migraine prophylaxis (anticonvulsants or antidepressants) or abortive treatment (triptans). It is a crucial point to mention that up to 36% of the patients utilized two or more medications, indicating a link between POTS and severe or unresponsive cases of Migraines. Further studies are needed to explore the possibility of Migraines in this specific patient population being secondary to the underlying Autonomic Dysfunction.

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Incidence of Insomnia in Postural Orthostatic Tachycardia (POTS) Patients

Pruthvi Goparaju, MBBS, Sami B. Alam, MD, Dutt Patel, MBBS, Muhammad Asad Fraz, MD, Nabihah Chaudhary, MBBS, Sabih Alam, MD, Amer Suleman, MD (American Psychiatric Association – 2019 Annual Meeting – San Francisco, CA)

Background: POTS is considered a form of Dysautonomia associated with a heterogeneous array of symptoms and many other co-morbidities. POTS is frequently misdiagnosed for other conditions because it commonly presents with concomitant symptoms that mimic those associated with those conditions. Many POTS patients come in having previously seen a Psychiatrist. Previous research data has shown how POTS can impair one's quality of life physically, mentally, and socially¹. The symptoms of POTS are vast because the Autonomic Nervous System plays an extensive role in regulating various functions and pathways throughout the body.

Sleep and the autonomic nervous system are closely related from an anatomical, physiological, and neurochemical point of view. Sleep disorders may cause or be associated with clinically relevant autonomic dysfunctions. Dysfunctions of cardiovascular and respiratory autonomic control have a significantly negative impact on prognosis of the associated sleep disorder and may represent a risk factor for the development of other chronic diseases or for life-threatening events².

The aim of this study is to determine the incidence of Insomnia present in POTS patients.

Method: As of 2018, 876 POTS patients were randomly selected from our clinic. Patients' electronic medical records were reviewed retrospectively for the diagnosis of depression. Inclusion criteria for POTS patients was a positive Tilt table test and abnormal Autonomic function tests; Insomnia based on DSM-V criteria (Pre-diagnosed from Psychiatric or other clinical settings fit to evaluate sleep disorders).

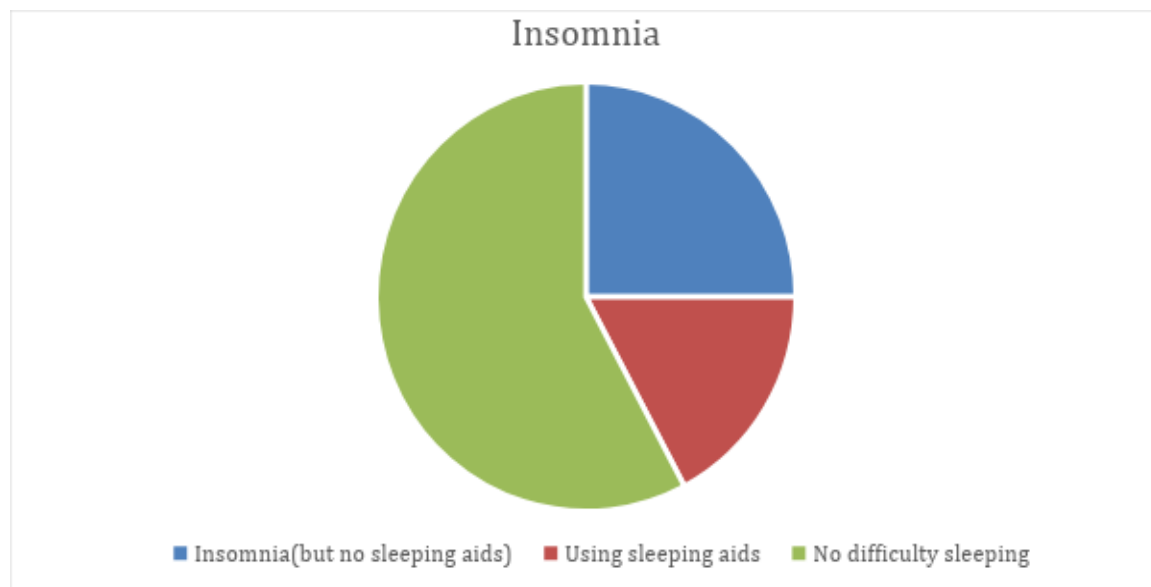
Results: Out of 876 patients, 86.3% are Female (756) and 13.6% are Male (120). 42.2% of those 876 patients are diagnosed with Insomnia (370); out of which 151 patients have been on sleeping aids. The other 219 patients have not been on sleeping aids. So 17% of those 876 patients are diagnosed with Insomnia and have been on sleeping aids (151). Out of the 120 male patients, 30% of those are diagnosed with Insomnia (36) and 12.5% used sleeping aids (15). Out of the 756 female patients, 44.1% of them are diagnosed with Insomnia (334) and 17.9% of them have been on sleeping aids (136). All POTS patients were asked about social and psychological factors on initial and subsequent follow up visits.



Out of the 151 patients who have been on sleeping aids, the following medications have been used:

Zolpidem– 26.4%(40)
 Trazadone–25.8%(39)
 Melatonin–20.5%(31)
 Temazepam–9.2%(14)
 Eszopiclone – 5.9%(9)
 Clonazepam–5.2%(8)
 Alprazolam– 1.9%(3)
 Others – 4.6%(7)

Conclusions: Insomnia can lead to poor quality of life which is one of the associated symptoms in POTS patients and detailed history and examination should be carried out for proper treatment and improvement of quality of life in POTS patients.



Reference:

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² Autonomic dysfunction in sleep disorders.

Pietro Cortelli MD PhD (Dr. Cortelli of the University of Bologna has no relevant financial relationships to disclose.)

Giovanna Calandra-Buonaura MD PhD (Dr. Calandra-Buonaura of the University of Bologna has no relevant financial relationships to disclose.)

Antonio Culebras MD, editor. (Dr. Culebras of SUNY Upstate Medical University at Syracuse received an honorarium from Jazz Pharmaceuticals for a speaking engagement.)



Prevalence of ADHD in Postural Orthostatic Tachycardia Syndrome (POTS) Patients

Dutt Patel, MBBS, Sami B. Alam, MD, Pruthvi Goparaju, MBBS, Muhammad Asad Fraz, MD, Nabihah Chaudhary, MBBS, Sabih Alam, MD, Amer Suleman, MD (American Psychiatric Association – 2019 Annual Meeting – San Francisco, CA)

Background: POTS is form of Dysautonomia associated with a heterogeneous array of symptoms and many other co-morbidities. POTS is frequently mistaken for other conditions because it commonly presents with concomitant symptoms that mimic symptoms associated with other psychiatric conditions. Symptoms like brain fog, lack of attention, and weakened memory and cognitive issues that are likely to be caused by Attention Deficit Hyperactivity disorder (ADHD), are many times also seen with POTS patients. In POTS patients however, treating the symptoms may not treat the underlying condition which is autonomic and neurogenic in nature. Many POTS patients come in having previously seen a Psychiatrist and already having been diagnosed with Attention Deficit Hyperactivity Disorder. POTS is relatively less common and one of the last differentials thought about, if at all.

The aim of this study is to determine the frequency of POTS patients diagnosed with ADHD and the medications prescribed. It is also to raise awareness about POTS for the future reference of psychiatrists.

Method: 876 POTS patients were randomly selected from our clinic. Patients' electronic medical records were reviewed retrospectively for diagnosis of ADHD having been previously diagnosed in a Psychiatric Setting.

Results: Out of 876 POTS patients, 85.5% are female (749) and 14.5% are male (127). 10.5% of those 792 patients are diagnosed with ADHD (92); out of which 88.0% are female (81) and 12.0% are male (11). 85.8% of patients diagnosed with ADHD were treated with Medication (79); out of which 90.2% were female (83) and 9.8% were male (9).



Out of the 151 patients who have been on sleeping aids, the following medications have been used:

- 9.0% of all POTS patients were on ADHD medication (79).
- 5.0% of all POTS patients were taking Adderall (44).
- 2.3% of all POTS patients were taking Vyvanse (20).
- 0.7% of all POTS patients were taking Methylphenidate [Ritalin] (6).
- 0.2% of all POTS patients were taking concerta(2).
- 0.1% of all POTS patients were taking evekeo(1).
- 0.1% of all POTS patients were taking tenex(1).
- 0.2% of all POTS patients were taking guanfacine(2).
- 0.2% of all POTS patients were taking strattera(2).
- 0.1% of all POTS patients were taking Dextroamphetamine(1).

Conclusions: POTS patients can present with symptoms of ADHD because of the overlap, but is rarely considered as a differential by Psychiatrists. These patients are prescribed ADHD medication, which may or may not alleviate symptoms. However there is also a risk that the medication may induce side effects which could possibly exacerbate POTS. Our data suggests that 1 in 10 POTS patients are diagnosed with ADHD and most of them (85.8%) do take medicine for the condition. Medicating patients without finding the underlying cause could possibly lead to unwanted drug dependence or unwanted side-effects. Detailed History, Physical examination, and appropriate referral should be carried out by the attending Psychiatrist. Further clinical studies are required to broaden the area of these discrepancies.



Genetic Testing for Secondary Causes in Dysautonomia Patients

Sabahat Usmani, MD, Sami B Alam, MD, Amer Suleman, MD (American Heart Association -Vascular Discovery: From Genes to Medicine 2019 Scientific Sessions)

Introduction: Dysautonomia is a broad term, which envelops various disorders of the Autonomic Nervous System including Postural Orthostatic Tachycardia Syndrome (POTS), Neurogenic Orthostatic Hypotension (nOH), Postural Orthostatic Intolerance (POI) and many others.

Genetic testing was conducted in Dysautonomia patients to screen for secondary causes like Amyloidosis and for Porphyrrias, which frequently present with symptoms indistinguishable from those of Dysautonomias such as arrhythmias, anxiety, and severe abdominal pain.

The aim of this study is to explore the prevalence of genetic markers in a Dysautonomic patient population for such conditions.

Method: 42 consenting patients were randomly selected to be genetically tested in collaboration with Alnylam Pharmaceuticals, using genetic panels developed by Invitae. 24 patients received both the full 120-gene Cardiomyopathy (CM) & Neuropathy (NP) Panels which includes Transthyretin (TTR) Amyloidosis testing, and the 4-gene Panel for Porphyrria; 18 patients received only the 4-gene Porphyrria panel. Saliva specimens were shipped to Invitae to process the genetic tests. A total of 124 genes were evaluated for sequence changes and exonic deletions/duplications.

Results: Of the total 17 patients that tested positive for genetic variants; 15 were POTS patients, 2 were nOH patients, and 1 was a POI patient. Porphyrria panel testing showed genetic variants in the CPOX gene in 2 patients. In one patient, further genetic testing of an immediate family member (Mother) with a history of similar symptoms revealed the CPOX variant. In the CM and NP Panel, 15 patients showed variants in the following 23 genes: DSP, HARS, MYBPC3, PMP22, RYR2, TTN, PRX, DNM2, LRSAM1, AGL, PLEKHG5, TMEM43, GAA, FLNC, NTRK1, GAN, TRPV4, DST, GDAP1, RBM20, SCN9A, CACNA1C and SBF2. The most frequent variants were RYR2 & FLNC from the Invitae CM Panel and NTRK1 & DST from the Invitae NP Panel.

Conclusion: RYR2 and FLNC genes are both associated with various forms of Cardiomyopathies. NTRK1 and DST genes are associated with different types of Hereditary & Sensory Autonomic Neuropathy. No more than 2 people tested positive for the same variant. While variants were observed in several genes, the pathogenicity of these variants has not been clearly established. Further large scale studies are needed to determine the significance, pathogenicity and familial hereditary patterns of these genes in the Dysautonomic population.



Prevalence of Thoracic Outlet Syndrome in Patients with Postural Orthostatic Tachycardia Syndrome

Arathi Kumar, MBBS, Telle Ang, Rishita Naga, MBBS, Vishak Kumar, MD, Sami B. Alam, MD, Belaal Sheikh, MD, Amer Suleman, MD (Society of Vascular Ultrasound – Annual Conference 2019 – Chicago, IL)

Introduction: Postural Orthostatic Tachycardia Syndrome (POTS) is defined as the presence of symptoms of orthostatic intolerance for at least 6 months accompanied by a heart rate increase of at least 30 beats/min within 5–30 minutes of assuming an upright posture and in the ABSENCE of Orthostatic Hypotension.¹

Thoracic outlet syndrome (TOS) refers to a constellation of symptoms resulting from neurovascular compression at the thoracic outlet, causing some combination of pain in the neck and upper extremity, weakness, sensory loss, paresthesia, swelling, and discoloration. Classification systems have used anatomical structures to subdivide TOS into venous TOS (VTOS)–compression of Subclavian vein, arterial TOS (ATOS)–compression of subclavian artery and neurogenic TOS (NTOS)–compressed brachial plexus.²

The aim of this study is to find the incidence of ATOS and/or VTOS in patients with POTS.

Method: Amongst 1142 patients with POTS, 40 were tested for TOS with imaging (arterial and venous doppler) to look for compromised arterial flow, venous flow or both and whether it was unilateral or bilateral. These 40 patients were also checked for a concurrent diagnosis of Ehlers–Danlos Syndrome as concurrent hypermobility, EDS, is overrepresented in patients with POTS.³

Results: 40 of the 1142 POTS patients (3.5%) were tested for TOS, out of which 20 patients (50%) were positive for it. In this group of 20, 5 patients (25%) had ATOS, 7 patients (35%) had VTOS and 8 patients (40%) had combined Arterial and Venous TOS. Also, in this group of 20, 13 patients (65%) had bilateral TOS and 7 patients (35%) had unilateral TOS. Amongst these 20 patients who tested positive for TOS, 14 patients (70%) also had a diagnosis of EDS.

Conclusion: The study revealed that half the patients with POTS tested for TOS, were positive for it and amongst these, VTOS and bilateral compression were most common. These positives also included a majority of EDS–patients and this could pave way for further research connecting EDS and TOS, supplementing existing literature that Thoracic outlet “symptoms” are common in EDS patients, and are often related to Thoracic Outlet Syndrome (TOS).⁴



JHBC

Journal of the Heartbeat Clinic



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Prevalence of Unilateral vs Bilateral Iliac Vein Compression in May-Thurner Syndrome

Fatima P. Siddiqi, MBBS, Sami B. Alam, MD, Sabahat Usmani, MD, Hemanshi Mistry, MD, Belaal Sheikh, MD, Amer Suleman, MD, Sabih Alam, MD (Society of Vascular Ultrasound – Annual Conference 2019 – Chicago, IL)

Introduction: May–Thurner syndrome (MTS) is a clinical condition in which the right common iliac artery overlies as well as compresses the left common iliac vein. This anatomical variant is present in over 20% of the population but is rarely diagnosed (1). Compression is considered significant if greater than 50% for a diagnosis of MTS. It causes pain and tightness upon activity, with swelling, hyperpigmentation, telangiectasias, or venous ulcerations being indicators on examination (2). The gold standard of diagnosis for MTS is venography, as it is diagnostic but can also be therapeutic. Other modalities include doppler ultrasound, helical abdominal CT, CT venography magnetic resonance venography and intravenous ultrasound (3). In terms of treatment, there are also several options available but catheter based solutions yield the best results (4). This applies specifically to a younger demographic where the correlation between iliofemoral thrombosis and post thrombotic syndrome is high. Stent placement in a patient with thrombotic MTS was first reported by Berger *et al.* (5). More reports have confirmed its effectiveness (6,7,8). Furthermore, one third of patients treated with thrombolysis for iliofemoral DVT who received stenting had considerably greater patency compared to those who did not undergo the procedure. (5). The Guidelines by the Society of Interventional Radiology and the Society of Vascular Surgery choose iliac venous stenting in the setting of external iliac vein compression from the studies that resulted. (9,10).

The purpose of this research is to explore effective diagnosis & treatment modalities for MTS based on its site of occurrence in a population with POTS. This retrospective study investigates the prevalence of Bilateral and Unilateral Iliac Vein Compression in randomly selected patients with May Thurner Syndrome.

Methods: A randomly selected sample of 1142 was taken from the population of current patients. This was conducted through the EMR at The Heartbeat Clinic in Medical City, McKinney. Patients were first filtered to those who suffered from autonomic nervous system disorders and further to those who suffered from Postural Orthostatic Tachycardia Syndrome (POTS) in a retrospective observation. Of the sample, 981 patients were female and 161 were male. Records were then analyzed for those who suffered from May Thurner Syndrome (47 patients were found to have MTS). The minimum level of compression required for inclusion in the sample was 50% (compression of both right and left iliac veins was included). The imaging modality of choice for MTS was a CT angiogram and this is what was analyzed to incorporate a patient into the interview list. Patients were interviewed via telephone interaction. They were asked if they had seen a vascular surgeon, proceeded with treatment and if they experienced an improvement or resolution of their symptoms. The use of anticoagulants and presence of DVTs was also studied. In conclusion, the primary outcome measure was presence of MTS unilaterally or bilaterally and the study was conducted over the course of November to December 2018.



Results: Of 1142 patients, 47 were confirmed as positive for May Thurner Syndrome. The mean age of those with autonomic system dysfunction was 35.9 years. The mean age for women with MTS was 32.16 ± 9.67 and 39.50 ± 13.44 for men with the syndrome. 45 of 47 patients were female, at 95.7% of the total population. 2 patients were found to have bilateral compression – at 4.26%, with the residual population suffering from unilateral compression (of the left iliac vein). 22 patients completed successful interviews. 12 out of 22 patients received a vascular surgery consult (54.55% of those interviewed). 3 out of 12 patients proceeded with treatment specifically for May Thurner Syndrome (0.25% of those who had a surgery consultation). All 3 patients saw significant improvement (100% of those of those who underwent surgery). Significant improvement refers to resolution of symptoms associated with May Thurner Syndrome. 0 patients were taking anticoagulants and 0 suffered from or had a history of DVTs. Secondary objectives for establishing the correlation between MTS and DVT were proven false.

Conclusion: The results were consistent with previous research, as 4.26% of the total population were diagnosed with bilateral compression in this study, which lands within the 2–5% range established by Knuttinen et al. (11). 95.74% of cases were confirmed to be confined to one side. The ~ 4.25% of rare Bilateral cases are explained by a high bifurcation of the aorta, causing the compression on both sides. The gender and age distribution was also proven to align with the belief that it is at least twice as frequent in women as compared to men (12) and in the predominantly female POTS population, it's close to completely dominated by female prevalence (95.7%). We conclude that the prevalence of bilateral iliac vein compression in patients with MTS is as rare as it is in the general population. In addition, those patients that were treated surgically (2 with stents, 1 with ablation) for MTS found that their POTS symptoms resolved (lightheadedness, dizziness, and valvular incompetence in both superficial and deep systems). We can thus infer that for POTS patients with May Thurner syndrome, it is not an asymptomatic compression of the iliac vein which was an incidental finding, but that iliac vein compression (greater than 50%) may further exacerbate or potentiate their POTS' symptoms. We conclude that the prevalence of bilateral iliac vein compression is as rare in the general population as it is in a POTS population.



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Utility of Autonomic Testing for the Diagnosis of Neurogenic Orthostatic Hypotension and Subsequent Management With Droxidopa

Sami B. Alam, MD, Wael Almardini, MD, Amer Suleman, MD (AHA Hypertension Scientific Sessions 2018 – Chicago, IL)

Case Report: A 62-year-old man with a history of Hodgkin lymphoma, lupus, type 2 diabetes mellitus, neuropathy, hypercholesterolemia, hypothyroidism, and orthostatic hypotension who was taking gabapentin, saxagliptin, pravastatin, levothyroxine sodium, omeprazole, prednisone, midodrine, and fludrocortisone was evaluated for syncope, collapse, and fluctuating blood pressure (BP). He reported a 2-year history of syncope, with progressive worsening over a 3-month period. Initially, his episodes of syncope were preceded by dizziness and nausea, but began to occur without warning. He also reported a 5-year history of orthostatic hypotension, with supine BP of 100/60 mmHg and standing BP of 50–60/40 mmHg. Treatment with midodrine (10 mg three times daily) and fludrocortisone (0.1 mg twice daily [BID]) did not relieve his symptoms. Initial cardiac evaluation revealed a systolic murmur, but was otherwise unremarkable. Autonomic function tests, including continuous electrocardiography, BP and heart rate monitoring at 2-min intervals, and transcranial Doppler, were performed in the supine and 80° head-up tilt positions. A BP drop from 112/68 mmHg to 76/60 mmHg occurred within 2 min of tilt, accompanied by dizziness and presyncope. Heart rate with deep breathing revealed a depressed baseline autonomic tone, and a Valsalva maneuver test showed a depressed Valsalva response (Valsalva ratio: 1.15). Taken together, these results indicated a diagnosis of neurogenic orthostatic hypotension (nOH). Treatment with droxidopa (100 mg BID) was initiated; this dose was titrated to 100 mg once daily due to hypertension, for which, a nitroglycerin patch (0.2 mg nightly) was prescribed. All other medications were continued. Treatment with droxidopa significantly improved his symptoms; he no longer loses consciousness upon positional change, but still experiences some symptoms of presyncope (eg, slight lightheadedness, dizziness). This has allowed greater integration into activities of daily living and a less strenuous routine for the patient and his caretakers. In conclusion, autonomic function testing in this patient effectively led to a timely diagnosis of nOH which, in turn, allowed for implementation of appropriate pharmacologic intervention to manage symptoms.



The Efficacy of Celiac Plexus Block as a Non-Invasive Procedure in Treating Median Arcuate Ligament Syndrome (MALS) Patients

A. Dada, MPH; S. Alam, MD; R. Naga, MBBS; S. Usmani, MD; M. Nasri, MD; W. Almardini, MD; S. Alam, MD; N. Noor, MD; H. Mistry, MD; A. Suleman, MD, FHRs, FAIS, FSCAI. (2019 American Neurogastroenterology and Motility Society Annual Meeting – Chicago, IL)

Introduction: The median arcuate ligament syndrome (MALS) is a rare condition characterized by chronic abdominal pain, nausea, vomiting, and weight loss. The pathophysiological origin can be explained by the compression of the celiac nerve plexus causing compression of the proximal celiac trunk by the median arcuate ligament. Standard diagnosis can be obtained through mesenteric ultrasounds by evaluating arterial velocities. Various procedures have shown to improve symptoms including celiac plexus blocks, decompression and revascularization. The purpose of this study is to determine the relationship between celiac velocity during the expiration phase and MALS symptoms after celiac plexus blocks.

Method: By reviewing the data collected from The Heartbeat Clinic's electronic medical records, we performed a retrospective study on 36 patients (pts) that had undergone the celiac plexus block procedure from June 2014 to February 2019. Only 28 out of the 36 patients, had a Mesenteric Ultrasound both pre- and post-procedure that were analyzed for changes in expiration celiac velocity. These 28 patients' records were also analyzed for patient reports of any subsequent symptomatic changes; and were then divided into 2 groups based on the duration of MALS Symptoms relief being short term (< 6 weeks) or long term (> 6 weeks). The patients were also categorized based on Celiac Expiration Velocity (cm/sec) following the procedure.

Results: Of the 28 patients, 5 were males (age 32.42 ± 4.92) & 23 were females (age 24.63 ± 5.24). 24 pts reported MALS Symptom relief being short term (< 6 weeks) and 4 pts had reported MALS Symptoms relief being long term (> 6 weeks). Of the 24 pts, 21 pts recorded a significant decrease in Celiac Artery Velocity with a mean decrease of $84 \text{ cm/s} \pm 9.3$. Only 4 pts reported no relief in symptoms and showed no significant changes in Celiac Artery Velocity.

Conclusion: Celiac plexus block provided significant symptom relief for MALS pts. There was a trend in the decrease of Celiac Artery Velocities and improvements of MALS symptoms.



Tachycardia Induced Cardiomyopathy in Patients with Ehlers-Danlos Syndromes (EDS)

Sabahat Usmani¹, Somia Anum¹, Mohammad Patwary², Sami Alam¹, Arathi Kumar¹, Zan Rehman², Amroo Dada¹, Amer Suleman¹. ¹The Heartbeat Clinic, McKinney, TX; ²UT Dallas, Richardson, TX (Heart Failure Society of America – 23rd Annual Scientific Meeting 2019 – Philadelphia, PA)

Introduction: POTS has been said to be a type of dysautonomia characterized by Tachycardia brought about by postural shifts in blood volume and circulation. This results in various symptoms including dizziness, lightheadedness, syncope, palpitations and tremulousness. POTS is often associated with EDS, a rare inherited connective tissue disorder pertaining to the production and processing of collagen. 1 in 3 POTS patients suffers from EDS.

POTS causes abnormal increases in heart rate. If left untreated, patients can suffer from Tachycardia Induced Cardiomyopathy (TIC) ^{1 2} indicated by increasing Left Ventricle End Diastolic Diameter (LVEDD).

Methods: Electronic health records of 1300 POTS patients were reviewed retrospectively. Female patients with EDS between the ages of 20–40 who had imaging records of 3 or more echocardiograms in the past 5 years, with no more than 1 year between consecutive echocardiograms were randomly selected. Our records identified 94 patients fulfilling the inclusion criteria. An assessment of the relation between associated Symptoms and corresponding Echocardiogram LVEDD values was made by observing patient reported Symptoms such as shortness of breath, chest pain, dizziness/lightheadedness, palpitations, and their overall feeling of well-being. Corresponding Functional Capacity values of each patient analyzed by reviewing Cardiopulmonary Exercise Stress Tests were also noted. Lastly, treatment modalities including drugs, exercise and I/V fluids were also monitored. A timeline with regards to changes in LVEDD was established to determine the effectiveness of various treatments.

Results: Of the 94 patients fulfilling the inclusion criteria: Mean age was 30 ± 6.3 yrs.

Mean LVEDD of patients reporting minimum symptoms was 4.02 ± 0.43 . Mean LVEDD of patients reporting maximum symptoms was 4.28 ± 0.45 . Classical two sample T-Test was performed to determine significance of the two means, p-value <0.0001 . Indicating the mean of LVEDD with least symptoms is significantly smaller than LVEDD with maximum symptoms at a very small level of significance. The average percentage decrease in LVEDD at minimum symptoms compared to LVEDD at maximum symptoms was 6.28%.



The average functional capacity (FC) of patients experiencing minimum symptoms was 69% while it was 63% for patients experiencing maximum symptoms. To compare the functional capacity values classical two sample T-Test was performed, $p\text{-value} = 0.006665$. Since $p\text{-value}$ is less than 0.01, it indicates that the mean of FC with least symptoms is significantly higher than FC with maximum symptoms at 1% level of significance. **The average Decrease in FC per 0.1 cm Increase in LVEDD was 2.55%**

Patients with LVEDD of 4.04 ± 0.48 were seen to have the highest FC >70%. Overall, 40 patients were seen to have >70% FC at LVEDD with the Lowest symptoms.

At >70% FC, 44 patients showed improvement with medication (Midodrine 20, Florinef 12, Beta Blockers 12), 25 patients with exercise and 11 on IV Fluids (including saline and/or albumin infusions).

Conclusion: Patients with POTS and EDS, tend to develop a worsening of symptoms as LVEDD progressively increases resulting in a Tachycardia Induced Cardiomyopathy. However, aggressive management of symptoms with medication and a dedicated exercise regimen can improve symptoms, decrease LVEDD and cause reversal of the disease process. From the evidence collected, it can be recommended that POTS patients' LVEDD values be maintained between **3.8 – 4.2 cm**, as this range was seen to produce the least amount of symptoms as well as higher functional capacities (>70%).

References:

1. <https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4144378/>
<https://www.ncbi.nlm.nih.gov/pubmed/15226218>



Split S1 in Postural Orthostatic Tachycardia Syndrome

Ramtej Atluri, MBBS, Sami B. Alam, MD, Sabahat Usmani, MD, Sabih Alam, MD, Wael Almardini, MD, Mohamad Nasri, MD, Nida Noor, MD, Hemanshi Mistry, MD, Amer Suleman, MD (American Society for Preventive Cardiology – 2019 Congress Meeting – San Antonio, TX)

Introduction: The Postural Orthostatic Tachycardia Syndrome (POTS) is a form of orthostatic intolerance that is more commonly seen in young women. POTS is a form of dysautonomia that is estimated to impact between 500,000 and 3,000,000 Americans, and millions more around the world. In POTS, due to hypovolemia or other mechanisms, there is decreased venous return to the heart, so to increase the cardiac output there is reflex tachycardia which commonly presents with symptoms such as lightheadedness, blurred vision, muscle weakness, nausea, and dizziness. These nonspecific symptoms along with palpitations, atypical chest pain, dyspnea, migraines, numbness or tingling, and exercise intolerance which could indicate dysautonomia have also been associated with Mitral Valve Prolapse (MVP). The combination of typical auscultatory features of MVP and any of these nonspecific symptoms is referred to as the Mitral Valve Prolapse Syndrome. In MVP the common auscultatory finding is of a mid-systolic click followed by a late systolic murmur heard best at apex. The first heart sound (S1) results from the closing of the mitral and tricuspid valves simultaneously so a single heart sound is usually heard. When the mitral valve closes significantly before the tricuspid valve, there is a split of the first heart sound allowing each valve to make a separate audible sound. Because of the decreased preload in POTS patients, the mitral valve closes before tricuspid valve and the split of S1 becomes audible which can be mistaken for the mid systolic click of MVP.

The aim of this study is to determine the incidence of a split of the first heart sound in patients with postural orthostatic tachycardia syndrome.

Methods: By retrospectively analyzing Electronic Medical Records, a randomized list of nine hundred and ninety nine (999) POTS patients were selected from our clinic (January 2014 to December 2018). We then reviewed the medical records of 999 POTS patients by referring to the physical examination for the auscultatory findings of the initial visit to look for incidence of a Split First Heart Sound and performed a collective analysis on the gathered data.

Results: Out of 999 patients, 85.9% are females (n=859) and 14% are males (n=140).

Out of 999 patients 168 patients (16.8%) had a split of the first heart sound.

Conclusion: The study reveals that the prevalence of a split of the first heart sound in POTS patients is 16.8%. Because of it being a dysautonomia, the presenting symptoms as well as the auscultatory findings in POTS and Mitral Valve Prolapse Syndrome can be similar, therefore POTS should be kept in consideration when seeing such especially in Females. There may be a possibility that before modern literature indicated so, POTS may previously have been misdiagnosed as Mitral Valve Prolapse Syndrome.