Clinical Pearls for P.O.T.S.

Peng-Sheng Chen, MD

Editor-in-Chief, Heart Rhythm journal
Burns & Allen Chair in Cardiology Research
Professor, Cardiology; Staff Physician III

COI: Medtronic Inc donated research equipment to my laboratory

Idiopathic postural orthostatic tachycardia syndrome: an attenuated form of acute pandysautonomia?

R Schondorf and P A Low, Neurology 1993

Reviewed records of Mayo autonomic reflex laboratory Age 20-51, exhibited tachycardia at rest or during head-up tilt test

- Usually women
- an acute onset of persistent lightheadedness and fatigue or gastrointestinal dysmotility
- ***In seven patients, a viral illness may have preceded the onset of symptoms***

POTS and COVID-19

Immunologic Research (2021) 69:205–211
https://doi.org/10.1007/s12026-021-09185-5

BRIEF REPORT

Postural orthostatic tachycardia syndrome (POTS) and other autonomic disorders after COVID-19 infection: a case series of 20 patients

Svetlana Blitshteyn[1] · Sera Whitelaw[2]

Received: 11 February 2021 / Accepted: 22 March 2021 / Published online: 30 March 2021

Dysautonomia

- Dysautonomia is the deviation from or interruption of the normal structure or function of the autonomic nervous system.
- *The most common cardiovascular diagnoses associated with dysautonomia are the postural orthostatic tachycardia syndrome (**POTS**) and the neurocardiogenic (vasovagal, reflex) syncope or presyncope (NCS), both diseases are associated with chronic orthostatic intolerance.*

Sheldon et al, HRS consensus statement 2015

POTS

- The prevalence of POTS is approximately 0.2% and affects approximately 500,000 people in the US alone.
- Peak onset 14 YO and over 75% of them are white women in childbearing age. (*In my clinic, 90% women and 80% white)*
- Dx criteria:
  - Orthostatic intolerance: palpitations, dizziness and syncope
  - An increase in heart rate (HR) of 30 bpm when moving from a recumbent to a standing position
  - The absence of orthostatic hypotension (< 10 mmHg reduction of BP when standing up)

Sheldon et al, HRS consensus statement 2015

22YOF, Active Standing Test

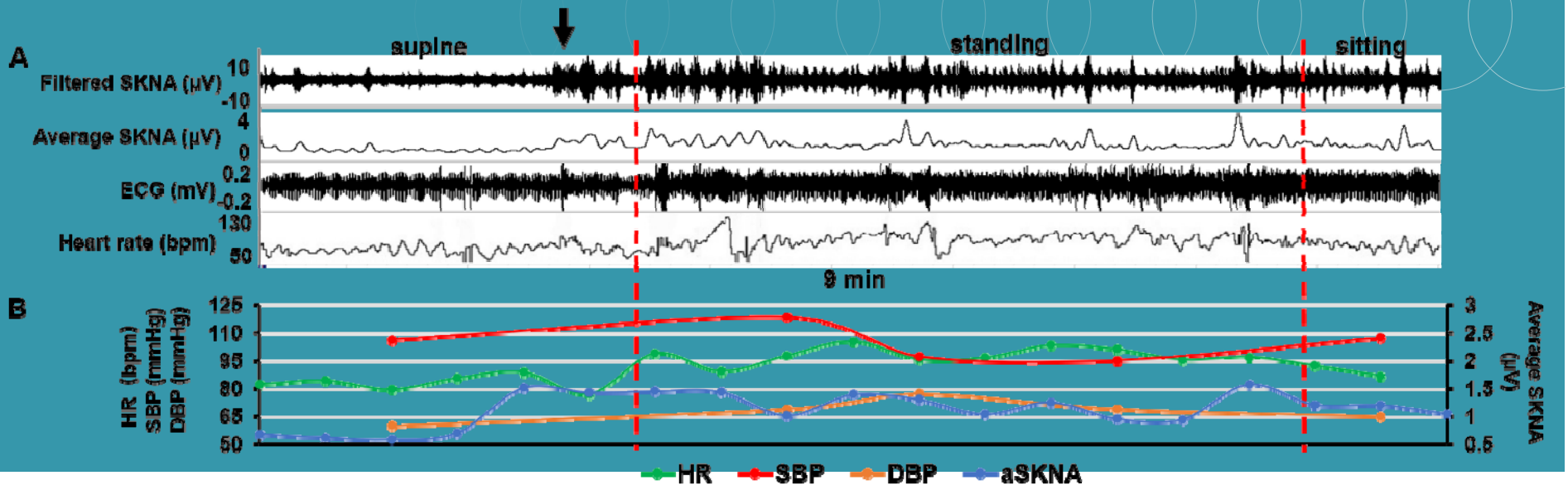
4. Orthostatic Test

	Time	Mark	ECG rhythm	HR (bpm)	BP (mmHg)
Supine	10:56	1		100	118 83
Standing up	10:59	2	/	148 /	116 / 91
Standing 1 min	11:01	3		114	127 / 87
2 min	11:02	4		115	124/78
3 min	11:03	5		116	127/88
Sitting down	11:05	6	/	96 /	128 / 85
sitting	11:06	7		105	121 / 67

CR 14April 2021

HR increased by 48 bpm, SBP did not change by > 10 mmHg

SKNA changes during active standing test



Liu et al, HRS abstract, 2021

POTS vs PSWT
postural symptoms without tachycardia

- **The 30 bpm increase of HR is not a good diagnostic criterion**

  Orthostatic tests are not reproducible

  Only 10% of the patients in my clinic has 30 bpm increase of HR upon standing

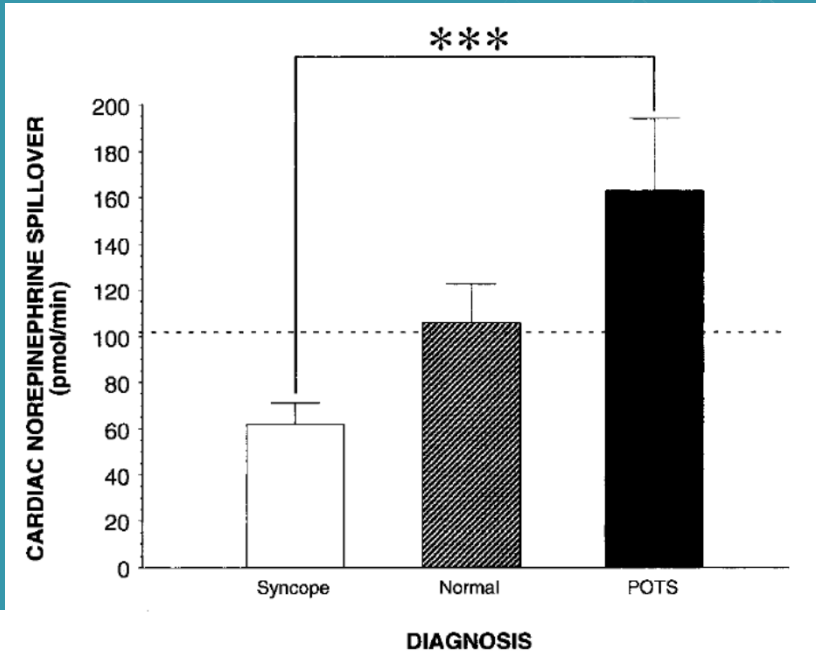
- **POTS vs PSWT (postural symptoms without tachycardia)**

Raj et al, Canadian CV society position statement, 2020

Cardiac Sympathetic Dysautonomia in Chronic Orthostatic Intolerance Syndromes

- 72 patients underwent right heart catheterization for measurements of cardiac norepinephrine spillover (rate of entry of norepinephrine into the coronary sinus)
- 40 underwent cardiac sympathoneural imaging by 6-[18F]fluorodopamine scanning.
- 34 underwent both procedures.

Cardiac Norepinephrine spillover (rates of entry of norepinephrine into coronary sinus plasma)

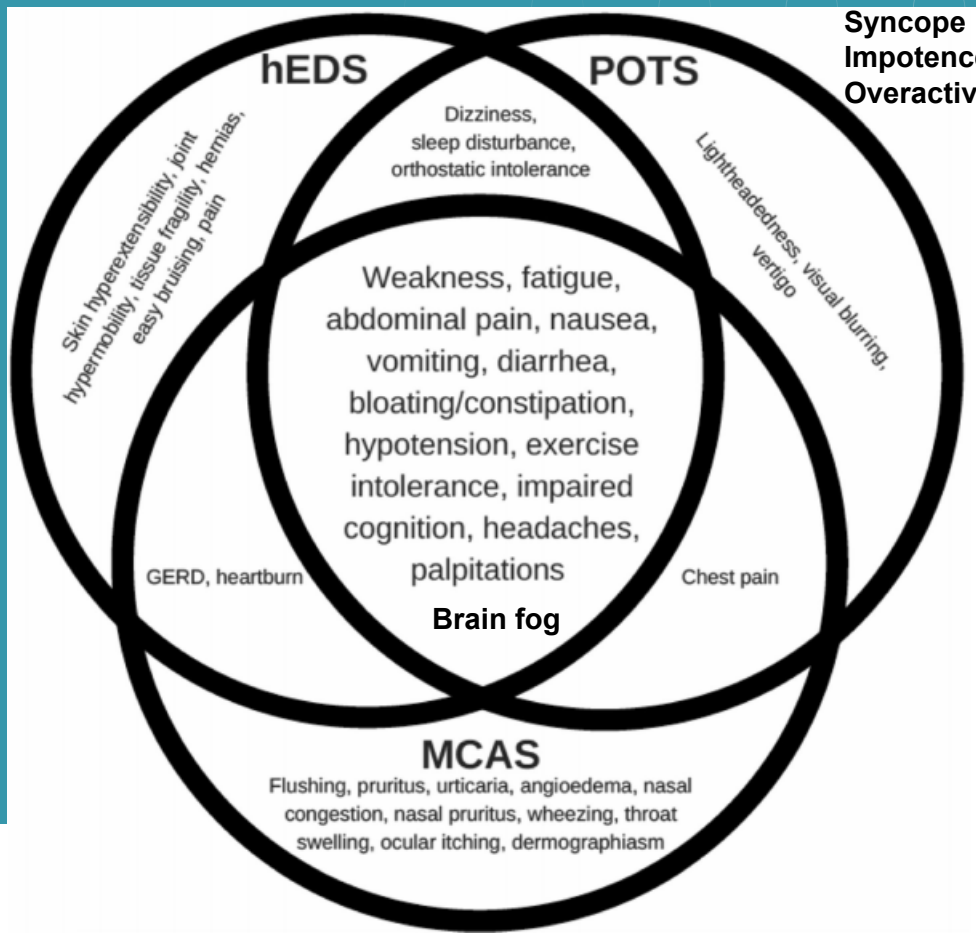


Conclusions—POTS and NCS differ in tonic cardiac sympathetic function, with increased cardiac norepinephrine release in the former and decreased release in the latter. (Circulation. 2002;106:2358-2365.)

Cardiac Sympathetic Dysautonomia in Chronic Orthostatic Intolerance Syndromes

TABLE 1. Patient Characteristics in POTS and Recurrent NCS

Finding	POTS	NCS	*P*
Mean age, y	39.7	37.6	...
Women/men	33/3	28/8	...
Orthostatic intolerance	35	28	0.01
Presyncope	29	36	0.005
Fatigue or weakness	28	23	...
Disabled	23	16	...
Chest pain or pressure	20	17	...
Syncope	20	30	0.01
Heat intolerance	18	16	...
Headache	17	21	...
Tilt-induced tachycardia	16/16	0/18	0.001
Palpitations	15	16	...
Allergies to drugs	15	15	...
Exercise intolerance	14	11	...
Arthralgia	14	10	...
Concentration decreased	13	18	...
Tilt-induced syncope	9 of 16	18 of 18	0.002
β-Blocker improved	8 of 14	5 of 13	...
Fludrocortisone improved	10 of 13	5 of 13	0.05
Midodrine improved	3 of 6	8 of 13	...
SSRI improved	2 of 5	1 of 7	...



Syncope
Impotence
Overactive bladder

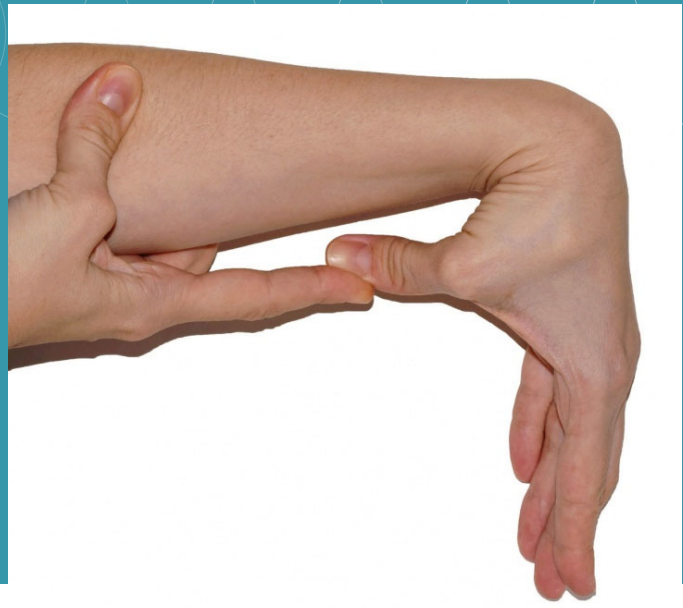
Management difficulties:

Beta blockers and EpiPen
Mestinon and chest pain (coronary spasm)
Co-existence of hyper- and hypotension
Co-existence of tachy and bradycardia
Convulsive syncope
Nitroglycerine and headache/migraine
Drug allergy and intolerance
Fluctuating course
Gastroparesis and dehydration
Complications of venous access ports
Deconditioning
Can't walk
Can't work

Kohn and Chang, Clinical Reviews in Allergy & Immunology 2020

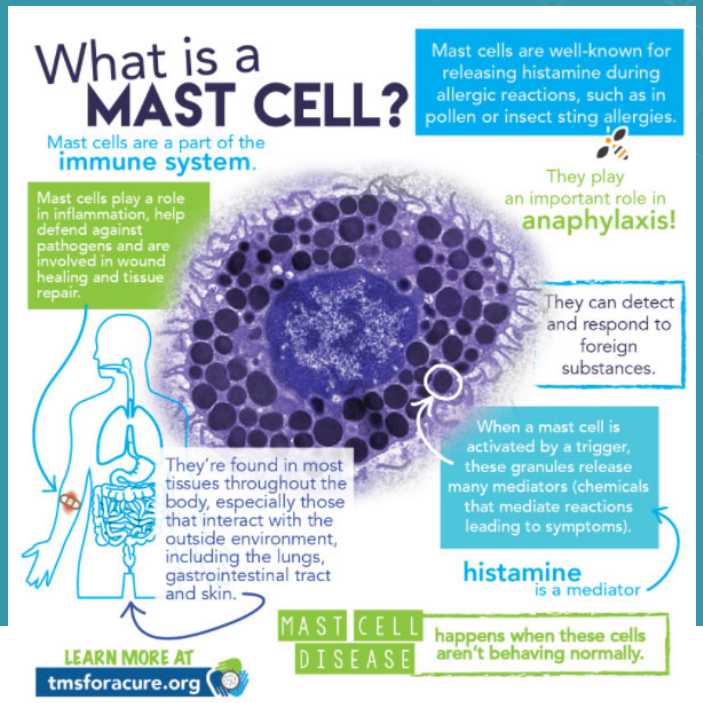
Ehlers-Danlos Syndrome (EDS)

- 1 in 2,500 to 1 in 5,000 people have EDS worldwide
- At least 19 genes are connected to the disorder
- Most common type is hypermobile EDS (hEDS). Genetic test for hEDS is usually negative.



Rehan, USC spine center

Mast cell activation syndrome



The mast cell disease society, Inc.

1. Episodic symptoms consistent with mast cell mediator release affecting two or more organ systems
2. A decrease in the frequency or severity; or resolution of symptoms with anti-mediator therapy: H1 and H2 histamine receptor antagonists, anti-leukotriene medications, or mast cell stabilizers (cromolyn sodium)
3. Evidence of an elevation in a validated urinary or serum marker of mast cell activation: Total serum tryptase is recommended as the markers of choice
4. Primary (clonal) and secondary disorders of mast cell activation ruled out.

(*Akin et al, J Allergy Clin Immunol 2010*)

Cardiovascular evaluation for POTS

- **Active standing test for HR/BP, plasma norepinephrine**
  - POTS vs PSWT
  - R/O orthostatic hypotension
  - Standing NE > 600 pg/ml and BP increase by ≥ 10 mmHg indicate hyperadrenergic POTS. (40% patients in my clinic).
- **7-day patch recording**
- **Echocardiogram**: cardiac function, aorta, IVC collapse (dehydration)
- **24-hr arterial blood pressure monitoring (ABPM)** (60% has 1 BP < 90/60)
- **Tilt table testing is optional (IIb)**

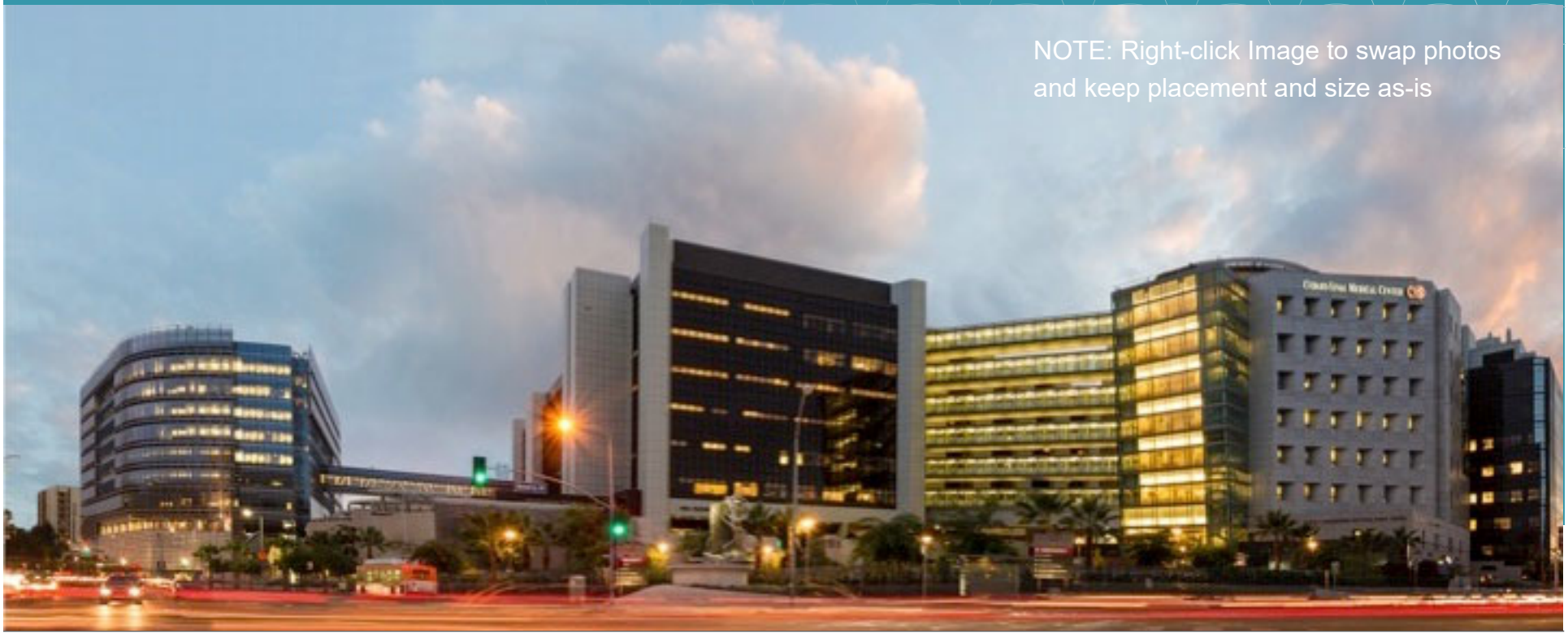
Non-Cardiovascular evaluation for POTS

- **MCAS:** hematologist/allergist
- **hEDS, sjogren's syndrome (dry eyes/month), join pains:** rheumatologist
- **Migraine:** neurologist
- **GI:** GI motility, gastroparesis management
- **Sleep;** rule out sleep apnea as a cause of fatigue and bad sleep
- **Urology:** overactive bladder
- **Endocrine:** Hashimoto’s

Management

- Deconditioning- exercise training (Levine protocol)
  - Cardiac rehab, Physical therapy
- Mast cell activation syndrome (MCAS)- H1 and H2 blockers, mast cell stabilizers
- Ehlers Danlos syndrome- physical therapy
- Hypovolemia- salt and water, i.v. infusion (2X/week)
- Hypotension- midodrine, droxidopa, fludrocortisone
- Tachycardia- propranolol 20 mg/d, ivabradine, mestinon
- Hyperadrenergic POTS- clonidine, ivabradine
- Autoantibodies- IVIG, plasmapheresis
- **QT prolongation**: droxidopa, ivabradine. **Pregnancy class D**: ivabradine

Sample photo page



NOTE: Right-click Image to swap photos and keep placement and size as-is

Cedars Sinai