POTS Syndrome
A Cardiologist’s Perspective

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Postural Tachycardia Syndrome: Beyond Orthostatic Intolerance

Diagnostic Criteria

- Increase in HR $\geq$ 30 bpm within 10 minutes of upright posture (tilt test or stand) from supine position (but $> 40$ bpm in pts $< 20$ yrs)

- Associated constellation of symptoms – worse on upright posture & improving with recumbence

- Chronic symptoms lasting $> 6$ months

- Absence of other disorders or medications causing orthostatic tachycardia

Overlapping Syndromes

Neurology versus Cardiology Literature ...

- Inappropriate Sinus Tachycardia
- Chronic Orthostatic Intolerance
  - Postural Orthostatic Tachycardia Syndrome
  - Idiopathic hypo-volaemia
  - Hyper-adrenergic syndrome
  - Sympatho-tonic orthostatic hypotension
  - Mitral valve prolapse syndromes

Brady PA, et al PACE 2005, 28:1112-21
Cardiology / FASS

- Patients can present to various ‘services’ ..... All need to be aware of POTS !

- ‘Patient experience’ of initial medical contact is often poor

- Poor initial experiences can confound / complicate later management

- Syndrome usually requires multi-disciplinary input for best outcome

- Very limited role for cardiac electrophysiologist ...
Key question for the cardiac electrophysiologist

Are this patient’s symptoms primarily due to a pathological cardiac arrhythmia?
Normal Sinus Rhythm

- Are the P-waves normal in SR?
- Is the heart structurally normal on imaging?

Atrial dilatation; Valve lesions; LVH; LV-dysfunction

No → Pathological Arrhythmias
Regular narrow-QRS tachycardia (SVT)

± pre-syncope ± chest discomfort ± breathlessness

- Discrete episodes / variable frequency
- Random onset
- Abrupt onset & offset
- Terminates with vagal manoeuvres & adenosine
- Asymptomatic between episodes
What is the AV-relationship?

Interacting with a sustained episode of arrhythmia – CSM / Adenosine

Adenosine iv

1:1 & terminates

3:1 & transient AV-block

What is the P-wave morphology in tachycardia?
DDx Sinus from Atrial tachycardias

Sinus tachycardia
- P waves have normal morphology
- Atrial rate 100-200 beats/min
- Regular ventricular rhythm
- Ventricular rate 100-200 beats/min
- One P wave precedes every QRS complex

Atrial tachycardia
- Abnormal P wave morphology
- Atrial rate 100-250 beats/min
- Ventricular rhythm usually regular
- Variable ventricular rate

Persistent ‘sinus tachycardia’ without cause .... Consider atrial tachy / atrial flutter / PJRT !!
### Is the tachycardia Sinus (as in POTS) or AT ...?

**Distinguishing generalisations ..... (?)**

<table>
<thead>
<tr>
<th></th>
<th>Sinus Tachycardia</th>
<th>Atrial Tachycardia</th>
</tr>
</thead>
<tbody>
<tr>
<td>Symptoms?</td>
<td>Similar</td>
<td>Similar</td>
</tr>
<tr>
<td>Is the patient ever fully well?</td>
<td>No</td>
<td>Yes, between episodes</td>
</tr>
<tr>
<td>Confounding other pathologies?</td>
<td>?</td>
<td>Cardiac (+) maybe</td>
</tr>
<tr>
<td>Patient age / gender?</td>
<td>Younger; M:F = 1:5</td>
<td>Older; M = F</td>
</tr>
<tr>
<td>Orthostatic feature consistently?</td>
<td>Yes</td>
<td>No</td>
</tr>
<tr>
<td>Tilt-testing / or similar</td>
<td>Positive consistently</td>
<td>Negative</td>
</tr>
<tr>
<td>Is the heart structurally normal?</td>
<td>Yes</td>
<td>Commonly abnormal</td>
</tr>
<tr>
<td>Are P-waves in SR normal?</td>
<td>Yes</td>
<td>Maybe abnormal</td>
</tr>
<tr>
<td>P-waves in SR vs in tachycardia</td>
<td>Identical</td>
<td>Usually different</td>
</tr>
<tr>
<td>Response to medications</td>
<td>Poor / Aggravation</td>
<td>Clinically useful</td>
</tr>
<tr>
<td>Role of ablation?</td>
<td>In excluding AT (?) Ablation contraindicated</td>
<td>Ablation when medication insufficiently effective</td>
</tr>
</tbody>
</table>
Atrial Tachycardias

How does ‘P-wave’ morphology compare to SR?

The change can be subtle – so need a 12-lead recording to compare!
Non-sinus ‘P-wave’-led tachycardia
(from mid crista terminalis RA at EP-study)
Is there a role for invasive EP-study

- To clarify the diagnosis
- To define the mechanism
- To target the circuit / focus for ablation
- To avoid / reduce anti-arrhythmic medications
- Ablation success is related to aetiology
Focal AT
- Paroxysmal, persistent or incessant
- May show ‘warm up’ phenomenon
- Heart may be normal
- May terminate with adenosine!

Macro-reentrant AT
- P-waves in SR may be abnormal
- Paroxysmal or persistent
- Heart usually scarred / abnormal
- AV block with adenosine but AT continues
Electro-anatomical Mapping of RA

- Real-time reconstruction of RA in 3D
- Based on local electrical signal amplitude & timing
- Locate source of rhythm (earliest location in sinus or arrhythmia)
- Is the source of the fast rhythm co-located with SAN?
Atrial tachycardia

Earliest site does not map to SAN location

Cranial end of crista terminalis or RAA – similar P-wave morphology to SR
Right Atrium .... & links with LA / RPVs !
Ablate and Pace for POTS. A 42-year-old woman with postural tachycardia syndrome (POTS) was admitted to our hospital with severe palpitations, light-headedness, and syncope. Several drugs had been administered previously, but all had been discontinued due to intolerable adverse effects or limited efficacy. One of the drugs, the \( I_f \) current inhibitor ivabradine, effectively slowed the patient's heart rate and relieved the symptoms, but was discontinued due to allergy. After unsuccessful sinus node ablation, atrioventricular node ablation and dual chamber pacemaker implantation was performed, which dramatically improved her symptoms and eliminated syncope. Atrioventricular node ablation could modify the cardiac autonomic balance and thereby suppressed the excessive orthostatic sympathetic activity. (J Cardiovasc Electrophysiol, Vol. pp. 1-4)
Short- & long-term outcomes of sinus node modification in pts with POTS & IST

Autonomic testing pre- & post-procedure
(Valsalva, deep breathing, tilt table testing, quantitative sudomotor axonal reflex testing)

Sinus rate can be effectively slowed by sinus node modification
Clinical symptoms are not significantly improved in pts with IAST & POTS

**Sinus node modification is not recommended in these patients**
New onset of POTS following ablation of AV-node re-entrant tachycardia

- Retrospective identification from Syncope & Autonomic Disorders Clinic.

- **N = 6** - previously healthy except for SVT
  EP-study proven AVNRT & successful slow pathway RF-ablation
  New onset of orthostatic intolerance consistent with POTS

- Initial symptom-free period (3-6 weeks)
  All 6 began to experience progressive severe fatigue
  Orthostatic tachycardia (5); Syncope (3); Pre-syncope (6)
  Symptom while upright & relieved by becoming supine.
  Each patient had experienced symptoms for > 6 months
  Repeat EP-testing for symptoms (3) – No AVNRT recurrence
  Head-up tilt POTS response ≤ 10 min (all 6) with clinical symptoms

- **POTS may be a complication of radiofrequency ablation of AVNRT**

Exercise in the postural orthostatic tachycardia syndrome

- Patients with the POTS have both orthostatic & exercise intolerance

- $V_{O2}$-peak is lower compared to sedentary healthy individuals

  This suggests **a lower physical fitness level in POTS patients**

  POTS pts have XS HR $\uparrow$ & SV $\downarrow$ for each level of absolute workload;
  But HR response for relative workload ($\%V_{O2}$-peak) is the same.

- **Exercise performance is improved after short-term exercise training**

  SV $\uparrow$ and HR $\downarrow$ at any given $V_{O2}$ during exercise after training
  Peak HR is the same but peak SV & CO are greater after training.
  HR recovery from peak exercise improved (improved autonomic circulatory control)

*Fu Q & Levine BD Auton Neurosci 2015, 188:86-89*
Exercise in the postural orthostatic tachycardia syndrome

Conclusions & Implications

■ POTS pts do not have an intrinsic abnormality of HR regulation on exercise

■ POTS tachycardia is due to a reduced stroke volume.

■ Cardiac remodelling & blood volume expansion associated with endurance training increase fitness and exercise performance in POTS patients.

Fu Q & Levine BD Auton Neurosci 2015, 188:86-89
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A “final common pathway” for overlapping patho-physiologies:

- Autonomic neuropathy of lower body
- Hypovolemia
- Increased sympathetic tone
- Mast cell activation
- Deconditioning
- Autoantibodies

POTS phenotype similar to other disorders:

- Chronic fatigue syndrome
- Ehlers-Danlos Syndrome
- Vasovagal syncope
- Inappropriate sinus tachycardia

Distinguishing POTS from Pathological Arrhythmias

- **Sinus tachycardia**
  - **Paroxysmal**
    - No obvious trigger: 24h Holter monitor, Electrophysiology study, SNRT
  - **Postural**
    - Head upright tilt test (positive): POTS
  - **Persistent**
    - No underlying cause: IST
    - Underlying cause present: NST

- Consider/exclude:
  - Atrial tachycardia
  - Atrial flutter
  - Atrioventricular re-entry tachycardia