What is PoTS?
The Newcastle approach

Julia Newton
What is CFS(ME)?

• Classified by WHO in ICD-10 as a neurological disorder G93.3

• Medical unexplained
  – Physiologically distinct from depression
  – Identifiable immunological, neurological, endocrine abnormalities that are consistent
What is CFS/ME?

- Severe debilitating fatigue causing interference with normal functions.
- Duration of at least 4 months
- No evidence for other medical or psychiatric problems.
- Typical history
- No pointers on examination to alternative diagnoses.
- Blood tests are normal
Potentially treatable clinical problems
Testing

Newton et al., CAR 2009
<table>
<thead>
<tr>
<th></th>
<th>CFS/ME</th>
<th>Controls</th>
<th>P</th>
</tr>
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<tbody>
<tr>
<td>N</td>
<td>64</td>
<td>64</td>
<td></td>
</tr>
<tr>
<td>Age mean ± SD</td>
<td>46 ± 12</td>
<td>48 ± 15</td>
<td>0.5</td>
</tr>
<tr>
<td>Males (%)</td>
<td>23 (36)</td>
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<td>Ns</td>
</tr>
<tr>
<td>FIS</td>
<td>97 ± 28</td>
<td>12 ± 20</td>
<td>&lt;0.0001</td>
</tr>
<tr>
<td>Hx of loss of consciousness (%)</td>
<td>27 (40%)</td>
<td>15 (23%)</td>
<td>0.04</td>
</tr>
<tr>
<td>HUT positive (in those able to tolerate the test)</td>
<td>19</td>
<td>7</td>
<td>0.004</td>
</tr>
<tr>
<td>Systolic OH</td>
<td>24</td>
<td>26</td>
<td>0.6</td>
</tr>
<tr>
<td>Delayed OH</td>
<td>2</td>
<td>0</td>
<td>0.5</td>
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Those with LOC - HUT was positive in 15 (56%) which is comparable to previous studies of the predictive value of head up tilt in those with unexplained syncope.

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Hollingsworth et al., EJCI 2010
The overlap between PoTS and CFS
A profile of patients with postural tachycardia syndrome and their experience of healthcare in the UK


Authors: Lesley Kavi, Michaella Nuttall, David A Low, Morwenna Opie, Lorna M Nicholson, Edward Caldow, Julia L Newton

Postural tachycardia syndrome (PoTS) is a recently recognised condition that usually affects
Results

• 92% female, 81% age 18-49
• 3 commonest symptoms:
  fatigue 91%
  presyncope/dizzy 90%
  palpitations 86%
• 58% - syncope/blackouts
What is PoTS?

• It is a syndrome and probably has many different causes and mechanisms
• Age 15-50
• 75% female to male
• Estimated prevalence 0.2%
• Described, defined and named in 1993
What is PoTS?

• Subset of orthostatic intolerance (defined as the provocation of symptoms on standing that are relieved by lying down) associated with the presence of excessive tachycardia on standing.

• Symptoms may be so severe that normal activities of life, such as bathing, housework, and even eating can be significantly limited.
How bad is it?

- PoTS patients have been reported to suffer from a degree of functional impairment similar to that seen in conditions such as chronic obstructive pulmonary disease and congestive heart failure, yet these patients are frequently misdiagnosed as having severe anxiety, panic disorder or CFS/ME.
How common is it?

• USA studies suggest that PoTS affects approximately 170 per 100,000 of the population and of this total, 25% are disabled and unable to work.

• Can affect any age group, but predominantly presents in young and middle age.

• PoTS does run in some families. The onset can be sudden or gradual. The quantity and severity of symptoms varies from day to day.
lightheaded
daughter
diarrhoea
abdominal pain
acrocyanosis
visual greying
exercise intolerance
brain fog
tremulous
sweating
sense of anxiety
nausea
**syncope**
chest pains
orthostatic headaches
poor sleep
coat hanger pain
abdominal pain
fatigue
palpitations

stickmancommunications.co.uk
Pathophysiology

• Unclear
• Relative hypovolaemia
• Autoimmunity
• Related conditions - JHS/EDS, CFS/ME, Sjogrens, Mitochondrial disease + more
Precipitants

- Viral illness, giving birth, or being exposed to great bodily stressors (i.e. surgery, trauma or chemotherapy).
- Teenagers sometimes develop the disorder during the years of rapid growth.
Diagnostic criteria for PoTS

• DEFINITION (HRS 2015 consensus)

1. Frequent symptoms of PoTS on standing
2. Associated increase in HR 30 bpm (40bpm in teens) *persistent
3. No drop in BP (but PoTS and VV not mutually exclusive)
A profile of patients with postural tachycardia syndrome and their experience of healthcare in the UK


Authors: Lesley Kavi, Michaela Nuttall, David A Low, Morwenna Ople, Lorna M Nicholson, Edward Caldow, Julia L Newton

Postural tachycardia syndrome (PoTS) is a recently recognised condition that usually affects younger women, who develop symptoms of orthostatic intolerance and a persistent tachycardia on standing upright. Healthcare professionals, patients and the national patient support group (PoTS UK) together created a survey, and the responses of 779 UK PoTS patients were analysed. The most
Obtaining a diagnosis

• PoTS diagnosis suggested by
  - cardiologist 34%
  - patient 20%
  - GP 7%

• Mean time from presentation to healthcare professional to diagnosis is 3.7 years (from symptom onset to diagnosis - 7 years)

• 48% psychiatric labeling - more common in women
Q20 If yes, what psychological or psychiatric disorder did they suggest as the cause of your PoTS symptoms? (tick all applicable)

Answered: 368  Skipped: 411

- Anxiety
- Panic disorder or panic attacks
- Depression
- Hypochondriac
- Other (please specify)
When should I suspect someone with fatigue?

- Syncope
- History of symptoms that are very postural in nature
- Postural dizziness or presyncope with meals or circumstances suggestive of drops in blood pressure
What do I recommend in clinic?

• Clinics for Research Evaluation and Service in Themed Assessments

• Fatigue – Monday morn

• CAV

• MDT
Avoid triggers

- Drugs – that lower BP, increase HR
- Prolonged standing/sitting
- Heat
- Alcohol
Activity management

- Graduated
- 6 weeks to work
- Pilates
- Swimming
- Recumbent bike
- Rowing
Conservative advise

- Fluid – 2.5 litres of water a day
- Consider increased salt
- Upto 5 cups of caffeine
- Counter manoeuvres
Conservative advise

- Fluid – 2.5 litres of water a day
- Consider increased salt
- Upto 5 cups of caffeine
- Counter maneouvers
- Tilt training
Medication

- Ivabradine
- B Blockers
- Midodrine
- Fludrocortisone
- Ca blockers
- Desmporessin
- Some antidepressants
- Clonidine
- Methylphenidate
- Erythropoeitin
- Octreotide
- Pyridostigmine
- Modanafil
- Bupropion
### Treatment

<table>
<thead>
<tr>
<th>Primary treatment</th>
<th>Combination</th>
<th>PoTs UK</th>
<th>PoTs Clinic</th>
</tr>
</thead>
<tbody>
<tr>
<td>No treatment</td>
<td>Alone</td>
<td>20 (24%)</td>
<td>17 (32.5%)</td>
</tr>
<tr>
<td></td>
<td>plus midodrine</td>
<td>4 (5%)</td>
<td>2 (4%)</td>
</tr>
<tr>
<td>Beta blocker</td>
<td>Alone</td>
<td>15 (18%)</td>
<td>10 (19%)</td>
</tr>
<tr>
<td></td>
<td>plus midodrine</td>
<td>4 (5%)</td>
<td>2 (4%)</td>
</tr>
<tr>
<td>Ivabradine</td>
<td>Alone</td>
<td>4 (5%)</td>
<td>7 (13%)</td>
</tr>
<tr>
<td></td>
<td>midodrine</td>
<td>0</td>
<td>2 (4%)</td>
</tr>
<tr>
<td></td>
<td>Fludrocortisone</td>
<td>1 (1%)</td>
<td>1 (2%)</td>
</tr>
<tr>
<td></td>
<td>Sertraline</td>
<td>1 (1%)</td>
<td>0</td>
</tr>
<tr>
<td></td>
<td>Duloxetine</td>
<td>1 (1%)</td>
<td>0</td>
</tr>
<tr>
<td></td>
<td>Beta blocker</td>
<td>0</td>
<td>1 (2%)</td>
</tr>
<tr>
<td>Diltiazem</td>
<td>Alone</td>
<td>1 (2%)</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Fludrocortisone</td>
<td>1 (1%)</td>
<td>0</td>
</tr>
<tr>
<td></td>
<td>Midodrine</td>
<td>0</td>
<td>1 (2%)</td>
</tr>
<tr>
<td>Verapamil</td>
<td>Alone</td>
<td>0</td>
<td>1 (2%)</td>
</tr>
<tr>
<td>Fludrocortisone</td>
<td>Alone</td>
<td>5 (6%)</td>
<td>2 (4%)</td>
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<td></td>
<td>Beta blocker</td>
<td>5 (6%)</td>
<td>2 (4%)</td>
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<tr>
<td></td>
<td>Midodrine</td>
<td>5 (6%)</td>
<td>0</td>
</tr>
<tr>
<td>Midodrine</td>
<td>Alone</td>
<td>12 (14%)</td>
<td>4 (7.5%)</td>
</tr>
<tr>
<td></td>
<td>Bupropion</td>
<td>1 (1%)</td>
<td>0</td>
</tr>
<tr>
<td>Octreotide</td>
<td>+ midodrine + fludro</td>
<td>2 (2.5%)</td>
<td>0</td>
</tr>
<tr>
<td>Pregabalin</td>
<td></td>
<td>1 (1%)</td>
<td>0</td>
</tr>
<tr>
<td>Clonazepam</td>
<td></td>
<td>1 (1%)</td>
<td>0</td>
</tr>
<tr>
<td>Antidepressant</td>
<td></td>
<td>3 (4%)</td>
<td>0</td>
</tr>
<tr>
<td>Salt only</td>
<td></td>
<td>2 (2.5%)</td>
<td>1 (2%)</td>
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</tbody>
</table>

- The commonest treatment either alone or in combination were beta blockers.
- Overall there were 21 different combinations described by PoTs patients.
- 24-32% were taking no treatment for their PoTs.
Fatigue Work is Supported by

Liver North

Northern CFS/ME Clinical Network

JRRG

ME Association