POTS UK
POSTURAL TACHYCARDIA SYNDROME

GP GUIDE TO PoTS

(Postural Tachycardia Syndrome)

WHAT IS PoTS?

PoTS was characterised in 1993, but previously existed under various other names including irritable heart, soldier's heart and idiopathic orthostatic intolerance. It is a heterogeneous group of disorders sharing similar characteristics.
On assuming upright posture, there is an excessive increase in heart rate associated with symptoms of orthostatic intolerance and sympathetic over-activity. There is brain hypoperfusion, usually in the absence of hypotension.

When humans adopt upright posture, approximately 500ml of blood drops into the abdominal cavity and limbs. A normal autonomic nervous system responds with immediate peripheral vasoconstriction and an increase in heart rate of up to 20bpm.

In POTS, it is considered that vasoconstriction is inadequate, resulting in pooling of blood, relative hypovolaemia and reduced venous return to the heart. Heart rate, inotropic status and, in some patients, catecholamine levels increase further to compensate. Dizziness and syncope can occur in the presence of normal BP; in fact some patients with PoTS have a hypertensive response to standing.

HOW COMMON IS PoTS?

The incidence in the UK is unknown. However, it is probably under-diagnosed due to lack of awareness and non-specific symptomatology. It is five times more common in women and tends to affect people age 15 to 50.

Patients may have some or all of the above symptoms. Most patients experience light-headedness, but 30% have blackouts.

Symptoms tend to be worse on standing or prolonged sitting and exacerbated by heat, food and alcohol.
Life expectancy is thought to be unaffected, but levels of disability can be equivalent to that found in congestive heart failure and COPD.

SYMPTOMS OF PoTS

Dizziness	Neck/shoulder pain (muscle ischaemia)
Syncope / pre-syncope	Palpitations (tachycardia / ectopics)
Orthostatic headache	Acrocyanosis (purplish hands/feet)
Fatigue	Visual greying or blurring
Poor memory	Weakness
Poor concentration	Insomnia
Sense of anxiety	Nausea
Exercise intolerance	
Tremulousness	
Sweating	
GI upset	

WHAT CAUSES PoTS?

PRIMARY PoTS

Abrupt onset may follow infection, pregnancy, surgery, immunisation or trauma. There is evidence of auto-immune aetiology.

'Developmental' PoTS affects teenagers (gradual onset around age 14 with symptoms peaking age 16) and 80% resolve within a few years.

Hyperadrenergic PoTS – in some, a genetic defect has been identified

SECONDARY PoTS

Deconditioning (e.g. after prolonged bed rest following illness)

Joint Hypermobility Syndrome (now Considered by most authorities as indistinguishable from, if not identical to, the Ehlers-Danlos-hypermobility type, formerly known as EDS type III); an inherited multi-system connective tissue disorder. See www.hypermobility.org

Other medical conditions – diabetes, amyloidosis, sarcoidosis, SLE * Poisons – alcohol, heavy metals * Cancer or chemotherapy.

HOW IS IT DIAGNOSED?

Diagnosis is usually made following a tilt table test. After resting, the patient is tilted head up by around 60 degrees for up to 45 minutes. In PoTS, heart rate increases by at least 30 bpm (40bpm in teenagers) or to >120 bpm. Symptoms are similar to those of hypotension and there is reduced cerebral perfusion, but by definition, blood pressure does not necessarily drop. However, there is an overlap with neurally mediated hypotension and 40% also have a drop in blood pressure.

A stand test can also be used to diagnose PoTS. Pulse rate / BP can be measured supine then after 2, 5 and10 minutes of standing still. Look for acrocyanosis (puffiness and purplish discolouration of feet/hands)

A sustained increase in heart rate of >30 bpm suggests PoTS. Stop if patient becomes very symptomatic. Some patients may faint during this test. Plasma noradrenaline levels are often elevated in the upright position in PoTS.

Other pathology must be excluded and underlying causes of PoTS considered.

Depending on history and symptoms, tests may include FBC, biochemical profile, calcium, thyroid function, serum cortisol, catecholamine levels, urine HIAAs, ECG, 24 hour BP, ECG and EEG.

DIAGNOSTIC CRITERIA FOR PoTS

1. **Sustained heart rate increase of 30 beats per minute or more within 10 minutes of standing or head up tilt (40 bpm age 12-19) in the absence of orthostatic hypotension**
2. **Standing heart rate is often 120 beats per minute or more within 10 minutes of standing or head up tilt**
3. **Orthostatic tachycardia may be accompanied by symptoms of cerebral hypo-perfusion and autonomic over-activity that are relieved by recumbence.**

Criteria not applicable for low resting heart rate.

MANAGEMENT

PHYSIOLOGICAL

Increased fluid (2-2.5 litres/day) and salt intake (2 to 4g ie up to 2/3 level teaspoon, Not in hyperadrenergic PoTS) * - increases blood volume

-Graduated exercise programme – swimming is ideal - improves calf muscle pump

-Small frequent meals low in refined carbohydrates - reduces diversion of blood to gut.

Elevate head of bed * - increases morning blood volume

Avoid prolonged standing and sitting * – reduces venous pooling.

Avoid heat and alcohol * – reduce vasodilatation.

Support tights – class 3, waist high * - reduce venous pooling.

Postural manoeuvres to avoid syncope eg elevate legs, cross legs and clench thighs and buttocks, clench a fist, rock up and down on tiptoes *- improves venous return.

DRUG THERAPY

Stop drugs that may cause orthostatic intolerance

Considered drug therapy if 3 – 6 months of physiological measures are ineffective.

Drugs used in PoTS are all unlicensed for this indication.

Some hospitals offer a shared care agreement.

Fludrocortisone **Desmopressin**	**Increase blood volume**
B Blocker **Ivabradine**	**Reduce heart rate and diastolic filling**
Midodrine	**Alpha -1 agonist / vasoconstrictor**
SSRIs / SNRIs	**Affects central control of HR and BP**
Clonidine	**Alpha-2 agonist / centrally sympatholytic**
Methylphenidate	**Vasoconstricts**
Erythropoeitin	**Increase red cell mass/vasoconstricts**
Octreotide	**Splanchnic vasoconstrictor**
Pyridostigmine	**Facilitates ganglionic neural transmission**

PSYCHOLOGICAL SUPPORT

CBT to help patients adjust to long-term illness and symptoms.

WHY IS PoTS UNDERDIAGNOSED?

Patients can repeatedly present with a multitude of symptoms including anxiety, often without clinical findings - typical 'heart-sink' patients.

Patients may not recognise the significance of symptoms or be reluctant to divulge them for fear of the clinician's response.

Blood pressure and pulse recordings are usually taken with patients seated when recordings may be normal.

There is little knowledge of PoTS within the medical community. It is often misdiagnosed as anxiety, panic attacks, vasovagal syncope, chronic fatigue syndrome or inappropriate sinus tachycardia.

WHY IS THIS IMPORTANT?

Diagnosis of PoTS is commonly delayed by several years.

PoTS causes considerable disability - patients can become wheelchair or bed bound. They are often unable to continue education or employment (25%)

Treatments are available and 90% of patients will respond.

Patients frequently receive psychiatric labels prior to correct diagnosis. Tools used to assess anxiety commonly rely on somatic anxiety symptoms (which may reflect high upright catecholamine levels). Using the Anxiety Sensitive Index (a cognitive-based measure of anxiety), PoTS patients tend to have lower anxiety levels than the general population.

WHAT SHOULD I DO IF I SUSPECT PoTS?

PoTS should be considered in patients (especially young women) with a combination of unexplained symptoms eg dizziness, syncope, fatigue, palpitations/tachycardia, headaches, exercise intolerance, anxiety.

Have a high index of suspicion in diabetes, chronic fatigue syndrome and joint hypermobility syndrome.

Stand test - this can be undertaken in a GP consulting room.

Exclude other pathology with biochemistry tests (see above) and ECG (in syncope or family history of sudden death, exclude prolonged QT interval).

Consider **24 hour BP/ heart rate monitoring** – normal recording does not exclude PoTS. (If equipment cannot be triggered by patient to record when symptomatic, PoTS, vasovagal syncope and orthostatic hypotension can be missed)

If PoTS is suspected, **referral** should be made to a physician with an interest in PoTS as diagnosis and management can be challenging. These consultants are often cardiologists with an interest in arrhythmias (i.e. cardiac electrophysiologists who may work in a syncope or blackout clinic). There are also some neurologists, medicine for elderly and neurovascular (autonomic) consultants with an interest in PoTS. There is a list of doctors with an interest in PoTS and syncope on the PoTS UK and STARS websites. (see below)

Further information for clinicians, patients and carers is available from:

www.potsuk.org
www.stars.org.uk
www.gpnotebook.co.uk
http://www.patient.co.uk/health/postural-orthostatic-tachycardia-syndrome

*currently no available medical evidence but generally accepted by experts.

Written by Dr Lesley Kavi
Approved by Professor M D Gammage
Updated 1/4/2014
Review date 1/4/2016
Version 2

Glossary for patients

arrhythmia - abnormal heart rhythm
auto-immune - when the immune system produces antibodies against the body's own tissues
autonomic - belonging to the nervous system that controls bodily functions that we do not have to think about
catecholamine - a chemical produced by the adrenal gland
CBT -cognitive behavioural therapy, a talking therapy
cerebral perfusion - blood supply within the brain
deconditioning - lack of physical fitness
heterogeneous - varied or diverse
hypoperfusion -reduced flow of blood / fluids through tissues
hypotension - abnormally low blood pressure
hypovolaemia - low volume of circulating blood
inotropic - ability of muscles to contract, usually related to heart muscle
multi-system - affecting several organs in the body
neutrally mediated hypotension - low blood pressure due to abnormality of the autonomic nervous system
noradrenaline - is a type of catecholamine-see above
orthostatic intolerance - symptoms that develop in the upright position and are relieved by lying down
orthostatic tachycardia - increased heart rate due to becoming upright
physiological measures - lifestyle changes that do not involve medication
prolonged QT interval - ECG abnormality that can sometimes lead to serious abnormal heart rhythms
sympathetic - belonging to the autonomic nervous system and involved in the 'fight or flight' response
syncope - brief loss of consciousness due to reduced blood supply to the brain
vasoconstriction - narrowing of blood vessels
vasodilatation - dilatation of blood vessels
vaso-vagal syncope - transient loss of consciousness caused by an abnormal functioning of the autonomic (parasympathetic) nervous system
venous return -return of blood through blood vessels towards the heart