

Request for Adoption of a Unique International Classification
of Disease Code for Postural Orthostatic Tachycardia
Syndrome: Request for Modification to
ICD-10-AM/CHI/ACS Classification 10: 13th Edition

on behalf of

The Australian POTS Foundation [APF]

and

The Australian Dysautonomia and Arrhythmia Research
Collaborative [ADARC]

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Executive summary

We are seeking an amendment to the International Classification of Diseases (ICD) 10th edition in Australia in order to provide a unique code for Postural Orthostatic Tachycardia Syndrome (POTS).

- POTS is a condition of the autonomic nervous system which largely affects women of child-bearing age but can be experienced by both men and women of any age. It is associated with high disability and low health related quality of life. ^(1, 2)
- Multiple international and an Australian landmark prospective study have demonstrated that POTS is the predominant phenotype in Long Covid with up to 79% of those with Long COVID meeting criteria for POTS. ⁽³⁻⁵⁾
- Australian patients report poor and inequitable access to diagnosis and care for autonomic dysfunction which is poorly recognised by clinicians. ^(2, 6)
- In the absence of a unique code, POTS is frequently allocated inappropriately to ‘other cardiac arrhythmias’ to which it does not relate, resulting in poor vision of the burden of the syndrome in Australia. ([NDIS participant diagnosis](#))
- The current lack of a unique ICD code for POTS, thwarts targeted research and health policy pathways and disincentivises diagnosis, management and treatment resulting in higher health utilisation and poorer health and economic outcomes.
- POTS has a recognised international diagnostic criterion. The World Health Organisation (WHO) identifies POTS as a medical condition and has allocated a unique code for POTS in the ICD-11th edition. However, this does not service the immediate and urgent need to track the burden in Australia while waiting for the adoption of ICD-11.
- **In October 2022, the Centre for Disease Control (CDC) approved an amendment to include POTS in the ICD-10-CM in the USA. USA clinicians may now use G90.A as the ICD-10 diagnostic code which allows tracking of the burden of this condition across multiple disease states including Long Covid.**
- Failure to include POTS in the Australian ICD-10 will lead to further delay in diagnosis, treatment, and blinds both government and researchers to the high health burden this condition places on the Australian Health Sector.
- The escalating presentation of those with POTS post Covid places an immediate need for urgent action on inclusion of POTS in the current iteration of the ICD register.
- A code amendment would allow full vision of the need to escalate the PBS approval of effective pharmacotherapy for POTS, leading to more equitable access to treatment of this disorder.

Overall, the emergency inclusion of POTS in the ICD-10, is necessary to improve recognition, diagnosis, and treatment of this debilitating condition. Allocation of a code would improve traceability and development of research and health policy strategies to manage the disorder in Australia.

What is POTS?

Postural orthostatic tachycardia syndrome (POTS) is a condition of the autonomic nervous system which largely affects women of child-bearing age but can be experienced by both men and women of any age.⁽⁷⁾

The autonomic nervous system controls many of the ‘unconscious’ functions of the body including heart rate and blood pressure, bladder regulation, gut movement and digestion, sweating, temperature control, stress responses (fight or flight). This explains why those with POTS often present with a vast array of seemingly unrelated symptoms such as fatigue, headache, gut disturbance, dizziness, pain, poor concentration and light-headedness.^(8,9)

Quality of Life

POTS is associated with high levels of social, economic and health burden. Many who experience the condition, find it difficult to work, engage in social activities or attend school. *25% of POTS patients report having to stop work or education due to the impact of the condition on their health.* Despite this, many still report a significant delay in diagnosis.⁽⁷⁾ Recent research from our group demonstrates that those with POTS have worse quality of life than most of the major chronic illnesses including chronic kidney disease, chronic cardiovascular disease, diabetes and heart disease.⁽²⁾

Prevalence

Estimating the incidence of POTS in Australia is challenging due to the lack of population-based studies. However, studies from other countries suggest that POTS is a relatively common condition, affecting up to 1-3% of the population. Moreover, the emerging evidence suggests that POTS is a common complication of Long COVID, with studies suggesting that 30%-80% of Long COVID patients may develop dysautonomia, including POTS.⁽⁵⁾ A recent study by the University of Adelaide, has demonstrated that 79% of Long Covid is attributable to POTS, based upon objective autonomic testing.⁽⁵⁾ Given the significant number of Australians who have been affected by COVID-19, the potential incidence of POTS in this population is a concerning public health issue that warrants further investigation and attention.

Aetiology

The pathophysiology of POTS is complex and diverse, and the underlying cause of the condition can vary widely among patients. Some of the known contributors to the development of POTS include hypovolemia or reduced blood volume, small nerve fibre neuropathy, dysregulation of G-protein coupled receptors, and viral infections such as Epstein-Barr virus and SARS-CoV-2.⁽¹⁰⁻¹³⁾ There is a high association between autonomic dysfunction and those with heritable connective tissue disorders including Ehlers-Danlos Syndrome.⁽¹⁴⁾

It is worth noting that many patients with POTS have a combination of these factors, and the interplay between them is not yet fully understood. Further research is needed to develop a comprehensive understanding of the pathophysiology of POTS and to identify effective treatment approaches.

Diagnostic criteria

Several organisations have issued international consensus statements on the diagnostic criteria for POTS in recent years. These include the Heart Rhythm Society (HRS), the European Federation of Autonomic Societies (EFAS), and the American Autonomic Society and American Academy of Neurology (AAS-AAN).^(9, 15)

The HRS published a consensus statement in 2015, which provided updated diagnostic criteria for POTS based on a comprehensive review of the available evidence. Professor Dennis Lau from the University of Adelaide [and co-author of this submission] was a contributing author to the HRS statement.⁽⁹⁾

The EFAS issued a similar consensus statement in 2018, which emphasised the need for a multidisciplinary approach to diagnosis and treatment. The AAS-AAN also published a consensus statement in 2019, which provided guidance on the clinical evaluation and management of POTS.

While there is some variation in the diagnostic criteria and recommendations across these consensus statements, they all emphasise the importance of a thorough clinical evaluation, including autonomic function testing, to confirm a diagnosis of POTS and rule out other potential causes of orthostatic intolerance.

International Diagnostic Criteria

- A sustained HR increment of not less than 30 beats/minute within 10 min of standing or head-up tilt. For individuals who are 12 to 19 years old, the required HR increment is at least 40 beats/minute; and
- An absence of orthostatic hypotension (i.e., no sustained systolic blood pressure [BP] drop of 20 mmHg or more in the first 3 minutes of standing); and
- Frequent symptoms of orthostatic intolerance. Symptoms may include lightheadedness, palpitations, tremulousness, generalised weakness, blurred vision, and fatigue; and
- Duration of symptoms for at least 3 months; and
- Absence of other conditions explaining orthostatic sinus tachycardia such as anaemia, dehydration or severe deconditioning caused by prolonged bed rest.

POTS is known to have a significant diagnostic delay with many individuals seeking review with multiple specialists yet still reporting delays of four years or more until diagnosis.

There is an absence of MBS items specific to autonomic testing and while Tilt Table Testing is reimbursable and occasionally used for POTS diagnosis, this test is cumbersome and expensive and is not routinely required to confirm diagnosis.⁽¹⁶⁾ An MBS item number specific to autonomic testing and in particular, pathology and diagnostic testing associated with small nerve fibre dysfunction, would allow for more efficient use of clinician and hospital resources.^(17, 18)

Treatment

There are several pharmacological and non-pharmacological treatment options available to reduce symptom burden and enhance functionality in those living with POTS.

Beta blockers are considered a first line treatment but may be contraindicated in many POTS patients due to the blood pressure lowering properties of the medication. However, other medications such as Ivabradine and Midodrine are effective but are not PBS approved for the condition, leading to inequitable access to appropriate treatment.

Consensus statements from several esteemed professional organisations including the Heart Rhythm Society, the Canadian Cardiovascular Society, the American Autonomic Society, and the POTS Working Group for the United States National Institutes of Health have detailed the benefit of pharmacotherapy in POTS.

Non-pharmacological interventions such as increasing salt and fluid intake, can also be helpful in increasing blood volume and improving orthostatic symptoms. In addition, compression garments, such as abdominal binders and compression stockings, can be used to improve venous return and reduce orthostatic symptoms in patients with POTS.

The below medications have been studied in POTS populations and demonstrated benefit.

- Midodrine, an alpha-1 agonist, is another medication that is commonly used to treat POTS by constricting blood vessels and increasing blood pressure. Currently, the TGA has approved Midodrine for the treatment of orthostatic hypotension only.
- Ivabradine, a selective sinus node inhibitor, has been shown to be effective in reducing heart rate without causing significant changes in blood pressure, making it a potential treatment option for patients with POTS.
- Propranolol, a beta-blocker, is also used to treat POTS by reducing the heart rate and improving orthostatic symptoms.
- Fludrocortisone, a mineralocorticoid, is another medication that can be used to treat hypovolaemia in patients with POTS.
- Desmopressin, a selective V2 agonist that promotes reduced urinary excretion thereby assisting with hypovolaemia.

Midodrine is currently in short supply in Australia, rendering individuals who are already struggling to function on a daily basis, unable to manage their symptom burden. We urgently call on the TGA to address this and implement an exemption to expand the current approved use for orthostatic hypotension to also include POTS.

Increasing International Government Focus

There has been growing international focus and recognition of POTS including:

- 2017: First US Congressional Briefing on POTS
- 2018: US Congress directs National Institutes of Health (NIH) to prepare report for Congress on state of POTS funding needs
- 2019: NIH held first international POTS Research Workshop
- 2020: NIH issued report on POTS to Congress

- 2021: NIH POTS Research Workshop panelists issued independent report – 2 peer-reviewed articles
- 2021: NIH issued Notice of Special Interest to stimulate research on diagnosis, treatment, mechanistic understanding of POTS
- 2021: NIH lists POTS funding in annual categorical spending report for the first time
- 2022: CDC enacted an amendment to the ICD-10-CM to allocate a unique code for POTS; G90.A [as of October 2022]

Inefficiencies of current code usage

1. Existing ICD-10-AM/ACHI code is too general or lacks specificity:
 - a. POTS is often coded to I49.8: Other specified cardiac arrhythmias. This includes Brugada syndrome, ectopic rhythm disorder, junctional rhythm disorder.
 - b. This is an inaccurate placement and description as POTS is not a cardiac arrhythmia and arrhythmias are not associated with POTS. Rather, in POTS sinus tachycardia is noted upon standing (orthostasis) with return to normal sinus rhythm or lower rate of sinus tachycardia when seated/recumbent.
 - c. Others have used a combination of R00.0 plus I95.1 or ACS 0005 syndromes.
2. **POTS has already been allocated a unique code (8D89.2) in the ICD-11 thus providing a precedent for inclusion in ICD-10.**
 - a. **POTS is classified under ‘Disorders of orthostatic tolerance’ in ICD-11**
3. Existing ICD-10-AM/ACHI code assignment is outdated due to advances in medical knowledge:
 - a. Advances in medical knowledge have led to a better understanding of POTS and its underlying pathophysiology. However, the current ICD-10-AM/ACHI codes for dysautonomia do not reflect this increased understanding. As a result, the current coding system may not accurately capture the clinical presentation of POTS, leading to potential misdiagnosis and inadequate treatment. The following have been identified as contributing aetiologies to POTS.
 - i. Inflammation, oxidative stress, and impaired blood vessel function.
 - ii. Alterations in the renin-angiotensin-aldosterone system (RAAS) including increased plasma renin activity and aldosterone levels
 - iii. Alterations in G-protein coupled receptors (GPCRs). Specifically, abnormalities in the function of adrenergic and muscarinic GPCRs have been identified in POTS patients.
 - iv. Evidence of decrease cerebral perfusion.
4. Disease, related health problem or procedure/intervention is currently not classified in ICD-10-AM/ACHI:
 - a. POTS has an internationally agreed upon diagnostic criteria as such it requires a specific code to accurately track its incidence and prevalence in Australia.

- b. Without a specific code, healthcare providers may be unable to accurately document the care provided to patients with POTS, leading to potential underreporting and inadequate reimbursement.
5. ICD-10-AM/ACHI/ACS indexing omissions:
- a. ICD-10-AM/ACHI/ACS indexing omissions can lead to potential misdiagnosis and inadequate reimbursement for healthcare providers. POTS is misclassified under other types of arrhythmias, leading to an inability to track the health burden of the condition.
 - b. A specific code for POTS would reduce the potential for indexing anomalies and omissions, leading to more accurate diagnoses and appropriate reimbursement.

IMPLICATION: The current ICD-10-AM/ACHI codes do not provide specific coding for POTS, and this lack of specificity has significant implications for patient care. Namely:

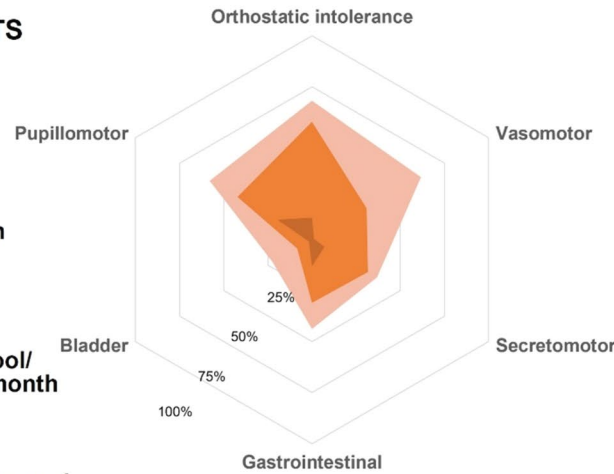
- Lack of appropriate service reimbursement, due to absence of specific MBS item numbers and poor awareness arising from lack of an ICD code, disincentivises diagnosis and treatment through both the public and private sector.
- Diagnostic delays drive higher healthcare utilisation and increased risk for overservicing of pathology and diagnostic studies.
- Delayed treatment impacts of workforce and educational absenteeism directly impacting on private and community economic status.

Conclusion

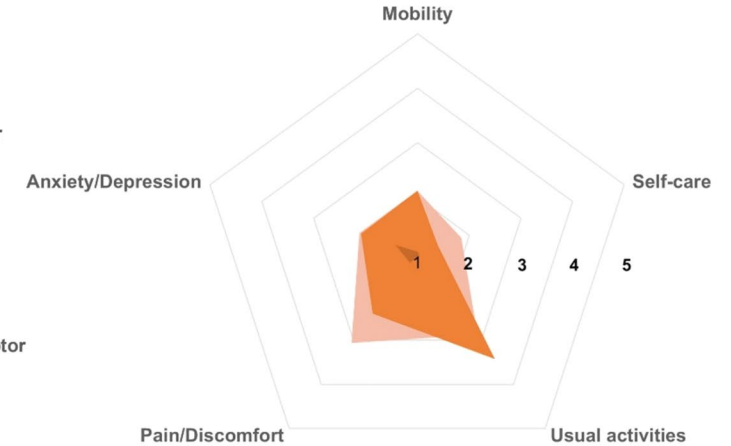
In conclusion, POTS is a debilitating condition with high association to poor functionality and reduced quality of life. As a result of the Covid pandemic the condition is increasing in prevalence. However, POTS is not currently widely recognised by clinicians resulting in extended diagnostic delay and significant burden on an already stretched healthcare system.

There are compelling reasons why Australia needs an International Classification of Disease amendment code for POTS. The existing ICD-10-AM/ACHI codes are too general, lack specificity, and the current coding system does not accurately capture the clinical presentation of POTS. POTS has unique clinical features that distinguish it from other types of dysautonomia, and specific MBS item numbers, addition of effective pharmacotherapy to the PBS, and an ICD code would allow for more timely diagnosis, improved management, reduced disability and greater understanding of the prevalence of POTS in Australia.

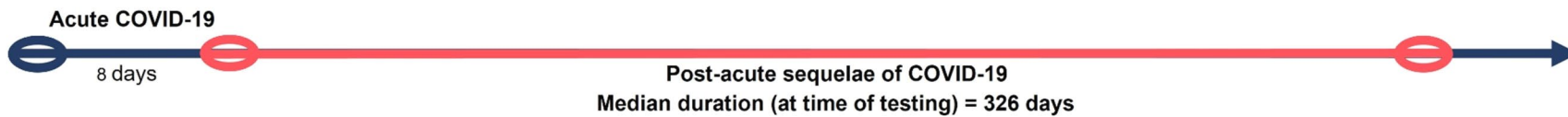
79% of PASC meet the criteria for POTS



■ POTS ■ PASC ■ CONTROL
0 to 100% indicates degree of severity



■ POTS ■ PASC ■ CONTROL
1 to 5 - no problems to extreme problems



Seeley et al. *Clinical Autonomic Research* (2023)

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