National Women's Health Advisory Council Submission: Experiences of Women with Postural Orthostatic Tachycardia Syndrome in Australia

on behalf of

The Australian POTS Foundation [APF]

and

The Australian Dysautonomia and Arrhythmia Research Collaborative [ADARC]

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Executive summary

- POTS is a condition of the autonomic nervous system which largely affects women of child-bearing age.
- The syndrome is associated with high disability, increased unemployment and low health related quality of life.
- Women with POTS have worse quality of life than most other major chronic illnesses including chronic kidney disease, chronic cardiovascular disease, diabetes and heart disease.(1)
- Australian women experience a diagnostic delay almost five times that reported in international cohorts and almost twice that of Australian males.
- International research shows female adolescents have a 503% increased risk of poor prognosis compared to males. (2)
- There is poor clinician awareness of autonomic dysfunction in Australia which is compounded by a lack of access to diagnostic testing.
- Australian women with POTS frequently report their symptoms are dismissed by health professionals despite frequent Emergency Department visits and multiple consultations with specialists.
- The current lack of a unique ICD code for POTS [in Australia], disincentivises diagnosis, management and treatment resulting in increased barriers to diagnosis and delayed treatment.
- Multiple studies have demonstrated that POTS is the predominant phenotype of Long Covid.(3) The escalating presentation of those with POTS post Covid places a need for urgent action to recognise the disorder and its significant impact on education and earning capacity for women who have the syndrome 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.(1, 4)

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.(5, 6)

Prevalence

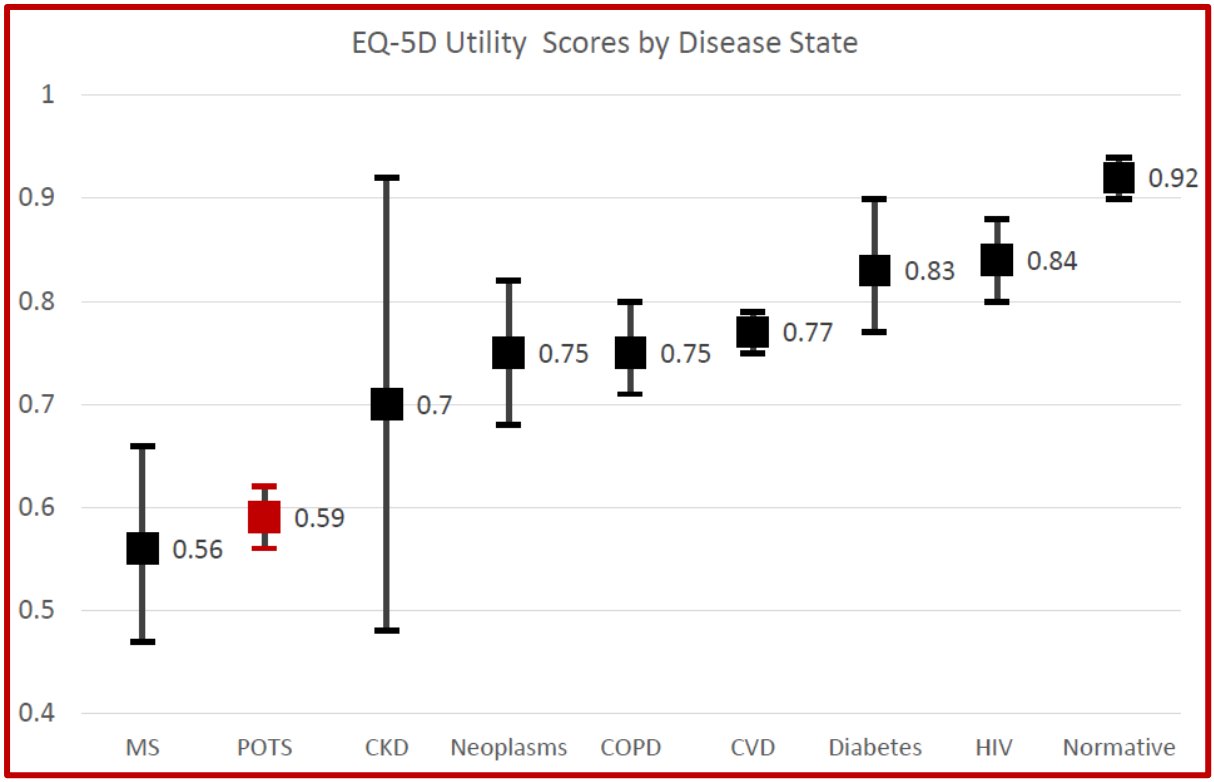
Estimating the incidence of POTS in Australia is challenging due to the lack of population-based studies. However, studies from other countries suggest that POTS is a relatively common condition, affecting up to 1-3% of the population. Moreover, the emerging evidence suggests that POTS may be a common complication of Long COVID, with some studies suggesting that 70% of Long COVID patients may develop dysautonomia, including POTS. A recent study by the University of Adelaide published in the American Journal of Medicine, has demonstrated that >70% of Long Covid is attributable to POTS, based upon objective autonomic testing.(3) Woman were the predominate sex affected by Long COVID in this study which is consistent with the majority of international studies into Long COVID. 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.

(7)

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.(4) Recent research from our group demonstrates that those with POTS have worse health-related quality of life [HrQoL] than people living with almost any of the highly funded, major chronic illnesses including chronic kidney disease, chronic cardiovascular disease, diabetes and heart disease. [Figure 1.] (1)

Figure 1. Comparisons of HrQoL between major chronic disease states and POTS



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MS-multiple sclerosis, CKD-chronic kidney disease, COPD-chronic obstructive pulmonary disease, CVD-cardiovascular disease, HIV-human immunodeficiency virus
Health Utility; 0 = Death and 1 = Full Health

Sex related outcomes in POTS.

The following statistics are taken from the Australian Postural Orthostatic Tachycardia Syndrome [POTS] Patient Registry [n = 457]

- Despite the female predominance of the syndrome [Figure 2], women universally experience a significantly longer diagnostic delay compared to men. (8) In Australia this diagnostic delay for females is almost twice that of males [Figure 3.]
- In total, 22% of adult women on the Australian POTS registry are unemployed or unable to work.
- In total, 61% report attending an Emergency Department [mean 6 times] for their POTS symptoms without a resulting diagnosis.
- On average, Australian women consult over 5 doctors before diagnosis.
- Australian women with POTS are more frequently mis-diagnosed with anxiety and depression than men [Figure 5.] despite:
    - Having more severe autonomic symptoms than men [Figure 4.]
    - Having a higher burden of multimorbidity [≥3 chronic health conditions] than men [Figure 6.]

Figure 2. Sex and race in POTS

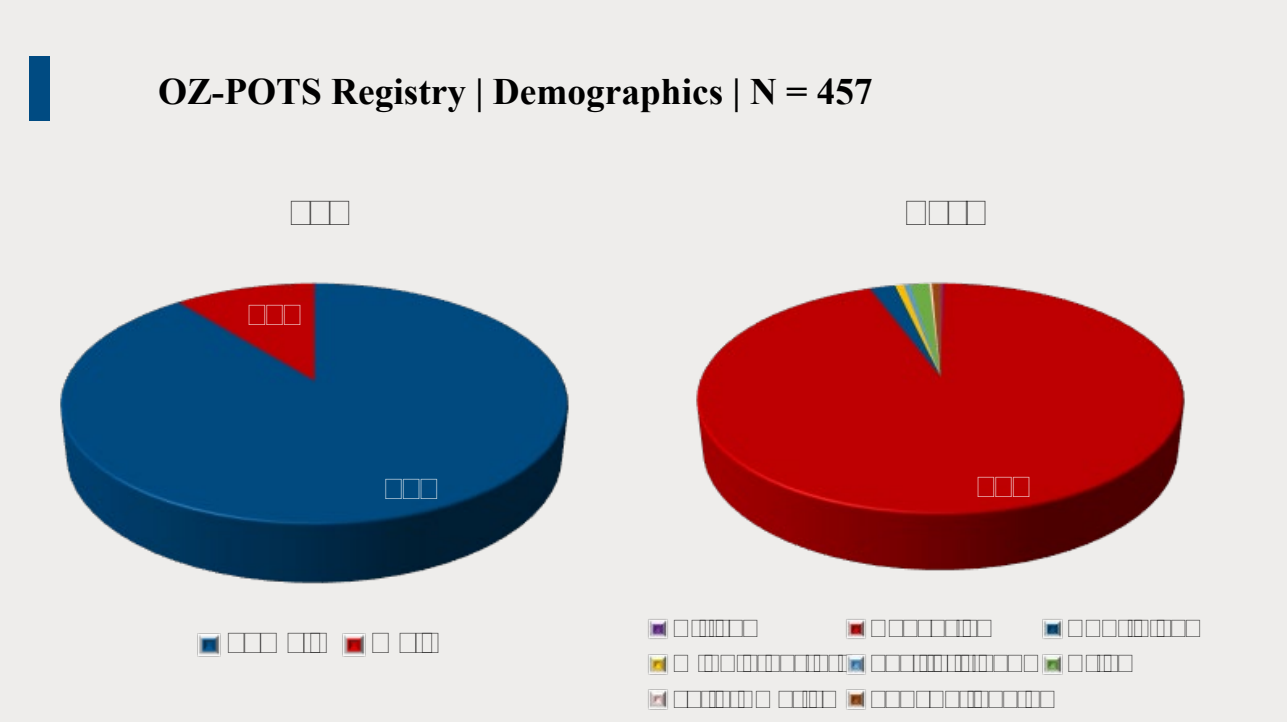
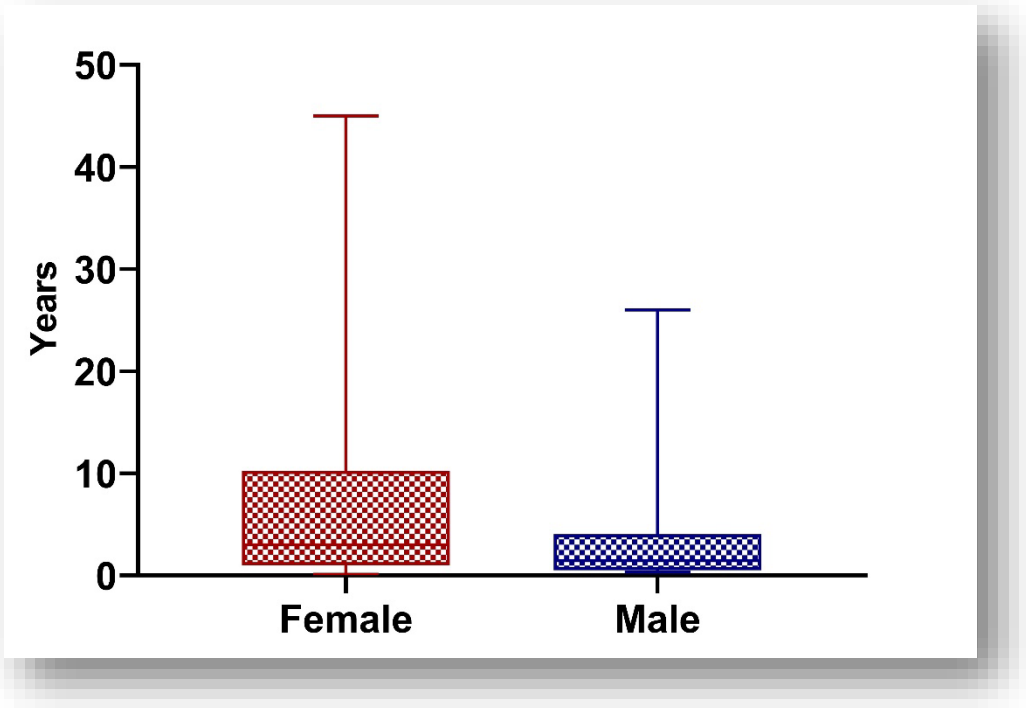
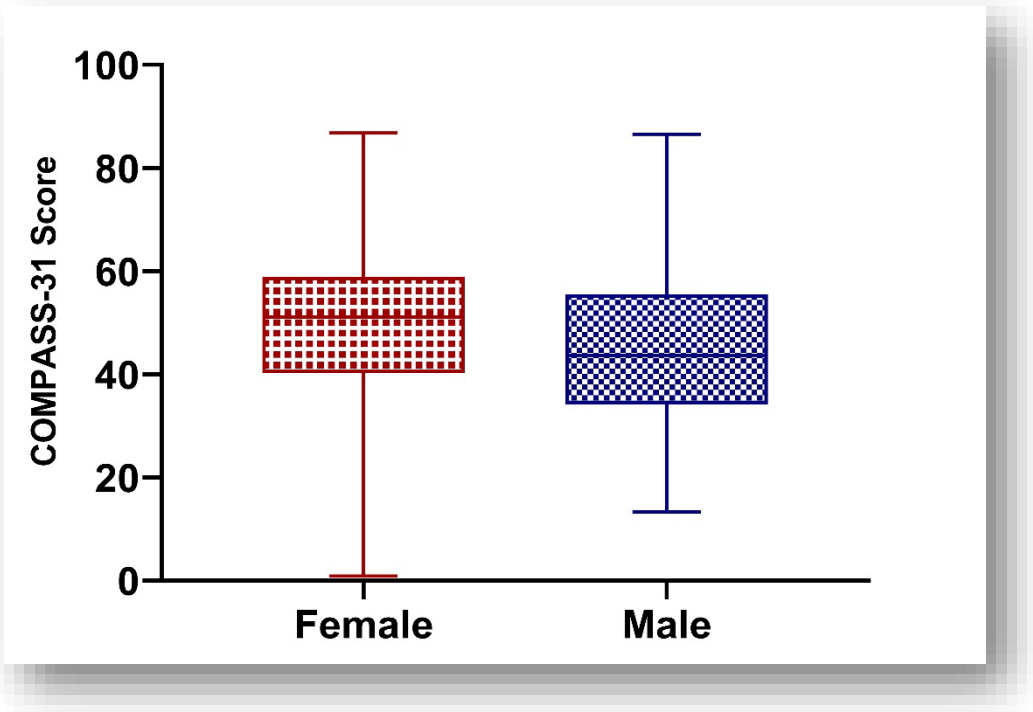


Figure 3. Sex differences in diagnostic delay in POTS [Australian POTS Register]



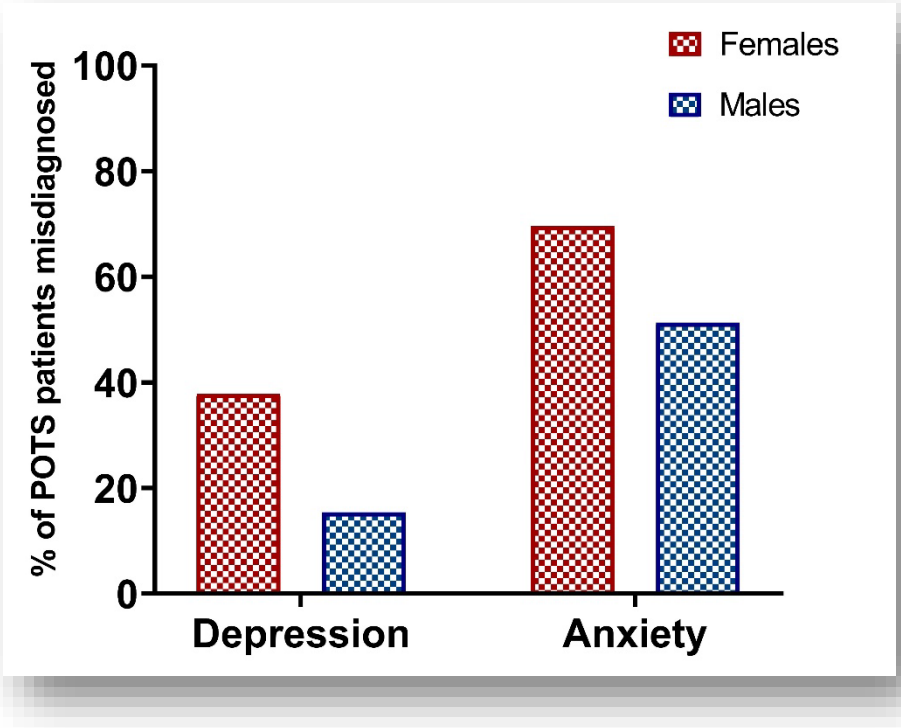
Mean diagnostic delay = Female 6.9 years [SD 8.8] versus Male 3.8years [SD 5.8]; p = .04

Figure 4. Composite autonomic symptom scores by sex



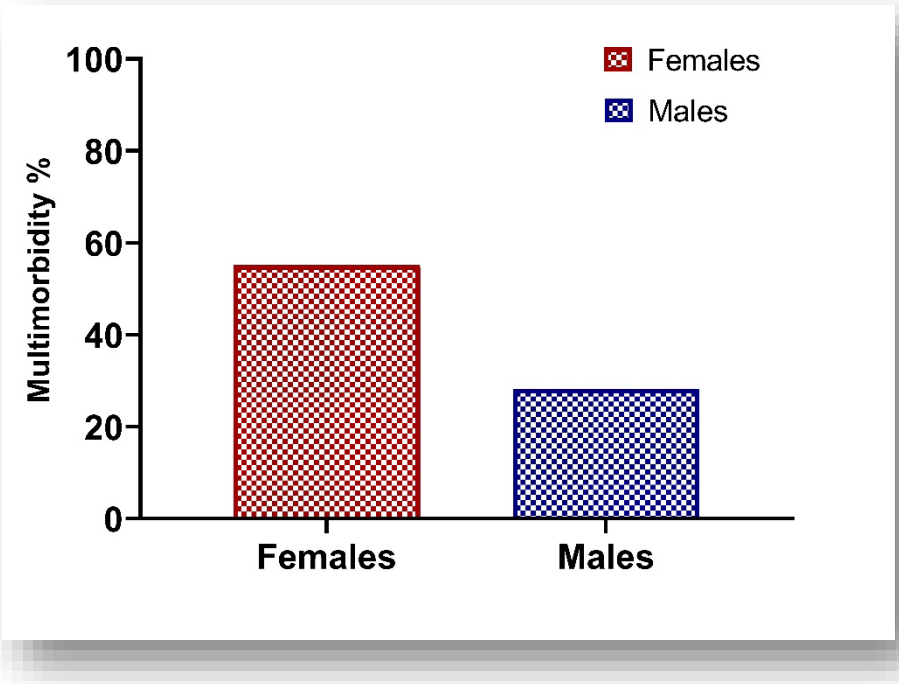
Higher score = worse autonomic symptoms
Mean COMPASS-31; Females 50 [SD 13] vs. Males 44 [SD15]; p = .01

Figure 5. Attribution of somatic POTS symptoms to psychiatric cause



Females more frequently report misattribution of POTS somatic symptoms to psychiatric cause. Depression [37.8% vs 15.4%; p = .005]; Anxiety [69.6% vs 51.3%; p = .020]

Figure 6. Multimorbidity % [presence of ≥3 comorbidities] by sex



Females experience higher levels of multimorbidity than males [55.1% vs 28.2%; p =.001]

The lived experience of the diagnostic odyssey of women with POTS

Below are the personal stories of women who attended our clinic and enrolled participants of the Australian POTS registry. When asked about their diagnostic journey, this is what they said.

> *"Exhausting, gas lit, you're a healthy young woman. You need to just get used to having a fast heart rate."*

> *"I was dismissed, gaslighted and ignored for 7 years prior to getting a diagnosis. I experienced years of being told there was nothing physically wrong with me, and that all of my symptoms were a result of anxiety or panic disorder. When CBT failed to treat my "anxiety", no one ever pushed further. An experience that stands out to me is the last time I saw the GP that was involved in my care for 7 years. It was following a 7-day hospital stay due to malnourishment as a result of severe, unrelenting nausea, where we finally realised that my heart rate was skyrocketing every time I stood up. My GP looked me in the eyes and told me to "stop attention seeking" and that she didn't care unless my heart rate was 200 at rest. It makes me incredibly angry how long I had to suffer before someone finally listened and took me seriously."*

> *"[I] was told that it was in my head and that I just didn't want to go to school. Was hospitalised and because they did not understand what was wrong with me, I had to see a physiologist. Had treatment which made my condition worse by making me exercise. So left hospital in a worse condition than originally went in. Still seeing professionals that don't understand or just think I get dizzy. No support for my wellbeing or my family.*

> *"[I was] brushed off by a paediatrician when [I] suggested POTS with "POTS is very controversial and it doesn't just come on like that". Other GPs and another paediatrician didn't know how to manage the condition once a diagnosis was received."*

> *"My experience with other health professionals was awful and detrimental to my well-being. No-one believed my symptoms, I was mis-diagnosed, I ultimately had to find my diagnosis and when I suggested it to one specialist, she basically mocked me and did not believe it was possible that I could have dysautonomia or POTS. "*

> *"Over a period of many years, I spent thousands of dollars on specialist appointments with general physicians, rheumatologists and cardiologists. I was told by a general physician that my debilitating fatigue was psychiatric. I was told by a cardiologist that my high heart rate was psychiatric and discouraged from seeking a Dysautonomia diagnosis as it was "almost impossible in Adelaide".*

"I found most of the GP's I saw to be dismissive. One gave me a script for Valium and advised me to "tell myself to calm down", despite explaining that I experienced tachycardia when standing and resolved when sitting. I did see some medical students (cardio) in hospital who were very good and first suggested POTS - this was dismissed by the actual cardiologist".

*"I saw in excess of 20 different doctors during my diagnosis journey. Many of these top ED doctors. I was dismissed for having anxiety mostly. Some doctors told me I was being a dramatic woman, some told me there wasn't anything wrong with me. Finally, I stumbled across my current GP .. She didn't dismiss me, and she did a standing heart rate and immediately she was like "hang on, this appears to be POTS". And made the referral to see **. My life changed from that moment on. I'm glad I took one more chance to see someone about what was going on."*

*"I had been dismissed as 'stressed' by numerous GPs. I had to insist that my standing heart rate be measured and then ask for a referral to Dr**. The GP who referred me made a ... derogatory comment about POTS being connected to eating disorders. I had seen 3 other cardiologists prior to diagnosis."*

"Most practitioners were redirecting me towards mental health diagnosis as they stated that I was experiencing panic attacks due to anxiety and stress. None of them were listening to my descriptions of the symptoms I was experiencing. One GP also suggested that it was all in my head and that I should just get out of bed and be more active."

"It was terrifying to have a body that wasn't working and Drs who thought I was crazy! I started to think I was crazy too! One specialist told me he couldn't spell dysautonomia. A cardiologist said she hadn't heard of POTS. A cardiac senior doctor told me "Don't come back to ER as there's nothing we can do for you; POTS isn't a real syndrome" .. and my "symptoms would vanish in six weeks" despite me having them for 6 months at that stage. My first introduction to POTS was during yet another ambulance trip and it was the female driver who asked me if I'd heard of POTS. She told me to suggest it to the ER doctor. He was young and open to my suggestion.

"One cardiologist told me my heart rate of 180 bpm seated was anxiety over seeing a piece of medical equipment in her office. I could not convince her that my heart was always beating that high! The ER staff knew nothing of autonomic issues, and I was sent home with no advice or help .. just told to come back if my heart was erratic again."

"I have been gaslit by doctors about my health for many years. I had given up but got to a point where I could no longer work. I looked for answers intensively for two years before being diagnosed. I had suggested POTS to my GP six months prior with heart rate data but was told I was being anxious. It was so, so hard to get diagnosed."

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"I could write journal on my experiences in the healthcare system so far, but it would just be unhinged and angry. I don't have enough time away from it to have emotional control so I think I can only say traumatising and dehumanising."

"Even though I have 10 years' experience as a specialist Registered ICU Nurse, NONE of my Doctors/Specialists/GP's believed my symptoms. My GP of 15 years diagnosed me instead with 'anxiety and depression'. After 10 different anti-depressants and nothing was working, I advocated that it was NOT mental health. My physical symptoms were not 'imagined or the cause of anxiety'. None of my GP's believed my reports of chest pain, palpitations or frequent falling downstairs and other severe syncope episodes. My newly acquired secondary hypertension went untreated for over 3 years, because I was accused of being 'anxious' when my BP was taken during Dr.'s visits. Even post being diagnosed 2x GP's did not believe POTS is an actual medical condition and declared that Dysautonomia does not exist. It's been the most horrendous experience. I literally thought I was dying with my symptoms, because of how ill and sick I've been and then when I sought medical attention told it was all psycho-somatically induced. It was not! I can greatly empathise and understand how some people could take their own lives or become chronically suicidal (none of which I am) if they are called 'crazy' or 'anxious' and accused of creating their own illness and physical symptoms, and medically gaslighted my other Dr.'s or specialists for years. I have no idea how some people have navigated their way through the health system to finally get a diagnosis years later, having no medical or nursing background! "

"It was torture and an absolute nightmare that led me to suffer severe medical trauma, lack of faith in medical professionals and medication trauma. Not to mention the enormous financial strain on numerous specialists, tests and psychological manipulation/medical gaslighting that put me at risk of serious harm."

"Downplaying of my symptoms, [they] told me that 'being lightheaded when you stand up is just a part of being a woman' and 'no one faints that often'.

*"Dealing with gaslighting and belittlement of my symptoms, unwillingness of GPs to research long Covid and associated illnesses and unwillingness to listen to patients was very disheartening. I was already quite sure that I was suffering POTS prior to coming to ***. The only useful thing my GP did was eventually give me a referral despite being sceptical it was needed".*

Conclusion

In conclusion, POTS is a debilitating condition which is highly associated with poor quality of life and increased unemployment. The condition has a high female preponderance however it is not currently widely recognised by clinicians resulting in extended diagnostic delay. Australian women experience significantly longer diagnostic delays than their male peers or indeed than females living outside Australia. Woman also experience a higher burden of symptoms and more comorbid chronic health conditions than their Australian male counterparts and are more likely to have their symptoms attributed to anxiety or depression. The lived experiences of women with POTS in Australia detail a diagnostic odyssey marked with disbelief, dismissal and clinical neglect. There is an urgent need for education, research and funding to support the improvement of diagnosis and access to care for women with POTS in Australia.

Appendix

POTS 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.(9-12) There is a high association between autonomic dysfunction and those with heritable connective tissue disorders including Ehlers-Danlos Syndrome. (13)

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).(6, 14)

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. (6)

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 light-headedness, 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.

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.

NB: Midodrine is currently in short supply in Australia, rendering individuals who are already struggling to function, unable to manage their symptom burden.

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