

Dear Policy Maker,

I wish to bring to your attention a poorly understood and widely unknown medical conditional from which I suffer; Idiopathic Hypersomnia (IH). This rare disease took from me my dream career in the service of our country and continues to be debilitating on a daily basis. Treatments are minimal at best and support is largely non-existent, because of a dearth of funding for research to develop treatments and identify efficacious supports for sufferers.

Nearly 20 years ago I was appointed to be an officer in the Australian Regular Army and began training to be a pilot. For as long as I have had career aspirations, I have wanted to be a military aviator. Many desire this and very few achieve it; I was told at one point that for every 2,500 pilot hopefuls that contact Defence Recruiting, only one applicant makes it to flying training. Clearly, the Australian Defence Force can afford to offer pilot positions to only the best whom apply, and fortunately I was one of them.

During training at Royal Military College – Duntroon (RMC-D), I began to notice it was quite difficult to stay awake in some classes. This made sense in the circumstances as the course was very intensive. I, and it seems the directing staff, put this down to a young man (I was the youngest of my officer intake) experiencing the rigours of full-time initial military training. In hindsight, this was the first warning bell.

Staying awake became more of a problem at Basic Flying Training School in Tamworth, NSW and after many weeks of ground school and a few weeks of flying, my excessive sleepiness could no longer be attributed to a new environment. The unit medical staff declared me temporarily medically unfit to fly and I was transferred to Headquarters 16th Brigade (Aviation) at Gallipoli Barracks in Enoggera, Queensland. At this unit I was closer to better-equipped medical facilities that could assess my wakefulness issues – it was expected that I would return to flying training in around three months. Sixteen months later, I was medically discharged from the army.

Defence medical officers did the best they could in the circumstances. During my final medical evaluation shortly before the decision to discharge me was made, I learned that the Royal Australian Air Force Chief of Aviation Medicine herself, (then) Group Captain Dr Tracy Smart, had to consult with the Israeli Air Force and United States Navy in an attempt to find reference cases against which to assess my clinical results, prognosis and diagnosis. This is to say that in the 90 year history of Australian military aviation there was no record of a case similar to mine, such is the rarity of the disease. Even the Civil Aviation Safety Authority could not provide guidance from a civilian aviation perspective. (Now) Air Commodore Dr Smart was published in a scientific journal on my (deidentified) case study.

The inordinate delay and difficulty the ADF had in determining my medical status was indicative of the lack of scientific understanding of Idiopathic Hypersomnia. Little has changed since then; the cause is not understood, diagnoses are slow to be made or misdiagnosed, treatments are not supported and IH sufferers are left to cope, or not, as best they can.

Every morning I wake with sleep drunkenness. The best way I could describe this is not as a strong desire to stay in bed and sleep a little longer, but that getting up is almost physically painful. I have lost count of the number of morning appointments I have missed because I have either entirely slept through my exceedingly elaborate alarm ritual or simply fallen back asleep literally within seconds of waking up. This experience is not alleviated by a longer- or better-night's sleep; it is a symptom of IH. The only difference between 4- or 10-hours sleep is the severity of the sleep drunkenness, and even then the correlation is not always present.

Not only is it a feat of Olympic effort to wake up and get out of bed in the morning; once up my mind is in a fog for a frustratingly long time. Frustrating for me and frustrating for my family: my wife is not allowed to tell me anything important within an hour of me waking up. I can't make decisions. I'm unable to play with my five-year-old son. I have to function on 'auto-pilot' until the fog clears, typically no less than an hour. This in itself is *extremely* limiting on my participation in society, especially since 'early birds' are so often seen as virtuous, hard-working, and thus rewarded. I have had far too many conversations with employers/managers about being late to work.

As if sleep drunkenness were not difficult enough to live with, the desire to sleep is ever-present throughout the day. 'Tired' is an entirely inadequate description for a sufferer of IH. 'Tired' is to IH as 'questionable decision making' is to Vladimir Putin. The feeling of tiredness is perpetual and unresponsive to behavioral interventions such as naps, longer overnight sleep, exercise and diet. A good day at work is one in which I have not involuntarily headbutted the keyboard (quite seriously - I'm not joking for effect).

By the time I pick up my son and return home, he is ready to play yet I simply *must* take a nap on the couch lest I pass out. No doubt any father would love to have the energy of their kindergartener, but I'm sure most are not wanting for sufficient physical capacity to give their kids just 15 minutes of their time.

Many things impact the levels of tiredness for all people, but I can assure you it is something else entirely for sufferers of IH. I have been in excellent health and fitness over the years, from daily physical training in the army to three days of squash and three days of weight training per week in civilian life, with the nutritional profile of a body builder. Fit, unfit, healthy, active, sedentary; it makes little difference to the symptoms of IH. This is why we need to obtain a better understanding of the causes, the most appropriate treatments, and supports.

I am no fool, yet I feel shackled to the ground floor of progress by the chains of hypersomnolence. I was identified as a gifted child in school; was one in 2,500 to make military flying training; at 20 years of age was the youngest in my officer training cohort at RMC-D where most were 25 or older; and scored 105 points on the ABC's *Hard Quiz* (at the time of writing, in 164 episodes of four contestants each (656 contestants), this has been achieved fewer than 10 times). Because of the impacts of Idiopathic Hypersomnia, these trifling and fleeting claims to glory are likely going to be the peak of my achievements. I've almost certainly

hit my career ceiling before reaching 30 years of age as it is just not possible to progress any further with the actual limitations on my concentration and prejudices relating to perceived effort/dedication.

I don't want this to be the end of my story but it will be if there isn't more awareness and support for Idiopathic Hypersomnia. You can help to change the situation for people like me who are living with Idiopathic Hypersomnia by spreading awareness, providing support to Hypersomnolence Australia (see below) and, contributing to the establishment of policies that will provide people with IH a more secure and optimistic future.

What is Idiopathic Hypersomnia (IH)?

Idiopathic Hypersomnia (sometimes referred to as Idiopathic Hypersomnolence in Australia) is a rare neurological sleep/wake disorder. Sufferers have an extraordinary need for excessive sleep yet, despite sleeping for 12 -16 hours (or more) in a 24 hour period, they wake up feeling like they have had no sleep at all.

This results in a range of extremely disabling symptoms including:

- Excessive Daytime Sleepiness (EDS) and the overwhelming need to sleep.
- Impaired vigilance or sustained attention.
- Cognitive dysfunction, especially linked to poor memory.
- Dependence on other people for awakening them.
- Long (>1 hour) unrefreshing naps and profound sleep inertia, known as sleep drunkenness (elucidated as prolonged and extreme difficulty waking up accompanied by confusion, disorientation, irritability, and poor coordination with an uncontrollable desire to go back to sleep. It can also include automatic behaviour with amnesia, ie: performing tasks without consciously knowing it and not remembering you have done them, eg; turning off alarm clocks or answering the phone). This difficulty in transitioning from sleep to wake (often referred to as the time it takes to "get going" in the morning) can last at least an hour but may be as long as 2 to 3 hours when severe.
- Despite extraordinary amounts of good quality sleep, people with IH never feel refreshed; they are in an almost constant state of sleepiness.

The pathogenesis of IH is unknown. An autosomal dominant mode of inheritance has been considered by researchers because around 50% of people with IH have at least one family member who also has symptoms. It appears IH may be more common in females. The prevalence of IH is thought to be 0.005%–0.3% but, due to lack of epidemiological studies, biological markers and misdiagnosis of the disease, the real prevalence is unclear.

Though the symptoms mainly begin in adolescence or young adulthood (typically between 15-30 years of age), it is not uncommon for there to be a delay of many years before an accurate diagnosis is made.

IH is a debilitating, lifelong condition often profoundly affecting work, education, relationships, and mental health. This is particularly the case for those without support.

The House of Representatives Standing Committee on Health, Aged Care and Sport inquiry into Sleep Health Awareness in Australia (2018) found that new drugs and novel medical technologies are urgently needed for people suffering from disorders of hypersomnolence. In the Chair's foreword of the report, Bedtime Reading, Committee Chairman Mr Trent Zimmerman said *"narcolepsy and associated conditions such as idiopathic hypersomnolence can have a debilitating impact on a person's quality of life. There is a need to improve the awareness of these conditions both within the community and among medical practitioners. Further consideration should be given to emerging treatment options that may provide some people experiencing these conditions the opportunity to improve their quality of life."*

Despite this, nothing has been done. There is no research being done in Australia. There is no formalised education or training on Idiopathic Hypersomnia in the sleep specialty curriculum. The Royal Australasian College of Physicians acknowledges the limitations in their training in the 'Sleep Medicine Advanced Training Curriculum', *"There are too few training posts in Australia and New Zealand that can provide broad exposure to and quality training in, the whole range of sleep disorders, particularly non-respiratory sleep disorders"*. And there are currently **no** medications on the PBS for IH. There are medications (including Pitolisant, Solriamfetol and Xyway) being used to treat the symptoms of disorders of hypersomnolence, including IH, in other parts of the world that we do not have access to in Australia. This is a significant problem because the medications which are available here are not suitable for many people. Side effects prohibit a large number of people from using them, they don't address many of the symptoms of IH and their long term inefficiency leaves some people without any options.

Hypersomnolence Australia (HA) has participated in The House of Representatives Standing Committee on Health, Aged Care and Sport Inquiry into Sleep Health Awareness in Australia and the more recent (2020) Inquiry into the Approval Processes for New Drugs and Novel Medical Technologies in Australia, with a particular focus on those for the treatment of rare diseases and conditions where there is high and unmet clinical need. HA has also approached the Australasian Sleep Association (ASA) on numerous occasions about the aforementioned issues, dating back to at least 2014. However, apart from having various ASA members agree that something should be done, nothing has been achieved. People with disorders of hypersomnolence understand that things take time however it is not acceptable for Australia to be so far behind the rest of the world.

People with IH desperately need more awareness, access to better treatment options and they need more support to manage their day to day lives.

Hypersomnolence Australia (HA) does its best to provide support, information, and advocacy for people with IH. However, they are extremely limited due to a lack of resources. HA does not receive funding from any source and they do not charge membership; they provide their services completely free. HA hosted an IH and Narcolepsy Education Day in Brisbane on 10th April, 2022. This was a very worthwhile event, attended by both medical professionals and members of the IH and Narcolepsy patient communities. The event was funded primarily by people in our small community. The general consensus on the Education Day was that patients often receive little or no information when they are initially diagnosed with IH. They are left feeling confused and wondering how they are going to manage the rest of their lives. HA would like to meet this need for information by producing an educational resource for doctors to give to patients when they are first diagnosed. This would be vetted by medical advisors and answer the many questions patients have from the outset. We would also like to curate more events like the Education Day but we can't do any of this without financial support.

Who is Hypersomnolence Australia?

Hypersomnolence Australia (HA) is the only not for profit organisation in Australia, and was the first in the world, to specifically represent and support the neurological sleep/wake disorder Idiopathic Hypersomnia (IH). Its primary purpose is to support the medical community work toward a better understanding of IH, effective treatment options, support for people affected by IH and raising awareness and educating others about hypersomnolence disorders. HA's goal is to change not just the process to diagnosis but also the level of care and services available to patients post diagnosis. However, HA relies solely on donations to survive. This means that the resources and support it can provide is limited. Please add your voice to our plight by engaging with our social media and consider making a donation to support our work. Learn more at <https://www.hypersomnolenceaustralia.org.au>.

References:

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3. Vernet C, Arnulf I, *Idiopathic hypersomnia with and without long sleep time: a controlled series of 75 patients,* Sleep 2009 Jun; 32(6): pp.753-9.
4. Lammers G, Bassetti CL, Dolenc-Groselje L, Jennum PJ, Kallweit U, Khatami R, Lecendreuxi M, Manconi M, Mayer G, Partinen M, Giuseppe P, Reading PJ, Santamaria J, Karel S, Dauvilliers Y. *Diagnosis of central disorders of hypersomnolence: A reappraisal by European experts.* Sleep Medicine, 2020.

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Written by Jeremy for the IH Awareness Project 2022

The IH Awareness Project has resulted in over 50 letters going out to politicians around Australia. The aim of the project is to create awareness of IH and the impact it has on the lives of those living with IH and their family. By sharing our stories and relevant information about IH with the politicians entrusted with representing us we hope to influence policy making in a meaningful way.

