

Dear Policy Maker,

I was diagnosed with a rare neurological disorder, Idiopathic Hypersomnia (IH) in 2019. I was 33 years old.

By this point in my life, I had already been diagnosed with several other chronic health issues which were mostly neurological in nature (Cervical Dystonia, Essential Tremor, Depression and Anxiety), however the diagnosis of IH would become the most life changing.

My partner jokes that I have a “sleeping” superpower... he always likes to tell people about the time he nearly called an ambulance as I slept uninterrupted for 3 days, and then went back for a further 12 hours after a snack and quick trip to the bathroom.

Being able to sleep for great lengths of time (anywhere) is shocking but is astoundingly one of the least impactful symptoms of this condition. The most frustrating symptom of IH for me is that no matter how much sleep I get, I never feel rested... I am constantly in a state of sleepiness. I never feel fully awake. I am fatigued and have a foggy brain, I can't think straight, and I become confused quite easily as I feel so sleep deprived. Although technically I'm not sleep deprived because I get more than enough good quality sleep. Sleep just never, ever, leaves me feeling refreshed, no matter how much I have.

Waking up is without a doubt the hardest (and most dreaded) part of my day as I sleep so deeply that waking up requires me to set multiple loud alarms, and ultimately rely on my partner to wake me and keep me awake. Sometimes, I wake with paralysis, meaning that my eyes (and mind) will wake up, but I can't move my body for several minutes.

When these symptoms first started many years ago, they were incredibly frightening and overwhelming as I did not know what was happening. Although I now understand what is going on, I'm filled with anxiety because it can be so unpredictable. It is also such a physically exhausting and emotionally draining way to start the day.

Once I am out of bed, I must be careful to get up and move slowly as I have low blood pressure which sometimes causes me to faint if I get up too quickly. This is coupled with something I have come to know as “sleep-drunkenness”, which has seen me walk into walls and doors, and fall down our stairs. I need to sit down often while getting ready as I become very fatigued very quickly. This is significantly worse in the mornings. My partner needs to be around to ensure that I do not fall back asleep as there have been several occasions when this has happened in the past.

I have a 9-year-old stepdaughter who stays with my partner and I on a week-on, week-off basis. Without doubt, it is much harder for me to manage my symptoms the weeks that we have her. I find that my self-care significantly drops below the low standard that it already is. I also struggle so much more with day-to-day activities including housework and shopping.

As a result of my poor health, I am unable to work a full-time job. I generally work 3 days per week (15 hours). On school days, I need to be up at 5am to get my stepdaughter to school for a 9am start.

Once I drop her to school, I have between 10am and 3pm (5 hours) before I need to pick her up from school. Regardless of what I need to do during this time I am consumed with sleepiness and fatigue.

To maximise my chances of getting a full night sleep I need to be in bed by 5pm, latest 6pm (a 'full night sleep' for me is approx. 11-12 hours) which means straight home from school, feed her dinner, and then leave her in the care of my partner until her bedtime. I have no time for myself or anything else.

On the weeks without my stepdaughter, I tend to spend the non-workdays asleep because I am utterly exhausted despite the enormous amount of good quality sleep I have at night. It takes me so long to wake up and get going in the mornings that on my workdays I wake at 7am to enable me to start work by 11am.

On the occasion where I need to work additional hours or additional days (inevitable when running a business), this takes time away from sleep and other activities. I find that many things that need to be done simply do not get done because I run out of time, I literally run out of wakefulness. When there are only 12 wakeful hours in a day and approximately 1/3 of this is consumed by my morning wake up routine (of sleep inertia and sleep drunkenness), there isn't much time for anything else.

My health is suffering but so is my quality of life. I need support but there is nothing available for people living with IH.

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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2. Trotti LM. *Waking up is the hardest thing I do all day: sleep inertia and sleep drunkenness*. *Sleep Med Rev*, 35, 2017, pp. 76-84.
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.
5. The House of Representatives Standing Committee on Health, Aged Care and Sport, inquiry into Sleep Health Awareness in Australia, *Bedtime Reading*, 2019.

Written by Sandy Rice 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.

