A Patients View on Reclassifying Idiopathic Hypersomnia to Narcolepsy Type-3

AKRAM, Umair <http://orcid.org/0000-0003-0150-9274>

A Patients View on Reclassifying Idiopathic Hypersomnia to Narcolepsy Type-3

AKRAM, Umair <http://orcid.org/0000-0003-0150-9274>

Title: A patients view on reclassifying idiopathic hypersomnia to narcolepsy type-3

Author: Umair Akram PhD[a,b*]

[a]Department of Psychology, Sociology and Politics, Sheffield Hallam University, Uk.
[b]Nuffiled Department of Clinical Neurosciences, University of Oxford, UK

Accepted Pre-Proof: SLEEP as a commentary

Institution work performed: Sheffield Hallam University, Uk.
Number of tables: 0
Number of Figures: 1
Abstract word count: n/a
Manuscript word count: 828
References: 14

COI: No conflicts of interest declared in relation to this letter.
Financial support declaration: This letter was not supported by funding.

***Corresponding Author:** u.akram@shu.ac.uk Department of Psychology, Sociology and Politics, Sheffield Hallam University, Collegiate Crescent, Sheffield, South Yorkshire, S10 2BP, UK.

As an individual diagnosed with idiopathic hypersomnia (IH), I read the recently proposed suggestions for reclassifying the central disorders of hypersomnolence[1] with great interest. Here, I allude to the article by Fronczek and colleagues[1] entitled "To split or to lump? Classifying the central disorders of hypersomnolence". Whilst I concur with the merging of narcolepsy type-2 (NT2) and idiopathic hypersomnia without long sleep duration, long sleeping individuals with IH remain overlooked.

Hypersomnias of central origin are characterised by marked deficits in the ability to remain awake at an appropriate time though the diurnal period. More typically, these disorders are quantified by the pathological nature of excessive daytime sleepiness (EDS), an increased propensity to fall asleep.[2-3] Like narcolepsy, idiopathic hypersomnia is an equally debilitating condition that commonly presents alongside additional ailments which include: long and unrefreshing naps, longer nocturnal sleep, difficulties in awakening, excessive sleep inertia, and impaired neuropsychological function.[4-5] For example, IH patients frequently self-report: sleep inertia (82.5%), unrefreshing nocturnal sleep (97.5%) and naps (73.8%) and multiple alarm use (57.2%).[4-5]

Unfortunately, from a diagnostic perspective, confirmation of IH largely relies on the objective quantification of EDS using the Multiple Sleep Latency Test (MSLT). Whilst the MSLT remains a useful tool for the diagnosis of narcolepsy type-1 (NT1), poor test-retest reliability limits adequate determination of NT2 and IH.[6-10] For example, in NT2 subjects completing the MSLT at two time points, only 18% of individuals consistently indicated NT2 with the second test changing diagnosis to idiopathic hypersomnia in 26% of cases or a negative MSLT in 57%.[11] Moreover, mixed evidence regarding the observation of low mean sleep-onset latency (SOL) in idiopathic hypersomnia further questions the utility of the MSLT in this population.[10] However, limitations of the MSLT when assessing idiopathic hypersomnia may not be a failure of the test itself, but rather a lack of understanding the aetiology of the disorder and consensus regarding the agreed criteria. That said, where mean SOL outcomes of ≤8 are not observed using the MSLT, the precedingly recorded objective sleep data (either by polysomnography or actigraphy) may confirm a diagnosis of IH. More specifically, where ≥ 11 hours of sleep time per 24-h period occurs at least three times over the course of a week. Nevertheless, it's recommended that clinicians should use their own judgment when presented with all possible data, particularly the use of additional soft criteria among those failing to meet the ≤8 sleep-onset latency.[12,13] Whilst a repeat MSLT is recommended in borderline patients, the aforementioned issues may limit its utility. Therefore, it is vital for the next revision of diagnostic criterion of IH to formally re-examine the ≤8-minute mean MSLT sleep-onset latency cut-off, multiple testing and examination of additional symptoms (i.e. unrefreshing naps, sleep inertia, cognitive functional deficits, sleep paralysis and hallucinations) to aid diagnosis.

Based partially on these observations, Fronczek and colleagues[1] propose the NT2 classification to be replaced by 'Narcolepsy Spectrum Disorder'. Here, IH with short sleep duration and NT2 would become merged where positive screening would no longer rely on the frequency of sleep-onset REM periods (SOREMPs). Certainly, this reclassification would benefit those who would otherwise be diagnosed with IH. Most notably, access to pharmacological treatments that are currently restricted to those with NT1 or NT2 such as (e.g. sodium oxybate). However, the categorical term 'Narcolepsy Spectrum Disorder' appears somewhat juxtaposed considering the linear divergence in symptom presentation between NT1, NT2 and IH. If central disorders of hypersomnolence are rebranded, then why not simplify re-categorisation. Rather than 'Narcolepsy Type-1', Narcolepsy Spectrum Disorder', and 'Idiopathic Hypersomnia', in line with the spectral nature of hypersomnolence disorders, the case for 'Narcolepsy Type-1, Type-2, and Type-3' is perhaps more compelling

(see Figure 1). Indeed, long sleeping individuals with IH would fail to benefit from the initially prosed reclassification.[1] Renaming the proposed IH classification to 'Narcolepsy Type-3' (NT3), would come with a number of benefits. First, like myself, many people with IH experience great stigma and difficulties in explaining the nature of the disorder to others.[14] Idiopathic hypersomnia is not a term that the majority of the general public are familiar with. I consistently find myself explaining "it's like narcolepsy" before an individual comes to the realisation that IH is indeed a sleep disorder. Second, a classification of NT3 may serve to increase research in this area with greater inclusion of the IH (or NT3) population in comparative studies of narcolepsy subtypes. Finally, reclassification of IH to NT3 may provide region-specific benefits (e.g. reimbursement of treatment offered in the US) and consideration of pharmacological options which in some countries are currently limited to those with NT1 and NT2.

To summarise, I would like to thank Fronczek and colleagues[1] for their important work highlighting the need to reclassifying the central disorders of hypersomnolence. Whilst the proposed merging NT2 and IH without long sleep duration is certainly overdue, it is important to also consider the remaining IH group. A more simplified classification should be considered moving forward, one which would include IH in the narcolepsy spectrum.

References

1. Fronczek R, Arnulf I, Baumann CR, Maski K, Pizza F, Trotti LM. To split or to lump? Classifying the central disorders of hypersomnolence. Sleep. 2020 Mar 20.
2. Carskadon MA, Dement WC. Normal sleep: an overview. Principles and practice of sleep medicine. Philadelphia: WB Saunders. 1994:16-25.
3. Curcio G, Casagrande M, Bertini M. Sleepiness: evaluating and quantifying methods. International Journal of Psychophysiology. 2001 Jul 1;41:251-63.
4. Bagai K, Malow BA. A novel approach to treating morning sleep inertia in narcolepsy. Journal of Clinical Sleep Medicine. 2010 Feb 15;6:77-8.
5. Trotti LM, Ong J, Plante D, Powell D, Bliwise DL. 0616 Self-Reported Sleep Inertia in the Hypersomnia Foundation Registry. Sleep. 2018 Apr;41(suppl_1):A228-9.
6. Mignot E, Lin L, Finn L, Lopes C, Pluff K, Sundstrom ML, Young T. Correlates of sleep-onset REM periods during the Multiple Sleep Latency Test in community adults. Brain. 2006 Jun 1;129:1609-23.
7. Pizza F, Franceschini C, Peltola H, Vandi S, Finotti E, Ingravallo F, Nobili L, Bruni O, Lin L, Edwards MJ, Partinen M. Clinical and polysomnographic course of childhood narcolepsy with cataplexy. Brain. 2013 Dec 1;136:3787-95.
8. Sauter C, Danker-Hopfe H. Multiple Sleep Latency Test. Somnologie-Schlafforschung und Schlafmedizin. 2013 Mar 1;17:28-33.
9. Trotti LM, Staab BA, Rye DB. Test-retest reliability of the multiple sleep latency test in narcolepsy without cataplexy and idiopathic hypersomnia. Journal of Clinical Sleep Medicine. 2013 Aug 15;9:789-95.
10. Vernet C, Arnulf I. Idiopathic hypersomnia with and without long sleep time: a controlled series of 75 patients. Sleep. 2009 Jun 1;32:753-9.
11. Ruoff C, Pizza F, Trotti LM, Sonka K, Vandi S, Cheung J, Pinto S, Einen M, Simakajornboon N, Han F, Peppard P. The MSLT is repeatable in narcolepsy type 1 but not narcolepsy type 2: a retrospective patient study. Journal of Clinical Sleep Medicine. 2018 Jan 15;14:65-74.
12. Bassetti C, Aldrich MS. Idiopathic hypersomnia. A series of 42 patients. Brain: a journal of neurology. 1997 Aug 1;120:1423-35.
13. American Academy of Sleep Medicine (2014). The International Classification of Sleep Disorders (3rd ed.). Darien, IL : American Academy of Sleep Medicine.
14. Kapella MC, Berger BE, Vern BA, Vispute S, Prasad B, Carley DW. Health-related stigma as a determinant of functioning in young adults with narcolepsy. PloS one. 2015;10. e0122478.

Figures

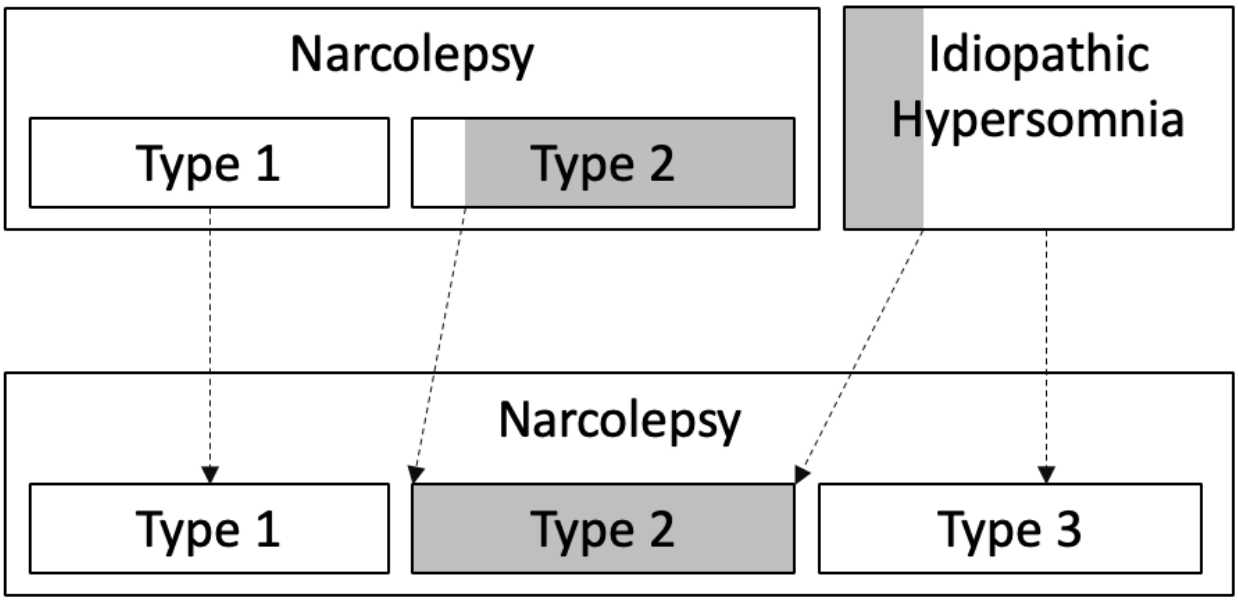

Figure 1. [Top row] Current International Classification of Sleep Disorders, third edition, versus proposed grouping. The ICSD-3 lists eight central disorders of hypersomnolence, including narcolepsy type 1, narcolepsy type 2, and idiopathic hypersomnia. **[Bottom Row]** Combination of those with narcolepsy type 2 and those with idiopathic hypersomnia without long sleep time into "Narcolepsy Type-2" as proposed by Fronczek and colleagues.[1] Proposed relabelling of Idiopathic hypersomnia with long sleep to "Narcolepsy Type-3".