



LETTER TO THE EDITOR

A patient's view on reclassifying idiopathic hypersomnia to narcolepsy type-3

Umair Akram^{1,2,*}

¹Department of Psychology, Sociology and Politics, Sheffield Hallam University, Sheffield, UK and ²Nuffield Department of Clinical Neurosciences, University of Oxford, Oxford, UK
Institution where work was performed: Sheffield Hallam University, UK.

*Corresponding author. Umair Akram, Department of Psychology, Sociology, and Politics, Sheffield Hallam University, Collegiate Crescent, Sheffield, South Yorkshire, S10 2BP, UK. Email: u.akram@shu.ac.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.” While 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 characterized by marked deficits in the ability to remain awake at an appropriate time through 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). While 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 etiology 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-hour 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]. While 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

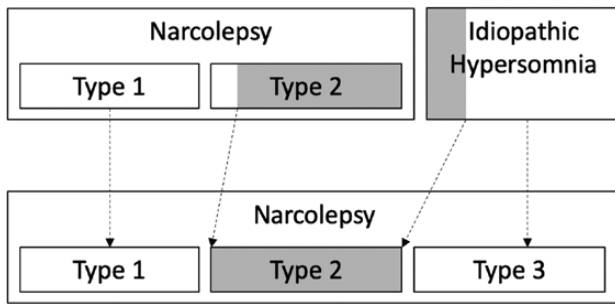


Figure 1. [Top row] Current International Classification of Sleep Disorders, third edition, versus proposed grouping. The ICSD-3 lists eight central disorders of hypersomnolence, including narcolepsy type 1, narcolepsy type 2, and idiopathic hypersomnia. [Bottom Row] Combination of those with narcolepsy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

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-categorization. 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 (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 realization 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 United States) and consideration of pharmacological options which in some countries are currently limited to those with NT1 and NT2.

To summarize, I would like to thank Fronczek and colleagues [1] for their important work highlighting the need to

reclassify the central disorders of hypersomnolence. While 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.

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