SHORT REPORT

A comparison of idiopathic hypersomnia and narcolepsy-cataplexy using self report measures and sleep diary data

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Abstract

Eighteen patients with idiopathic hypersomnia (IH) were compared with 50 patients with the narcoleptic syndrome of cataplexy and daytime sleepiness (NLS) using self report questionnaires and a diary of sleep/wake patterns. The IH group reported more consolidated nocturnal sleep, a lower propensity to nap, greater refreshment after naps, and a greater improvement in excessive daytime sleepiness since onset than the NLS group. In IH, the onset of excessive daytime sleepiness was predominantly associated with familial inheritance or a viral illness. Two variables—number of reported awakenings during nocturnal sleep and the reported change in sleepiness since onset—provided maximum discrimination between the IH and NLS groups. Confusional arousals, extended nap or nocturnal sleep, autonomic nervous system dysfunction, low ratings of medication effectiveness, or side effects of medication were not associated differentially with either IH or NLS.

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Idiopathic hypersomnia (IH) has been distinguished from narcolepsy on the basis of the presence of prolonged rather than short diurnal sleep periods and the absence of both cataplexy and episodes of rapid eye movement at the onset of sleep. Several studies have associated IH with longer nocturnal and nap sleep, less refreshing naps, more confusional arousals, and deeper sleep with fewer awakenings than in the narcoleptic syndrome (NLS).¹⁻⁴ Stimulant medication may be less effective and poorly tolerated by patients with IH.^{5,6}

Three clinical variants of IH have been described.^{1,7} (1) Familial history of excessive daytime sleepiness with symptoms suggesting autonomic nervous system instability. (2) Postinfective onset (commonly infectious mononucleosis). (3) No familial or postinfective history.

Published data comparing patients with IH and patients with NLS outside the sleep laboratory are scarce. Using self report questionnaires and sleep diary data our aim was to determine the extent of group differences and which variables best discriminated between IH and NLS.

Patients and methods

DIAGNOSTIC CRITERIA

Idiopathic hypersomnia

Patients were selected with a complaint of excessive daytime sleepiness without cataplexy and with no evidence of any medical, psychological, drug related, or respiratory disorder. All patients met diagnostic criteria ascertained from questionnaire responses: Epworth sleepiness scale score⁸ > 13; duration of excessive daytime sleepiness > five years, profile of mood states depression-dejection scale score within one SD of outpatient norms,⁹ no cataplexy,¹⁰ no clinical evidence of sleep apnoea or upper airway resistance syndrome, neck circumference < 16.5 inches, snoring amount and volume moderate level or less, no suggestion of chronic insomnia, no excessive alcohol intake, no other medical condition that may contribute to excessive daytime sleepiness, and no head injury within 12 months of onset of excessive daytime sleepiness. In addition, all patients with IH met minimal and additional criteria of the International Classification of Sleep Disorders (ICSD) for IH.⁴

NARCOLEPTIC SYNDROME

All patients with NLS met the minimal ICSD criteria⁴ for narcolepsy and had unequivocal cataplexy as established through both clinical interview and their score on the postural atonia rating scale.¹⁰

RESPONSE RATE AND PATIENTS

Idiopathic hypersomnia

The questionnaires (see later) were circulated to 209 patients who had attended the sleep clinic at the Maudsley Hospital, London over a period of 10 years and whose primary diagnosis was hypersomnia of unknown origin. Sixty three returns were obtained and 42 patients returned evaluable questionnaires. Twenty four of these were excluded as their responses were outside the defined criteria.

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Table 1 Mean (SD) ratings by patients with IH and patients with NLS (n = 50)

	IH (n = 18)	NLS (n = 50)
Confusion on awakening		
1-5: never-always	2.7 (1.1)	2.9 (0.9)
Confusion from night sleep		
1-5: never-always	2.1 (0.9)	2.3 (1.0)
Refreshed from naps		
1-5: never-always	2.5 (1.2)	3.2 (1.1)*
Refreshed in morning		
1-5: never-always	2.8 (1.2)	2.9 (1.2)
Sleepiness five years ago		
1-5: lot worse-lot better	3.4 (1.2)	2.9 (1.1)
Sleepiness at onset		
1-5: lot worse-lot better	3.5 (1.6)	2.7 (1.7)*
Medication effectiveness		
1-5: not at all-extremely	3.2 (1.1)	3.8 (1.1)
Medication side effects		
1-5: not at all-extreme	2.5 (1.6)	2.3 (1.3)
Arousal threshold during sleep		
0-100: easy-hard to wake	60.0 (29.5)	53.2 (30.1)
Epworth sleepiness score		
0-24: no propensity-very high	18.7 (3.1)	20.3 (2.7)*
Estimated TV nap duration (minutes)	50.9 (67.8)	31.0 (44.3)
Estimated afternoon nap duration (minutes)	75.8 (54.5)	65.4 (45.4)
Body weight (pounds)	156.8 (28.6)	177.8 (32.0)*
Insomnia score		
0-100: like "a log"-severe	37.6 (38.9)	47.7 (30.4)

*P < 0.05; t test. 1-5, 0-100, 0-24 indicate intervals on response scale.

The final IH sample (eight male, 10 female) had an average age of 45.61 (SD 17.06, range 18-70) years.

Narcoleptic syndrome

The questionnaires were circulated to 132 patients with NLS from the same clinic who had previously participated in a questionnaire study. Fifty eight questionnaires were returned complete and 50 patients aged 70 or less were included to obtain an age matched sample. The NLS sample (24 male, 26 female) had a mean age of 52.18 (SD 15.12, range 15-70) years.

QUESTIONNAIRES

The questionnaires consisted of three parts. (1) A five page questionnaire required responses on a five point scale (100 mm line) or a yes/no answer. (2) The sleep diary about sleep, medication, and alcohol intake completed for three consecutive days. Stimulant medication was to be avoided as much as pos-

sible. (3) Profile of mood states⁸ checklist to be completed after breakfast.

DATA ANALYSIS

Student's *t* tests, χ^2 analyses, and logistical discriminant analysis were used as appropriate. Sleep diaries that indicated stimulant use were excluded and the sleepest day was chosen for analysis.

Results

EXCESSIVE DAYTIME SLEEPINESS ONSET, DURATION, AND SEVERITY

The IH and NLS groups were similar in terms of reported duration of excessive daytime sleepiness (23.4 and 29 years) and age of onset of excessive daytime sleepiness (22.3 and 23.1 years). The propensity to fall asleep (Epworth) was slightly lower in the IH group than the NLS group ($t(66) = 2.1$, $P = 0.04$). Patients with IH reported a slight improvement in severity of excessive daytime sleepiness after onset, whereas patients with NLS reported a slight deterioration ($t(63) = 2.4$, $P = 0.017$). Table 1 shows all questionnaire mean values.

Those reporting a first degree relative with excessive daytime sleepiness were in the minority (0.38 IH; 0.24 NLS). No familial member with sleep paralysis or NLS was reported by the IH group, but such members were present in the NLS group (0.25 and 0.10 respectively). Glandular fever (or an illness of similar symptoms) in the six month period to onset of excessive daytime sleepiness was reported more often by the IH group (0.31) than the NLS group (0.16). There was no overlap between the six patients with IH who reported such an illness before the onset of excessive daytime sleepiness and the six patients with IH reporting familial excessive daytime sleepiness.

DAYTIME SLEEP-WAKE BEHAVIOUR

Patients with IH were less likely to feel refreshed after a nap than patients with NLS ($t(63) = 2.22$, $P = 0.03$). Confusion on waking from naps or night sleep did not differ between groups; nor did the extent of refreshment felt in the morning. Half of each group said they were most alert in the morning (0.56 IH; 0.48 NLS). The average duration of each nap was similar in both groups (see tables 1 and 2).

NOCTURNAL SLEEP-WAKE BEHAVIOUR

The sleep diary (table 2) showed that nocturnal sleep in the IH group was characterised by significantly fewer awakenings ($t(40) = 3.7$, $P = 0.001$) and reduced wake duration ($t(40) = 2.2$, $P = 0.015$) than in the NLS group. Other nocturnal sleep variables did not differ between groups.

Most parasomnias were noted in similar proportions in both groups. These included muscle jerking during sleep (0.56; 0.50), sleep talking (0.43; 0.58), and sleep walking (0.13; 0.14). However, NLS was associated with more night terrors (0.52; 0.31) and nightmares (0.64; 0.19) than IH, with nightmares showing a significant group difference ($\chi^2 = 7.49$, $df =$

Table 2 Mean (SD) and range for sleep diary variables comparing patients with IH and patients with NLS (all stimulant free)

	IH (n = 13)	NLS (n = 23)
Number of naps	2.4 (1.2) 0-4	3.4 (1.7) 0-8
Total nap time (min)	70.7 (61.8) 0-240	113.1 (91.3) 30-420
Average nap duration (min)	36.6 (32.7) 5-120	37.3 (26.6) 10-105
Nocturnal sleep latency (min)	15.2 (12.3) 1-45	11.8 (13.1) 1-40
No of nocturnal awakenings	1.3 (1.22) 0-3	3.3 (1.8)** 1-8
Duration nocturnal wake (min)	7.7 (10.7) 0-40	34.4 (49.7)* 2-240
Nocturnal time in bed (min)	521.0 (138.9) 395-810	491.7 (86.9) 300-780
Nocturnal total sleep time (min)	498.2 (139.5) 340-780	447.6 (94.4) 225-900
Minutes asleep over 24 hours	568.5 (183.8) 400-1020	567.5 (128.45) 359-877

*P < 0.05; **P < 0.005; t test.

1, $P = 0.006$). More patients with NLS (0.32) reported breath holding during sleep than patients with IH (0.06).

STIMULANT DRUG AND ALCOHOL INTAKE

The groups' ratings of effectiveness of stimulant medication and medication side effects did not differ. Caffeine and alcohol intake did not differ between groups.

AUTONOMIC NERVOUS SYSTEM DYSFUNCTION

About equivalent proportions of patients with IH and patients with NLS reported moderate to severe headaches (0.38; 0.52), migraines (0.00; 0.12), very cold hands and feet (0.3; 0.44), and fainting spells (0.13; 0.08). Such dysfunctional responses were not differentially clustered in the six patients with IH reporting familial excessive daytime sleepiness. These symptoms were not related to stimulant use.

BODY WEIGHT

Mean body weight was less in the IH group than the NLS group ($t(60) = 2.27$, $P = 0.027$). Use of antiepileptic drugs was not a significant factor affecting body weight within the NLS group.

MOOD

The profile of mood states factors of tension, depression, anger, vigour, fatigue, and confusion showed no differences between the groups.

DISCRIMINANT ANALYSIS

Relevant interval and nominal variables were submitted to a stepwise logistical regression. Variables that best discriminated groups were the number of awakenings during nocturnal sleep and reported change in sleepiness level since excessive daytime sleepiness onset. These two items correctly classified 86% of patients. The discriminant function was developed using 75% of the sample and tested on the remaining 25% (assuming a 1:5 probability of IH versus NLS occurring in the population³). This cross validation method upheld the validity of the initial discrimination with 90% of the test group being correctly classified.

Discussion

The day-night sleep pattern was different between patients with IH and those with NLS. Propensity to daytime sleepiness, nap frequency, and refreshment after naps was lower in patients with IH than those with NLS. Nocturnal awakenings were less frequent in the IH group than in the NLS group. Patients with IH in this study reported improvement in excessive daytime sleepiness after the initial onset. This contrasts with earlier reports of symptom stability over decades.¹ The main

differences in sleep-wake patterns found between the patients with IH and patients with NLS were in the number of nocturnal awakenings and in the reported sleepiness change since excessive daytime sleepiness onset. These two variables correctly discriminated 86% of the sample.

As in previous studies of IH, three distinct subgroups were identified: familial history, postinfective onset, and a group reporting neither. By contrast with previous reports,^{6,7} symptoms of autonomic nervous system dysfunction were no more prevalent in IH than NLS and were not clustered into any IH subgroup.

The differential diagnosis between IH and NLS is sometimes difficult, especially when there is a long period between the onset of excessive daytime sleepiness and cataplexy. This study suggests that there is little value in focusing on issues such as confusional arousals, perceived depth of sleep, or longer duration of nocturnal or nap sleep. The most important discriminators of the two sleep disorders as disclosed by this study will typically only become evident over time. The frequent nocturnal awakenings typical of NLS often only appear several years after onset of excessive daytime sleepiness^{11,12} while the rating of change since onset requires a period for retrospection. It is unlikely that IH will be a single diagnostic entity. Those patients without a definite familial or postinfective onset may need detailed clinical evaluations to exclude upper airway resistance syndromes, atypical depression, or drug abuse, all conditions which sometimes mimic IH.

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- 1 Roth B. *Narcolepsy and hypersomnia*. Basel: Karger, 1980.
- 2 Parkes JD. Idiopathic hypersomnia. In: *Sleep and its disorders*. London: Saunders, 1985:267-73.
- 3 Chanellas F, Nougier J, Tafti M, Billiard M. Nighttime and daytime sleep in idiopathic central nervous system hypersomnolent subjects in comparison with narcoleptic subjects. *Sleep Res* 1989;18:211.
- 4 Billiard M. Other hypersomnias. In: Thorpy MJ, ed. *Handbook of sleep disorders*. New York: Marcel Dekker, 1990:353-71.
- 5 American Sleep Disorders Association Diagnostic Classification Steering Committee. *The international classification of sleep disorders: diagnostic and reading manual*. Rochester: ASDA, 1990.
- 6 Chokroverty S. Sleep, breathing and neurological disorders. In: Chokroverty S, ed. *Sleep disorders medicine*. London: Butterworth-Heinemann, 1994:319-21.
- 7 Guilleminault C. Disorders of excessive sleepiness. *Ann Clin Res* 1985;17:209-19.
- 8 Johns MW. A new method for measuring daytime sleepiness: the Epworth sleepiness scale. *Sleep* 1991;14:540-5.
- 9 McNair DM, Lorr M, Droppleman LF. *Profile of mood states manual*. San Diego, CA: Educational and Industrial Testing Service, 1992.
- 10 Parkes JD, Dahlitz MJ, Clift S, Chen S. What is cataplexy? *J Sleep Res* 1994; 3 (suppl 1):192.
- 11 Broughton RJ. Narcolepsy. In: Thorpy MJ, ed. *Handbook of sleep disorders*. New York: Marcel Dekker, 1990:197-216.
- 12 Billiard M, Bessett A, Cadilhac J. The clinical and polygraphic development of narcolepsy. In: Guilleminault C, Lugaresi E, eds. *Sleep/wake disorders: natural history, epidemiology, and long-term evolution*. New York: Raven Press, 1983:171-85.