ELSEVIER

Sleep Medicine 6 (2005) 359–362

SLEEP MEDICINE

www.elsevier.com/locate/sleep

Brief communication

Continuous positive airway pressure for sleep-related breathing disorders in multiple system atrophy: long-term acceptance

Imad Ghorayeb[a,*], Farid Yekhlef[b], Bernard Bioulac[a], François Tison[b]

[a]*Service d'Explorations Fonctionnelles du Système Nerveux, Hôpital Pellegrin, Place Amélie Raba-Léon, 33076 Bordeaux Cedex, France*
[b]*Service de Neurologie, CHU de Bordeaux, Bordeaux, France*

Received 4 August 2004; received in revised form 26 August 2004; accepted 12 October 2004

Abstract

Background and purpose: To assess the long-term acceptance of non-invasive nasal continuous positive air pressure (CPAP) in multiple system atrophy (MSA) patients with polysomnographic (PSG)-confirmed sleep apneas and/or nocturnal stridor.

Patients and methods: Sleep-related breathing disorders were investigated by PSG in 22 MSA patients in whom stridor and sleep-related respiratory disturbances were clinically suspected. Patients in whom the first PSG disclosed either a sleep apnea/hypopnea index (AHI) ≥ 10 or stridor with or without apneas underwent a second PSG for CPAP titration.

Results: Three patients presented with an obstructive sleep apnea syndrome without stridor, whereas 15 patients presented stridor occurring alone or accompanied by apneas. Twelve patients pursued CPAP. Two severely disabled patients died a few days after CPAP initiation, and five discontinued CPAP because of discomfort. One patient died after 17 months of follow-up. Since the onset of CPAP, the four remaining patients reported more efficacious sleep and improved daytime alertness. These patients had significantly less severe disease at the time of CPAP initiation. Age, disease duration, the presence of sleep complaints, excessive daytime somnolence (EDS) and AHI did not account for CPAP compliance.

Conclusion: The severity of motor impairment at the initiation of treatment appears to be the most significant limiting factor for CPAP long-term acceptance.

Keywords: Multiple system atrophy; Stridor; Sleep apnea; Continuous positive airway pressure

1. Introduction

In multiple system atrophy (MSA), the diffuse neurodegenerative process results in a more aggressive course, dopaminergic drug resistance, and more frequent sleep disorders and excessive daytime somnolence (EDS) compared to patients with Parkinson's disease [1]. Often associated with sleep-related breathing disorders, nocturnal stridor appears to be of poor prognosis associated with increased risk of sudden death during sleep [2]. Tracheostomy and vocal cord surgery are the invasive options usually proposed to suppress stridor, and recently botulinum toxin has also been proposed [3]. However, only non-invasive continuous positive air pressure (CPAP) has been shown to be effective in suppressing both stridor and sleep apneas [4]. We investigated sleep-related breathing disorders by means of audio–video monitored polysomnography (PSG) in 22 MSA patients in whom nocturnal stridor and sleep-related respiratory disturbances were clinically suspected, and studied the effect and long-term acceptance of CPAP in patients with PSG-confirmed sleep apneas and/or nocturnal stridor.

2. Patients and methods

The University of Bordeaux 2 ethical committee approved the study. A total of 22 patients fulfilling the MSA clinical diagnostic criteria for 'possible' or 'probable' MSA [5] were

* Corresponding author. Tel.: +33 5 56 79 55 13; fax: +33 5 56 79 61 09.
E-mail address: imad.ghorayeb@umr5543.u-bordeaux2.fr (I. Ghorayeb).

1389-9457/$ - see front matter © 2004 Elsevier B.V. All rights reserved.
doi:10.1016/j.sleep.2004.10.002

selected for sleep studies. Recruitment was based on the patients, bed partners, or caregivers reports of loud snoring, nocturnal stridor, defined as a strained high-pitched sound, and/or observation of apneic events during sleep. Six patients showed diurnal stridor. Laryngoscopy was not performed systematically as nocturnal stridor may be present in patients with normal vocal cord mobility on laryngoscopy during wakefulness [2,6]. All patients underwent standard PSG with audiovisual recording, which allowed reliable detection of stridor. Demographic data, disease duration and severity (Hoehn and Yahr scale), the presence of sleep complaints and EDS (Epworth Sleepiness Scale) were collected.

A second PSG for CPAP titration was performed in patients in whom the first PSG evaluation disclosed either an apnea/hypopnea index (AHI) ≥ 10, or stridor with or without apneas. Patients and family members were followed up at month 1 and then every 3 months. The mean age, disease duration and severity, the mean AHI, the presence of sleep complaints and EDS were compared between patients with satisfactory compliance to CPAP (≥ 3 h of nightly use) and those who could not tolerate this treatment using Student's *t*-test.

3. Results

Mean age was 65.55 ± 8.38 years; 12 were men and 10 were women. Twenty-seven percent were MSA-C, 73% were MSA-P, mean disease duration was 4.9 ± 2.4 years, and mean disease severity was 4 ± 1.1.

All patients' baseline PSG showed sleep fragmentation, reduction of total sleep time (mean $= 329 \pm 134$), and reduction of sleep efficiency (mean $= 63 \pm 18\%$). Periodic limb movements were seen in 73% of the patients and REM sleep behaviour disorder in 64%.

Of the total group, three patients presented with a typical obstructive sleep apnea syndrome (OSAS) (mean AHI = 37) without stridor, whereas 15 presented stridor occurring alone (AHI < 10) or accompanied by apneas (AHI ≥ 10). Seven out of these 15 patients with stridor presented predominantly obstructive apneas whereas apneas of the central type were predominantly seen in one patient and mixed apneas in another (Table 1).

Of these 18 patients, 13 agreed to initiate CPAP, including three with isolated stridor. At the end of the titration night, and although there was no immediate subjective improvement, stridor and apneic events had totally abated in all patients. The mean CPAP pressure was 6.9 ± 0.6 cmH_2O (range, 6–8). All of these 13 patients agreed to pursue nasal CPAP at home; their mean AHI was 23. The five remaining patients declined further follow-up.

One patient died before home initiation of CPAP, two died while sleeping 2–3 days after CPAP initiation, and four patients discontinued CPAP usage after the first week of treatment initiation because of discomfort. Of these four patients, two had isolated stridor and two had a severe AHI mainly of the obstructive type (mean AHI = 55), but only one patient reported sleep complaints and EDS at inclusion (Table 2). To date, three of these four patients have died.

Of the six remaining patients, one discontinued CPAP after 10 months of follow-up because of discomfort, although he initially reported subjective sleep benefit and improved quality of life under treatment. Another patient died after 17 months of follow-up because of pneumonia. The four remaining patients and their spouses reported better sleep and improved vigilance. The best compliance was observed in two patients with a mild form of MSA-C on the first evaluation PSG. The mean duration of follow-up for these four remaining patients was 24.75 months (range 12–37 months) and the mean nightly use was 4 h 42 (Table 2).

Patients with good compliance with CPAP had a significantly less severe disease at the time of CPAP initiation (mean Hoehn and Yahr stage $= 2.7 \pm 0.96$ vs. 4.8 ± 0.45, $P = 0.02$). Age, disease duration, AHI, the presence of sleep complaints and EDS did not differ significantly between both groups.

4. Discussion

We studied the long-term acceptability of CPAP treatment in a large cohort of patients with MSA presenting either isolated obstructive apneas (AHI ≥ 10), or isolated nocturnal stridor, or both. As results come from only 12 out of the initial 22 patients the validity of our study is reduced. On the other hand, studies like this in a relatively rare disease like MSA will inevitably suffer from the inclusion of low numbers of patients.

Eighty-two percent of our patients showed a large variety of sleep-related breathing disorders. Among these, obstructive apneas appear to be the most common [7,8] even in the absence of stridor. Sleep-related breathing disorders can cause significant morbidity and may be associated with increased mortality [9]. MSA patients may be at additional risk because severe bradykinesia, resulting in a supine

Table 1
Number (percentage) of patients with or without stridor presenting or not a predominant pattern of apneic events on polysomnography (only patients with an apnea/hypopnea index ≥ 10 are shown)

Number of patients (percentage)	Without apnea	With apnea predominantly of the obstructive type	With apnea predominantly of the central type	With apnea predominantly of the mixed type
With stridor	6 (27%)	7 (32%)	1 (4.5%)	1 (4.5%)
Without stridor	4 (18%)	3 (14%)	0	0