Use of Hypoglossal Nerve Stimulation for Obstructive Sleep Apnea in Adults With Intellectual and Developmental Disabilities

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ABSTRACT

Introduction: The objective of this study was to assess the safety, efficacy, and patient satisfaction of hypoglossal nerve stimulation (HNS) therapy as an alternative to continuous positive airway pressure therapy for obstructive sleep apnea in adults with intellectual and developmental disabilities (IDDs), including trisomy 21 and cerebral palsy.

Methods: We conducted an observational study of 4 adults with IDDs and moderate to severe obstructive sleep apnea who underwent implantation of an HNS device. Patient demographic characteristics, device usage, obstructive sleep apnea and daytime sleepiness outcomes, and patient satisfaction scores were retrospectively reviewed and analyzed.

Results: The mean adherence rate was 92.3%. The mean [SD] pretreatment apnea-hypopnea index was 39.0 [16.7] events per hour, which decreased to 8.5 [4.7] event per hour after treatment, representing a mean [SD] reduction of 77.5% [12.9%]. Epworth Sleepiness Scale scores decreased in 2 patients, were unchanged in 1 patient, and increased in 1 patient. Patient-reported satisfaction was high, with mean [SD] scores of 5.0 [0.0] for comfort, ease of deactivation, and effectiveness and 4.5 [0.6] for ease of activation.

Conclusions: Treatment of obstructive sleep apnea with HNS was associated with decreased apnea-hypopnea index values and was well tolerated among adults with IDDs, suggesting that it may be a viable alternative to continuous positive airway pressure therapy for patients who cannot tolerate conventional treatment. However, the variable Epworth Sleepiness Scale findings and small sample highlight the need for larger studies with long-term follow-up to further evaluate the safety and effectiveness of HNS in this population.

INTRODUCTION

Obstructive sleep apnea (OSA) is a prevalent sleep disorder that affects individuals of all ages. This condition is marked by repeated episodes in which the upper airway is either completely or partially blocked during sleep. Notably, the incidence of OSA is higher among patients with intellectual and developmental disabilities (IDDs), such as trisomy 21 and cerebral palsy, than in the general population. The prevalence of OSA ranges from 30% to 80% among individuals with trisomy 21 and as high as 54% among those with cerebral palsy.[1-5] OSA symptoms are generally more severe in patients with IDDs, and complications occur more frequently, negatively affecting quality of life.[2,6]

For many patients with IDDs, continuous positive airway pressure (CPAP) therapy is the first-line recommended treatment for moderate to severe OSA. Although up to 80% of patients who initiate CPAP therapy tolerate it well,[7] studies including patients with IDDs are limited, and a considerable proportion cannot adhere to CPAP therapy.[8] Reasons for poor adherence include anxiety, claustrophobia, skin irritation, and mask-related discomfort. Additionally, sensory sensitivities, cognitive impairments, and anatomic differences may complicate mask fitting.

Hypoglossal nerve stimulation (HNS) therapy is a novel alternative to CPAP for the treatment of OSA and has demonstrated promising results in the general population,[9] although it has not been extensively studied in adults with IDDs. With HNS therapy,

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a surgically implanted device engages the hypoglossal nerve during sleep. Most HNS devices employ a dual-component system consisting of a sensing electrode positioned between the intercostal muscles and a stimulation lead wrapped around the anterior branches of the hypoglossal nerve. This configuration allows for the delivery of electrical impulses to the muscles responsible for tongue protrusion during inspiration, which effectively mitigates upper airway obstruction.

Two previously published case series highlighted the efficacy of HNS for adolescents with trisomy 21 who had severe OSA despite adenotonsillectomy and were intolerant of positive airway pressure therapy.[10,11] HNS treatment was associated with an acceptable adverse event profile and significant improvements in both therapy response and quality of life for 42 adolescents.[10] Additionally, HNS was well received and effective.[11] Therefore, HNS may represent a promising treatment option for patients with IDDs who cannot tolerate CPAP therapy.

Despite recent advancements in HNS therapy, research regarding its safety and efficacy in adults with IDDs remains limited. Therefore, we assessed HNS treatment in adults with IDDs and moderate to severe OSA who could not tolerate CPAP therapy. Specifically, we evaluated the safety and efficacy of HNS, as well as the device's operational parameters, patient usage, adherence, and satisfaction. These findings may clarify the role of HNS as a treatment option for adults with IDDs and OSA.

METHODS

Ethical Considerations and Approval

This study was conducted in accordance with the ethical principles of the Declaration of Helsinki and was deemed exempt by our institutional review board. The retrospective nature of the study ensured that no interference with patient care occurred, and all data were anonymized to protect patient confidentiality. All patients or their surrogate decision-makers provided authorization for the research use of their health records.

Study Design and Participants

We retrospectively reviewed the electronic health records of adult patients with IDDs who underwent implantation of an HNS device (Inspire II Implantable Pulse Generator, Model 3024; Inspire Medical Systems, Inc) within our regional health system in the Midwestern United States from November 2020 through January 2024. Inclusion criteria included adults (aged ≥18 years) with an IDD who underwent HNS device implantation for management of moderate to severe OSA, as determined by polysomnography or equivalent sleep study. Patients were identified using the following *Current Procedural Terminology* codes: 64582, 64568, and 0466T. IDDs were identified using the following search terms: "cognitive dysfunction," "Down syndrome," "cerebral palsy," "intellectual disability," "developmental disorder, unspecified," and "chromosomal deletion." Exclusion criteria included age younger than 18 years, absence of an IDD, mild OSA or another sleep disorder unrelated to OSA, or no HNS device implantation procedure.

OSA severity was quantified with apnea-hypopnea index (AHI), measured as events per hour and categorized as mild (5 to <15), moderate (15 to <30), or severe (≥30). Daytime sleepiness was assessed using the Epworth Sleepiness Scale (ESS), with scores of 11 or higher indicating excessive daytime sleepiness. ESS scores were used to assess symptom burden but were not used to determine treatment eligibility. HNS was pursued only after shared decision-making, confirmation of eligibility according to established criteria, and discussion of alternative therapies, including oral appliance therapy, which were deemed unsuitable because of disease severity or tolerability limitations.

Data Collection

Data were manually abstracted from health records accessed through the pulmonary, critical care, and sleep medicine departments. Abstracted variables included demographic characteristics (ie, age, sex, and body mass index [BMI] values), adverse events, and follow-up outcomes. Outcome measures included quantitative and qualitative assessments of HNS efficacy and patient satisfaction, device usage data (pause duration and voltage requirements), and immediate and long-term adverse events associated with the device. ESS scores and patient satisfaction data were collected via patient self-report and/or family member proxy report, depending on cognitive status.

Data Analysis

To evaluate HNS efficacy, we analyzed AHI values, ESS scores, and device usage data. AHI and ESS scores were obtained from assessments performed within 60 days before HNS device implantation (pretreatment). The device was activated 30 days after implantation. Posttreatment ESS scores were recorded 90 days after implantation, and posttreatment AHI values were recorded 120 days after implantation.

Patient satisfaction was assessed using a 4-item questionnaire administered 4 months after implantation. Responses were scored on a 5-point Likert scale, with 1 indicating poor and 5 indicating excellent.

All data analyses were descriptive. Data are presented as counts and percentages or as mean [SD] and median (range). The percentage decrease in AHI values was calculated as the difference between pretreatment and posttreatment values divided by the pretreatment value. Adherence was calculated as the percentage of nights during a 30-day period in which the HNS device was used for more than 4 hours

RESULTS

We identified 4 adult patients with IDDs and moderate to severe OSA who underwent HNS implantation at our regional hospital during the study period; 3 (75%) were men and 1 (25%) was a

woman. One patient had trisomy 21 (Down syndrome), 1 had Asperger syndrome, 1 had cerebral palsy, and 1 had Koolen-de Vries syndrome. The mean [SD] age was 44.8 [5.9] years, with a median (range) of 44 (39-52) years. The mean [SD] BMI value was 32.0 [1.5], with a median (range) of 32.2 (30.1-33.5).

Usage and Adherence

The patient with Koolen-de Vries syndrome and the patient with Asperger syndrome were able to turn on and use the HNS device independently, whereas the patients with Down syndrome and cerebral palsy required assistance from family members. All patients used the device consistently each night, with a mean [SD] nightly usage of 6.6 [0.9] hours. The mean adherence rate was 92.3%, and the median adherence rate was 90.0%.

Operational Parameters

The implanted HNS devices required relatively low stimulation voltages and had minimal pause durations (Table 1). The mean [SD] voltage requirement was 2.2 [0.7] V, with a median (range) of 1.9 (1.8-3.3) V. The mean [SD] pause duration was 0.25 [0.50] second, with a median (range) of 0.00 (0.00-1.00) second.

AHI Outcomes

The mean [SD] pretreatment AHI was 39.0 [16.7] events per hour, whereas the mean [SD] posttreatment AHI was 8.5 [4.7] events per hour (Table 2). This constituted a mean [SD] decrease of 77.5% [12.9%]. The median (range) pretreatment AHI was 31.5 (29.0-64.0) events per hour, compared with 8.5 (4.0-13.0) events per hour after treatment.

ESS Scores

For the patient with Koolen-de Vries syndrome, ESS data were primarily self-reported and corroborated by her parents. The patient with Asperger syndrome completed the ESS independently. For the patients with Down syndrome and cerebral palsy, ESS data were obtained through family-member proxy reports.

The mean [SD] ESS score decreased from 10.5 [7.3] before treatment to 5.8 [4.9] after treatment (Table 3). Median (range) scores were 12.5 (0.0-17.0) before treatment and 5.0 (1.0-12.0) after treatment, highlighting substantial variability among the patients. Two patients (1 with Koolen-de Vries syndrome and 1 with cerebral palsy) had decreased ESS scores following HNS device implantation, whereas the score was unchanged in the patient with trisomy 21 and increased in the patient with Asperger syndrome.

Patient-Reported Satisfaction

For the patient with Koolen-de Vries syndrome, satisfaction data were reported primarily by her parents. The patient with Asperger syndrome provided satisfaction data independently. For the patients with Down syndrome and cerebral palsy, satisfaction data were obtained through family-member proxy reports.

Table 1. Operational Parameters of the Implanted HNS Device

Patient	Mean Pause Duration, s	Voltage Requirement, V
1	0.00	1.9
2	0.00	1.8
3	0.00	1.8
4	1.00	3.3
Mean [SD]	0.25 [0.50]	2.2 [0.7]
Median (range)	0.00 (0.00–1.00)	1.9 (1.8-3.3)

Abbreviation: HNS, hypoglossal nerve stimulation; s, second; V, volts.

Table 2. AHI Values Before and After HNS Therapy

		AHI, events/h		
Patient	IDD	Pretreatment	Posttreatment	Decrease, %
1	Trisomy 21	64.0	13.0	79.7
2	Asperger syndrome	31.0	4.0	87.1
3	Cerebral palsy	32.0	5.0	84.4
4	Koolen-de Vries syndrome	29.0	12.0	58.6
Mean [SD]		39.0 [16.7]	8.5 [4.7]	77.5 [12.9]
Median (range)		31.5 (29.0–64.0)	8.5 (4.0–13.0)	82.1 (58.6–87.1)

Abbreviations: AHI, apnea-hypopnea index; HNS, hypoglossal nerve stimulation; h, hour; IDD, intellectual and developmental disability.

Table 3. ESS Scores Before and After HNS Therapy

		ESS score	
Patient	IDD	Pretreatment	Posttreatment
1	Trisomy 21	12.0	12.0
2	Asperger syndrome	0.0	1.0
3	Cerebral palsy	17.0	7.0
4	Koolen-de Vries syndrome	13.0	3.0
Mean [SD]		10.5 [7.3]	5.8 [4.9]
Median (range)		12.5 (0.0–17.0)	5.0 (1.0–12.0)

Abbreviations: ESS, Epworth Sleepiness Scale; HNS, hypoglossal nerve stimulation; IDD, intellectual and developmental disability.

No short-term or long-term adverse events, including infection, pain, hematoma or seroma, pneumothorax, device migration and lead issues, stimulation-related discomfort, neuropraxia, or transient tongue paresis, were noted.

All patients reported a high level of satisfaction with the HNS device. Mean [SD] scores were 5.0 [0.0], the maximum possible score, for comfort, ease of deactivation, and effectiveness compared with CPAP therapy (Table 4). The only questionnaire item that did not receive the maximum score from all patients was ease of activation, which had a mean [SD] score of 4.5 [0.6].

DISCUSSION

In this small observational study, we investigated the use of HNS for the treatment of moderate to severe OSA among adults with IDDs. Our findings underscore the potential of HNS as an effec-

tive and patient-preferred alternative to CPAP therapy, particularly for individuals with IDDs, who have a higher prevalence and severity of OSA and often experience challenges using conventional OSA therapies.

Table 4. Posttreatment Patient Satisfaction Scores[a]

	Patient				
Question	1	2	3	4	Mean [SD]
How comfortable is the device?	5.0	5.0	5.0	5.0	5.0 [0.0]
How easy is it to activate the device?	5.0	4.0	5.0	4.0	4.5 [0.6]
How easy is it to deactivate the device?	5.0	5.0	5.0	5.0	5.0 [0.0]
How effective is the device compared to CPAP?	5.0	5.0	5.0	5.0	5.0 [0.0]

Abbreviation: CPAP, continuous positive airway pressure.

[a]Responses were reported with a 5-point Likert scale in which a score of 1.0 represents poor and a score of 5.0 represents excellent.

The considerable reduction in AHI values is consistent with previous evidence supporting the effectiveness of HNS in mitigating upper airway obstruction during sleep, not only in the general population but also in patients with trisomy 21 and potentially other IDDs.[12-14] Importantly, the high patient satisfaction scores relating to comfort, ease of use, and perceived effectiveness highlight the potential of HNS to improve quality of life in this patient population.

The first reported case of HNS device implantation in an adolescent involved a 14-year-old patient with Down syndrome and a baseline AHI of 49 events per hour.[12] After device implantation, the AHI decreased to 3.4 events per hour. The patient's voltage requirement was 1.4 V, similar to the voltage requirements observed in our cohort (mean, 2.2 V; median, 1.9 V).

In a large cohort of adults, adherence to HNS was generally high, with mean nightly use of approximately 5 to 6 hours at 12 months after implantation.[15] Early acclimatization is also typically favorable in neurotypical adults, with an objective early-use study reporting average nightly use exceeding 7 hours during the first month after activation.[16] Factors that may have reduced effective nightly use included stimulation-related discomfort, particularly during titration to higher therapeutic amplitudes, sleep disruption or arousals, difficulty acclimating to therapy, insomnia symptoms, and perceived lack of benefit when residual sleep-disordered breathing persisted.

Few studies have evaluated HNS efficacy in patients with Down syndrome. Although these patients often require substantial caregiver involvement, adherence can be high when patients and families are well supported. In a multicenter trial including adolescents with Down syndrome, mean [SD] nightly HNS use was 9.0 [1.8] hours, and 95.2% of patients used the device for at least 4 hours per night on 70% or more of nights.[10] Similarly, in a small prospective trial including adults with Down syndrome, objective adherence exceeded 4 hours on nearly all nights shortly after HNS activation, with a median nightly use of more than 8 hours.[17] Those findings suggest that caregiver engagement may mitigate certain adherence barriers and underscore the importance of counseling regarding acclimatization, troubleshooting, and realistic expectations for incremental programming changes over time.

The therapeutic efficacy and tolerability of HNS are closely related to device programming parameters—particularly stimulation amplitude (voltage), which is adjusted to promote tongue protrusion and improve airway patency while minimizing arousals and discomfort. In clinical practice, higher amplitudes may be required to optimize control of obstructive events; however, higher settings may be less comfortable and contribute to interruptions in use during sleep and reduced adherence. Common early, typically transient adverse effects include tongue or oral discomfort and stimulation-related sensations. Among a cohort of adolescents with Down syndrome, temporary tongue or oral discomfort was the most frequently reported complication.[10] Although no published studies have demonstrated that higher HNS voltage settings directly contribute to tongue discomfort or pose challenges with tolerance and adherence, this remains a potential concern, particularly in patients with IDDs. Nevertheless, the minimal pause durations and low-voltage requirements observed in our cohort suggest that HNS was well tolerated and operationally efficient in this population.

ESS scores decreased after HNS device implantation in 2 patients. The patient with Koolen-de Vries syndrome had a substantial decrease, from 13 to 3, and the patient with cerebral palsy had a decrease from 17 to 7. However, the patient with trisomy 21 had no change in ESS score, and the patient with Asperger syndrome had a marginal increase from 0 to 1. These variable findings suggest that ESS may not be a fully reliable measure of daytime sleepiness in patients with IDDs because of differences in baseline characteristics and the subjective nature of self-reported or proxy-reported assessments. Additionally, the small sample size limited our ability to identify consistent changes. Therefore, these findings should be interpreted with caution. Future studies with larger cohorts and objective measures of daytime sleepiness and alertness are needed to better characterize the effects of HNS on daytime symptoms in patients with IDDs.

Although our findings are promising, several limitations warrant consideration. First, the retrospective design limits the generalizability of the results. Second, the absence of a control group and reliance on self-reported satisfaction measures may have introduced bias. Finally, the relatively short follow-up period precludes insights into long-term efficacy, adherence, and potential sequelae of HNS therapy. Despite these limitations, our findings provide valuable preliminary data regarding the use of HNS in a population with unique therapeutic challenges. Future research should aim for larger, randomized controlled trials with long-term fol-

low-up data to validate the efficacy and safety of HNS in adults with IDDs. Additional research examining the effects of HNS on other aspects of sleep quality, daily functioning, and the economic implications of adopting this technology in clinical practice may also be valuable.

CONCLUSIONS

The preliminary findings of this observational study suggest that HNS may be an effective and well-tolerated treatment alternative for moderate to severe OSA in adults with IDDs. High patient-reported satisfaction, favorable adherence, and improvements in clinical outcomes suggest that HNS may be a reasonable option for patients who cannot tolerate CPAP therapy. However, larger prospective studies with control groups and long-term follow-up are needed to confirm these findings and to establish HNS as a standard treatment for OSA in this population.

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