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SCIENTIFIC INVESTIGATIONS

Adherence to positive airway pressure therapy in patients with Down syndrome: assessing cloud-based monitoring data

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Study Objectives: Obstructive sleep apnea is common in Down syndrome (DS) with many patients prescribed positive airway pressure (PAP) therapy. This study evaluates PAP adherence and identifies factors influencing adherence.

Methods: Retrospective analysis of electronic health records and cloud-based PAP therapy data from patients with DS at Rady Children's Hospital, San Diego, CA. Cloud data were evaluated cross-sectionally at 30- and 90-night post-clinic visit and longitudinally in patients with ≥ 2 90-night data downloads. Outcomes included adherence (percentage of nights with ≥ 4 hours of use) and usage (percentage of nights with any PAP usage). The impact of demographic and PAP therapy factors (eg, mask leak) on these outcomes was also assessed.

Results: Forty-seven patients with DS with cloud-based PAP therapy data over a 90-night period and 46 over a 30-night period were analyzed. The mean age was 17.7 ± 4.6 years (21 females). Median adherence was significantly higher at 30 nights (56.7%, interquartile range: 0.0, 90.8%) than at 90 nights (34.4%, interquartile range: 0.0, 86.7%) ($P < .05$). Median usage did not differ between the 30-night and 90-night periods. Demographic characteristics and PAP therapy parameters were not associated with adherence or usage. Among the longitudinal cohort (n = 32), median adherence was 69.7% (interquartile range: 19.2, 90.0%), and median usage was 78.2% (interquartile range: 45.2, 95.7%). Compared to an age- and sex-matched cohort without DS, patients with DS demonstrated higher PAP adherence ($P < .05$).

Conclusions: Cross-sectional and longitudinal analyses reveal that many patients with DS successfully adhere to PAP therapy, challenging the misconception that they struggle with adherence and proving they may be as successful, if not more, than non-DS patients.

Keywords: obstructive sleep apnea, Down syndrome, positive airway pressure, adherence

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BRIEF SUMMARY

Current Knowledge/Study Rationale: Obstructive sleep apnea is highly prevalent among individuals with Down syndrome. Due to the high risk of residual obstructive sleep apnea following conventional surgical treatments, many patients are prescribed positive airway pressure therapy, but little is known about their adherence to this therapy over time.

Study Impact: Our study demonstrates that many patients adhere to positive airway pressure therapy, with a subset achieving near-perfect adherence and sustaining this adherence longitudinally. These results provide evidence supporting positive airway pressure as an effective therapy for managing obstructive sleep apnea in individuals with Down syndrome.

INTRODUCTION

Down syndrome (DS) stands as the foremost prevalent genetic chromosomal disorder, characterized by the primary cause of nondisjunction of chromosome 21 during cell division. Its reported incidence affects approximately 1 in 700 live births.[1] The phenotypic spectrum of DS includes intellectual disability, congenital heart disease, hypothyroidism, obesity, and sleep-disordered breathing.[2]

Among children with DS, obstructive sleep apnea (OSA) emerges as a prevalent potentially lifelong comorbidity, with reported rates ranging from 50–79%, markedly surpassing the 2–3% prevalence reported in the general pediatric population.[3] OSA onset in patients with DS typically begins during childhood, primarily attributed to adenotonsillar hypertrophy, the major risk factor for pediatric OSA.[4] However, distinct clinical features of DS such as macroglossia, midface hypoplasia, and hypotonia,[5] which intensify with age, contribute to the sustained high prevalence of OSA throughout the lifespan. Of note, clinical symptoms in children with DS do not reliably indicate the presence or absence of OSA,[6] necessitating diagnostic polysomnography screening for all children with DS by the age of 4 years.[7]

The primary treatment modality for OSA in children with DS remains adenotonsillectomy (AT), acknowledged as the first-line intervention for pediatric OSA.[4] However, both our

research and that of others have demonstrated that while AT often leads to improvement in OSA, it frequently falls short of achieving a cure.[8–12] Moreover, there are inherent surgical risks associated with AT in patients with DS.[13,14] Given the limitations of AT and its modest efficacy in children with DS, positive airway pressure (PAP) therapy emerges as a common alternative for this population. PAP stands as one of the most effective treatment options for pediatric OSA, yet challenges such as adherence and the complexity of pressure titration persist.[15,16] The adherence to PAP therapy among children with DS remains underexplored, with the long-term efficacy of PAP therapy in this population remaining undefined.

This study endeavors to (1) evaluate both cross-sectional and longitudinal adherence to PAP therapy and (2) pinpoint risk factors, encompassing demographic variables and factors associated with PAP therapy (such as mask leak or PAP pressures), which are hypothesized to influence adherence to PAP therapy in children with DS.

METHODS

Study population

This retrospective study aimed to identify individuals diagnosed with DS, indicated by an International Classification of Disease-10th Revision code (Q90.x), within the electronic health records (EHR) of the Center for Healthy Sleep at Rady Children's Hospital, San Diego, CA. Moreover, eligible individuals were required to have an additional International Classification of Disease code for PAP dependence (Z99.89) or a physician's order for a PAP device or related supplies (such as mask, filter, tubing) at any documented point within the EHR. Subsequently, following the identification process within the EHR, only patients with recorded data in the cloud-based PAP adherence platforms, namely Airview or Care Orchestrator online databases, were considered eligible for inclusion. Patients lacking data in these online databases or those utilizing PAP devices with no capability to transmit data to these platforms were excluded from the study cohort. This retrospective study was approved by the University of California, San Diego Institutional Review Board (#181769).

Study outcomes

The study focused on 2 primary outcomes: adherence and usage. Adherence referred to the percentage of nights where PAP device usage was equal to or greater than 4 hours per night, while usage indicated the percentage of nights with any duration of PAP device usage. In the absence of established thresholds for PAP adherence in children, we evaluated adherence across the entire population by using the median adherence of all patients as a reference point. Patients with adherence below the median were deemed nonadherent.

Risk factors associated with PAP adherence

To evaluate the factors influencing PAP adherence outcomes as mentioned earlier, we analyzed demographic characteristics obtained from the EHR, comprising sex, age, race, and ethnicity. Additionally, we explored the influence of cloud-based PAP parameters such as device pressures, residual apnea-hypopnea index, and median device leak on PAP adherence outcomes.

PAP usage

We assessed PAP usage in patients with DS through 2 distinct approaches. Initially, we conducted a cross-sectional evaluation of PAP adherence in all enrolled individuals over 2 duration periods: 30 nights and 90 nights following their most recent sleep clinic visit before June 2021. The selection of June 2021 as the cutoff date was influenced by the Philips device recall.[17] Our primary analyses examined the impact of demographic risk factors and cloud-based PAP parameters on adherence against the backdrop of 30-night and 90-night usage windows.

In addition, we delved into a longitudinal exploration of PAP adherence within this cohort, analyzing serial 90-night adherence data reports spanning from the initiation of PAP therapy up to June 2021.

Statistical analyses

Comparisons between normally distributed continuous variables (eg, age) and categorical variables (eg, adherent vs nonadherent were performed using 2-tailed *t* tests, and chi-square was used to compare binary categorical variables (eg, male sex, ethnicity, preferred language). Non-normally distributed variables (eg, residual apnea-hypopnea index, body mass index) were compared using nonparametric Mann-Whitney *U* tests. Adherence rates were compared using unpaired 2 tailed *t* test. A *P* value of $< .05$ was considered statistically significant. Statistical data analyses were performed using the software IBM Statistical Package for Social Sciences version 28.0.1.1 (Armonk, NY).

RESULTS

From the EHR, we identified 73 patients with DS who were using PAP therapy. Cloud-based data were accessible for 47 of these patients (64%) over a 90-night period and for 46 patients (63%) over a 30-night period. A demographic overview of the study cohort is presented in **Table 1**. The mean age of the patients was 17.7 ± 4.6 years, with 21 females (45%). Most patients were using continuous PAP or auto-adjusting continuous PAP devices. Additional demographic characteristics are detailed in **Table 1**.

The analysis of median adherence revealed higher rates at 30 nights post-clinic visit compared to 90 nights post-clinic visit: 56.7% (interquartile range [IQR]: 0.0, 90.8%) vs 34.4% (IQR: 0.0, 86.7%), respectively ($P < .05$). **Figure 1** provides a graphical representation of adherence at 30 nights and 90 nights, illustrating that approximately one-third of patients were highly adherent, another third were nonusers, and the remaining third exhibited partial adherence.

There was no significant difference in median usage between the 30-night and 90-night periods following the clinic visit: 58.2% (IQR: 13.0, 93.9%) vs 62.0% (IQR: 13.0, 94.4%), respectively (P = nonsignificant).

When assessing adherent patients (defined as those using PAP for 4 or more hours greater than the median), we observed

Table 1—Demographic summary of study population (n = 47).

Sex	
Female	21
Male	26
Age (mean ± SD) (years)	17.7 ± 4.6
BMI (mean, IQR) (kg/m^2)	26.63 (20.9, 31.7)
Ethnicity	
Hispanic	34
Non-Hispanic	13
Primary language	
English	28
Spanish	19
Insurance	
Public	31
Private	16
Device	
CPAP	27
BPAP	5
APAP	15

APAP = autotitrating positive airway pressure, BMI = body mass index, BPAP = bilevel positive airway pressure, CPAP = continuous positive airway pressure, IQR = interquartile range, SD = standard deviation.

that 23 out of 46 patients (50%) were adherent at 30 nights post-clinic visit, and 23 out of 47 patients (49%) were adherent at 90 nights post-clinic visit. Across both 30-night and 90-night intervals, no demographic risk factors were identified as being associated with PAP adherence. Among PAP-based parameters, no factors were significantly associated with adherence at either 30- or 90-nights post-clinic visit. However, there was a trend suggesting that higher expiratory positive airway pressure and/or lower residual apnea-hypopnea index were associated with greater adherence (**Table 2**).

We also conducted a longitudinal assessment of PAP usage and adherence. Out of the 47 individuals with accessible cloud-based adherence data, 32 individuals (68.1% of the cohort) had longitudinal data spanning 2 or more consecutive 90-night PAP adherence reports. A demographic summary of these 32 individuals is presented in **Table S1** in the supplemental material.

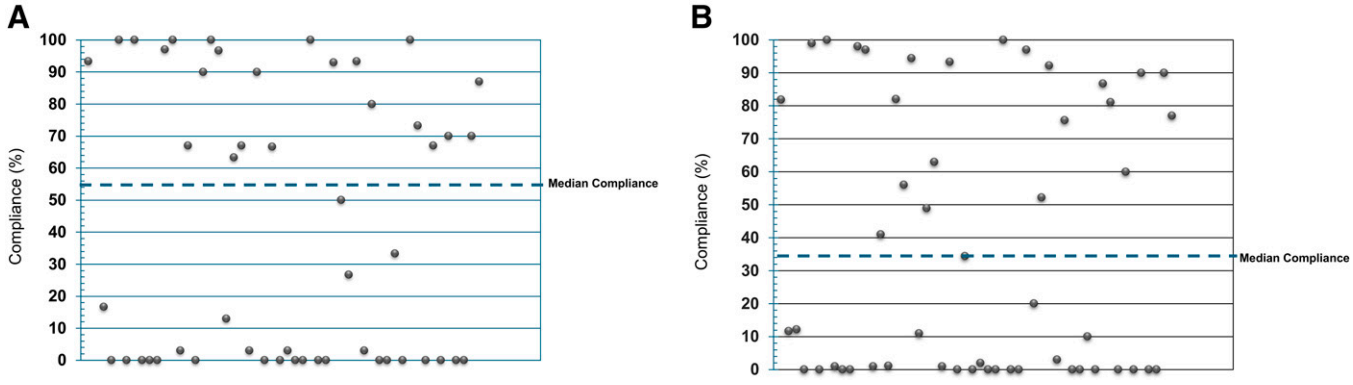
The median adherence rate was 69.7% (IQR: 19.2, 90.0%), and the median usage rate was 78.2% (IQR: 45.2, 95.7%). A graphical representation of longitudinal adherence and usage for this cohort is depicted in **Figure 2**. The median hourly usage of PAP in this longitudinally followed cohort was 6.40 hours (IQR: 3.2, 7.9 hours). Additionally, when assessing Centers for Medicare and Medicaid Services adherence—defined as using PAP for at least 4 hours per night on 70% of nights during a consecutive 30-night period within the initial 90 nights—this was observed in 19 out of 32 individuals (59%).

When comparing the mean ± standard deviation adherence rate of 54.5 ± 12.1% of the longitudinal cohort to both (1) an age- and sex-matched population of patients prescribed PAP without DS or other congenital abnormalities, including cerebral palsy (see **Table S2** in the supplemental material for matching data) and (2) previously published data,[16] patients with DS demonstrated significantly higher PAP adherence at 90 nights (**Figure 3**) compared to all groups with exception to the group of children aged 12–15 years (n = 0.08) from previously published data.[16]

DISCUSSION

Our study on patients with DS using PAP therapy, through both cross-sectional and longitudinal analyses, confirmed several key findings. While many patients with DS faced challenges in consistently using PAP, including some who discontinued its use, a marked number exhibited what we would term "super usage," with adherence rates of 90% or higher (eg, 13 of 46 or 28% in the cross-sectional analysis of 30 nights following clinic visit). Notably, when compared to patients at our center without congenital or syndromic conditions using PAP, or to findings

Figure 1—Cross sectional PAP therapy adherence.



(A) Adherence or the percentage of nights where PAP device usage was equal to or greater than 4 hours per night over 30 consecutive nights following sleep clinic visit. **(B)** Adherence or the percentage of nights where PAP device usage was equal to or greater than 4 hours per night over 90 consecutive nights following sleep clinic visit. PAP = positive airway pressure.

Table 2—Risk factors associated with positive airway pressure adherence at 30-night and 90-night intervals.

	30-Night Interval			90-Night Interval		
	Nonadherent (n = 23)	Adherent (n = 23)*	*P*	Nonadherent (n = 23)	Adherent (n = 24)**	*P*
Demographic Characteristics						
Age	17.3 (4.8)	17.6 (4.3)	NS	17.8 (4.9)	17.5 (4.3)	NS
Male sex	13 (56.5)	12 (52.2)	NS	14 (60.9)	12 (50)	NS
Hispanic ethnicity	16 (69.6)	15 (65.2)	NS	15 (65.2)	16 (66.7)	NS
Spanish preferred language	9 (39.1)	9 (39.1)	NS	8 (34.8)	10 (41.7)	NS
BMI (kg/m^2)	25.4 (19.7, 30.2)	29.6 (22.3, 34.9)	NS	25.4 (20.9, 31.5)	26.7 (21.5, 33)	NS
Device Data						
EPAP	7.9 (5, 11.1)	9.4 (7.8, 10)	.103	7.9 (5, 9.9)	8 (6, 10)	.160
rAHI (events/h)	6.4 (4.1, 12.6)	1.7 (1.4, 3.1)	.173	6.3 (2.9, 10.6)	1.6 (1.3, 3.2)	NS
Median leak (lpm)	2.1 (0.3, 4.6)	3.9 (0.3, 7)	NS	4.9 (0.9, 7.2)	2.7 (0.4, 5.3)	NS

*Median adherence % of nights use in the 30-night adherence: 56.7% (used for cutoff at 30-night interval); **Median adherence % of nights use in the 90-night adherence: 34.4% (used for adherence cutoff of 90-night interval). BMI = body mass index, EPAP = expiratory positive airway pressure, lpm = liters per minute, NS = nonsignificant, rAHI = residual apnea-hypopnea index.

from a previously published big-data study on pediatric PAP adherence,[16] we found that patients with DS in fact demonstrated even higher adherence at 90 nights. At minimum, these results challenge the notion that individuals with DS cannot successfully use PAP therapy. In fact, our longitudinal analysis showed that most patients using PAP therapy experience improvements over time. Notwithstanding, we do remark that our cross-sectional analysis revealed a decline in adherence rather than usage when comparing PAP reports for 90 nights vs 30 nights following the clinic visit. While we cannot explicitly explain why this occurred, we speculate that the proximity of the 30-night period to the clinic visit emphasizes the importance of regular follow-up and close contact with the health care team. This likely helps ensure continued patient education and motivation, which may explain the higher adherence seen in the immediate 30-night follow-up compared to the 90-night follow-up.

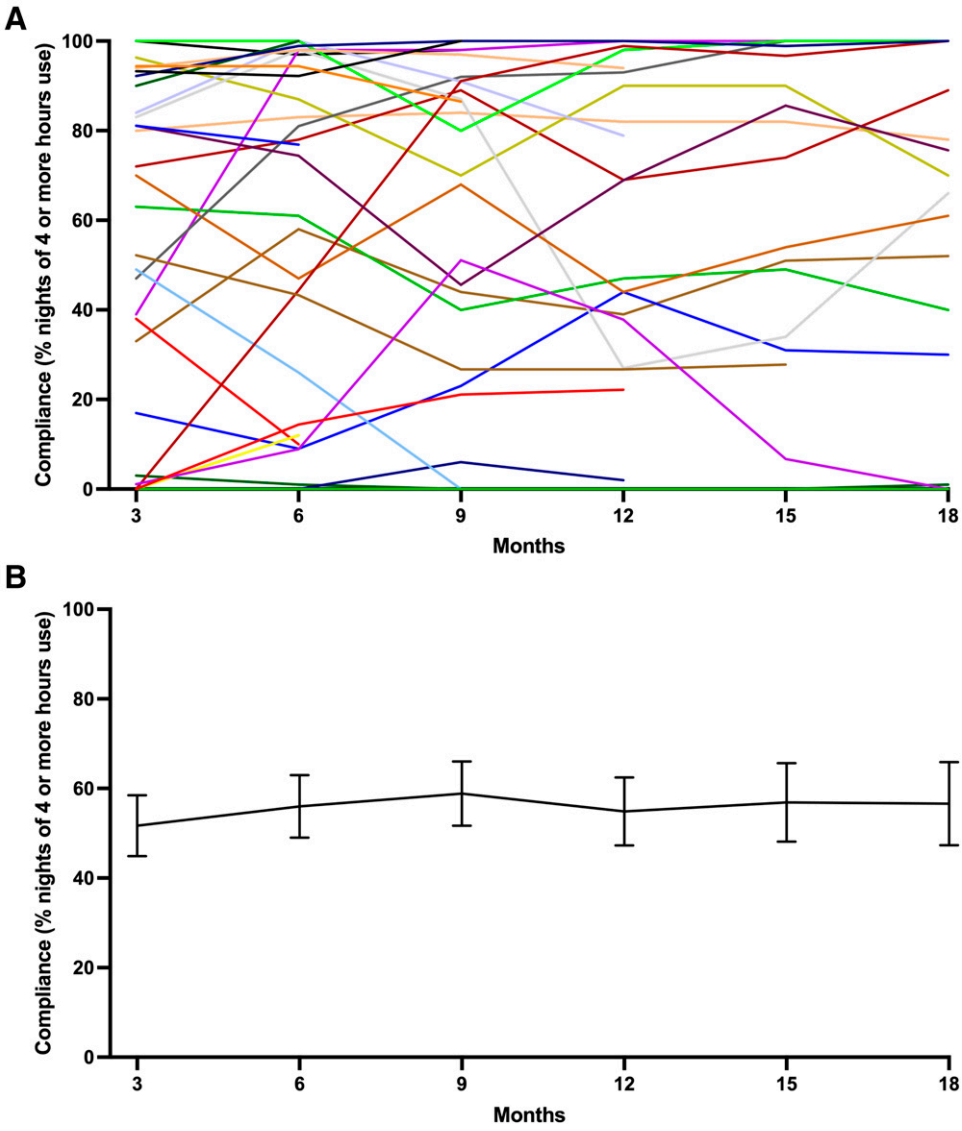
Pediatric OSA management primarily involves surgical interventions, with AT being the most common procedure. However, it is well recognized that this approach is insufficient in nearly 20% of cases,[18] highlighting the need to explore alternative therapies for treating pediatric OSA.[19] Studies in children with DS also consistently show that conventional surgical approaches often fall short in the management of OSA.[8–12] Other more intensive surgical approaches for managing OSA in DS include tongue reduction, uvulopalatopharyngoplasty, maxillomandibular advancement, and lingual tonsillectomy, often tailored to address specific anatomical abnormalities causing airway obstruction. Additionally, the surgical placement of a hypoglossal nerve stimulator, recently approved for children with DS aged 13 and older,[20] has emerged as a viable adjunctive treatment.[21,22] While these secondary surgeries offer significant promise for treating OSA in DS, it is important to note that they are potentially invasive and carry substantial risks.

Our study provides valuable insights, indicating that PAP therapy is highly effective in many patients with DS and suggests a strong likelihood of success in this population. Although few studies have investigated OSA across the lifespan in patients with DS, the progressive hypotonia of upper airway muscles with age suggests that PAP or hypoglossal nerve stimulation may eventually be the only therapies with sustained effectiveness, although this concept remains unknown. Furthermore, the impact of OSA treatment on altering the life course of patients with DS is considerably understudied. The possibility that OSA therapy could slow or prevent the progression of cardiovascular disease or delay the onset of Alzheimer's disease and dementia—conditions highly prevalent in DS—raises critical questions for patients and families about the importance of treating OSA to improve long-term health outcomes. Our findings support the rationale for developing clinical trials to evaluate how PAP therapy can enhance the life course of patients with DS by effectively managing OSA.

Previous studies have shown that PAP therapy is effective in treating OSA in children with DS, including improvements in health.[23,24] However, research specifically focusing on PAP usage and adherence in this population remains limited. Kang et al[25] found that, compared to typically developing children, 103 children with developmental disabilities—including 31% with DS—demonstrated higher PAP usage at 3 and 6 months, with up to 90% of nights utilizing the device. Notably, 18 of the 103 children dropped out, potentially due to nonadherence. A key limitation of the study is that adherence, defined as the percentage of nights with at least 4 hours of PAP use, was not reported. Nonetheless, PAP usage was higher and remained higher over time in children with developmental disabilities. The study also identified higher income and repeat PAP titrations as positive predictors of increased PAP usage in these children.

Other studies have similarly reported that children with developmental disabilities are more likely to adhere to continuous PAP therapy.[26] While the reasons for higher adherence in these children remain unclear, factors such as increased parental

Figure 2—Longitudinal PAP therapy adherence.



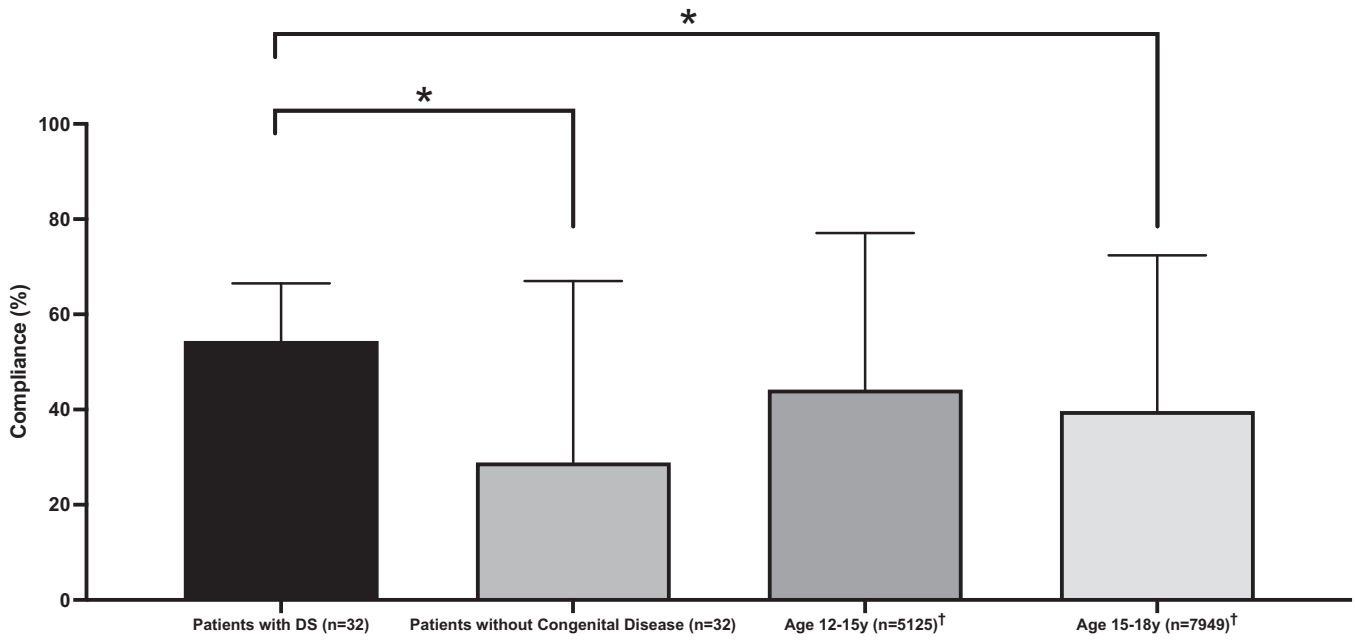
(A) Longitudinal adherence of individual patients with Down syndrome (n = 32). **(B)** Aggregated longitudinal adherence of all patients with Down syndrome (n = 32). Expressed as mean ± standard error of the mean. PAP = positive airway pressure.

involvement in care may play a major role. Additionally, children with disabilities, including DS, often require more intensive medical support, including frequent clinical follow up visits, which may contribute to the observed improvements in PAP adherence over time in both our findings and those of Kang et al.[25] Regular follow-up for children using PAP could help address common issues like mask fit, air leaks, PAP levels, and device mode adjustments, including considerations for bilevel PAP. Timely intervention for symptoms such as nasal congestion may further optimize adherence. These follow-up visits also reinforce education, emphasizing the rationale behind PAP therapy, while providing strategies for better adherence, such as behavioral modification and positive reinforcement through rewards. Routine follow-up is a critical component of adherence to therapy.[27]

Xanthopoulos et al[28] have highlighted considerable heterogeneity in parental perceptions and experiences in caring for children with DS who use PAP. Variability exists in initial education about PAP and adherence strategies, including the use of visual aids, as well as in caregiver experiences with the medical care team. Given the dynamic nature of PAP adherence, the authors emphasize the importance of ongoing communication and partnership with families to optimize outcomes, underscoring the role of regular follow-up. This notion is especially pertinent in children with DS, where the complexity of care may necessitate more frequent and comprehensive follow-up visits, potentially explaining higher adherence rates in children with DS compared to controls.

A key factor in improving PAP adherence in children is the presence of family members who also use PAP therapy and can

Figure 3—Comparing PAP therapy adherence in patients with DS to that of other populations.



Adherence with PAP therapy in a longitudinal cohort of patients with DS compared to other populations. †PAP adherence data for patients aged 12–15 years and 15–18 years derived from a previously published study[16]; *$P < .05$. DS = Down syndrome, PAP = positive airway pressure.

serve as role models. Puri et al[29] found that among 56 children, 32% of whom had developmental disabilities, those with a family member using PAP showed significantly higher usage hours and a trend toward greater adherence compared to children without a family member using the therapy.

The findings of these studies underscore the critical role that family members and caregivers play in supporting children on PAP therapy. This involvement may help explain why patients with DS often demonstrate higher usage and adherence, as parental engagement is integral to their care.

Our study has several limitations. Although we evaluated the relationship between certain demographic factors, the retrospective design did not allow us to assess important variables such as parental experiences, knowledge of PAP therapy, previous exposure to PAP, or the use of behavioral modification techniques at home, including visual aids, which may influence PAP adherence. Additionally, we lacked information on other potentially important demographic factors, such as parental education level or socioeconomic status. Additionally, our sample was drawn from a single center, limiting the generalizability of our findings, particularly given the existing variability in access to pediatric sleep medicine, clinical care, and education provided by durable medical equipment companies. The small sample size further constrained our ability to perform adequately powered multivariate logistic regression analyses to assess the impact of factors like mask leaks and PAP pressures, both of which have been identified as significant predictors of adherence in larger studies using big data approaches.[16] Future studies should adopt multicenter designs to increase sample size and improve generalizability. Finally, a key limitation of studies assessing PAP adherence longitudinally is the selection bias inherent in focusing on patients who continue using PAP across all adherence levels. Patients who discontinue PAP due to nonusage are excluded from the analysis, resulting in the absence of their longitudinal data and potentially inflating adherence rates. Despite this, our longitudinal analysis with a smaller cohort (n = 32) showed that a subset of patients consistently adhered to PAP therapy.

CONCLUSIONS

Through both cross-sectional and longitudinal analyses, our findings strongly demonstrate that many patients with DS successfully adhere to PAP therapy. These results align with the DISCOVER study, where Svensson et al[30] reported that among 33 adults with DS and OSA, cross-sectional analysis revealed that PAP adherence (≥ 4 hours of use) was achieved on 60% of nights, comparable to the 62.6% seen in 132 control adults over 1.3 years of follow-up. In contrast to the DISCOVER study, our research included children, was conducted at a children's hospital, and featured longitudinal analyses. Together, these findings challenge the misconception that patients, including children with DS struggle with PAP therapy. In fact, they are as successful—if not more—than those without DS. Given these observations, by identifying patients with DS who are super users a priori, our study lays the groundwork for future research to explore whether PAP therapy can effectively improve long-term health outcomes in DS, including cognitive function and the prevention of Alzheimer's disease.

ABBREVIATIONS

AT, adenotonsillectomy
DS, Down syndrome
EHR, electronic health record
IQR, interquartile range
OSA, obstructive sleep apnea
PAP, positive airway pressure

REFERENCES

1. Parker SE, Mai CT, Canfield MA, et al; National Birth Defects Prevention Network. Updated national birth prevalence estimates for selected birth defects in the United States, 2004-2006. *Birth Defects Res A Clin Mol Teratol.* 2010;88(12):1008–1016.
2. Desai SS. Down syndrome: a review of the literature. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod.* 1997;84(3):279–285.
3. Lee CF, Lee CH, Hsueh WY, Lin MT, Kang KT. Prevalence of obstructive sleep apnea in children with Down syndrome: a meta-analysis. *J Clin Sleep Med.* 2018;14(5):867–875.
4. Marcus CL, Brooks LJ, Draper KA, et al; American Academy of Pediatrics. Diagnosis and management of childhood obstructive sleep apnea syndrome. *Pediatrics.* 2012;130(3):576–584.
5. Simpson R, Oyekan AA, Ehsan Z, Ingram DG. Obstructive sleep apnea in patients with Down syndrome: current perspectives. *Nat Sci Sleep.* 2018;10:287–293.
6. Nehme J, LaBerge R, Pothos M, et al. Predicting the presence of sleep-disordered breathing in children with Down syndrome. *Sleep Med.* 2017;36:104–108.
7. American Academy of Pediatrics Committee on Genetics. Health supervision for children with Down syndrome. *Pediatrics.* 1994;93(5):855–859.
8. Hurvitz MS, Lesser DJ, Dever G, Celso J, Bhattacharjee R. Findings of routine nocturnal polysomnography in children with Down syndrome: a retrospective cohort study. *Sleep Med.* 2020;76:58–64.
9. Farhood Z, Isley JW, Ong AA, et al. Adenotonsillectomy outcomes in patients with Down syndrome and obstructive sleep apnea. *Laryngoscope.* 2017;127(6):1465–1470.
10. Nation J, Brigger M. The efficacy of adenotonsillectomy for obstructive sleep apnea in children with Down syndrome: a systematic review. *Otolaryngol Head Neck Surg.* 2017;157(3):401–408.
11. Thottam PJ, Choi S, Simons JP, Kitsko DJ. Effect of adenotonsillectomy on central and obstructive sleep apnea in children with Down syndrome. *Otolaryngol Head Neck Surg.* 2015;153(4):644–648.
12. Shete MM, Stocks RM, Sebelik ME, Schoumacher RA. Effects of adeno-tonsillectomy on polysomnography patterns in Down syndrome children with obstructive sleep apnea: a comparative study with children without Down syndrome. *Int J Pediatr Otorhinolaryngol.* 2010;74(3):241–244.
13. Goldstein NA, Armfield DR, Kingsley LA, Borland LM, Allen GC, Post JC. Postoperative complications after tonsillectomy and adenoidectomy in children with Down syndrome. *Arch Otolaryngol Head Neck Surg.* 1998;124(2):171–176.
14. Bower CM, Richmond D. Tonsillectomy and adenoidectomy in patients with Down syndrome. *Int J Pediatr Otorhinolaryngol.* 1995;33(2):141–148.
15. Watach AJ, Xanthopoulos MS, Afolabi-Brown O, et al. Positive airway pressure adherence in pediatric obstructive sleep apnea: a systematic scoping review. *Sleep Med Rev.* 2020;51:101273.
16. Bhattacharjee R, Benjafield AV, Armitstead J, et al; medXcloud group. Adherence in children using positive airway pressure therapy: a big-data analysis. *Lancet Digit Health.* 2020;2(2):e94–e101.
17. Philips I. https://www.usa.philips.com/a-w/about/news/archive/standard/news/press/2021/20210614-philips-issues-recall-notification-to-mitigate-potential-health-risks-related-to-the-sound-abatement-foam-component-in-certain-sleep-and-respiratory-care-devices/. Published 2021. Accessed 2024.
18. Bhattacharjee R, Kheirandish-Gozal L, Spruyt K, et al. Adenotonsillectomy outcomes in treatment of obstructive sleep apnea in children: a multicenter retrospective study. *Am J Respir Crit Care Med.* 2010;182(5):676–683.
19. Ehsan Z, Ishman SL, Soghier I, et al. Management of persistent, post-adenotonsillectomy obstructive sleep apnea in children: an official American Thoracic Society clinical practice guideline. *Am J Respir Crit Care Med.* 2024;209(3):248–261.
20. FDA. https://www.fda.gov/medical-devices/recently-approved-devices/inspire-upper-airway-stimulation-p130008s089. Published 2023. Accessed 2024.
21. Diercks GR, Wentland C, Keamy D, et al. Hypoglossal nerve stimulation in adolescents with Down syndrome and obstructive sleep apnea. *JAMA Otolaryngol Head Neck Surg.* 2018;144(1):37–42.
22. Holtzlander B, Rodman C, Manchanda S, et al. A scoping review highlighting the need for outcomes research in hypoglossal nerve stimulation for patients with Down syndrome. *J Clin Sleep Med.* 2024;20(7):1217–1219.
23. Nehme J, LaBerge R, Pothos M, et al. Treatment and persistence/recurrence of sleep-disordered breathing in children with Down syndrome. *Pediatr Pulmonol.* 2019;54(8):1291–1296.
24. Hill EA, Fairley DM, Williams LJ, Spanò G, Cooper S-A, Riha RL. Prospective trial of CPAP in community-dwelling adults with Down syndrome and obstructive sleep apnea syndrome. *Brain Sci.* 2020;10(11):844.
25. Kang EK, Xanthopoulos MS, Kim JY, et al. Adherence to positive airway pressure for the treatment of obstructive sleep apnea in children with developmental disabilities. *J Clin Sleep Med.* 2019;15(6):915–921.
26. Hawkins SM, Jensen EL, Simon SL, Friedman NR. Correlates of pediatric CPAP adherence. *J Clin Sleep Med.* 2016;12(6):879–884.
27. Brookhart MA, Patrick AR, Schneeweiss S, et al. Physician follow-up and provider continuity are associated with long-term medication adherence: a study of the dynamics of statin use. *Arch Intern Med.* 2007;167(8):847–852.
28. Xanthopoulos MS, Nelson MN, Eriksen W, et al. Caregiver experiences helping children with Down syndrome use positive airway pressure to treat obstructive sleep apnea. *Sleep Med.* 2023;107:179–186.
29. Puri P, Ross KR, Mehra R, et al. Pediatric positive airway pressure adherence in obstructive sleep apnea enhanced by family member positive airway pressure usage. *J Clin Sleep Med.* 2016;12(7):959–963.
30. Svensson M, Ekström M, Sundh J, Ljunggren M, Grote L, Palm A. Adherence to CPAP therapy in Down syndrome: the population-based DISCOVERY study. *J Clin Sleep Med.* 2023;19(3):453–458.

SUBMISSION & CORRESPONDENCE INFORMATION

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DISCLOSURE STATEMENT

All authors have seen and approved the manuscript. This work was conducted at Rady Children's Hospital (RCHSD), San Diego, CA and University of California San Diego Health (UCSD), San Diego, CA. Dr. Malhotra reports a philanthropic donation to the University of California, San Diego by ResMed Inc. All other authors do not report any conflicts of interest related to this work.

Table S1—Demographic summary of study population assessed for longitudinal analysis of PAP adherence (n=32).

Number of patients	32
Number of 90-night PAP reports	6.3±3.9
Age (years)	19±4.01 (9.6-26.2)
BMI (kg/m^2)	28.20±9.98
Sex	
Female	14 (44%)
Male	18 (56%)
Race	
White	14 (44%)
Hispanic	8 (25%)
Asian	4 (12%)
Other	6 (19%)
Device type	
CPAP	15 (47%)
Bilevel PAP	5 (16%)
APAP	12 (37%)
CMS Medicare compliance	
Yes	19 (59%)
No	13 (41%)

No statistical differences were observed between the non-compliant and compliant groups. NS – not statistically significant ($P > 0.2$).

BMI – body mass index; EPAP – expiratory positive airway pressure; rAHI – residual apnea-hypopnea index; lpm – liters per minute

Table S2—Comparison of patients with Down syndrome and without congenital syndromes.

	Patients with DS (n=32)	**Patients without Congenital Syndromes (n=32)**	***P***
Age (y)	17.7±4.6	17.2±4.1	NS
Sex			NS
Female	14	14	
Male	18	18	

DS – Down syndrome; NS – not statistically significant ($P > 0.2$).