An In-patient Model for Positive Airway Pressure Desensitization: A Report of 2 Pediatric Cases

Kelli-Lee Harford PhD, Supriya Jambhekar MD, Gulnur Com MD, Lisa Bylander RRT-NPS, Kristi Pruss RRT RPSGT, Jana Teagle, and Wendy Ward PhD

Application of positive airway pressure is frequently indicated in pediatric patients with a diagnosis of obstructive sleep apnea. Adherence to equipment use is often less than optimal and can be more challenging when working with children with special needs. An in-patient protocol was designed utilizing various techniques and strategies from the medical adherence literature and applied to 2 cases. This protocol utilizes specialists from various disciplines, including respiratory therapists, psychologists, physicians, nurses, and child life therapists, as well as parental involvement. This paper outlines this protocol using 2 case studies. Both patients successfully used their equipment for greater than 4 hours at night by the end of their hospital stay of 4 days and maintained or advanced these gains at follow-up. These 2 cases suggest that more research should be conducted to further evaluate the effectiveness of similar programs. *Key words: pediatric; PAP; sleep apnea.* [Respir Care 2012;57(5):802–807. © 2012 Daedalus Enterprises]

Introduction

Obstructive sleep apnea (OSA) is a sleep related breathing disorder consisting of partial and/or complete upper-airway obstruction that is associated with a disruption in ventilation and disturbance of sleep. Prevalence of OSA in pediatric populations ranges from 0.7% to 10.3%.[1] OSA is associated with a number of negative sequelae in pediatric patients, including restless sleep, behavior problems, neurocognitive abnormalities, growth problems, and cardiovascular complications.[1] In children, tonsillar and/or adenoidal enlargement is the main cause of OSA and surgery (ie, tonsillo adenoidectomy/uvulopalatopharyngoplasty is often the first line of management for OSA). However, there are a substantial percentage of children with OSA who either do not have any surgical options, are poor surgical candidates, or who have residual OSA after surgery. Positive airway pressure (PAP) is frequently indicated in this group of pediatric patients with a diagnosis of OSA. Use of these devices improves ventilation and oxygenation and has been associated with an improvement in sleep quality, daytime symptoms, quality of life, and overall health.[2,3] Although the majority of these studies have been with adults, research on PAP use in pediatric populations also indicates improvement in the negative effects of OSA with PAP use.[4,5]

While the efficacy of PAP has been established, adherence to this equipment is relatively poor.[6] In pediatric populations, adherence rates range from 50% to 100% and vary based on the population studied, the criteria for assessing and measuring adherence,[7–9] and participation of the sample in behavioral interventions. Evidence of effective clinical programs to improve pediatric adherence is particularly sparse. In children, factors that have been associated with non-adherence include age, with 6–12-year-olds most accepting and 1–5-year-olds and adolescents demonstrating the greatest difficulty. Use of an oro-

Dr Harford is affiliated with the Department of Orthopedics and Rehabilitation, University of Wisconsin Hospital and Clinics, Madison, Wisconsin. Drs Jambhekar, Com, and Ward are affiliated with the Department of Pediatrics, University of Arkansas for Medical Sciences, Little Rock, Arkansas. Ms Bylander and Ms Pruss are affiliated with the Department of Respiratory Care Services; and Ms Teagle is affiliated with the Department of Child Life and Education Services, Arkansas Children's Hospital, Little Rock, Arkansas.

The authors have disclosed no conflicts of interest.

Correspondence: Kelli-Lee Harford PhD, Department of Orthopedics and Rehabilitation, University of Wisconsin Hospital and Clinics, 1675 Highland Avenue, MC 2424, Madison WI 53792. E-mail: kharford@uwhealth.org.

DOI: 10.4187/respcare.01231

nasal mask and equipment that is generally uncomfortable have also been associated with non-adherence.[8]

Factors that have been associated with non-adherence include severity of disease,[8] side effects, and pressure intolerance.[9] These factors may be compounded in children with special needs, and research has indicated that adherence may be particularly challenging when working with children within this population.[7]

Education has been shown to be effective in improving adherence.[8] Other strategies that may help to improve adherence include optimizing the fit of the equipment and effectively incorporating family members. There is also some existing research suggesting that by using behavioral therapy principles, PAP treatment can successfully be implemented in pediatric populations in both in-patient and out-patient settings.[8–10] Previous authors have used nursing, meter reading, and parent report to assess adherence to PAP treatment, in both in-patient and out-patient settings, and provided a detailed task analysis for PAP desensitization as well as behavioral guidelines for families when incorporating PAP equipment at home.[9,10] Others have assessed adherence using compliance cards in an out-patient setting.[7] Here we present 2 patients to illustrate an in-patient protocol using behavioral techniques to address non-adherence with PAP after unsuccessful out-patient treatment. Adherence was measured subjectively by parent report and objective download of the patients' compliance cards on a daily basis.

Case Report 1

Patient 1 was a 28-month-old male toddler who had undergone a tonsillectomy, adenoidectomy, and uvuloplasty, due to severe OSA. Due to persistence of clinical symptoms and overnight polysomnograph findings (apnea-hypopnea index 12 events/h, rapid-eye-movement index 27 events/h), CPAP treatment with a pressure of 6 cm of H_2O was recommended, but despite the family's efforts for 6 months, the child continued to be very resistant to the CPAP equipment and adherence was essentially zero. Patient 1 also exhibited poor growth and appetite. On admission it was noted that the family was skeptical about the protocol and reluctant to give any medication to their child. The family's concerns about the protocol and the medication were addressed by the team, including the utility of the medication in assisting with sleep onset and PAP desensitization. The parents appeared more receptive to the intervention following this discussion, and provided consent to proceed with the protocol, and received a prescription for an antihistamine.

Case Report 2

Patient 2 was a 4-year-old female with CHARGE syndrome (coloboma of the eye, heart defect, atresia of the choanae, retardation of growth and/or development, genital and/or urinary abnormalities, and ear abnormalities and deafness) associated with tracheomalacia, hearing disability, visual impairment, and severe OSA (apnea-hypopnea index 12 events/h), who had initially utilized CPAP at a pressure of 8 cm of H_2O for 1 year and then refused to use her equipment for more than 6 months due to aggression/behavioral problems and increased overall sensory sensitivity, per parent report. In addition to OSA, this patient also demonstrated problems with sleep initiation and maintenance. She was on a medication regimen of melatonin, clonidine, and an antihistamine at home, to help with sleep, but this had not been successful in helping with adherence. These medications were continued during her in-patient admission and immediately after discharge.

Discussion

These 2 patients were identified during clinical team meetings as being non-adherent after intensive out-patient educational and behavioral interventions were unsuccessful. These out-patient interventions included: initial education about the need for PAP following the child's sleep study; opportunities to ask questions about the treatment; equipment fitting and education about the equipment; nursing support by telephone; and regular follow-up by a multidisciplinary team including physicians, nurses, respiratory therapists, and psychologists every 2–3 months to monitor adherence. During these visits, if needed, families met with each member of the team individually to problem solve obstacles to adherence and discuss behavioral interventions such as desensitization procedures and incentive programs as potentially beneficial interventions. In vivo modeling was provided as appropriate.

After reviewing the patient's chart and assessing suitability for the in-patient protocol, based on their unsuccessful treatment during out-patient clinic visits and substantial need for PAP treatment, the patients were admitted to the hospital for the purposes of addressing PAP use. An in-patient PAP desensitization protocol involving a multidisciplinary approach was developed and implemented with these 2 patients. Team members included a pulmonologist, respiratory therapists, psychologists, child life specialists, and nurses. Each team member met with the child and parents at least once per day, and frequently multiple times per day, to facilitate implementation of the desensitization protocol. The majority of these visits were conducted individually, but some of these visits, particularly during the first day of hospitalization were conducted with 2 or more providers. Parental involvement was also emphasized during the hospital stay. The specific roles of the providers and the family during the in-patient stay are outlined below:

- The physician was responsible for the initial evaluation; daily rounds to monitor progress; consults for respiratory therapy, child life, and psychology on admission; providing an order for sleep inducing medication when needed; communicating with the family and team regarding implementation of the appropriate action; and assisting in the continuing education of the parent.
- The psychologist designed an individualized desensitization plan in collaboration with the family and other team members, based on the child's cognitive level, preexisting sensory or psychological problems, and observations of family dynamics and interactions. In addition, the psychologist, in collaboration with the other team members, demonstrated the desensitization techniques to the parents, and was involved with the implementation and modification of the plan as needed.
- The respiratory therapist, who is highly experienced with children and familiar with the adherence program, provided education and assistance with the PAP machine, such as optimizing the best fitting interface, adjusting the machine pressures, setting the alarms, and downloading the patient's compliance card, and was involved with the implementation of the plan.
- The child life therapist provided assistance with distractors and/or reinforcers and was involved with implementation of the plan.
- Nursing assisted with the administration of medication as needed and monitoring the patient at night for equipment related problems.
- Families were initially greeted by the physician and psychologist for an assessment and to discuss the desensitization program based on their individual needs.

Since the family's understanding and acceptance of the program was viewed as the key to overall success at home, their participation in the in-patient desensitization was required, and problems related to the procedures were addressed. After the initial meetings with the physician and psychologist, team members met with the family based on their availability and the needs of the family each day.

Patients were seen 3–6 times per day by members of the team during their in-patient stay for implementation of the desensitization plan. The desensitization plan involved the following components to varying degrees: parent and patient education regarding OSA and how PAP works; problem-solving fit and comfort issues related to the equipment; and assessing/intervening as necessary. Team members met with each patient initially every 2 hours during the day while they were awake, performing various activities specific to each patient. Team members engaged in providing reinforcement and modeling to improve acceptance of the mask and headgear. Both patients required sensory desensitization to the feel of the mask and the sounds and feel of the air when the machine was turned on, as they both exhibited anxiety related to the equipment. To help facilitate this, distraction techniques were incorporated during the shaping procedure to help decrease anxiety and increase tolerance of the equipment. Longer time periods for adherence were encouraged by providing reinforcement for wearing the mask for short periods of time and gradually increasing the length of time that the child was expected to wear the equipment. The patients were provided with familiar cues to help them anticipate the length of time that they were expected to wear the equipment. The mask was replaced gently but firmly, in a supportive environment, if it was removed too early. Shaping procedures were slowed to a more manageable level of progression if patients continued to take the mask off prematurely. The details of how these procedures were implemented for each patient are outlined below.

Patient 1 enjoyed rolling a ball back and forth between himself and his caregivers. He was allowed to do this enjoyable activity only as long as he would comply with the various desensitization activities. The mask and headgear were initially placed very loosely on the patient. As he began to tolerate this for longer periods, the headgear was tightened and he was allowed to experience the sensation of air flow blowing on his face, but not yet attached to the mask. As he began to tolerate this, the tubing was attached to the mask with PAP flow going through the mask. These interventions were conducted in a very playful environment, with frequent praise. After the first day of desensitization the patient was provided with an antihistamine for mild sedation and he was easily placed on his PAP and slept well. The next day, similar play desensitization was done while the child was awake, and there were no problems with initiating PAP that night and throughout the remainder of his hospitalization.

Patient 1 demonstrated at least 8 hours of usage per night by the end of the hospital stay of 4 days (Fig. 1). Both parents stayed with the child during the hospitalization. After the first night of CPAP use, his parents noted that he was more energetic, alert, and happy, with significantly increased appetite, and felt assured that he had not exhibited snoring or gasping for breath during the night. This observation seemingly encouraged his parents and motivated them to be consistent with the machine use.

Patient 2 had CHARGE syndrome, with many sensory issues, including deafness and partial blindness, as well as hypersensitivity to anything touching her face. The patient's mother informed us that one activity her child found pleasurable was having lotion applied to her arms and legs. With this patient the headgear, mask, and air flow were introduced in a similar manner to patient 1, but only continuing with the application of lotion as long as she was calm and not fighting the PAP equipment. At night, the

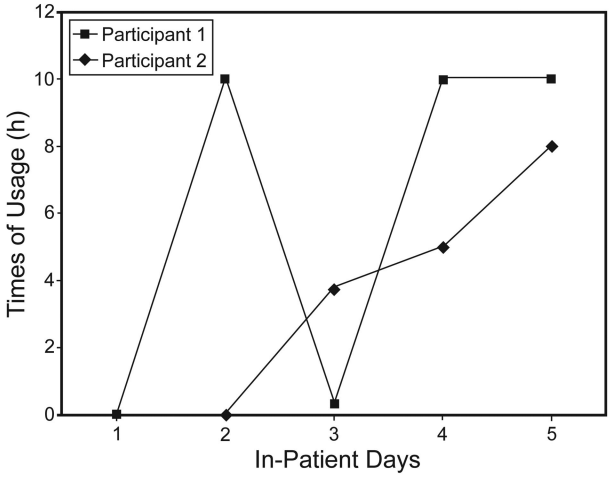

Fig. 1. Positive airway pressure usage during inpatient stay.

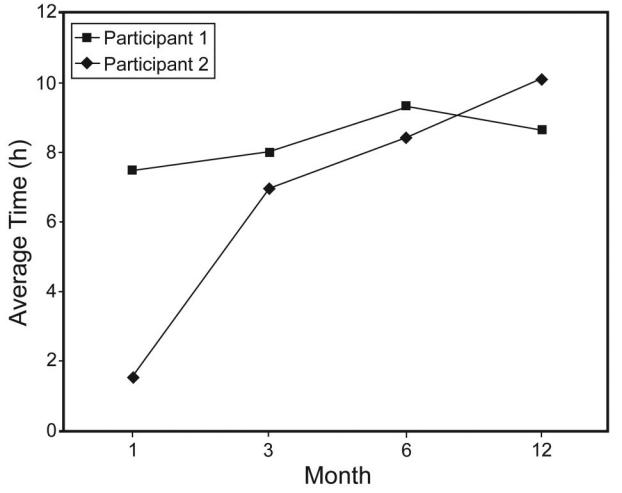

Fig. 2. Positive airway pressure usage at follow-up.

patient was provided with her usual medications (melatonin, clonidine, and an antihistamine). As the patient became sleepy, her PAP equipment was placed by a team member as her mother applied lotion to the patient. The patient slept well and the process was repeated the next day. On night 3 of admission, the patient became very combative with PAP placement, and pulled out her gastrostomy tube. She spent less than 2 hours on PAP. The next day desensitization activities resumed, with improved compliance for the rest of the admission (see Fig. 1).

At least one parent remained with the child throughout the entire hospitalization. Parents were taught the techniques used by the team members during each session and were encouraged to implement them at other times of the day and after discharge. At the beginning of the in-patient stay, interventions were done primarily by staff. However, as parents learned these techniques and began to feel more confident in their ability to implement them, they began to assume more of the responsibility for implementing these interventions. This helped parents feel more confident in their ability to implement these interventions at home and thus to be able to generalize the success obtained in the hospital setting to their home environment. The patients were discharged from the hospital when they accepted the PAP with ease throughout the night; when the parents felt comfortable continuing to help their children utilize PAP at home using the strategies learned in the hospital; and when improved nighttime symptoms and daytime functioning were observed during the in-patient stay.

Patients were then followed every 2–3 months in an out-patient multidisciplinary PAP adherence clinic manned by a sleep physician, psychologist, and respiratory therapist. Adherence was measured by objective download of the patients' compliance cards.

Since there is no pediatric measure for successful PAP usage in children, we used the adult criteria of $>$ 4 hours of PAP usage per night, which has also used by others[8] in their study of adherence in pediatric patients for the assessment of long-term success. Other adherence criteria of $\geq$ 3 hours have also been used.[5] While the measure of adherence used in this study may not accurately reflect differences in sleep needs in pediatric populations, it is the currently accepted standard of practice in the sleep clinic and consistent with previous research.

Table 1. Growth Information for Patient 1

	Weight, kg	Weight Percentile	Height, cm	Height Percentile
Before admission	11.9	10th	89	25th
1 year after	17.0	95th	104	90th

Long-term follow-up was assessed at approximately 3 months, 6 months, and 12 months for both patients. Patient 1 had 80% usage of $>$ 4 hours/night at 3 month follow-up. At 7 month follow-up he had 95% usage for $>$ 4 hours/night. Usage was maintained at 12 month follow-up, and in fact demonstrates $>$ 6 hours/night of usage on average after 3 months at home (Fig. 2). In addition, patient 1 showed improved growth following the intervention (Table 1).

Patient 2 had 91% usage of $>$ 4 hours/night at 3 month follow-up. At 7 month follow-up she had 94% usage for $>$ 4 hours/night. Usage was maintained at 12 month follow-up, and again shows $>$ 6 hours of usage per night on average (see Fig. 2). While medication was thought to assist with initial acceptance, she was weaned off medication over time, and maintained substantial usage rates. Patient 2 also showed improved weight following the intervention, although an accurate height could not be obtained because of behavioral problems during check-in (Table 2).

Table 2. Growth Information for Patient 2

	Weight, kg	Weight Percentile	Height, cm	Height Percentile
Before admission	17.1	25th	112	90th
1 year after	22.3	90th	Unable to obtain correct height	–

This intensive in-patient program achieved adherence in these 2 children who had previously failed out-patient efforts. When followed in an intensive out-patient program, compliance was maintained at short-term and long-term follow-up. These results demonstrate that patients who may not initially have been successfully managed in an out-patient setting may respond to an intensive in-patient program. However, given that only 2 patients are presented, it is not yet clear whether the in-patient protocol would be equally effective in all non-adherent pediatric PAP users, and further research is necessary to make a definitive statement about its effectiveness in all patients. However, the authors are cautiously optimistic, as the 2 patients presented were particularly challenging due to development delays, hearing and visual impairments, and behavioral resistance.

A number of factors should be considered when utilizing this protocol. Care should be taken when deciding which patients may be appropriate for in-patient versus out-patient treatment. The protocol should also be appropriate for the child's underlying condition and should be tailored based on the family's cultural background and understanding of the condition. For example, case 1 was a Hispanic child whose parents were very protective and wary of suggested interventions, including CPAP and medication. Therefore they were very resistant to the treatment. However, after seeing their child was more alert, interactive, and playful, and displayed more appetite after using CPAP in the hospital, these parents seemingly had a greater appreciation of the benefits of CPAP for their child.

Furthermore, although the intervention measured adherence using the standard of $>$ 4 hours of PAP use nightly, which has also been used by other researchers,[8] given the sleep needs of young children, a longer sleep duration is likely more beneficial. As the long-term follow-up data show, the 2 children studied did typically demonstrate on average between 6–10 hours of sleep with their PAP equipment during the majority of the period studied, and this may indicate that once they are able to achieve 4 hours on PAP, they typically are able to tolerate PAP for the majority of their sleep duration. Additionally, both children showed improved growth, when compared with their growth before the intervention.

One limitation of this intervention is that, for some patients and their families, the stress related to an in-patient hospitalization may outweigh the benefit of this intervention, and alternative out-patient options may need to be explored. Additionally, given the cost and intensity of services necessary for this in-patient protocol, out-patient treatment will likely continue to be the first line of treatment for the majority of patients requiring PAP. Furthermore, not all institutions will be able to provide the intense level of services required for this in-patient protocol. Because there have been relatively few reported studies detailing desensitization protocols, it is difficult to compare this study to others with regard to efficacy of techniques and efficiency with regard to cost and personnel usage. Although smart card downloads were used in this study, future studies may benefit from use of other objective measures of daily functioning, such as emotional and behavioral rating scales, which may be affected by PAP adherence. While these 2 cases demonstrate the potential effectiveness of this in-patient program for some patients, further research should be conducted to further evaluate the effectiveness and feasibility of similar programs.

Even with these limitations, these case studies do suggest that there may be an important role for multidisciplinary teams in adherence to PAP, and that these approaches should be encouraged and further researched. In addition, as discussed, the current adherence criteria of greater than 4 hours of use nightly may not accurately reflect the sleep needs of pediatric patients. Future research is indicated to determine appropriate criteria that can be used to assess adherence in future research. As additional approaches are published, comparisons of these programs with regard to effectiveness and efficiency would be an important step to developing a gold standard for PAP desensitization that can be widely used and perhaps assist in securing appropriate reimbursement for these multidisciplinary teams.

REFERENCES

1. Schechter MS; Section on Pediatric Pulmonology, Subcommittee on Obstructive Sleep Apnea Syndrome. Technical report: diagnosis and management of childhood obstructive sleep apnea syndrome. Pediatrics 2002;109(4),e69.
2. Brostrom A, Stromberg A, Ulander M, Fridlund B, Martensson J, Svanborg E. Perceived informational needs, side effects and their consequences on adherence: a comparison between CPAP treated patients with OSAS and healthcare personnel. Patient Educ Couns 2002;74(2):228-235.
3. Tsara V, Kaimakamis E, Serasli E, Katsarou Z, Christaki P. Health related quality of life in Greek patients with sleep apnea-hypopnea syndrome treated with continuous positive airway pressure. Sleep Med 2009;10(2):217-225.
4. Hsiao KH, Nixon GM. The effect of treatment of obstructive sleep apnea on quality of life in children with cerebral palsy. Res Developmental Disabil 2008;29(2):133-140.
5. Marcus CL, Rosen G, Davidson Ward SL, Halbower AC, Sterni L, Lutz J, Stading PJ, et al. Adherence to and effectiveness of positive

airway pressure therapy in children with obstructive sleep apnea. Pediatrics 2006;117(3):e442-e451.
6. Weaver TE. Adherence to positive airway pressure therapy. Curr Opin Pulm Med 2006;12(6):409-413.
7. O'Donnell AR, Bjornson CL, Boh SG, Kirk VG. Compliance rates in children using noninvasive continuous positive airway pressure. Sleep 2006;29(5):651-658.
8. Uong EC, Epperson M, Bathon SA, Jeffe DB. Adherence to nasal positive airway pressure therapy among school-aged children and adolescents with obstructive sleep apnea syndrome. Pediatrics 2007; 120(5):e1203-e1211.
9. Koontz KL, Silfer KJ, Cataldo MD, Marcus CL. Improving pediatric compliance with positive airway pressure therapy: the impact of behavioral intervention. Sleep 2003;26(8):1010-1015.
10. Silfer KJ, Kruglak D, Benore E, Bellipanni K, Falk L, Halbower AC, et al. Behavioral training for increasing preschool children's adherence with positive airway pressure: a preliminary study. Behav Sleep Med 2007;5(2):147-175.