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Case Report

Utilization of Positional Therapy for Management of Severe OSA in a Pediatric Patient

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Abstract

We report a longitudinal case of an obese subject presenting at age two (BMI percentile > 99) with symptoms of OSA. Polysomnogram revealed severe OSA (AHI 37.1/hour) with abnormal oxygenation and ventilation. Despite extensive desensitization to PAP, the subject was intolerant to PAP. A tracheostomy was placed due to a failed PAP attempt. Although some children treated with tracheostomy for OSA have medical complexities, this child had no underlying neurologic or craniofacial syndromes. Tracheal de-cannulation occurred following a systematic approach including multiple polysomnograms and PAP desensitization. However, due to inability to resolve OSA with PAP and pressure intolerance, reinsertion of tracheostomy was considered. A second tracheostomy was avoided at age 17 (BMI percentile > 99) by utilizing a commercially available positional device in conjunction with PAP (AHI 3.4/hour, normal oxygenation and ventilation).

Although the utilization of PAP in conjunction with a positional device proved effective in this case, there is limited literature documenting concurrent use, and no current literature describes this practice in children. Further research is needed to determine if the combination of a positional device along with PAP is an effective treatment option in pediatric patients with difficult to treat OSA.

ABBREVIATIONS

ADP: Adherence Program; AHI: Apnea Hypopnea Index; AI: Arousal Index; BPAP: Bi-Level Positive Airway Pressure; CPAP: Continuous Positive Airway Pressure; EPAP: Expiratory Positive Airway Pressure; E_TCO_2: End Tidal Carbon Dioxide; IPAP: Inspiratory Positive Airway Pressure; OSA: Obstructive Sleep Apnea; PAP: Positive Airway Pressure; POSA: Positional Obstructive Sleep Apnea; PSG: Polysomnogram; rAHI: Apnea Hypopnea Index during Rapid Eye Movement; REM: Rapid Eye Movement; SpO_2: Oxygen Saturation Measured by Pulse Oximeter

INTRODUCTION

Obstructive sleep apnea syndrome is defined as complete or partial upper airway obstruction during sleep that is disruptive to normal oxygenation, ventilation, and sleep patterns [1]. Primary snoring can be prevalent in up to 27% of children, with Obstructive Sleep Apnea (OSA) occurring in 1-5% [1,2]. Adeno-tonsillectomy is many times a curative treatment. However, residual OSA is found in some children [1]. While additional surgery options or an oral appliance are sometimes treatment options for this subset of children, the standard treatment of OSA is Positive Airway Pressure (PAP) therapy [1]. PAP therapy can consist of either Continuous Positive Airway Pressure (CPAP) or Bi-Level Positive Airway Pressure (BPAP). While effective, adherence to PAP therapy is difficult in adults and children alike [1,3,4]. For patients with severe OSA, more invasive treatments, including tracheostomy, are sometimes required to successfully resolve OSA when other treatments have been ineffective [5,6]. We are reporting a case of a pediatric patient intolerant to BPAP who successfully avoided a second tracheostomy by utilizing a commercially available positional device in conjunction with BPAP at lower pressures.

CASE REPORT

This is a pediatric case report of a now adult black male with morbid obesity (body mass index 52.9 kg/m^2) and an extensive history of severe OSA. At the age of 2 years and 9 months, obesity (BMI percentile > 99), snoring, snorting, witnessed apneas, and oxygen desaturations were identified during a hospital stay for an asthma exacerbation. CPAP was attempted during sleep, without success, due to pressure intolerance. BPAP was initiated and otolaryngology was consulted. Tonsils were 4+ and adenoids were 80% obstructing. An adeno-tonsillectomy was scheduled, and the subject was discharged home with BPAP settings of Inspiratory Positive Airway Pressure (IPAP) 18 cmH_2O, Expiratory Positive Airway Pressure (EPAP) 6 cmH_2O and a backup respiratory rate 20 breaths per minute. Three months later at 3 years of age,

he underwent an adeno-tonsillectomy (removal of 4+ tonsils and 80% obstructing adenoids) which resulted in resolution of symptoms as reported at the surgical follow up visit, and BPAP was discontinued at that time.

At 4 years of age, the patient returned to the otolaryngology clinic, and it was reported that snoring had returned along with struggling to breathe during sleep, respiratory pauses, restless sleep, and the need to sleep in an upright position. Physical exam noted that he was markedly overweight with boggy nasal turbinates, a large tongue, and a long, bulbous uvula. A Polysomnogram (PSG) was performed at 5 years, 11 months of age. The study had been ordered to consider the possibility of the use of CPAP/BPAP, but the child was very anxious and upset during the hookup. Consequently, it was decided to only intervene with supplemental oxygen if needed. After 360 minutes of total sleep time, supplemental oxygen was added during the remaining 117.6 minutes of sleep. During the study, the child had mild to moderate snoring, demonstrated paradoxical breathing, and gasping. Fifty percent of the time, substernal and suprasternal retractions were noted. Without oxygen, this study revealed an overall Apnea- Hypopnea Index (AHI) of 37.2 per hour, AHI during Rapid Eye Movement Sleep (rAHI) 92.4 per hour, an Arousal Index (AI) 60.3 per hour, lowest oxygen saturation (SpO_2) 57%, and maximum end tidal carbon dioxide (E_TCO_2) 63 mm Hg. With supplemental oxygen, the overall AHI was 65.8 per hour. Almost all events were obstructive with some true apneas recorded. The patient was admitted for inpatient desensitization to BPAP and recommended to follow up in the otolaryngology clinic as quickly as possible to discuss treatment options with continued CPAP/BPAP to be considered as a possibility even with surgical treatment, due to the severity of his OSA. Because of the redundant pharyngeal tissue and flat uvula, an uvulopalatopharyngoplasty with a revision adenoidectomy, tongue-based suspension using a repose bone screw system, and bilateral inferior turbinate trim surgery was performed.

Due to the severity of OSA, continued symptoms, and oxygen desaturations following surgery, he was discharged home on BPAP. Unfortunately, despite being hospitalized to acclimate to PAP in addition to working at home with desensitization to the pressure, his adherence was extremely poor. Eight months following surgery, he attended otolaryngology clinic follow up visit and reported BPAP intolerance, including caregivers needing to replace the mask up to four times per night. Symptoms noted while wearing BPAP included improved but residual snoring, restless sleep, sleeping in an upright position, respiratory pauses, nocturnal enuresis, and frequent arousals. Daytime symptoms included excessive daytime somnolence, poor school performance, behavioral problems, and morning headaches. He was also noted to have acanthosis nigricans, and hypertension. He repeated kindergarten. Discussions were had with the family regarding alternate treatments, specifically tracheostomy since his sleep apnea was severe and he was intolerant to PAP. A plan was made to continue working on desensitization, but also to schedule tracheostomy. If he became tolerant of PAP during the interim, tracheostomy would be deferred.

A split-night PSG was performed. The patient was started on a CPAP pressure of 4cmH_2O, increased up to 10 cmH_2O, and then switched to BPAP up to an IPAP of 16 cmH_2O and EPAP of 12 cmH_2O. Off BPAP, AHI was 124.3 per hour, low SpO2 46%, high E_TCO_2 62 mm Hg, and AI 202 per hour. On BPAP, AHI was 80.7 per hour, low SpO_2 50%, high E_TCO_2 54 mm Hg, and AI 161.8 per hour. OSA events and oxygenation could not be controlled adequately on any BPAP settings studied. The child was noted to pull on the equipment throughout the night and removed the mask multiple times, especially every time the pressure increased. Due to the severity of OSA and inability to tolerate BPAP, a tracheostomy was performed at 6 years, 2 months of age.

At the one month follow up post tracheostomy, the patient's caregivers reported no apneas, no bedwetting, and an improvement in diet and exercise. He had lost 15 pounds since the previous clinic visit. Six months following tracheostomy, a PSG with uncapped tracheostomy was performed. During the study, there was only 1 scorable central apnea, obstructive index was zero, a total AHI 0.1 per hour, low SpO_2 90%, highest E_TCO_2 53 mm Hg and AI 4.0 per hour.

The patient continued to do well with no issues or signs of OSA. At 10 years and 5 months of age, a PSG with tracheostomy capped to evaluate readiness for tracheal de-cannulation was performed. During this PSG, the first 131 minutes of sleep were recorded with the tracheostomy uncapped, and this portion of the study was normal. After 131 minutes of recording, including the first rapid eye movement sleep cycle, the tracheostomy was capped according to protocol procedure. During this portion of the PSG, the AHI was 41.7 per hour, low SpO_2 was 82%, high E_TCO_2 62 mm Hg and AI was 30.1 per hour. It was determined that OSA remained too severe for tracheal de-cannulation at this time.

At age 12 years and 6 months, tracheal de-cannulation was once again considered, and another PSG with the tracheostomy capped was performed as previously described. On the capped portion of the study, there was significant sleep disruption, oxygenation and ventilation were very abnormal and AHI was elevated at 55.1 per hour. It was again determined that the patient was not ready for tracheal de-cannulation. At this point, the patient and caregiver were interested in attempting PAP again. The plan was for him to enroll in the sleep center's positive airway pressure Adherence Program (ADP) to acclimate to CPAP with the tracheostomy open, transition to CPAP while capping the trach, and eventually possible de-cannulation, if tolerated.

The ADP is led by respiratory therapists and sleep behavioral psychologists with support from the sleep providers. Children participating in this program receive an individualized desensitization plan and are encouraged to improve PAP adherence while in the program and beyond program completion. This patient trialed both nasal and full-face interfaces and was introduced to CPAP up to 10cm H_2O with tracheostomy both capped, uncapped and with a Passey Muir valve in place. The patient reported feeling more comfortable with CPAP while

trach was capped. The family was instructed to practice daytime desensitization with pressure and tracheostomy capped during the daytime and caregivers to check on the patient during the night. During this time, he started wearing the BPAP regularly and achieved good adherence with the tracheostomy open and while capped. Eight months after entering the ADP, the tracheostomy was removed; however, once de-cannulated, he continued struggling with BPAP use, possibly due to a tracheo-cutaneous fistula. Total PAP usage was adequate and in the 90+ percent range, however he was unable to achieve adequate pressure due to a large leak. The patient was scheduled for surgery, and a tracheostomy scar revision was performed.

At 16 years of age, PSG was repeated as a PAP titration. CPAP was started at 6 cmH_2O, quickly titrated up to 20 cmH_2O, and then switched to BPAP with IPAP 20 cmH_2O, EPAP 16 cmH_2O. Due to a large leak, an oronasal mask with chin strap was used. He was titrated for events on BPAP as high as IPAP 30 cmH_2O and EPAP 25 cmH_2O. He spent approximately the last half of the study on BPAP settings of IPAP 30 cmH_2O, EPAP 25 cmH_2O. During this time, he had the lowest AHI exhibited on the positional device which was 31 per hour. This study revealed persistence of OSA on all BPAP pressures attempted, with apnea hypopnea indices as high as 242 per hour on lower PAP settings. It was determined that re-insertion of the tracheostomy seemed to be the only option for management of his OSA. Upon closer analysis of the PSG, OSA events appeared to be managed fairly well on BPAP settings of IPAP 30 cmH_2O, EPAP 25 cmH_2O while in a lateral body position, but no adequate pressure could be identified for control of OSA in the supine body position. It was determined that if he would sleep in a lateral body position while using BPAP, OSA events may be controlled. The patient was provided with a commercially available positional device to keep him on his sides while wearing BPAP (Figure 1) with settings of IPAP 30 cmH_2O, EPAP 25 cmH_2O and a full-face PAP interface. This was a commercially available positional device that consisted of a chest belt and an air bladder across the back (Figure 2). The device prevented the patient from lying in the supine body position. A BPAP titration PSG performed with the positional device starting with IPAP 14 cmH_2O, EPAP 14 cmH_2O and titrated to IPAP 22 cmH_2O, EPAP 18 cmH_2O. This study revealed sleep efficiency of 88%. Rapid Eye Movement (REM) sleep was increased at 30% of total sleep time, AI 6.9 per hour, AHI 3.4 per hour, rAHI 3.1 per hour, and the lowest SpO2 was 86%. BPAP was titrated at various pressures. Events were well controlled, and oxygenation was normal on settings of IPAP 20 cmH2O, EPAP 16cmH_2O. The patient was instructed to use these settings and a repeat PSG with BPAP titration would be performed in 6 months using the positional device to determine if BPAP could be titrated to lower pressures (Table 1).

At the follow up PSG, the plan was to start at IPAP 20 cmH_2O, EPAP 16 cmH_2O and decrease pressure until lowest pressure was achieved that resolved obstructive events and normalized oxygenation and ventilation. Sleep was of poor quality with multiple prolonged awakenings during the night of the study, which reduced the sleep efficiency to 68%. There was a total of 364 minutes of sleep. Adequate REM sleep (87 minutes) was studied during the night. He did well on BPAP IPAP 18 cmH2O, EPAP 14 cmH_2O, with good control of OSA (AHI 5.0 per hour), normal oxygenation (mean oximetry 97.2%) and normal ventilation (mean end tidal carbon dioxide 42.3 mm Hg). It was recommended to update to these settings and repeat the PSG with the positional device again in 6 months to one year (Figures 1,2).

At 19 years of age (BMI percentile > 99), the patient transitioned to an adult sleep provider. At the last visit with the adult sleep center, the patient reported he was still using BPAP with the positional device. The most recent results of an in-lab BPAP titration with use of the positional device performed at the adult center included an AHI 134 per hour off BPAP, with pressures titrated to IPAP 17cmH_2O, EPAP13 cmH2O with excellent response during titration. On a BPAP pressure of IPAP 17 cmH_2O, EPAP 13 cmH_2O, he had an AHI of 0 per hour. A six-month adherence report was obtained and showed usage > 4 hours per night at 92.4% and total use 97.2%.

DISCUSSION

Positional Obstructive Sleep Apnea (POSA) is defined as obstructive sleep apnea with a higher number of respiratory events in the supine body position, but specific criteria such as supine to non-supine AHI ratio is debated [7]. In adult patients with difficult to treat POSA where optimal pressures were unable to be achieved during titration, successful management of OSA was obtained by utilizing positional therapy in conjunction with PAP therapy [8]. In children, there is no universally accepted

Figure 1: How the positional device was positioned on the patient during the polysomnogram.

Figure 2: Photograph of the positional device.

Table 1: Summary of polysomnograms performed.

Type of Study	PSG possible intervention		PAP Titration		PSG trach	PSG Trach capped protocol		PSG Trach capped protocol		PAP Titration	PAP Titration	PAP titration
Age	5y		6y		7Y	10y		12y		16y	17y	17y
BMI Percentile	U/A		> 99		> 99	> 99		97		99	> 99	> 99
Height (cm)			121		127.8	140.8		151.6		165.5	165.6	165.6
Weight (kg)			42		49	55.9		60.8		108.2	111.7	111.7
	Off O2	On O2	Off BPAP	On BPAP	uncapped	uncapped	capped	uncapped	capped		**Positional device**	**without positional device**
AHI	37.2	65.8	124.3	80.7	0.1	0.5	41.7	1.2	55.1	56.2	3.4	5.9
rAHI	92.4	119.7	130	NA		NA	74.5	NA	71.2	15	3.1	1.4
Supine AHI	Data U/A	Data U/A	Data U/A	Data U/A	Data U/A	0.5	59.9	0	37.3	71.3	4	2.5
Supine rAHI	Data U/A	Data U/A	Data U/A	NA	Data U/A	NA	Data U/A	NA	64.9	31.1	2.8	0
Arousal Index	60.3	135.2	202	161.8	4	7.1	30.1	8.8	43.3	51.6	6.9	9.2
Mean Oxygen Saturation (%)	87 - 99%	95 - 100%	70 - 98%	80 - 97%	94 - 100%	94 - 98%	93 - 100%	97 - 100%	96 - 100%	95 -99%	97.6-100%	96.7 - 100%
Oxygen Nadir	57%	68%	46%	50%	90%	91%	82%	92%	81%	82%	86%	82%
Percentage of Sleep Time Spent with Oxygen ≤ 85%	3.9%	2.4%	33.3%	15.3%	0%	0%	0.1%	0%	0.4%	0.2%	0%	0%
Percentage of Sleep Time Spent with Oxygen ≤ 90%	3.9%	2.4%	33.3%	15.3%	0%	0%	0.1%	0%	2.6%	6.5%	0.2%	0.9%
Maximum End Tidal Carbon Dioxide (mmHg)	63	64	62	54	52	51	62	53	56.3	55.4	52.7	48.9
Mean End Tidal Carbon Dioxide (mmHg)	42-59	42-59	34-52	32-51	44-47	45-47	48-51	46-50	42.5	35.2-55.4	43.4-52.7	43.5-48.9
Percentage of Time Spent with End Tidal Carbon Dioxide > 50 mmHg	4.4%	23%	4.6%	4%	0.2%	0.3%	45.7%	0.2%	14.6%	0.8%	1.3%	0%

definition of POSA, so prevalence is unclear [7].

Causes of PAP intolerance varies among users, with pressure intolerance being a common complaint. Bhattacharjee, et al., suggest that children less than 6 years of age and adolescents ages 15 to 18 may need additional interventions and support than other ages to optimize PAP adherence [3]. In our report of this case, the subject was initially very young and the OSA was unable to be controlled on any pressure titrated. Prior to tracheostomy, the reported subject was only followed and managed in the otolaryngology clinic rather than the pediatric sleep clinic. During the treatment of this subject, our organization experienced a change in electronic medical record systems, which resulted in some raw data reports being unavailable with only dictated summaries of those reports available after the merge. This is one limitation of the retrospective nature of this case report. However, the authors surmise that positional therapy alone would not have prevented the subject from initial tracheostomy as AHI values noted on the polysomnogram with tracheostomy capped were severely abnormal both in the supine and non-supine body positions. In addition to this, tracheostomy was performed at age six and positional therapy alone may have been difficult in this age. The tracheostomy was performed due to pressure intolerance. Utilization of neither PAP (as suggested by the residual events on PAP titration study), nor the positional device alone (as patient had severe OSA in non-supine body positions that would not have been treated by positional device alone) resolved the OSA in this subject, and concurrent use of both treatments prevented a second tracheostomy. By utilizing a commercially available positional device simultaneously with PAP therapy, BPAP pressures were able to be reduced to a level that was tolerated by the patient, while also successfully treating OSA by preventing supine sleep [9,10].

Use of a commercially available positional device may help management of difficult to treat OSA before proceeding to more invasive management, such as tracheostomy. There is a large amount of literature regarding positional OSA and the use of positional devices for treatment of OSA in adults; however, such literature documenting use of concurrent treatment is limited, with no studies in the pediatric population [4,10,11]. The authors of the current study had limited experience using positional therapy in clinical practice, however due to the severity of OSA in this patient, it was decided to attempt this therapy to prevent another tracheostomy. Goyal, et al. were able to utilize positional therapy in conjunction with PAP to

successfully treat adult patients that previously failed titration in the supine position [8]. In addition, in these patients, OSA was resolved using lower pressures while the patients were kept in the lateral position [8]. OSA in adults is thought to be more severe in the supine position due to multiple factors including effect of gravity, anatomy of the upper airway including the relative size and shape of the tongue, soft palate, lateral pharyngeal fat pads in relation to the surrounding bony structures, as well as upper airway collapsibility, interacting with each other [9]. Other factors like lung volume and the compensatory capacity of the upper dilator muscles are also thought to be important [9]. This could contribute to why patients may require lower pressures in the lateral position, and also why our patient responded well to concomitant use of PAP and a positional device. Adherence to PAP therapy and positional devices independently is poor, so education as to the importance of treating the OSA and explaining more invasive treatments is necessary. Utilization of a positional device concurrently with PAP therapy may be an effective alternative for patients with difficult to treat OSA. Such use may prevent more invasive treatments such as tracheostomy, thereby improving quality of life. While Xiao, et al., reported that a positional device was effective in treating OSA in a small sample of children intolerant to PAP therapy [4], to the authors' knowledge, this is the first report of the utilization of both a positional device in conjunction with PAP therapy to treat OSA in a pediatric patient who had tracheostomy. Additional research including randomized, controlled trials, is needed to determine the validity of this concomitant therapy in pediatric patients with OSA.

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