Official Case Reports Journal of the Asian Pacific Society of Respirology
Respirology Case Reports
OPEN ACCESS
APSR

Nocturnal respiratory failure in a child with congenital myopathy – management using average volume-assured pressure support (AVAPS)

Natalie Gentin[1,2], Bruce Williamson[1,2], Ganesh Thambipillay[1,2] & Arthur Teng[1,2,3]

[1]Department of Sleep Medicine, Sydney Children's Hospital, Sydney, New South Wales, Australia.
[2]School of Women and Children's Health, Faculty of Medicine, University of New South Wales, Sydney, New South Wales, Australia.
[3]School of Medicine, University of Tasmania, Tasmania, Australia.

Keywords
Average volume-assured pressure support, myopathy, nocturnal hypoventilation, non-invasive ventilation, pediatric.

Correspondence
Natalie Gentin, Department of Sleep Medicine, Sydney Children's Hospital,High Street Randwick, NSW 2031, Australia.
E-mail:
Natalie.gentin@sesiahs.health.nsw.gov.au

Received: 11 May 2015; Revised: 12 July 2015; Accepted: 14 July 2015.

Respirology Case Reports **2015; 3(3): 115–117**

doi: 10.1002/rcr2.117

Abstract

This is a case report of the effective use of bi-level positive airway pressure support (BPAP) using the volume-assured pressure support feature in a pediatric patient with a congenital myopathy and significant nocturnal hypoventilation. Our patient was started on nocturnal nasal mask BPAP but required high pressures to improve her oxygen saturations and CO_2 baseline. She was then trialed on a BPAP machine with the volume-assured pressure support feature on. The ability of this machine to adjust inspiratory pressures to give a targeted tidal volume allowed the patient to be on lower pressure settings for periods of the night, with the higher pressures only when required. She tolerated the ventilation well and her saturations, CO_2 profiles, and clinical condition improved. This case report highlights the benefits of the volume-assured pressure support feature on a BPAP machine in a child with a neuromuscular disorder.

Introduction

Volume-assured pressure support is a feature on bi-level positive airway pressure support (BPAP) machines that deliver a consistent preset target volume by automatically adjusting the inspiratory pressure support within a set range. Therefore, varying pressure requirements in different sleep stages can be provided.

We present a case of a patient in whom the volume-assured pressure support feature was effective in managing nocturnal respiratory failure.

Case Report

Our patient is a 3-year-old girl with multiminicore myopathy (a recessively inherited congenital myopathy).

At 7 months old she was noted to have axial weakness, gross motor delay, and poor weight gain. She had frequent lower respiratory tract infections and required antibiotics, oxygen, and chest physiotherapy.

She was referred to the Sydney Children's Hospital (SCH) Neuromuscular Clinic due to her significant weakness and respiratory issues at 12 months old. The diagnosis was confirmed by electromyography and muscle biopsy. She had a weak cough, poor secretion clearance, gastroesophageal reflux, and probable recurrent aspiration pneumonia. She was started on nasogastric feeds and then a percutaneous endoscopic gastrostomy tube was inserted. She was referred to the SCH Department of Sleep Medicine at 24 months old on the basis of her diagnosis and possibility of nocturnal hypoventilation.

Following review, a routine overnight polysomnogram (PSG) was organized and this was carried out at the age of 31 months using the Compumedics Grael system (Melbourne, Australia). Oxygen saturation was <92% for 25% of the study and there were frequent desaturations to a nadir

of 72%. In REM sleep, there were multiple episodes of poor respiratory effort, desaturation, and CO_2 retention ($TcCO_2$ range 50–72 mmHg). The apnea–hypopnea index was 12.2 per hour mainly due to central hypopneas.

BPAP was initiated with a BiPAP Harmony machine (Philips Respironics, Murrysville, PA, USA) with settings of inspiratory positive airway pressure (IPAP) 10 cm H_2O, expiratory positive airway pressure (EPAP) 4 cm H_2O, and respiratory rate (RR) 25 per minute in the spontaneous timed (ST) mode. A Maskmedic Paediatric Concept™ (Maskmedic Pty Ltd, Balmain, NSW, Australia) size 2 nasal mask was used.

A BPAP titration study was carried out with these settings. There were still significant desaturations and CO_2 retention to a maximum $TcCO_2$ of 92 mmHg despite increasing the IPAP and EPAP to 20 and 6 cm H_2O, respectively. The patient triggered all breaths and synchronized well with the machine. There were significant differences in ventilatory requirements in different sleep stages. The final settings were ST mode, IPAP 20 cm H_2O, EPAP 6 cm H_2O, RR 22 per minute, and inspiratory time 1 sec.

Five days later, she had another BPAP titration study with the Philips Respironics Trilogy machine with average volume-assured pressure support (AVAPS) in the ST mode. The targeted tidal volume (TV) was 80 mL (8 mL/kg). The EPAP was fixed at 4 cm H_2O. The minimum IPAP ($IPAP_{min}$) was 11 cm H_2O. The initial maximum IPAP ($IPAP_{max}$) was 15 cm H_2O. There was good synchrony. The $IPAP_{max}$ was increased to achieve the target TV as this was not carried out with an IPAP of 15 cm H_2O. $TcCO_2$ ranged from 54 to 67 mmHg. The lowest oxygen saturation was 86%. In NREM sleep, the IPAP required to reach the TV was 13–15 cm H_2O and in REM sleep 16–17 cm H_2O.

The final settings were EPAP 4 cm H_2O, $IPAP_{min}$ 11 cm H_2O, $IPAP_{max}$ 17 cm H_2O, AVAPS on with target TV of 80 mL, AVAPS rate of change of 2 cm H_2O/minute, RR 22 per minute, and inspiratory time 1 sec (see Table 1).

She was reviewed 2 months later. She was tolerating the airway support well. She had not had further episodes of pneumonia and was more active and alert. Her compliance was 98% with average daily use of 5.5 h. The low usage was most likely due to humidification issues and improved with correction of humidification. Machine download showed that she had varied IPAPs to achieve her preset TV with an $IPAP_{max}$ of 17 cm H_2O at some point each sleep (range was 11–17 cm H_2O; see Fig. 1).

Discussion

Many patients (especially those with neuromuscular disorders) have varying pressure requirements in different stages of sleep. The volume-assured pressure support feature allows the BPAP machine to adjust the IPAP to achieve a set TV. This means that patients can be managed with lower pressures for most of the night but with increased pressures when needed. This then improves comfort, compliance, and ventilation [1, 2].

There have been some small studies looking at volume-assured pressure support in adults. The patient groups were adults with obesity hypoventilation syndrome and chronic obstructive pulmonary disease (COPD). Improved sleep quality and normalization of $TcCO_2$ with volume-assured pressure support was shown, but possibly with a decrease in sleep efficiency [1–3]. The volume-assured pressure support feature has been shown to be beneficial in acute respiratory failure in patients with COPD and hypercapnic encephalopathy [4]. There are almost no studies of volume-assured pressure support in children. There is one case report of a teenager with congenital central hypoventilation

Table 1. PSG parameters and BPAP settings.

	Sleep efficiency (%)	AHI (/h)	Baseline SaO_2 (%)	SaO_2 nadir (%)	CO_2 range (mmHg)	Final settings
Diagnostic study	93	12.1	97	72	54–74	
Conventional BPAP	82	0	99	77	58–92	IPAP: 20 cm H_2O EPAP: 6 cm H_2O Rate: 22 bpm Mode: S/T
BPAP with AVAPS	72.9	0	99	88	50–66	Target TV: 80 mL IPAP min: 8 cm H_2O IPAP max: 17 cm H_2O EPAP: 4 cm H_2O Rate: 20 bpm Mode: S/T with AVAPS

AHI, apnea hypopnea index; AVAPS, average volume-assured pressure support; BPAP, bi-level positive airway pressure support; EPAP, expiratory positive airway pressure; IPAP, inspiratory positive airway pressure; PSG, polysomnogram; TV, tidal volume.

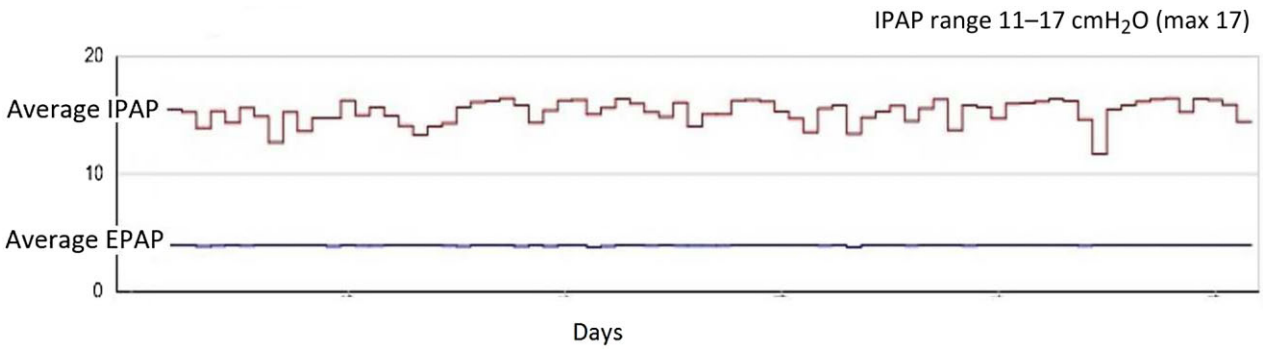

Figure 1. Daily variation in IPAP requirements.

syndrome who was successfully transitioned from ventilation via tracheostomy to non-invasive ventilation – BPAP with AVAPS [5]. In PSGs performed following initiation, she had normal sleep architecture with normal oxygen saturations and $TcCO_2$.

This case highlights the potential benefits of BPAP with volume-assured pressure support such as AVAPS in managing nocturnal hypoventilation in pediatric neuromuscular disorders with possible extension to other conditions requiring nocturnal ventilatory support. As most studies to date have been carried out in adults with different diseases to children, further studies looking at the volume-assured pressure support feature in children is warranted.

Disclosure Statements

No conflict of interest declared.

Appropriate written informed consent was obtained for publication of this case report and accompanying images.

References

1. Storre JH, Seuthe B, Fiechter R, et al. 2006. Average volume-assured pressure support in obesity hypoventilation, a randomized crossover trial. Chest 130:815–821.
2. Murphy PB, Davidson C, Hind MD, et al. 2012. Volume targeted versus pressure support non-invasive ventilation in patients with super obesity and chronic respiratory failure: a randomised controlled trial. Thorax 67:727–734.
3. Crisafulli E, Manni G, Kidonias M, et al. 2009. Subjective sleep quality during Average Volume Assured Pressure Support (AVAPS) ventilation in patients with hypercapnic COPD: a physiological pilot study. Lung 187:299–305.
4. Briones Claudett KH, Briones Claudett M, Chung Sang Wong M, et al. 2013. Noninvasive mechanical ventilation with average volume assured pressure support (AVAPS) in patients with chronic obstructive pulmonary disease and hypercapnic encephalopathy. BMC Pulm. Med. 13:12.
5. Vagiakis E, Koutsourelakis I, Perraki E, et al. 2010. Average volume-assured pressure support in a 16-year-old girl with congenital central hypoventilation syndrome. J. Clin. Sleep Med. 6:609–612.

