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Volume-assured pressure support mode for noninvasive ventilation: can it improve overnight adherence in children with neuromuscular disease?

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Abstract

Purpose Volume-assured pressure support in noninvasive ventilation (VAPS-NIV) is a newer mode providing automatic pressure support adjustment to ensure a constant alveolar ventilation. Previous studies have shown that NIV effectiveness depends on patient adherence and tolerance. The aim of this study was to determine the adherence and efficacy of VAPS-NIV compared to spontaneous-time (S/T) mode in pediatric patients with neuromuscular disease (NMD).

Methods This was a prospective observational study. Children with NMD who utilized NIV at home for ≥ 3 months were recruited from the Long-term ventilation clinic at The Hospital for Sick Children, Toronto, Canada, from July 1, 2015, to July 1, 2019. Baseline characteristics, date of initiation of NIV, and pulmonary function tests were recorded. Polysomnogram (PSG) data and adherence were recorded and analyzed comparing VAPS and S/T modes.

Results Twenty children with NMD (17 male, 85%) were enrolled. The mean (SD) age at initiation of NIV was 11.6 ± 4.6 years. The median (IQR) duration of ventilation was 1.36 (0.80–2.98) years. The mean average daily usage and the median daily usage for VAPS mode and S/T mode were 8.4 ± 1.6 versus 7.2 ± 2.5 h ($p = 0.012$) and 8.6 ± 1.4 versus 7.8 ± 2.1 h ($p = 0.022$), respectively. There was no difference in sleep architecture, gas exchange, or parent proxy report of NIV tolerance between S/T and VAPS modes.

Conclusion VAPS was associated with an improvement in adherence to therapy in children with NMD compared to S/T mode. Longitudinal studies are required to evaluate long-term clinical outcomes using VAPS mode in children with NMD.

Keywords Volume-assured pressure support mode · noninvasive ventilation · pediatric neuromuscular disease · patient adherence

Abbreviations

AASM	American Academy of Sleep Medicine
AVAPS	Average volume-assured pressure support ventilation
Bi-level PAP	Bi-level positive airway pressure
BMI	Body mass index
BUR	Back up rate
CAHI	Central apnea-hypopnea index
CCHS	Congenital central hypoventilation syndrome
COPD	Chronic obstructive pulmonary disease
CSA	Central sleep apnea
DMD	Duchene muscular dystrophy
ECG	Electrocardiogram
EEG	Electroencephalogram
EMG	Electromyogram
EOG	Electrooculogram
FEF_{25-75}	Forced expiratory flow at 25% to 75% of forced vital capacity
FEV_1	Forced expiratory volume in 1 s
FVC	Forced vital capacity
IPAP	Inspiratory positive airway pressure
IQR	Interquartile range
iVAPS	Intelligent volume-assured pressure support

The study was conducted at the Hospital for Sick Children (SickKids) in Toronto, Canada.

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MEP	Maximal expiratory pressure
MIP	Maximal inspiratory pressure
MV	Minute ventilation
NIV	Noninvasive ventilation
NMD	Neuromuscular disease
NREM	Non-rapid eye movement
OAHI	Obstructive apnea-hypopnea index
OHS	Obesity hypoventilation syndrome
OSA	Obstructive sleep apnea
PLM	Periodic limb movement index
PSG	Polysomnogram/polysomnography
REM	Rapid eye movement
SDB	Sleep-disordered breathing
SD	Standard deviation
SpO_2	Oxygen saturation
S/T	Spontaneous-time
$TcCO_2$	Transcutaneous CO_2
TST	Total sleep time
VAPS-NIV	Volume-assured pressure support in noninvasive ventilation
VAS	Visual analogue scales
VT	Tidal volume

Introduction

Sleep-disordered breathing (SDB) is a broad term encompassing abnormalities in respiratory pattern and gas exchange during sleep [1, 2]. Children with neuromuscular disease (NMD) have a high risk of developing SDB. The prevalence of SDB in NMD is estimated to be more than 40%, a 10-fold greater occurrence than in the general population [3]. The failure to achieve adequate ventilation and gas exchange during sleep can lead to sleep disruption and consequent daytime symptoms [3–6].

Noninvasive ventilation (NIV) is frequently prescribed for SDB in individuals with NMD. Spontaneous-time (S/T) mode is one of the more common modes of NIV, which delivers pressurized air via a nasal/facemask to improve minute ventilation (MV) by augmenting the inspired tidal volume on a breath-by-breath basis through application of an inspiratory and expiratory pressure [7]. NIV corrects nocturnal hypoventilation, alleviates associated symptoms, prevents pulmonary exacerbations and improves quality of life [8–11].

However, the use of S/T mode does not adapt to the increased need for respiratory support during rapid eye movement (REM) sleep in patients with NMD as the pressures are fixed over the course of the night [7]. Additionally, it is not able to auto-titrate the degree of respiratory support provided over time with disease progression, which is a common trajectory with many NMDs. Although NIV is an effective treatment option for SDB, adherence rates are suboptimal at approximately 60–70% [12–14]. Non-adherence to NIV results in significant healthcare service utilization such as repeated hospital admissions for pulmonary exacerbations, prolonged intensive care unit stays, repeat polysomnograms (PSG), multiple clinic visits, and telephone calls [4, 15, 16].

Volume-assured pressure support in noninvasive ventilation (VAPS-NIV) is a newer mode of ventilation in which the pressure support is automatically modulated during the course of the night to ensure a constant alveolar ventilation to reflect the changing lung mechanics and patient effort during sleep [17]. A potential advantage of this NIV mode would be the auto-titration of pressures such that the higher pressures that are needed during REM sleep are not administered during non-rapid eye movement (NREM) sleep which may be contributing to poor adherence. In adults, VAPS-NIV has been shown to significantly improve nocturnal hypoventilation [18] and MV [19] in individuals with obesity hypoventilation syndrome (OHS) as well as NMD. It has also been shown to improve adherence as compared to standard NIV in a mixed cohort of patients with NMD [17].

In contrast, there is a dearth of literature for pediatric VAPS with no published studies evaluating pediatric patients with NMD. Therefore, our aim was to (1) determine if VAPS-NIV is an effective treatment mode for pediatric patients with NMD requiring NIV and (2) evaluate if adherence to NIV is improved with VAPS-NIV as compared to standard S/T mode.

Materials and methods

Study design and setting

This was a prospective observational study which was approved by the Research Ethics Board at The Hospital for Sick Children (SickKids), University of Toronto, Canada (REB No 1000049656). The study participants were recruited from the Long-term Ventilation clinic at The Hospital for Sick Children, Toronto, Canada from July 1, 2015 to July 1, 2019.

Study population

Eligibility criteria Inclusion criteria: (1) a diagnosis of NMD; (2) use of NIV at home for ≥ 3 months; (3) using NIV at least 4 h or more a night; (4) body weight ≥ 30 kg (VAPS mode is designed for use with this weight range). Exclusion criteria: (1) intercurrent illness at the time of patient recruitment; (2) failure to consent to the study.

Study procedures

There were three study visits that all occurred during regularly scheduled outpatient clinic visits. Visit 1: Study participant consent, demographic data collection, adherence determined by ventilator data download, subjective tolerance to NIV (S/T

mode), and pulmonary function testing were performed. Visit 2: The patient was initiated on VAPS mode during an overnight PSG. Visit 3: This visit occurred 3 months after the study participant was prescribed VAPS-NIV. During this visit, adherence and subjective tolerance to VAPS-NIV were documented.

Data collection methods

Demographics and medical history

We collected study participant information including age, gender, height, weight, body mass index (BMI) (calculated as weight (kg)/height $(m)^2$), primary diagnosis, comorbidities, date of initiation of NIV, and ambulatory status.

Adherence to NIV

We reported adherence to S/T mode or VAPS mode using data downloads for the previous 3 months of NIV use. At study enrollment, the download for the previous 3 months while using S/T mode was obtained. We also obtained the data download for the 3 months after changing to VAPS mode. We reported the number of used days ≥ 4 h, percentage of used days ≥ 4 h, total days used, average daily usage (hours), median daily usage (hours), median leak (L/min), % spontaneous triggered breath, median respiratory rate (/min), median tidal volume (VT) (ml/kg), and median MV (L/min) using the ResScan™ Smart Card technology (ResMed, Australia).

Tolerance of NIV

We used a 10-cm visual analogue scale (VAS) (score from 0 to 10 for each question (0 = negative and 10 = positive) for each of 3 questions related to comfort of breath, ease of falling asleep, and ease of ventilator use in both child self-report and parental proxy report (Appendix 1).

Respiratory assessment

Study participants also completed spirometry (VIASYS, Cardinal Health) to determine the forced vital capacity (FVC) and respiratory muscle strength (VIASYS, Cardinal Health) measurements to evaluate the maximal inspiratory pressure (MIP) and maximal expiratory pressures (MEP).

NIV prescription

The Stellar 150® (ResMed Inc., Sydney, Australia) was used for both S/T mode and VAPS mode for all patients. This NIV device was previously provided to all patients by a centralized Ventilator Equipment Pool in Ontario, Canada, as per standard clinical care prior to data collection for the study. NIV settings were titrated to optimize the control of SDB and patient-machine synchrony. The titration studies were performed by two experienced respiratory therapists (DM, AH) that are also trained sleep technologists. The RTs were present throughout the entire night of the PSG. The initial starting settings for the VAPS mode titration studies were informed based on the recommended settings for standard NIV (i.e. S/T mode).

Polysomnogram

All PSGs included 8–10 h of overnight monitoring and NIV was titrated to control the patient's SDB, optimize gas exchange and determine optimal settings for home use. The PSGs were conducted and analyzed in accordance with the American Academy of Sleep Medicine (AASM) guidelines of sleep and associated events with a computer software system [20]. All PSGs were interpreted by two study authors (RA, SA).

The computer software system used for the PSG was Natus (Natus Medical Incorporation, San Carlos, CA, USA). Overnight PSG montages consist of a 6-lead electroencephalogram (EEG) (C3, C4, F3, F4, O1, and O2), two bilateral electrooculogram (EOG) leads referenced to A1 or A2, one submental and two tibial electromyograms (EMG), and one electrocardiogram (ECG) lead. Respiratory measurements were evaluated by chest wall and abdominal movement using inductance pneumography and flow signals using the ResMed VPAP Tx system, oxygen saturation (SpO_2) and heart rate using a Masimo pulse oximeter (Irvine, CA), and transcutaneous carbon dioxide measurement ($TcCO_2$) using a Sentec carbon dioxide sensor (Therwil, Switzerland). Video and audio recordings were recorded. Sleep architecture was assessed by standard techniques. PSG information included sleep onset latency, REM onset latency, total sleep time (TST), sleep efficiency, time spent in each sleep stage (minutes and percentage), and number and classification of arousals and snoring.

All respiratory events were scored in accordance with the AASM pediatric scoring rules for all children younger than 18 years of age. Recorded respiratory data included counts and indices of the following events: obstructive apneas, obstructive hypopneas, central apneas, central hypopneas and mixed apneas during sleep. An obstructive apnea event was defined as a reduction in airflow more than 90% from baseline for at least 90% of the entire respiratory event with chest and/or abdominal movement throughout the entire event, for a duration of at least 2 baseline breaths. Obstructive apnea-hypopnea index (OAHI) was defined as the number of obstructive apnea, mixed apnea and hypopnea episodes/hour during sleep. Hypopneas were defined as decrease in airflow of at least 30% from baseline for a duration of at least 2 respiratory breaths, associated with a minimum 3% desaturation, arousal or

awakening. A mixed apnea was defined as a reduction in airflow more than 90% from baseline for at least 90% of the entire respiratory event, for minimum duration of 2 baseline respiratory breaths, and associated with absent inspiratory effort in the initial portion of the event, followed by resumption of inspiratory effort before the end of the event. The AASM pediatric obstructive sleep apnea (OSA) severity scoring criteria was used for children: mild OSA = OAHI ≥ 1.5 to < 5/h; moderate OSA = OAHI ≥ 5 to < 10/h; and severe OSA = OAHI ≥ 10/h. A central apnea was defined as the absence of chest and/or abdominal movement related to an absence in airflow more than 20 s, or lasting more than 2 baseline respiratory cycles if it was associated with an arousal, an awakening or oxygen desaturation of at least 3%. Significant central sleep apnea (CSA) was defined as a central apnea-hypopnea index (CAHI) of at least 5 or more events/hour of sleep. Nocturnal hypoventilation was referred to $TcCO_2$ level greater than 50 mmHg for equal to or greater than 25% of TST [20].

Statistical analysis

The study participants' data was summarized using descriptive statistics. Baseline characteristics, PSG statistics, adherence, tolerance and pulmonary function data were reported as mean (standard deviation; SD) for normally distributed continuous variables and as median (interquartile range; IQR) and frequency (percent) for skewed continuous variables and categorical variables, respectively. Normally distributed, continuous variables were compared using Student's *t* tests and skewed continuous variables were compared using Wilcoxon pair signed-rank test. A *p* value less than 0.05 indicated statistical significance. Data analysis was carried out using IBM Statistical Package for the Social Sciences (SPSS) for Windows, Version 23.0 (IBM Corp, Armonk, NY).

Results

We enrolled 23 children with NMD. We excluded 3 patients. One patient passed away after visit 1. A second patient did not meet the inclusion criteria as he had non-adherence to NIV (S/T mode). The third patient could not tolerate VAPS mode during the PSG and the PSG was not successful. Twenty children with NMD were included in this study. Patient demographics and clinical characteristics are summarized in Table 1. There were 17 (85%) males. The mean (SD) age of NMD children was 14.1 ± 3.4 years old. The median (IQR) duration of ventilation use was 1.36 (0.80–2.98) years. The study participants started to use NIV with S/T mode and VAPS mode at mean (SD) age 11.6 ± 4.6 and 14.4 ± 3.3 years old, respectively. The most common NMD diagnosis among the study participants was Duchenne muscular dystrophy (DMD) (60%).

The comparison of PSG data for S/T mode versus VAPS mode are presented in Table 2. We found that there were no significant differences in sleep architecture and sleep quality between the two modes.

We compared the 90-day data downloads for the cohort when using S/T mode and VAPS mode (see Table 3). The mean (SD) average daily usage and the median daily usage for VAPS mode and S/T mode were 8.4 ± 1.6 versus 7.2 ± 2.5 h (p = 0.012) and 8.6 ± 1.4 versus 7.8 ± 2.1 h (p = 0.022), respectively. The median (IQR) of percentage of usage ≥ 4 h in VAPS mode and S/T mode was 100.0 (89.5–100.0) versus 87.0 (60.8–97.5) % (p = 0.011), respectively. There was no difference in median leak, % spontaneous triggered breaths, median respiratory rate, median VT or median MV between the two modes of NIV.

We also evaluated the patient and proxy reported tolerance by using a 10-cm VAS for comfort of breath, ease of falling asleep, and ease of ventilator use between use of the two modes of NIV (see Table 4). There were no reported differences in all tolerance data.

Discussion

This is the first pediatric study to report on the use of VAPS mode versus conventional S/T mode in a cohort of children with NMD. Our study demonstrated that children with NMD on nocturnal ventilatory support with VAPS-NIV mode had better adherence to therapy as compared to when they were using S/T mode, which was evidenced by the statistically significant increase in mean and median daily usage hours and median percentage of usage ≥ 4 h. We also found that there were no differences in sleep architecture and parent proxy report of tolerance data when using S/T versus VAPS mode.

Children with NMD are at risk of SDB [2, 21]. Upper airway muscle weakness increases the passive upper airway collapsibility causing obstruction, while reduced intercostal and diaphragm strength can contribute to instability in the control of breathing as well as shallow breathing resulting in central events [16, 21, 22]. The reduced muscle strength and ongoing progression of muscle weakness as well as other potential factors including scoliosis and obesity results in inadequate MV during sleep and subsequent nocturnal hypoventilation [4, 16, 21].

NIV is the most commonly prescribed treatment for children with NMD and SDB. NIV has become the therapeutic standard for patients with NMD because of the demonstrated benefits on survival, sleep quality and quality of life [8, 9, 23–25]. NIV S/T mode is flow triggered and the patient cycles through a pre-set inspiratory and expiratory pressures. VAPS

Table 1 Demographic and clinical characteristics of the study population

Demographic and clinical characteristics		Neuromuscular patients (n = 20)
Age (years)		14.1 ± 3.4[a]
Male (*n* (%))		17 (85)
Weight (kg)		43.90 ± 16.44[a]
Height (cm)		138.45 ± 17.29[a]
Body mass index percentile		85.5 (64.00–94.75)[b]
Body mass index Z-score		1.08 (0.36–1.61)[b]
Duration of ventilation (years)		1.36 (0.80–2.98)[b]
Age at the time of bi-level PAP S/T mode initiation (years)		11.64 ± 4.57[a]
Age at the time of bi-level PAP VAPS mode initiation (years)		14.37 ± 3.29[a]
Diagnosis	Duchene muscular dystrophy (*n* (%))	12 (60)
	Spinal muscular atrophy type 2 (*n* (%))	2 (10)
	Myotonic dystrophy (*n* (%))	2 (10)
	Congenital muscular dystrophy (*n* (%))	1 (5)
	SCN4A myopathy (*n* (%))	1 (5)
	SEPN1 myopathy (*n* (%))	1 (5)
	Ullrich congenital muscular dystrophy (*n* (%))	1 (5)
Comorbidities	Osteoporosis or osteopenia (*n* (%))	8 (40)
	Scoliosis (*n* (%))	7 (35)
	Developmental delay (*n* (%))	3 (15)
	Cardiomyopathy (*n* (%))	3 (15)
Pulmonary function test	FEV_1 (%)	59.8 ± 33.1
	FEV_1 (L)	1.3 ± 0.7
	FVC (%)	58.4 ± 31.5
	FVC (L)	1.5 ± 0.8
	FEV_1/FVC%	87.2 ± 9.6
	FEF_{25-75} (%)	62.6 ± 39.2
	FEF_{25-75} (L)	1.8 ± 0.9
	MIP (cmH_2O)	39.9 ± 13.7
	MEP (cmH_2O)	36.5 ± 11.3

[a] Mean (SD)

[b] Median (IQR)

cm, centimeter; *kg*, kilograms; *m*, meter; *NIV*, noninvasive ventilation; *S/T mode*, spontaneous-time mode; *VAPS mode*, volume-assured pressured support mode

mode is a new hybrid (both volume- and pressure-presets) mode of NIV in which the pressure support is automatically adjusted during sleep to guarantee a constant alveolar ventilation [17].

Despite the many advantages of NIV use identified, both short- and long-term adherence often requires considerable ongoing healthcare provider engagement [26]. One published study conducted by Kelly and colleagues demonstrated an improvement in adherence to therapy with VAPS mode in adult patients with chronic obstructive lung disease (COPD), OHS, chest wall disorders, and muscle weakness. Their cohort of patients were randomized to VAPS or standard ventilation first and then received the crossover treatment. VAPS-NIV was shown to improve adherence as compared to standard NIV with a median (IQR) hours of usage (5:40 (4:42–6:49) versus 4:20 (2:27–6:17) hh:mm/night, $p = 0.004$) [17]. The improvement in usage hours found in Kelly and colleagues' study mirrors the findings in our pediatric study.

To date, there are no published studies evaluating VAPS in comparison to S/T mode in children with NMD. In a pediatric cohort of children with Congenital Central Hypoventilation Syndrome (CCHS), NREM peak $TcCO_2$ has been shown to be lower in VAPS mode, median (IQR) (43.0 (40.0–46.0) versus 46.5 (45.0–48.0) mmHg, $p < 0.05$) [27]. Furthermore, there is only one case report of initiation of VAPS-NIV in a 3-year-old girl with congenital myopathy who was successfully transitioned from conventional Bi-level positive airway pressure (Bi-level PAP) to Average volume-assured pressure support ventilation (AVAPS) mode. She was tolerating the device well and her compliance was 98% with average daily usage of 5.5 h [28].

Table 2 PSG characteristics for NIV S/T mode versus VAPS mode

PSG variables	S/T mode (n = 20)	VAPS mode (n = 20)	p value
Total sleep time (minutes)	364.4 ± 77.4	360.5 ± 57.3	0.836
Sleep efficiency (%)	81.8 ± 13.0	83.5 ± 11.1	0.615
Total sleep time in REM (%)	15.2 ± 6.9	14.9 ± 4.9	0.850
Arousal index (event/h)	13.8 ± 5.5	13.1 ± 7.3	0.542
OAHI (event/h)	2.9 (0.0–4.1)[a]	0.8 (0.0–2.7)[a]	0.234
CAHI (event/h)	1.0 (0.0–2.3)[a]	0.7 (0.0–1.9)[a]	0.794
PLM index (event/h)	0.0 (0.0–9.1)[a]	0.0 (0.0–5.4)[a]	0.508
Desaturation index (event/h)	3.2 (1.6–7.8)[a]	2.3 (0.6–7.1)[a]	0.825
Nadir SpO_2 (%)	88.2 ± 5.9	90.2 ± 5.6	0.329
Mean SpO_2 (%)	97.7 ± 0.82	97.5 ± 1.2	0.513
NREM mean $TcCO_2$ (mmHg)	39.3 (36.5–44.5)[a]	38.1 (36.5–41.1)[a]	0.212
NREM max $TcCO_2$ (mmHg)	44.7 (40.8–50.9)[a]	43.9 (41.5–46.7)[a]	0.444
REM mean $TcCO_2$ (mmHg)	38.4 (36.4–46.2)[a]	39.1 (37.4–42.7)[a]	0.156
REM max $TcCO_2$ (mmHg)	41.3 (39.4–49.4)[a]	42.3 (39.6–46.4)[a]	0.305
% total sleep time with $TcCO_2 \geq 50$ mmHg (%)	0.0 (0.0–0.3)[a]	0.0 (0.0–0.0)[a]	0.237

All data were reported in mean (SD) for continuous variable

[a] Median (IQR)

CAHI, central apnea-hypopnea index; *VAPS mode*, volume-assured pressured support mode; *NREM*, non-rapid eye movement; *OAHI*, obstructive apnea-hypopnea index; *PLM*, periodic limb movement; *PSG*, polysomnography; *REM*, rapid eye movement; *SpO_2*, oxygen saturation; *S/T mode*, spontaneous-time mode; *$TcCO_2$*, transcutaneous carbon dioxide

We demonstrated no significant difference in sleep characteristics, VT, MV and tolerance data between S/T and VAPS mode which is similar to the findings in the few adult studies with heterogenous populations that exist. Ekkernkamp and colleagues performed a randomized, open-label, two-treatment, two-period, crossover study in 14 adults with COPD receiving NIV to treat chronic hypercapnic respiratory failure. They compared high intensity S/T mode providing high inspiratory positive airway pressure (IPAP) and back up rate (BUR) with intelligent volume-assured pressure support (iVAPS) mode [29]. They did not find any significant differences in sleep characteristics between iVAPS and high intensity S/T mode. Another study by the same primary author studied 27 patients with COPD requiring NIV. There was no difference in MV between high intensity S/T mode and iVAPS ($p = 0.25$) [30]. Ambrogio et al. evaluated 28 adults with chronic respiratory insufficiency and also found no differences in PSG metrics or arterial blood gases between conventional pressure support and AVAPS therapy[19].

There are some notable limitations in our study. Firstly, VAPS mode was limited to children with a body weight ≥

Table 3 Adherence data for NIV S/T mode versus VAPS mode

Download variables	S/T mode (n = 20)	VAPS mode (n = 20)	p value
Percentage of days with ≥ 4 h usage (%)	87.0 (60.8–97.5)[a]	100.0 (89.5–100.0)[a]	0.011[b]
Average daily usage (hour)	7.2 ± 2.5	8.4 ± 1.6	0.012[b]
Median daily usage (hour)	7.8 ± 2.1	8.6 ± 1.4	0.022[b]
Median leak (L/min)	20.7 ± 21.7	17.2 ± 10.3	0.444
% spontaneous triggered breaths	9.0 (5.0–20.0)[a]	17.0 (5.7–25.5)[a]	0.256
Median respiratory rate (/min)	18.0 ± 2.6	18.4 ± 2.6	0.270
Median tidal volume (ml/kg)	8.1 ± 3.2	7.6 ± 2.3	0.352
Median minute ventilation (L/min)	5.9 ± 2.3	5.4 ± 1.4	0.186

All data were reported in mean (SD) for continuous variable

[a] Median (IQR)

[b] Statistical significance $p < 0.05$

VAPS mode, volume-assured pressured support mode; *kg*, kilograms; *L*, liter; *ml*, milliliter; *S/T mode*, spontaneous-time mode

Table 4 Tolerance data for NIV S/T mode versus VAPS mode

Tolerance data of NIV		S/T mode (n = 20)	VAPS mode (n = 20)	p value
Child self-report	Comfort of breath	3.0 ± 2.3[a]	2.1 ± 1.7[a]	0.383
	Ease of falling asleep	3.2 ± 1.7[a]	2.1 ± 1.4[a]	0.095
	Ease of ventilator use	7.0 ± 3.6[a]	6.9 ± 4.2[a]	0.933
Parents	Comfort of breath	2.9 ± 2.2	1.6 ± 1.1	0.109
	Ease of falling asleep	2.4 ± 1.6	2.2 ± 1.8	0.770
	Ease of ventilator use	7.3 ± 3.4	7.9 ± 3.6	0.604

All data were reported in mean (SD) for continuous variable

[a] There were 13 children with NMD reporting the tolerance data for NIV

VAPS mode, volume-assured pressured support mode; *NIV*, noninvasive ventilation; *S/T mode*, spontaneous-time mode

30 kg. Therefore, our results are not generalizable to younger children with NMD. Secondly, our results might be biased as we excluded one patient that could not tolerate VAPS mode and another patient that was not adherent to NIV. Lastly, all the children in our cohort used NIV (S/T mode) first for 3 months followed by VAPS mode for 3 months. A subsequent randomized controlled trial with longer term follow up would be advantageous to further investigate our initial findings.

Conclusions

In conclusion, VAPS mode improved adherence in children with NMD compared to the traditional S/T mode of NIV. In addition, our results suggest that it is equally efficacious as traditional S/T mode. Therefore, it may be useful for clinicians to consider VAPS mode as a therapeutic option when prescribing NIV to further increase adherence while ensuring maintenance of optimal respiratory support.

Supplementary Information The online version contains supplementary material available at https://doi.org/10.1007/s11325-021-02288-1.

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Authors' contributions All authors contributed to the study conception and design. Material preparation, data collection, and analysis were performed by Kanokkarn Sunkonkit (KS), Munazzah Ambreen (MA), Cora Mocanu (CM), Adam Qazi (AQ), and Reshma Amin (RA). The first draft of the manuscript was written by KS and RA and all authors commented on previous versions of the manuscript. All authors read and approved the final manuscript.

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Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

Ethics approval This study was approved by the SickKids Research Ethics Board (REB No 1000049656).

Consent to participate Informed consent was obtained from legal guardians or the parents.

Consent for publication The legal guardians or the parents have consented to the submission of the data to the journal.

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