Home Noninvasive Ventilation in Pediatric Patients With Neuromuscular Diseases: One Size Fits All

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BACKGROUND: Home noninvasive ventilation (NIV) improves disease courses of patients with respiratory insufficiency due to neuromuscular diseases. Data about appropriate ventilator settings for pediatric patients are missing. METHODS: In this retrospective study, ventilator settings of 128 subjects with neuromuscular disease aged 0–17 y with NIV were compared between 4 age groups (< 1 y, 0–5 y, 6–11 y, and 12–17 y). Additionally, correlations of ventilator settings with age and vital capacity were investigated in an ungrouped approach. RESULTS: Ventilator backup rate decreased significantly with age, leading to significant backup rate differences between all groups except the oldest two. Median (interquartile range) backup rates were 36 (11.5), 24 (4), 20 (4), and 20 (3) breaths/min in groups 1–4, respectively. Median [IQR] expiratory positive airway pressures (4 [0.5], 4 [0], 4 [0], 4 [1] cm H_2O, respectively) and median [IQR] inspiratory positive airway pressures (12 [1.5], 12 [5], 12 [2.3], and 14 [4] cm H_2O, respectively) showed no significant differences. However, correlation analyses indicated an increase of inspiratory positive airway pressure with age and decreasing FVC, as well as an increase of backup rates with decreasing FVC. CONCLUSIONS: Similar NIV settings fit all age groups of pediatric subjects with neuromuscular disease. Thus, we propose an expiratory positive airway pressure of 4–5 cm H_2O, an inspiratory pressure delta of 8–10 cm H_2O, and an age-oriented backup rate as a starting point for NIV titration. Patients with advanced disease stages might require slightly higher inspiratory positive airway pressures and backup rates. *Key words: chronic respiratory insufficiency; Duchenne muscular dystrophy; spinal muscular atrophy; home ventilation; neuromuscular disease.* [Respir Care 0;0(0):1–●. © 0 Daedalus Enterprises]

Introduction

Neuromuscular diseases (NMD), such as Duchenne muscular dystrophy (DMD) and spinal muscular atrophy (SMA), are a group of mostly genetic diseases leading to progressive muscle weakness. Most of these diseases manifest during infancy, and, despite heterogeneity in the underlying pathogenesis, involvement of the respiratory system is similar.

Respiratory muscle weakness reduces FVC and leads to respiratory insufficiency regardless of a primarily healthy lung, although diaphragmatic and accessory ventilatory muscle involvement varies in different NMDs.[1-3] Initially, respiratory insufficiency occurs during sleep and slowly progresses to diurnal respiratory failure in the later course of the disease. Chest wall deformities (especially scoliosis) and increasing rigidity due to stiffness of muscles and connective tissue contribute to the progression of respiratory insufficiency.[4,5] Furthermore, ventilation inhomogeneity,

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cough insufficiency with chronic mucus retention, and in some cases episodes of aspiration cause infection and inflammation in the lower airways, damaging bronchial and pulmonary tissue and leading to a secondary structural lung disease in advanced disease stages.[6-8] Noninvasive home ventilation (NIV) is the treatment of choice for patients with NMD and respiratory insufficiency, and it vastly improves their morbidity and mortality as well as their quality of life.[9-11] Despite the widespread use of NIV in patients with NMD, the choice of ventilator settings is currently not well supported by scientific evidence, especially for pediatric patients.[12] Therefore, the quality of NIV in patients with NMD almost completely relies on the local experience of the establishing center. With this investigation of our tertiary care referral center cohort of home-ventilated pediatric patients with NMD, we aim to provide information on adequate ventilator settings in these patients.

Methods

This study was a single-center, cross-sectional, retrospective analysis performed in the Department of Pediatric Pulmonology and Sleep Medicine, Children's Hospital, University of Duisburg-Essen, in Essen, Germany. Local patient registries of patients attending our pediatric outpatient department for home ventilation from 2004 to 2018 were screened for patients up to 17 y of age with NIV due to respiratory insufficiency caused by NMD. Older patients, patients with other causes of respiratory insufficiency (eg, bronchopulmonary dysplasia, interstitial lung disease, airway anomalies) and patients receiving invasive ventilation (via tracheostomy) were not included in the analyses. Datasets from eligible subjects were extracted from the registries and the subjects' medical records. Ventilator settings (eg, mode, pressure levels, backup rate, interface) were collected from routine inpatient visits (mostly for sleep studies) in clinically stable situations as determined by the study team (eg, no signs of respiratory tract infection, no recent acute respiratory failure or failure to wean from invasive ventilation). Subjects with no or insufficiently documented ventilator settings were excluded from further analyses. Ventilator pressure levels and backup rates of included subjects were compared to age and FVC in spirometry from the same time point. FVC percent of predicted values were uniformly (re-)calculated according to reference values from the Global Lung Function Initiative 2012.[13] When available, data from respiratory polygraphy (ie, S_{pO_2} mean, S_{pO_2} min, and apnea-hypopnea index) and capillary blood gas analyses (HCO_3- and capillary P_{CO_2}) under the respective reported ventilator settings were included to provide data on sufficiency of the ventilator settings. Polygraphies were recorded with Embletta Gold (Embla Systems, Broomfield, Colorado) or SOMNOtouch RESP recorder

QUICK LOOK

Current knowledge

Noninvasive home ventilation improves disease courses of patients with respiratory insufficiency due to neuromuscular diseases. Data about appropriate ventilator settings for pediatric patients are lacking.

What this paper contributes to our knowledge

We present a retrospective analysis of ventilation parameters of 128 home-ventilated subjects with neuromuscular diseases. All subjects received pressure-limited ventilation, and similar pressure levels were found to fit all age groups.

(SOMNOmedics, Randersacker, Germany) and analyzed with Embla RemLogic (Embla Systems) and Domino (SOMNOmedics) analysis software, respectively. Standard polygraphy at our center includes analysis of nasal flow, snore sound, S_{pO_2}, and respiratory inductance plethysmography of thorax and abdomen. Continuous capnometry is not usually performed for routine ventilation check-ups. There were no extra analyses of polygraphy for this study. Two statistical approaches were taken for data analysis: group-based comparison after stratification of all subjects by age into 4 groups (group 1: < 1 y; group 2: 1–5 y; group 3: 6–11 y; group 4: 12–17 y), and collective (non-grouped) correlation analyses. Non-parametric tests were chosen because the datasets showed no clear normal distribution: the Mann-Whitney U test was used for pairwise group comparisons; the Kruskal-Wallis H test was used for determination of group dependence; the Spearman correlation was used for correlation analyses. A P value < .05 was considered statistically significant. This study was approved by the local ethics committee of the University of Duisburg-Essen. Subjects' or legal guardians' consent was not sought for this retrospective data analysis.

Results

Local registries contained 600 patients in total; 472 patients were excluded from analyses (81 due to invasive ventilation via tracheostomy, 74 due to age > 17 y, 113 due to causes of home ventilation other than NMD, 56 due to absence of mechanical ventilation, 148 due to missing information on ventilation parameters). A total of 128 subjects were included in the analyses: 41 with SMA (16 with Type 1, 24 with Type 2, and 1 with Type 3), 27 with DMD, 25 with congenital myopathies, 19 with congenital muscular dystrophies, 8 with other NMD, and 8 with unspecified NMD (for a detailed list of diagnoses, see Table 1). All subjects received pressure-limited ventilation modes. Subject

Table 1. Distribution of Neuromuscular Diseases in the Study Cohort

Neuromuscular Diseases	Subjects, n
Total	128
Duchenne muscular dystrophy	27
Spinal muscular atrophy type 2	24
Spinal muscular atrophy type 1	16
Unspecified congenital muscular dystrophy	13
Unspecified congenital myopathy	8
Unspecified neuromuscular disease	8
Nemaline myopathy	5
Collagen type VI-related myopathy (Ullrich)	4
Congenital myasthenic syndrome	4
Centronuclear myopathy	3
Merosin-deficient congenital muscular dystrophy	3
Rigid spine muscular dystrophy	3
Spinal muscular atrophy type 3	1
BAG3-positive myopathy	1
Myotonic dystrophy	1
Selenoprotein-associated myopathy	1
Central core myopathy	1
Congenital muscular dystrophy type 1c	1
Minicore-multicore myopathy	1
Pompe's disease	1
Mucolipidosis	1
Cross-linked myotubular myopathy	1

study datasets are summarized in Table 2. Spirometry (FVC) data were available for 88 subjects. Respiratory polygraphy data were available for 88 subjects, with S_{pO_2} mean and S_{pO_2} min being available for all 88 subjects and apnea-hypopnea index available for 50 subjects. Capillary blood gas analyses were available for 98 subjects, with pH and capillary P_{CO_2} being available for all 98 subjects and HCO_3- for 65 subjects. The ventilation parameters of the 4 defined age groups are plotted in Figure 1. Median (interquartile range [IQR]) values of the backup rate were 36 (11.5), 24 (4), 20 (4), and 20 (3) breaths/min for groups 1, 2, 3, and 4, respectively. Backup rate differed significantly between all groups except groups 3 and 4 (Table 2, Fig. 1). Inspiratory positive airway pressure (IPAP) was 12 (1.5), 12 (5), 12 (2.3), and 14 (4) cm H_2O, and expiratory positive airway pressure (EPAP) was 4 (0.5), 4 (0), 4 (0), and 4 (1) cm H_2O for groups 1, 2, 3, and 4, respectively. There were no significant differences between either group for EPAP and IPAP (Table 2, Fig. 1). These findings were confirmed with the Kruskal-Wallis H test, which showed group dependence for backup rate ($P < .001$) but not for EPAP ($P = .39$) and IPAP ($P = .11$). However, the ungrouped correlation analyses showed a positive correlation between age and IPAP ($r = 0.242$, $P = .006$) (Fig. 2A). Negative correlations were found for age as compared to backup rate ($r = -0.432$, $P < .001$) (Fig. 2A) and for FVC percent of predicted as compared to backup rate ($r = -0.324$, $P = .002$) and IPAP ($r = -0.313$, $P = .003$), respectively (Fig.

2B). FVC negatively correlated with age ($r = -302$, $P = .002$) (data not shown). Available datasets (median [IQR]) for S_{pO_2} mean (96.6% [2.8]), S_{pO_2} min (90% [8]), apnea-hypopnea index (0.8 [3.4]), pH (7.39 [0.04]), HCO_3- (24.8 mmol/L [2.5]), and P_{CO_2} (40.3 mm Hg [7]) were indicative of adequate ventilator settings (Table 2). All subjects received pressure control ventilation modes. Five had a predefined target tidal volume, and 6 subjects had a predefined alveolar target volume. NIV interfaces were either oronasal masks ($n = 68$) or nasal masks ($n = 60$). Nasal masks were used significantly more often for subjects with SMA (27 of 41, 65.9%) than for subjects with DMD (10 of 27, 37.0%) ($P = .02$ with the chi-square test).

Discussion

In this retrospective analysis of 128 pediatric subjects with NMD receiving NIV in one of the largest specialized German centers, we have noted that all age groups require similar pressure levels on NIV. The exclusive use of pressure-limited ventilation modes in our cohort does not necessarily reflect a universal approach, as other centers might have good experiences with volume control ventilation modes in patients with NMD. In a small, short-term study of 13 adult subjects with different NMDs, pressure control, pressure support, and volume control ventilation modes have been shown to have similar effects on alveolar ventilation and unloading of respiratory muscles,[14] but the lack of large prospective studies comparing the different home ventilation strategies for patients with NMDs leaves a crucial evidence gap, especially for children.[15]

It is our understanding that home ventilation for patients with NMDs is supposed to maintain physiological pulmonary gas exchange and provide muscular relief by minimizing a patient's breathing efforts. This includes minimizing triggered breaths, because triggering ventilator breaths requires respiratory effort. Therefore, we use pressure-limited ventilation modes with the backup rate titrated to a (physiological) level just above the patient's breathing frequency so that EPAP and IPAP minimize muscular efforts for each breath and prevent the development of atelectasis.[12,16] We occasionally add tidal or alveolar volume targets to cover short episodes of sleep stage-dependent hypopnea/hypoventilation, such as that due to upper-airway obstruction or (suspected) chest wall rigidity.

In our cohort, stratification was by age and not by NMD because, even though genetic NMDs are a heterogeneous group, involvement of the respiratory tract is relatively similar between the different diseases, characterized by a primarily healthy lung impaired by insufficient ventilation and ineffective mucus clearance due to respiratory muscle weakness.[17] Consistent with the progressive natural course of genetic NMDs, the risk of respiratory insufficiency and

Table 2. Subject Characteristics and Study Datasets for Each Study Group

Parameter	Age Groups				All
	< 1 y	1–5 y	6–11 y	12–17 y	
Subjects, n	7	19	28	74	128
Median age, months	6 (3.5; 3–10)	30 (13; 12–66)	105.5 (29.5; 73–142)	179 (29.8; 144–214)	155.5 (95.5; 3–214)
Neuromuscular disease, n	SMA: 3	SMA: 12	SMA: 10	SMA: 16	SMA: 41
	Other: 4	Other: 7	DMD: 1	DMD: 26	DMD: 27
			Other: 17	Other: 32	Other: 60
Median backup rate, /min	36 (11.5; 22–45)	24 (4; 16–30)	20 (4; 14–30)	20 (3; 14–26)	20 (4; 14–45)
Median EPAP, cm H_2O	4 (0.5; 4–6)	4 (0; 4–6)	4 (0; 3–6)	4 (1; 2–8)	4 (1; 2–8)
Median IPAP, cm H_2O	12 (1.5; 10–17)	12 (5; 8–18)	12 (2.3; 8–20)	14 (4; 7–22)	14 (4; 7–22)
Median S_{pO_2}, %	$n = 5$	$n = 11$	$n = 19$	$n = 53$	$n = 88$
	97 (3; 92–99)	98 (1.1; 94–99)	96.1 (2.6; 92–99)	96.4 (2.5; 91.4–99)	96.6 (2.8; 91.4–99)
Median S_{pO_2} min, %	$n = 5$	$n = 11$	$n = 19$	$n = 53$	$n = 88$
	87 (6; 80–90)	86 (7.5; 82–92)	88 (9; 76–94)	90 (6.5; 57–96)	90 (8; 57–96)
Median apnea-hypopnea index	$n = 1$	$n = 4$	$n = 15$	$n = 30$	$n = 50$
	8.1	3.358 (1.13; 2.4–6)	0.6 (2.2; 0–1.3)	0.7 (2.4; 0–15)	0.8 (3.4; 0–15)
Median FVC, L	NA	$n = 2$	$n = 23$	$n = 63$	$n = 88$
		0.28 (0.1; 0.2–0.3)	0.6 (0.6; 0.2–1.6)	0.7 (0.7; 0.3–2.0)	0.7 (0.71; 0.2–2.0)
Median FVC, % of predicted	NA	$n = 2$	$n = 23$	$n = 63$	$n = 88$
		30 (2; 28–32)	29 (29; 10–68)	23 (21.5; 8–60)	24.5 (23.3; 8–68)
Median pH	$n = 6$	$n = 12$	$n = 23$	$n = 57$	$n = 98$
	7.39 (0.06 7.30–7.49)	7.41 (0.04; 7.31–7.43)	7.39 (0.05; 7.30–7.48)	7.39 (.04; 7.33–7.52)	7.39 (.04; 7.30–7.52)
Median HCO_3-, mmol/L	$n = 4$	$n = 8$	$n = 16$	$n = 37$	$n = 65$
	26 (2.65; 24.7–32.3)	23.9 (1.6; 2.6–27.6)	24.6 (3.2; 21.7–28.5)	24.9 (1.9; 2.8–29)	24.8 (2.5; 2.6–32.3)
Median capillary P_{CO_2}, mm Hg	$n = 6$	$n = 12$	$n = 23$	$n = 57$	$n = 98$
	41.1 (4.65; 3.6–43.1)	43.8 (3.1; 39.5–53.0)	41.1 (9.8; 32.6–51.0)	39.5 (5.1; 31.0–52.8)	40.3 (7; 3.6–53.0)

For age, ventilator settings, respiratory polygraphy parameters, and blood gas analyses, median is given with interquartile range (IQR) and range. When data were not available for all subjects, the number of available datasets is indicated (n).
SMA = spinal muscular atrophy; DMD = Duchenne muscular dystrophy; IPAP = inspiratory positive airway pressure; EPAP = expiratory positive airway pressure; NA = not available

subsequent necessity of ventilation support rises with increasing age during childhood, leading to the uneven age distribution seen in our study cohort. The association of lower FVC percent of predicted highlights the progressive nature of NMDs with higher age in our cohort. In the 4 defined groups in this study, backup rate decreased significantly with age, which we interpret as (intended) mirroring of the physiological decrease of the breathing frequency of a growing child. We did not find any significant differences in the settings for EPAP or IPAP between the groups, yet the correlation analysis indicated a tendency for higher IPAPs with increasing age and deteriorating respiratory function. As would be expected, lower FVCs were also associated with higher backup rates. This apparent need for higher IPAPs and backup rates for more advanced disease stages, as indicated by lower FVCs, likely reflects advanced muscular weakness with the concomitant need for a higher ventilation support to maintain sufficient gas exchange. Increased rigidity of the thoracic cage and the possible development of a secondary structural lung disease might be contributing factors in this context.[4,6,8] We nevertheless consider the clinical importance of the observed higher

settings for IPAP and backup rate in advanced disease stages to be small and thus do not recommend an indiscriminate increase of IPAP or backup rate settings in patients with declining lung function.

We did not include inspiratory time in our analyses because of the variability and thus bad comparability of this parameter in different ventilators and ventilation modes. To clarify, inspiratory time is either fixed or set with a range in different ventilation modes (eg, fixed in pressure control ventilation or with a range in pressure support ventilation), making it unsuitable for comparison between individuals, especially because of the variability of inspiratory time over time in a single patient ventilated with a set range of inspiratory times. In our experience, inspiration time should cover about 40% of the breathing cycle, resulting in a near-physiological inspiratory-expiratory ratio of 1:1.5-2. We also excluded inspiratory triggers from the analyses because they can be either be flow- or pressure-dependent in different ventilators and ventilation modes and thus cannot be compared directly. In general, we normally choose highly sensitive triggers in patients with significant neuromuscular impairment.
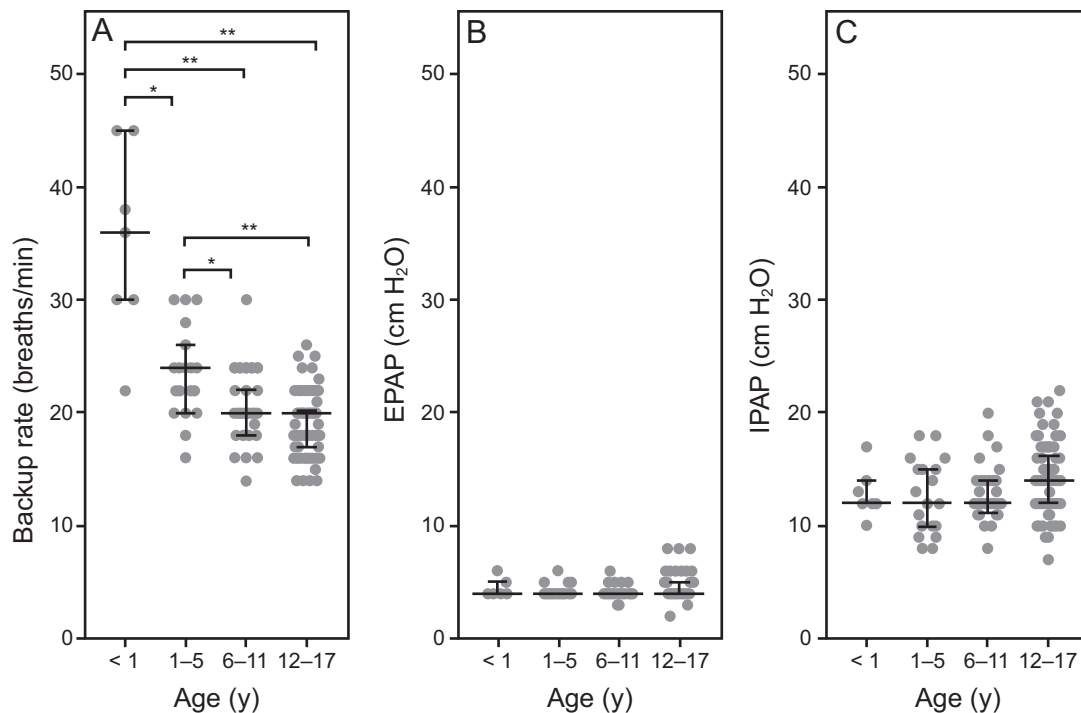
Fig. 1. Scatter plots for backup rate, expiratory positive airway pressure (EPAP), and inspiratory positive airway pressure (IPAP) for each study group are shown with median and interquartile range. Significant differences between the groups are marked by asterisks (*$P < .01$; **$P < .001$).
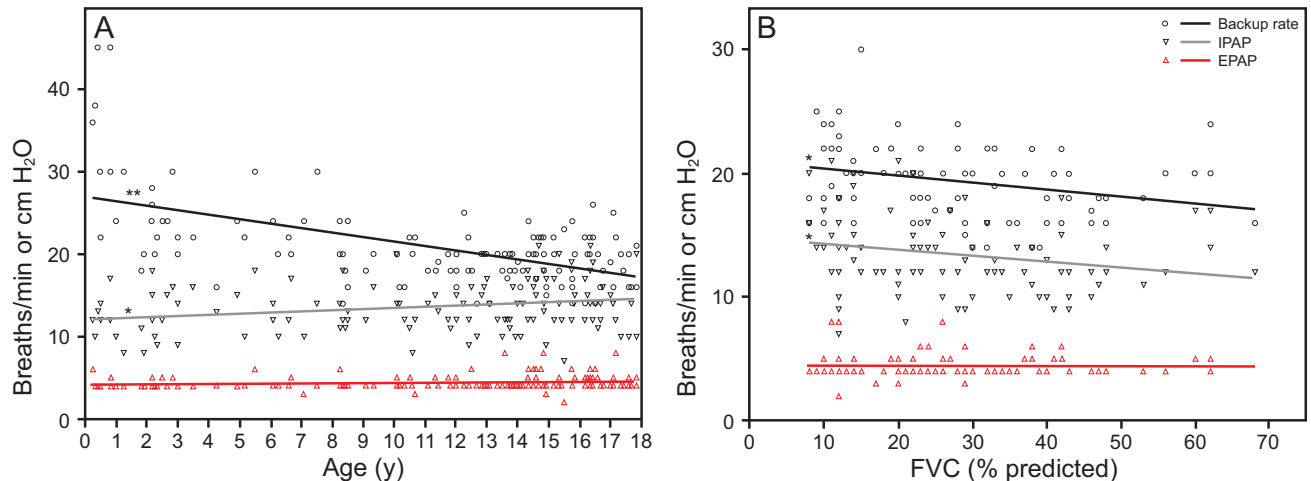

Fig. 2. Ventilator settings with linear regression lines plotted against (A) age and (B) FVC of home ventilated subjects with neuromuscular disease ($n = 128$). A significant positive correlation was found for age and IPAP ($r = 0.242$, $P < .01$), and significant negative correlations were found for age and backup rate ($r = -0.432$, $P < .001$), FVC and backup rate ($r = -0.324$, $P < .01$), and FVC and IPAP ($r = -0.313$, $P < .01$). Significant correlations are marked by asterisks (*$P < .01$; **$P < .001$). EPAP = expiratory positive airway pressure; IPAP = inspiratory positive airway pressure

In our cohort, only oronasal masks and nasal masks were used as NIV interfaces, indicating a low need for less common NIV interfaces such as total face masks or helmets. Generally, nasal masks are preferred over oronasal masks for NIV in pediatric patients with NMDs because they maintain the patient's ability to communicate and reduce aerophagia and the risk of aspiration compared to oronasal masks.[12,18] Subjects with DMD required oronasal masks significantly more often than subjects with SMA in our cohort. This might indicate a more frequent failure of nasal masks in patients with DMD compared to patients with SMA, due, for example, to a higher prevalence of mouth leaks.[19]

All of the findings of this study have to be interpreted against the background of the single-center retrospective design of the study. We tried to quantify sufficiency of the reported ventilator settings by inclusion of respiratory polygraphy and blood gas parameters, but some datasets were incomplete and the included parameters cannot prove ideal ventilation settings per se, especially in the absence of data from continuous capnometries. Data were missing particularly in older cases, where the original measurements were no longer available and data were taken from indirect sources, such as doctor's notes. Finally, the choice of interfaces, ventilation modes, and ventilation parameters are likely to be influenced by personal and local preferences.

Conclusions

We demonstrate that all age groups of pediatric subjects with NMDs receiving NIV required similar ventilator pressure levels in clinically stable situations, whereas the adequate setting for the backup rate of the ventilator reflects the physiologically decreasing breathing frequency of a growing child. Despite the given limitations of a single-center retrospective study, the remarkable overall similarity of ventilation parameters in the different study groups renders the establishment of NIV relatively easy in pediatric patients with NMDs. When starting pressure-limited NIV in patients with NMDs, we propose setting the EPAP to 4–5 cm H_2O and the IPAP to 12–15 cm H_2O (inspiratory pressure delta 8–10 cm H_2O) with an age-oriented backup rate in the initial ventilator settings, and adjusting the settings individually for the patient from this starting point. Patients whose disease is in the advanced stages (ie, more severe muscular impairment and possible structural lung damage) might require slightly higher IPAPs and backup rates. The findings of this study may serve as guidance but should not supersede the necessary careful individual assessment and monitoring of each child with respiratory insufficiency due to NMD.

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