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# Home noninvasive ventilation in pediatric patients: Does one size fit all?

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**Keywords:**

adherence; continuous positive airway pressure; national survey; noninvasive ventilation; pediatrics; settings.

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# Abstract

## Background:

A French national survey showed that 1447 children were treated with long-term continuous positive airway pressure (CPAP) or noninvasive ventilation (NIV) in 2019. Data about the ventilatory settings for children are scarce. The aim of the study was to report the CPAP/NIV settings from the survey according to the patients' age and disorders.

## Methods:

CPAP and NIV settings were compared between 5 age groups (<1, 1-5, 6-11, 12-17 and  $\geq 18$  years), and 6 disease categories (upper airway disorders; neuromuscular disease, NMD; disorder of the central nervous system; cardiorespiratory disorder; congenital bone disease, CBD; and other).

## Results:

Age correlated positively with constant CPAP pressure ( $r=0.364$ ,  $p<.0001$ ), and negatively with CPAP adherence ( $r=-0.173$ ,  $p<.0001$ ). Mean age at CPAP initiation, CPAP pressures and adherence did not differ between disorders. Regarding NIV, mean inspiratory positive airway pressure (IPAP) increased with age ( $r=0.152$ ,  $p=.0001$ ), whereas respiratory rate (RR;  $r=-0.593$ ,  $p<.0001$ ) and adherence to NIV decreased with age ( $r=-0.154$ ,  $p=.0002$ ). NIV settings were quite similar between disease categories, with the CBD group having the highest IPAP, and NMD group having the lowest expiratory positive airway pressure and RR. Adherence tended to be higher with NIV than CPAP.

## Conclusions:

CPAP pressure and IPAP increase with age, while settings seem quite similar between diseases. Even if our study provides some information about CPAP/NIV settings, they should always be individually adapted according to the severity of the disease.

# 1. Introduction

Home continuous positive airway pressure (CPAP) and noninvasive ventilation (NIV) are increasingly used in children worldwide [1-6]. Our recent 2019 national survey confirmed this trend, and showed a more than 14-fold increase in the number of children treated with long-term CPAP/NIV in France between 2000 and 2019 [2]. One thousand four hundred and forty-seven patients were included in this study, with a mean age of  $9.8 \pm 5.8$  (range 3 months–26 years) years. The underlying disorders of the patients were: upper airway disorders (UA, 46%), neuromuscular disorders (NMD, 28%), disorders of the central nervous system (CNS, 13%), cardiorespiratory disorders (Cardioresp, 7%), and congenital bone diseases (CBD, 4%), and other disorders (Other, 3%). This study reported the mean CPAP and NIV settings according to the disease categories, as well as the mean adherence to treatment over the last month. However, CPAP/NIV settings were not compared between disease categories or according to the age of the patients. Some studies reported the ventilatory settings for CPAP [7-13] and NIV [7,14-18] in children. However, studies reporting the settings by age or disease categories are scarce [8,11,18]. CPAP/NIV initiation is usually based on objective criteria, however other criteria including the patient's respiratory status and underlying disease, lung function data and/or other clinical parameters may also play a role [1,19]. Actually, the initiation criteria are still not validated even though some recommendations exist [1,20]. The American Academy of Sleep Medicine recommends a titration polysomnography to set the optimal CPAP or NIV pressures for patients with obstructive sleep apnea [21], however this may not be easily feasible in all centers. A recent ERS statement on pediatric long-term noninvasive respiratory support gathered some guidance for CPAP/NIV settings [1].

The aim of the present study was to report the CPAP/NIV settings data from the French national survey according to the age of the patients and the disease categories.

## 2. Methods

This study was a sub-analysis of the national cross-sectional survey performed in 2019 among all the centers of the French national pediatric NIV/CPAP network, which gathers 28 pediatric university centers distributed among 24 cities [2]. The detailed methodology of the survey has been published [2]. In summary, all the centers filled in an anonymous questionnaire for every child aged  $\leq 18$  years old treated since at least 3 months with CPAP or NIV on June the 1st, 2019. Some young adults aged 18-26 years who were still followed in these centers since childhood were also included. The questionnaire comprised the following information: date of birth, gender, primary and secondary diagnosis, investigations performed before CPAP/NIV initiation, parameters that led to the decision of CPAP/NIV treatment, CPAP/NIV duration, mean objective adherence (CPAP/NIV usage) to treatment during the last month obtained from the device built-in software, type of interface, and CPAP/NIV device. The study was approved by the local institutional board (Comité d'Ethique de Necker Enfants Malades, CENEM, n° CENEM19-5-BF) on March the 7th 2019, and all patients aged  $> 6$  years and all parents gave their informed consent. This sub-analysis focused on the settings of CPAP and NIV according to the patients' age and disease categories.

Comparisons between the settings and adherence to treatment between the different subgroups were performed using the One-Way Analysis of Variance, or the Kruskal-Wallis One Way Analysis of Variance on Ranks in case of non-parametric data. The correlations between 2 variables were assessed using the Pearson correlation test. Adherence to CPAP/NIV was analyzed according to the recommended sleep time for age [22]. We considered a recommended sleep time of 14, 12, 10, 9 and 8 hours for the age categories <1, 1-5, 6-11, 12-17 and ≥18 years, respectively. We have evaluated the number of patients with a CPAP/NIV usage equal or over the recommended sleep time, and we have calculated the ratio between the CPAP/NIV usage and the recommended sleep time for each patient. Statistically significant difference was considered for a *p* value < 0.05.

Table 1. Settings according to the age categories.

	< 1 year (n=106)	1-5 years (n=336)	6-11 years (n=368)	12-17 years (n=539)	≥ 18 years (n=74)	p value
<b>CPAP (n)</b>	<b>63</b>	<b>181</b>	<b>188</b>	<b>194</b>	<b>23</b>	
Age CPAP (years)	0.3 ± 0.2 (0.2-0.9)	1.7 ± 1.6 (0.2-5.8)	6.6 ± 2.9 (0.2-11.7)	12.0 ± 3.4 (0.2-17.3)	14.9 ± 4.0 (5.0-21.7)	-
<i>Constant</i> CPAP (cmH <sub>2</sub> O)	n=63 6.9 ± 1.4 § (5-11)	n=177 7.4 ± 1.5 (4-15)	n=150 8.0 ± 1.8 (4-14)	n=138 8.8 ± 1.9 (6-15)	n=13 9.1 ± 1.6 (7-12)	<0.001
<i>Autoset CPAP</i> Min pressure (cmH <sub>2</sub> O)	n=0 -	n=4 7.0 ± 1.4 (6-8)	n=38 7.0 ± 1.7 (4-12)	n=56 7.1 ± 1.7 (4-10)	n=10 7.1 ± 1.9 (4-9)	-
Max pressure (cmH <sub>2</sub> O)	-	12.0 ± 0 (12)	12.3 ± 1.9 (8-16)	13.3 ± 2.0 (9-17)	11.7 ± 2.7 (8-16)	-
Adherence CPAP (hours/night)	8.4 ± 4.6 † (0-18)	7.1 ± 3.1 (0-15)	6.8 ± 2.6 (0-12)	6.2 ± 2.8 (0-12)	6.0 ± 2.7 (1-9)	<0.001
≥ recommended sleep time (n, %)	7 (11%)	8 (4%)	24 (13%)	39 (20%)	9 (39%)	-
< 4 h/night (n, %)	9 (14%)	25 (14%)	28 (15%)	37 (19%)	5 (22%)	-
among which no use (n, %)	4 (6%)	8 (4%)	4 (2%)	10 (5%)	0	
≥ 16 h/night (n, %)	5 (8%)	0	0	0	0	
<b>NIV (n)</b>	<b>43</b>	<b>155</b>	<b>180</b>	<b>345</b>	<b>51</b>	
Age NIV (years)	0.3 ± 0.2 (0.2-0.8)	1.6 ± 1.4 (0.2-5.9)	5.4 ± 3.2 (0.2-11.2)	11.1 ± 3.9 (0.2-17.2)	15.5 ± 3.5 (4.9-23.8)	-
IPAP (cmH <sub>2</sub> O)	12.1 ± 3.2 * (7-22)	13.4 ± 3.7 (7-30)	13.9 ± 3.6 (8-25)	14.1 ± 3.4 (8-28)	15.3 ± 3.8 (9-26)	<0.001
EPAP (cmH <sub>2</sub> O)	5.3 ± 1.1 (4-8)	5.2 ± 1.5 (0-11)	5.0 ± 1.4 (2-10)	5.2 ± 1.5 (0-12)	4.8 ± 1.8 (0-8)	0.278
RR (b/min)	27.9 ± 7.5 ** (10-40)	24.5 ± 6.5 (10-40)	19.8 ± 4.3 (10-35)	16.6 ± 3.6 (10-30)	17.4 ± 4.8 (10-30)	<0.001
Target volume (n, %)	3 (7%)	15 (10%)	28 (16%)	50 (15%)	19 (37%)	-
Adherence NIV (hours/night)	9.5 ± 3.3 (1-18)	8.7 ± 3.0 (0-22)	8.3 ± 3.5 (0-24)	7.7 ± 3.1 § (0-24)	8.2 ± 2.3 (1-13)	<0.001
≥ recommended sleep time (n, %)	3 (7%)	15 (10%)	61 (34%)	137 (40%)	33 (65%)	-
< 4 h/night (n, %)	2 (5%)	7 (5%)	16 (9%)	32 (9%)	2 (4%)	-
among which no use (n, %)	0	3 (2%)	6 (3%)	4 (1%)	0	
≥ 16 h/night (n, %)	2 (5%)	3 (2%)	3 (2%)	5 (1%)	0	

Data are given as mean ± standard deviation and ranges, or number and proportions.

For CPAP, 3 data were missing regarding the age. For NIV, 4 data were missing regarding the age, and 5 regarding the mode of NIV and settings.

For NIV, IPAP, EPAP are RR were analysed only in patients without target volume.

§ Significantly different between ages < 1 and 6-11, and 12-17 and ≥ 18 years; significantly different between ages 1-5 and 12-17, and ≥ 18 years.

† Significantly different between ages < 1 and 12-17 and ≥ 18 years; significantly different between ages 1-5 and 12-17, and ≥ 18 years.

\* Significantly different between ages < 1 and 6-11, and 12-17 and ≥ 18 years.

\*\* Significantly different between all categories except between ages < 1 and 1-5 years, and between ages 12-17 and ≥ 18 years.

§ Significantly different between ages 12-17 and < 1, and 1-5 years.

Abbreviations: CPAP, continuous positive airway pressure; IPAP, inspiratory positive airway pressure; EPAP, expiratory positive airway pressure; RR, respiratory rate; NIV, noninvasive ventilation; Min, minimal; Max, maximal.

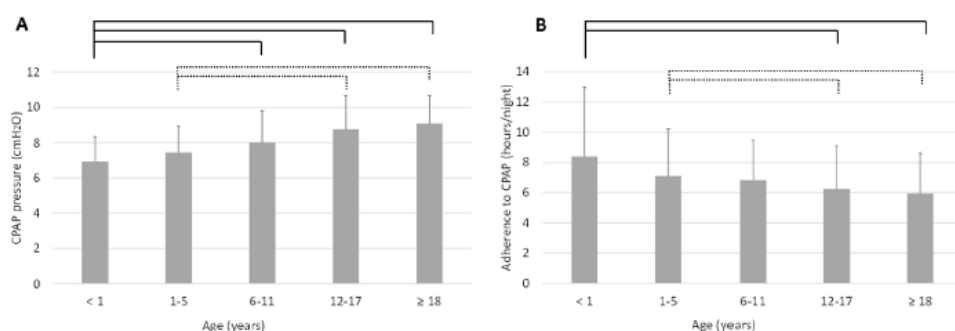
### 3. Results

Among the 1447 patients, 4 patients were excluded from the subsequent analysis because they were treated by adaptive servo-ventilation, and 8 because of missing data regarding the type of ventilation. Among the 652 patients treated with CPAP, 544 (83%) were on constant CPAP and 108 (17%) on autotset CPAP. Autotset CPAP was mainly used in the UA group. Regarding NIV, 783 patients were set on different NIV modes, with 123 (16%) patients set on a hybrid mode with target volume. NIV modes with target volume were used mainly in children >6 years old (Table 1).

#### 3.1. Settings according to the age categories

Regarding constant CPAP, there was a trend for an increase in mean CPAP pressure with age (Table 1 and Figure 1A). Mean CPAP pressure was significantly lower in children aged <1 year compared with the other age categories. There was also a significant positive correlation between CPAP pressure and age ( $r=0.364$ ,  $p<0.001$ ) (Online Figure 1A).

**Figure 1. Mean constant CPAP pressures and adherence to CPAP according to age.** A: mean constant CPAP pressure was significantly different between < 1 and 6-11, and 12-17 and  $\geq 18$  years old, and between 1-5 and 12-17, and  $\geq 18$  years old. B: mean adherence to CPAP was significantly different between < 1 and 12-17 and  $\geq 18$  years old, and between 1-5 and 12-17, and  $\geq 18$  years old. Abbreviations: CPAP, continuous positive airway pressure.

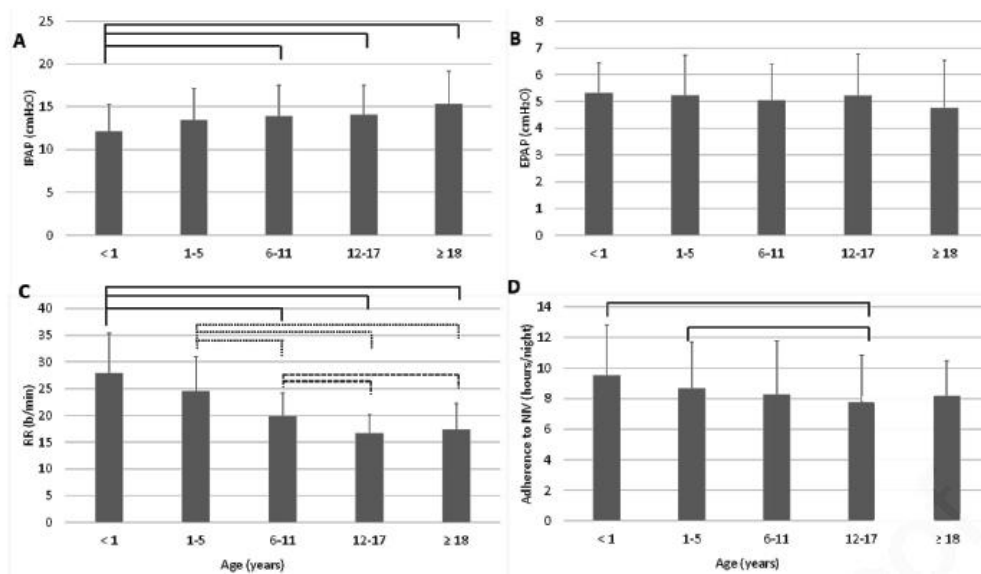


Adherence to (constant and autotset) CPAP had a wide distribution among the ages. Mean adherence to CPAP decreased significantly with age, with children aged <5 years having a higher adherence as compared to children aged >12 years (Table 1 and Figure 1B). A significant weak negative correlation was observed between adherence to CPAP and age ( $r=-0.173$ ,  $p<0.001$ ) (Online Figure 1B). Online Figure 2A shows the number of patients on CPAP with an adherence below or above the recommended sleep time for age. Only a minority of patients had an adherence to CPAP corresponding to or greater than their recommended sleep time. And for these patients, the ratio between the adherence to CPAP and the recommended sleep time slightly decreased with age (Online Figure 2B).

**Figure 2. Mean IPAP, EPAP, RR and adherence to NIV according to age.** A: mean IPAP in children with no hybrid NIV modes with target volume was significantly different between < 1 and 6-11, and 12-17 and  $\geq 18$  years old. B: mean EPAP in children with no hybrid NIV modes was not significantly different between age categories. C: mean RR in children with no hybrid NIV modes was significantly different between all categories except between < 1 and 1-5 years old, and between 12-17 and  $\geq 18$  years old. D: mean adherence to NIV in all children on NIV was significantly different between 12-17 and < 1, and 1-5 years old. Abbreviations: IPAP,



inspiratory positive airway pressure; EPAP, expiratory positive airway pressure; RR, respiratory rate; NIV, noninvasive ventilation.



Regarding NIV, among the patients who did not have a NIV mode with a target volume, mean inspiratory positive airway pressure (IPAP) and respiratory rate (RR) varied significantly with age (Table 1 and Figures 2A and 2C). Indeed, mean IPAP was significantly lower in children aged <1 year as compared with the children aged >6 years. Mean RR tended to decrease with age, with children aged <5 years having a higher RR as compared with older children. The correlation analyses showed a significant slight increase of IPAP with age ( $r=0.152$ ,  $p=0.001$ ), and a decrease of RR with age ( $r=-0.593$ ,  $p<0.001$ ) (Online Figures 4A and 4B).

Mean adherence to NIV (including all NIV modes) also significantly varied with age (Table 1 and Figure 2D). However, only the children aged 12-17 years had a lower adherence as compared with the children aged <5 years. Adherence to NIV appeared to be higher than adherence to CPAP, regardless of age (Table 1). A significant weak correlation was observed between adherence to NIV and age ( $r=-0.154$ ,  $p=0.002$ ) (Online Figure 4C). Online Figure 2C shows the number of patients on NIV with an adherence below or above the recommended sleep time for age. A high number of children aged >12 years had a NIV adherence corresponding to or above their recommended sleep time. The ratio between adherence to NIV and the recommended sleep time remained quite stable over age (Online Figure 2D).

### 3.2. Settings according to the disease categories

CPAP was mainly used in the UA group (80%). Mean age at CPAP initiation, mean constant CPAP pressure and adherence did not differ between the disease categories (Table 2 and Online Figure 3A).

For the UA group on constant CPAP (n=430), there was a trend for an increase in mean CPAP pressure with age (Online Table 1). Mean CPAP pressure was similar in children aged <1 and 1-5 years, but significantly lower than in older children.

Table 2. Settings according to the disease categories.

	UA (n=667)	NMD (n=396)	CNS (n=178)	Cardioresp (n=94)	CBD (n=58)	Other (n=41)	p value
<b>CPAP (n)</b>	<b>523</b>	<b>25</b>	<b>41</b>	<b>21</b>	<b>29</b>	<b>13</b>	
Age at CPAP initiation (years)	6.6 ± 5.23 (0.2-18.7)	6.0 ± 6.0 (0.2-15.5)	7.7 ± 5.7 (0.2-21.7)	3.2 ± 5.1 (0.2-18.0)	5.3 ± 4.4 (0.2-16.3)	7.2 ± 5.3 (0.3-15.7)	0.556
<i>Constant</i> CPAP (cmH <sub>2</sub> O)	n=430 8.0 ± 1.8 (4-15)	n=21 8.0 ± 2.1 (5-12)	n=38 7.2 ± 1.4 (5-11)	n=18 7.1 ± 1.5 (6-10)	n=27 7.9 ± 1.3 (6-11)	n=10 6.6 ± 1.1 (5-8)	0.050
<i>Autoset CPAP</i> Min pressure (cmH <sub>2</sub> O)	n=93 7.1 ± 1.7 (4-12)	n=4 8.3 ± 1.3 (7-10)	n=3 5.0 ± 1.4 (4-6)	n=3 6.0 ± 0 (6)	n=2 8	n=3 6.0 ± 0 (6)	-
Max pressure (cmH <sub>2</sub> O)	12.8 ± 2.2 (8-17)	13.8 ± 1.7 (12-16)	12.0 ± 0 (12)	11.3 ± 1.2 (10-12)	12	11.0 ± 1.4 (10-12)	-
Adherence CPAP (hours/night)	6.8 ± 3.2 (0-18)	7.6 ± 2.8 (0-13)	6.4 ± 2.6 (0-10)	7.8 ± 3.3 (2-16)	6.9 ± 2.7 (2-12)	6.1 ± 1.7 (4-9)	0.355
≥ recommended sleep time (n, %)	51 (10%)	2 (8%)	1 (2%)	3 (14%)	3 (10%)	0	-
< 4 h/night (n, %)	90 (17%)	2 (8)	6 (15%)	2 (10%)	5 (17%)	0	-
among which no use (n, %)	23 (4%)	1 (4%)	2 (5%)	0	0	0	
≥ 16 h/night (n, %)	4 (1%)	0	0	1 (5%)	0	0	
<b>NIV (n)</b>	<b>144</b>	<b>371</b>	<b>137</b>	<b>73</b>	<b>29</b>	<b>28</b>	
Age at NIV initiation (years)	6.5 ± 5.4 (0.2-23.2)	8.0 ± 5.5 (0.2-21.2)	7.2 ± 5.4 (0.2-19.4)	7.5 ± 6.5 (0.2-23.8)	6.4 ± 5.1 (0.2-17.1)	8.1 ± 5.6 (0.3-16.8)	0.053
IPAP (cmH <sub>2</sub> O)	13.9 ± 3.2 (8-24)	13.8 ± 3.8 (7-28)	13.6 ± 3.7 (8-30)	13.4 ± 3.2 (7-22)	15.9 ± 3.6 \$ (7-23)	14.2 ± 2.9 (10-21)	0.048
EPAP (cmH <sub>2</sub> O)	6.2 ± 1.4 * (4-10)	4.7 ± 1.1 ** (0-10)	5.2 ± 1.7 (0-12)	5.3 ± 1.6 (2-10)	6.3 ± 2.1 (3-10)	5.2 ± 1.3 (4-9)	<0.001
RR (b/min)	20.3 ± 6.5 (10-40)	18.8 ± 5.4** (10-50)	20.2 ± 5.7 (10-40)	20.5 ± 6.8 (10-40)	22.3 ± 5.9 (14-40)	20.8 ± 6.7 (12-40)	0.004
Target volume (n, %)	26 (18%)	55 (15%)	16 (12%)	12 (16%)	9 (31%)	5 (18%)	-
Adherence NIV (hours/night)	8.2 ± 3.1 (0-24)	8.0 ± 3.1 (0-24)	8.1 ± 3.4 (0-23)	8.9 ± 3.5 (0-20)	8.1 ± 2.2 (4-12)	8.1 ± 2.2 (3-13)	0.442
≥ recommended sleep time (n, %)	27 (19%)	77 (21%)	28 (20%)	22 (30%)	6 (21%)	5 (18%)	-
< 4 h/night (n, %)	11 (8%)	26 (7%)	16 (12%)	5 (7%)	0	1 (4%)	-
among which no use (n, %)	2 (1%)	5 (1%)	4 (3%)	2 (3%)	0	0	
≥ 16 h/night (n, %)	3 (2%)	6 (2%)	2 (1%)	2 (3%)	0	0	

Data are given as mean ± standard deviation or (ranges).

One patient with NIV did not have the diagnosis reported in the survey.

\$ Significantly different between CBD and CNS.

\* Significantly different between UA and NMD, CNS, and Cardioresp.

\*\* Significantly different between NMD and CBD.

Abbreviations: CPAP, continuous positive airway pressure; IPAP, inspiratory positive airway pressure; EPAP, expiratory positive airway pressure; RR, respiratory rate; NIV, noninvasive ventilation; UA, anomaly

For the UA group on (constant and autoset) CPAP, mean adherence did not significantly vary according to age, even if there was a trend for a lower adherence in older children (Online Table 2). The NIV settings were quite similar between disease categories in patients with no NIV mode with a target volume (Table 2 and Online Figure 3B). Only the CBD group had a higher mean IPAP as compared with the CNS group (p=0.048). The NMD group had the lowest expiratory positive airway pressure (EPAP) and RR. Patients with UA, CBD and other disorders had a high mean EPAP. NIV modes with a target volume were used preferentially in patients with CBD, while the proportion of patients with these modes were quite similar between the other disease categories (Table 2).

Mean adherence to NIV was similar between disease categories. Adherence to NIV seemed to be slightly higher than adherence to CPAP among the different disease categories. If we consider patients with UA, NMD or CNS, mean IPAP was significantly lower only in children with NMD aged <1 year as compared with older NMD children (Online Table 3). There was a tendency for higher IPAP with age in the NMD patients. For the UA, NMD or CNS groups, mean adherence to NIV did not vary significantly with age (Online Table 4).

## 4. Discussion

Our sub-analysis of the 2019 French national survey on 1447 children treated with CPAP/NIV shows that constant CPAP pressure and IPAP increase with age. Adherence to CPAP and NIV was higher in younger children, even though CPAP/NIV adherence appeared to be close to the recommended sleep time even in older children. Constant CPAP pressures did not vary between the different disease categories, contrarily to IPAP and EPAP. Mean IPAP increased with age in children with NMD.

Our 2019 French survey reported the CPAP/NIV settings according to the disease [2], but did not perform comparisons and did not consider age categories. The aim of the present study was therefore to provide information on ventilatory settings in patients treated with home CPAP/NIV, according to age and disease categories. Indeed, only a few studies reported the CPAP or NIV settings in children according to age [11,18], and to our knowledge, no study gathered NIV settings according to the underlying disease. Indeed, only one study reported the CPAP settings by age categories in 94 patients with different disorders [11]. The authors found that a median CPAP pressure of 8 cmH<sub>2</sub>O was required (range 4-20 cmH<sub>2</sub>O), and that CPAP pressure was independent of age or diagnosis. Another study reported the settings of NIV in 128 children with different NMD, and showed that similar NIV settings fit all age groups, except respiratory rate (RR) which is age-related [18]. The ERS Statement on pediatric long-term noninvasive respiratory support noted that mean therapeutic CPAP pressure is usually set at  $8 \pm 3$  cmH<sub>2</sub>O, but that a minimal CPAP pressure has not been validated [1]. Our findings showed that mean constant CPAP was  $8 \pm 2$  cmH<sub>2</sub>O for the whole population, which is in line with the Statement. However, mean CPAP pressure varied between 6.6 and 9.1 cmH<sub>2</sub>O according to the age and/or underlying disorder. Contrarily to Marcus et al. [11], we observed that young children were treated with a lower CPAP pressure than older children. We also observed a positive correlation between CPAP pressure and age.

Our contrasting findings may be explained by the higher number of patients, as there was a trend for a higher CPAP pressure with age in the study by Marcus et al. [11]. Regarding NIV, the ERS Statement noted that therapeutic IPAP usually ranged between 10-14 cmH<sub>2</sub>O and EPAP 4-6 cmH<sub>2</sub>O, with starting pressures of 4 cmH<sub>2</sub>O for EPAP and 8 cmH<sub>2</sub>O for IPAP [1]. EPAP can be set at 0 cmH<sub>2</sub>O when using a single limb circuit with an expiratory valve, or a double limb circuit, while it should be set at least at 4 cmH<sub>2</sub>O with single limb circuit to guarantee a correct carbon dioxide washout, even though some ventilators propose minimum values of 2-3 cmH<sub>2</sub>O. RR was usually set at two to three breaths below the patient's physiological or spontaneous breathing rate, and ranged between 12-18 breaths/min [1]. We found similar values with IPAP ranging between 12-16 cmH<sub>2</sub>O, EPAP 5-6 cmH<sub>2</sub>O, but a higher RR ranging between 17-28 breaths/min. IPAP increased with age while RR decreased with age. The reduction in RR reflects the physiological decrease of the RR with growing age. Contrarily to Steindor et al. [18], we found different IPAP values according to age in the 311

patients with NMD, with younger children having lower pressures. Indeed, they did not find significantly different mean IPAP according to age in 128 children with NMD, however they found a moderate positive correlation with age. Patients with CBD had the highest IPAP values, which may be explained by the fact that these patients may present a more restrictive disorder, with a more rigid chest wall and therefore small lung volumes. This underlines the need for individually tailored NIV settings in order to achieve satisfactory tidal volumes.

Adherence appeared to decrease with age, even though the values remained over 8 hours, in particular in patients on NIV. Interestingly, older patients on NIV had an adherence close to their recommended sleep duration. This may be explained by the profiles of patients requiring NIV as compared with CPAP, and the potential disease severity and clinical outcomes. Numerous studies reported data on adherence, especially in children treated with CPAP [1]. However, there is still no validated definition of good adherence in children. Therefore, the use of CPAP/NIV close to the recommended sleep time for age, or at least close to the actual real sleep time for age [23], should be the objective, especially in ventilator-dependent children. Several strategies exist to improve adherence to CPAP/NIV which should be adapted according to the child's age and underlying disorder [24-26]. Twenty-six children were not using CPAP at all, and 13 children were not using NIV (Table 1), i.e. about 3% of the study cohort. We do not have the data regarding the children who dropped out CPAP or NIV during the 2019 survey, however, we can extrapolate that probably at least 5% of the population will drop out treatment, mainly adolescents treated by CPAP, children with behavioral issues, and/or younger children whose parents are not involved in their treatment [27,28].

Regarding the interfaces used, the majority of patients used a nasal interface (79%), whatever the diagnostic group or type of ventilation [2]. Nasal masks were used at a younger age ( $6.4 \pm 5.5$  years old) as compared to oronasal masks ( $9.8 \pm 4.5$  years old), nasal prongs ( $10.6 \pm 4.8$  years old) or total face masks ( $9.6 \pm 5.5$  years old). We did not detail these results, as it was not the scope of this study. Issues related to the interfaces are still common and comprises pain, skin injuries, but also more serious side effects such as skin necrosis, midface hypoplasia and maxillary retrusion [29-31]. Therefore, a regular follow-up is required to rule out or monitor these potential complications.

Our study has some limitations. First, some data were missing regarding the different items, we however did not detail all the numbers for more clarity in the tables. Second, we did not analyze the settings according to the different CPAP or NIV modes. Third, we did not report the CPAP/NIV settings according to the severity of the disease within the subgroups of disorders. Fourth, we did not report the type of ventilators according to the age, as we believed this would not have impacted our findings. Of note, the type of ventilators according to the subgroups of disorders are presented in the national survey [2]. Finally, we used the recommended sleep time for age to assess the adherence to CPAP/NIV instead of the real sleep time for age. Moreover, we did not use range values for the recommended sleep time but a single value per categories of age, therefore some patients may have been misclassified.

## **5. Conclusions**

Our findings can give some guidance on common CPAP/NIV settings according to the patient's age and disease category. However, we recommend to individually adapt the CPAP/NIV settings according to the severity of the disease and the tolerance of the patient.

Close monitoring is mandatory to ascertain the efficacy of CPAP/NIV, and to adapt the settings and/or interfaces to the evolution of the patient.

### **Ethics approval and consent to participate**

The study was approved by the local institutional board (Comité d’Ethique de Necker Enfants Malades, CENEM, n° CENEM19-5-BF) on March the 7th 2019, and all patients aged > 6 years and all parents gave their informed consent.

### **Consent for publication**

All patients aged > 6 years and all parents gave their informed consent.

### **Availability of data and materials**

Datasets are available through the corresponding author upon reasonable request.

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### **CRedit authorship contribution statement**

**SK:** literature search, data collection, study design, analysis of data, manuscript preparation, review of manuscript; **LG:** review of manuscript; **AA:** data collection, study design, manuscript preparation, review of manuscript; **FB:** data collection, review of manuscript; **PB:** data collection, review of manuscript; **JT:** data collection, review of manuscript; **NS:** data collection, review of manuscript; **MBE:** data collection, review of manuscript; **JM:** data collection, review of manuscript; **II:** data collection, review of manuscript; **CS:** data collection, review of manuscript; **MEL:** data collection, review of manuscript; **AB:** data collection, review of manuscript; **CM:** data collection, review of manuscript; **JB:** data collection, review of manuscript; **BMB:** data collection, review of manuscript; **RR:** data collection, review of manuscript; **ES:** data collection, review of manuscript; **GL:** data collection, review of manuscript; **ABe:** data collection, review of manuscript; **EH:** data collection, review of manuscript; **IP:** data collection, review of manuscript; **SB:** data collection, review of manuscript; **FG:** data collection, review of manuscript; **JMe:** data collection, review of manuscript; **JMo:** data collection, review of manuscript; **MCR:** data collection, review of manuscript; **SM:** data collection, review of manuscript; **MLL:** data collection, review of manuscript; **RH:** data collection, review of manuscript; **MP:** data collection, review of manuscript; **LLC:** data collection, review of manuscript; **PB:** data collection, review of manuscript; **BD:** data collection, review of manuscript; **AM:** data collection, review of manuscript; **PH:** data collection, review of manuscript; **CMe:** data collection, review of manuscript; **MJ:** data collection, review of manuscript; **EG:** data collection, review of manuscript; **CP:** data collection, review of manuscript; **AP:** data collection, review of manuscript; **PG:** data collection, review of manuscript; **LGC:** data collection, review of manuscript; **EF:** data collection, review of manuscript; **ABa:** data

collection, review of manuscript; **PC**: data collection, review of manuscript; **ABr**: data collection, review of manuscript; **MO**: data collection, review of manuscript; **GuL**: data collection, review of manuscript; **LC**: data collection, review of manuscript; **GA**: data collection, review of manuscript; **BF**: data collection, review of manuscript.

### **Declaration of competing interest**

The authors declare no conflicts of interest.

### **Highlights**

- Studies reporting CPAP/NIV settings by age or disease categories are scarce.
- Age correlated positively with constant CPAP pressure.
- CPAP pressures and adherence do not differ between disorders.
- Mean IPAP increases, while respiratory rate and adherence to NIV decrease with age.
- NIV settings are quite similar between disease categories.

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