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# **Non-invasive respiratory support in children and young adults with complex medical conditions in pediatric palliative care**

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**Keywords:**

Paediatrics; Pain; Palliative Care; Respiratory Medicine; Sleep.

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# Abstract

## **Objective:**

Dyspnoea and sleep-disordered breathing (SDB) are common in children with life-limiting conditions but studies on treatment with non-invasive ventilation (NIV) or continuous positive airway pressure (CPAP) are scarce. The aim of the study was to describe children treated with long-term NIV/CPAP within a paediatric palliative care programme in France.

## **Methods:**

Cross-sectional survey on children and young adults with complex medical conditions treated within the French paediatric NIV network with long-term NIV/CPAP. Characteristics of the patients were analysed and patient-related outcome measures of NIV/CPAP benefit were reported.

## **Results:**

The data of 50 patients (68% boys), median age 12 (0.4-21) years were analysed. Twenty-three (46%) patients had a disorder of the central nervous system and 5 (10%) a chromosomal anomaly. Thirty-two (64%) patients were treated with NIV and 18 (36%) with CPAP. NIV/CPAP was initiated on an abnormal Apnoea-Hypopnoea Index in 18 (36%) of the patients, an abnormal nocturnal gas exchange alone in 28 (56%), and after an acute respiratory failure in 11 (22%) of the patients. Mean objective NIV/CPAP adherence was  $9.3 \pm 3.7$  hours/night. NIV/CPAP was associated with a decrease in dyspnoea in 60% of patients, an increase in sleep duration in 60% and in sleep quality in 74%, and an improvement in parents' sleep in 40%.

## **Conclusions:**

In children with life-limiting conditions, long-term NIV/CPAP may be associated with relief of dyspnoea, an improvement of SDB and an improvement in parents' sleep.

# INTRODUCTION

The number of children with complex and life-limiting conditions and managed within a paediatric palliative care (PPC) programme is increasing. Indeed, due to improvement in care and innovative therapies, especially for rare and complex diseases, the number of children surviving with chronic disorders and multiple comorbidities is increasing. A study performed in 2018 during a 6-month period in a respiratory intermediate care unit of a tertiary paediatric university in Italy identified 130 children with medical complexity managed within a PPC programme.<sup>1</sup> One hundred and ten (85%) patients had a rare disease, 42 (33%) a neuromuscular disorder and 33 (25%) a central nervous system disorder. A national survey performed in Spain in 2019 identified 164 children managed within 14 PPC teams, with oncological, neurological or neuromuscular diseases being the most common diseases.<sup>2</sup>

PPC is a complex mosaic of activities that aim to relieve suffering and provide comfort to patients and their families, addressing their physical, psychological, spiritual, social and ethical needs. The International Standards for PPC were published in 2022 and state that the main goal is to prevent or eradicate distressing symptoms in order to optimise the quality of life of the child and his family.<sup>3</sup> The child's physical symptoms should be assessed, treated and monitored according to the patient's individual response, age, culture, and whether care is performed at home or in a hospital or hospice.

Dyspnoea and sleep problems are among the most common symptoms reported by children with life-limiting conditions and have a major impact on the child's and family's quality of life.<sup>4</sup> Among the 164 children managed by a PPC in Spain, dyspnoea was the most common symptom, reported by 63% of the patients, followed by pain (50%), excessive secretions (48%) and sleep problems (47%).<sup>2</sup> This high prevalence of dyspnoea and sleep problems contrasts with the lack of tools and guidelines to screen and treat these symptoms. An online questionnaire completed by 23 PPC experts from 13 countries on symptoms commonly encountered in PPC showed that 50% of the experts found it 'difficult' or 'very difficult' to find specific literature and evidence on managing sleep disorders.<sup>5</sup> Sleep disorders were ranked among the top three difficult symptoms or problems, on the same level as delirium, dystonia and neuroirritability.<sup>5</sup>

The aim of non-invasive ventilation (NIV) or continuous positive airway pressure (CPAP) is to correct or improve sleep-disordered breathing (SDB) and dyspnoea.<sup>6 7</sup> However, the literature on the use of NIV/CPAP in PPC is scarce. Of the 130 children with complex disease managed by a respiratory team in Italy, 47 (36%) were treated with long-term NIV >12 hours/day and 26 (20%) with invasive ventilation. In the Spanish survey, 12% of the 164 patients were treated with NIV with the same number being treated with invasive ventilation. However, no information is available on the criteria that led to the initiation of NIV, the tolerance and adherence of the patients, and the benefit of NIV with regard to patient-related outcome measures (PROMs) such as quality of sleep and quality of life. As such, the benefit of NIV in children in PPC with regard to the improvement in dyspnoea, quality of sleep and quality of life was listed as one of the research priorities by the recent European Respiratory Society Statement on paediatric long-term non-invasive respiratory support.<sup>7</sup>

The aim of the study was to describe the characteristics of children and young adults with a life-limiting condition, treated with long-term NIV/CPAP in France between December 2022 and March 2023.

## MATERIAL AND METHODS

The study was a national cross-sectional survey performed among the French national paediatric NIV network which gathers 28 paediatric university centres.<sup>8</sup> From 1 December 2022 to 31 March 2023, the referring paediatrician of the centre filled in an anonymous questionnaire for every child treated since at least 3 months with NIV or CPAP within a PPC or equivalent. Only patients belonging to life-limiting or life-threatening conditions of Category 3 ('Progressive conditions without curative treatment options') or Category 4 ('Irreversible but non-progressive conditions causing severe disability leading to susceptibility to health complications and likelihood of premature death') were included.<sup>9</sup> Patients with neuromuscular disease were excluded as they represent a specific disease entity belonging to Category 2 (Conditions where premature death is inevitable).<sup>9</sup> As not every hospital has a specific PPC, patients were included if their care comprised at least two of the following multidisciplinary decisions:

- ▶ 'Limitation of care'
- ▶ 'Do-not-intubate' decision
- ▶ 'Comfort care only'
- ▶ Management by a PPC
- ▶ Management by a paediatric pain team
- ▶ Presence of an advanced directive.

Some young adults, aged 18–21 years, who were followed within the NIV paediatric network since childhood and initiated on NIV/CPAP before the age of 18 years, and who were not yet transitioned to an adult centre, were also included.

The survey was developed by the members of the NIV network and comprised the same information collected on a previous national survey on long-term paediatric NIV/CPAP such as age, gender, main diagnosis, investigations performed before NIV/CPAP initiation, parameters that led to the decision of NIV/CPAP treatment, NIV/CPAP duration, mean objective NIV/CPAP adherence during the last month obtained from the device built-in software, type of interface, NIV/CPAP settings and the prescription of additional respiratory technologies at home.<sup>8</sup> Within the context of PPC, additional information on medical treatment and non-respiratory technologies was also gathered. As every patient was seen at least every 3 months on a routine basis, PROMs about the subjective benefit of NIV/CPAP were asked by the referring paediatric pulmonologist to the parents and patients >6 years when possible (family/patient), during a routine follow-up visit (table 1). All responses were centralised and analysed by the NIV centre of Necker hospital. All the families/ patients gave their informed consent.

<b>Table 1 Patient-related outcomes on the subjective benefit of non-invasive ventilation/continuous positive airway pressure (NIV/CPAP)</b>		
	Yes	No
Is NIV/CPAP associated with a decrease in dyspnoea in your child?		
Is NIV/CPAP associated with an improvement in sleep duration in your child?		
Is NIV/CPAP associated with an improvement in sleep quality in your child?		
Is NIV/CPAP associated with an improvement in your own sleep duration or quality?		
Optional open response(s)		

## RESULTS

The data of 50 patients (68% boys), median age 12 (0.4–21) years, were analysed ([table 2](#)). The patients were followed by 15 of the 28 centres with one to seven patients per centre, with 13 centres having no PPC patient treated with long-term NIV/CPAP at the time of the survey. All patients had complex and rare diseases with the most common diseases being disorders of the central nervous system (n=23, 46%), chromosomal anomalies (n=5, 10%) or peripheral neurological diseases (n=3, 6%) ([table 2](#)). Three patients were older than 18 years but were managed since childhood by a paediatric centre (one patient with Kenny Caffey syndrome, one patient with encephalopathy and one patient with fibrodysplasia ossificans progressiva). The patients had a median of three (two to six) PPC decisions with ‘limitation of care’ for 98% of the patients, ‘do-not- intubate’ for 72%, ‘comfort care only’ for 46%, with an advanced directive being available for 70% of the patients ([table 3](#)). Sixty-six per cent were managed by a PPC and 50% by a paediatric pain team. Opioids were prescribed to 36% of patients, analgesics to 48%, anxiolytics to 30% and melatonin to 20% for sleep problems ([table 3](#)). Sixty per cent of the patients had oxygen at home, 62% a pulse oximeter and 62% a suction equipment ([table 3](#)). Fifty per cent of the patients had an airway clearance device at home and 68% of the patients had a gastrostomy. Forty-six patients lived at home, 23 attended a specialised medical institution, 4 attended a specialised boarding school and 3 attended a normal school with a school life assistant.

A poly(somno)graphy (P(S)G) was performed prior to NIV/ CPAP initiation in 44% of patients while 33% had only a nocturnal pulse oximetry (SpO<sub>2</sub>) with a transcutaneous carbon dioxide pressure (PtcCO<sub>2</sub>) recording ([table 4](#)).

The decision of NIV/CPAP initiation was taken on objective criteria in 78% of the patients ([table 4](#)): a high Apnoea-Hypopnea Index in 36% of patients, an abnormal nocturnal SpO<sub>2</sub> and PtcCO<sub>2</sub> alone in 56% of patients, or one of these last two parameters in 8% of the patients. In 22% of the patients, NIV/CPAP was initiated during an acute respiratory failure, severe inspiratory dyspnoea (8%) or witnessed apnoeas (8%).

Sixty-four per cent of the patients were treated with NIV and 36% with CPAP. Mean CPAP level was 8±1.5 cm H<sub>2</sub>O ([table 5](#)). For NIV, mean inspiratory and expiratory pressures were 16±5 cm H<sub>2</sub>O and 7±2 cm H<sub>2</sub>O, respectively, with a mean backup rate of 21±5 breaths/min. Mean objective adherence was excellent with a mean use of 9.3±3.7 hours/night ([table 5](#)).

None of the patients used his NIV/CPAP less than 4 hours/night and only one patient used NIV >16 hours/day. The majority of patients (78%) used a nasal interface.

Median (range) age, years	12 (0.4–21)
Male/female, n (%)	34 (68%)/16 (32%)
Diagnosis	
Central nervous system disorder	
Encephalopathy (unknown origin)	4
MeCP2 mutation	3
Cerebral malformation	2
Rett syndrome	2
Perinatal anoxia	2
Cerebral tumour	2
Lennox-Gastaut syndrome	2
Metachromatic leucodystrophy	1
Woxx deficiency	1
Kleefstra syndrome	1
Joubert syndrome	1
Ceroid lipofuscinosis	1
Sulphate oxidase deficiency	1
Chromosomal anomaly	
Pallister-Killian syndrome	1
Other chromosomal anomaly	4
Peripheral neurological disease	
Neuroaxonal dystrophy	1
Sensitivo-motor neuropathy	1
Congenital myasthenia with sequelae of cardiac arrest	1
Mitochondrial cytopathy	3
Mucopolysaccharidosis type II	3
Polymalformative syndrome	2
Fibrodysplasia ossificans progressiva	2
Mucopolipidosis type II	1
Kenny-Caffey syndrome	1
CHARGE syndrome	1
Prader-Willi syndrome	1
Down syndrome	1
Pulmonary atresia with pulmonary hypertension	1
Immune deficiency	1
Congenital methemoglobinemia	1
Heart-lung transplantation after rhabdoid tumour	1

PROMs were available for 86% of the parents. Sixty per cent of these family/patients, reported that NIV/CPAP was associated with a decrease in their child's dyspnoea, 60% of parents reported an improvement in their child's sleep duration, 74% an improvement in their child's sleep quality, and 40% of the parents reported an improvement in their own sleep (figure 1). NIV/CPAP was also associated with a decrease in hospitalisations for respiratory episodes (n=12), in daytime fatigue with an increase in alertness (n=6), a decrease in respiratory secretions (n=2), a decrease in epileptic episodes (n=1), the withdrawal of daytime oxygen therapy (n=1) and an increase in weight (n=1). The improvement after NIV/CPAP did



not differ according to the initiation after objective measurements or after a clinical evaluation only. Five (10%) patients died during the 4 months of the data collecting period.

<b>Palliative care decisions</b>	
'Limitation of care'	49 (98%)
'Do-not-intubate' decision	36 (72%)
'Comfort care only'	23 (46%)
Management by a paediatric palliative care team	33 (66%)
Management by a paediatric pain team	25 (50%)
Advanced directive	35 (70%)
<b>Drugs</b>	
Opioids	18 (36%)
Analgesics	24 (48%)
Anxiolytics	15 (30%)
Melatonin	12 (22%)
<b>Nutritional support</b>	
Nasogastric tube	3 (6%)
Gastrostomy	34 (68%)
<b>Respiratory equipment</b>	
Home oxygen therapy	30 (60%)
Home pulse oximeter	31 (62%)
Suction equipment	31 (62%)
<b>Airway clearance device</b>	
Intermittent positive pressure breathing	7 (14%)
Mechanical insufflator-exsufflator	11 (22%)
Intrapulmonary percussive ventilation	7 (14%)

## DISCUSSION

This study is the first to show that long-term NIV/CPAP may be associated with a subjective improvement in patient's dyspnoea, sleep duration and sleep quality, and an improvement in parents' sleep, in children and young adults with complex life-limiting disorders and managed within a PPC or equivalent. All patients had rare and complex disorders, mainly of the central nervous system and an excellent objective adherence to NIV/CPAP.

This study explores a major area that is largely underdiagnosed and undertreated in children managed within a PPC. Fifty children were identified in this cross-sectional study, which represents approximately 3.5% of the total number of children treated with long-term NIV/CPAP in France, or 4.7% when excluding children with neuromuscular disease.<sup>8</sup> This number is larger than the 19 children included in the Spanish study but comparable to the 47 children in the single-centre Italian study.<sup>1 2</sup> This may be explained by the fact that the Italian study and the present study were performed by paediatric pulmonologists who have been concerned with SDB and its management. However, given the extremely high prevalence of dyspnoea, SDB and sleep problems in children in PPC,<sup>2</sup> it is probable that SDB is under-evaluated.

**Table 4** Investigations performed prior to NIV/CPAP initiation and NIV/CPAP initiation criteria

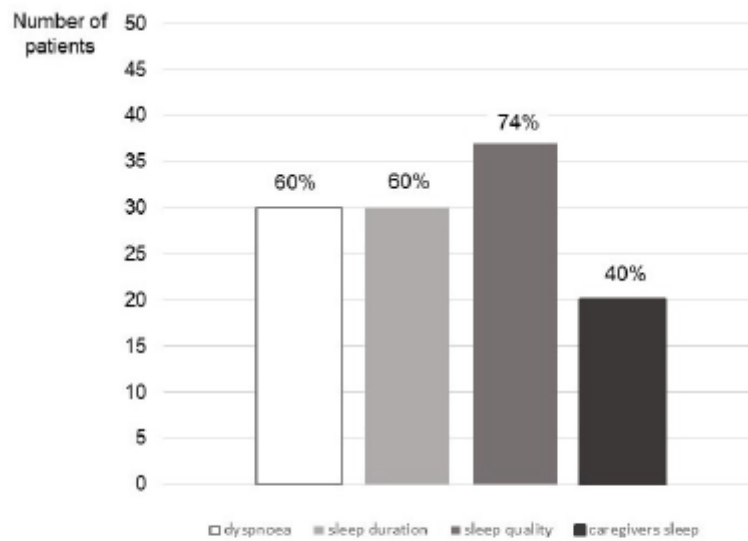
Investigations prior to NIV/CPAP initiation, n (%)	
PG/PSG	22 (44%)
Nocturnal SpO <sub>2</sub> + PtcCO <sub>2</sub> recording alone	16 (33%)
Nocturnal SpO <sub>2</sub> recording (alone)	3 (6%)
NIV/CPAP initiation criteria, n (%)	
Apnoea-Hypopnoea Index	18 (36%)
Nocturnal SpO <sub>2</sub> + PtcCO <sub>2</sub> recording alone	28 (56%)
Nocturnal SpO <sub>2</sub> recording (alone)	2 (4%)
Nocturnal PtcCO <sub>2</sub> recording alone	2 (4%)
Clinical criteria	
Acute respiratory failure requiring admission to the PICU	11 (22%)
Inspiratory dyspnoea	4 (8%)
Witnessed apnoeas	4 (8%)
Life-threatening respiratory event	1 (2%)
Before arthrodesis	1 (2%)

CPAP, continuous positive airway pressure; NIV, non-invasive ventilation; PG, respiratory polygraphy; PICU, paediatric intensive care unit; PSG, polysomnography; PtcCO<sub>2</sub>, transcutaneous carbon dioxide pressure; SpO<sub>2</sub>, pulse oximetry.

**Table 5** Duration of NIV/CPAP, NIV/CPAP adherence, equipment and settings

Duration of NIV/CPAP treatment, years	4.2±3.0 3 (0.5–12)
Adherence (hours/night)	9.3±3.7 10 (3–20)
<4 hours/night (n, %)	0 (0%)
≥16 hours/night (n, %)	1 (2%)
Interfaces	
Nasal (n, %)	39 (78%)
Nasobuccal (n, %)	9 (18%)
Total face (n, %)	2 (4%)
NIV, n (%)	
IPAP, cm H <sub>2</sub> O	16±5 14 (8–24)
EPAP, cm H <sub>2</sub> O	7±2 6 (4–10)
BUR, breaths/min	21±5 20 (15–32)
CPAP, n (%)	
CPAP level, cm H <sub>2</sub> O	8.0±1.5 8 (6–14)

Data are given as mean±SD and median and range or number (percentage).  
BUR, backup rate; CPAP, continuous positive airway pressure; EPAP, expiratory pressure; IPAP, inspiratory pressure; NIV, non-invasive ventilation.



**Figure 1** Patient-related outcomes in dyspnoea, sleep duration, sleep quality and parents' sleep reported by the family/patient. White bar: number of patients with an improvement in dyspnoea, light grey bar: number of patients with an improvement in sleep duration, dark grey bar: number of patients with an improvement in sleep quality, black bar: number of main caregivers with an improvement in sleep quality.

The diagnosis of SDB is challenging in children with life-limiting disorders. The patients are usually uncomfortable, the patient and the family may be reluctant to perform sophisticated exams like a PSG, and the cooperation of the child may be difficult as many children have disorders associated with a developmental delay and poor cooperation. This gives weight to simplified tools able to monitor sleep duration and sleep quality. Respiratory polygraphy may be an alternative to PSG.<sup>7</sup> Abnormal nocturnal gas exchange may also justify NIV or CPAP on its own.<sup>7</sup> Actigraphy has also been shown to be a valuable tool to assess sleep behaviour, sleep efficiency and abnormal breathing in eight children in PPC.<sup>11</sup> But NIV or CPAP may also be indicated in some clinical situations such as acute respiratory failure.<sup>7</sup> In the present study, NIV/CPAP was initiated on clinical criteria in 22% of the patients. A more systematic assessment of SDB is highly recommended in this population, at least on clinical symptoms, as the main objective of NIV/CPAP in PPC is to relieve dyspnoea and SDB symptoms.

The medical complexity of the patients is reflected by the underlying disorders and the multiple devices prescribed at home. Indeed, 62% of the patients had a disorder of the central nervous system, a chromosomal anomaly or a peripheral neurological disease (table 2). Sixty-eight per cent of the patients had nutritional problems requiring a gastrostomy. More than half of the patients had also 'as needed' oxygen therapy, guided by punctual SpO<sub>2</sub> measurements by means of a pulse oximeter. This high dependence on respiratory and non-respiratory Technology is comparable to the experience of other paediatric centres.<sup>1 2</sup>

One of the major interests of our study is that NIV/CPAP was associated with a decrease in dyspnoea in 60% of the patients with an increase in sleep duration and sleep quality. These results are in line with the excellent objective NIV/CPAP adherence in patients of whom a majority have a developmental delay and behaviour problems. This high NIV/CPAP adherence may be an additional, indirect marker of the patients' comfort with NIV/CPAP.

Parental care for a child with a life-limiting condition is a complex physical and mental burden. Indeed, a recent review observed that caregiver health was negatively impacted in 84% of cases with pain and sleep problems being the most common problems,<sup>4</sup> underlining the urgent need of interventions aimed at improving physical and mental health in caregivers.<sup>4</sup> There seems also to be an association between patient-reported severity of symptoms and caregiver symptoms with sleep symptoms being among the most common complaints.<sup>12</sup> The improvement of dyspnoea and SDB by means of long-term NIV/CPAP in selected children with life-limiting conditions may thus be one tool to decrease the caregivers' burden.

As the study was performed within the French paediatric NIV network, the recruitment of the patients may be biased. Indeed, in the literature, the majority of PPC children have neurological diseases, oncological and haematological cancers, heart disease, or a variety of congenital and chromosomal abnormalities. <sup>1 2 4 11 13–15</sup> The patients were cared by 15 centres with a limited number of patients per centre. Only 66% of the patients were managed by an official PPC. Indeed, as not every participating centre had a dedicated PPC, we made the choice to list a series of multidisciplinary decisions associated with a limitation of care. As this was a cross-sectional study including only patients treated with NIV/CPAP since at least 3 months, we have no information on the patients who were not proposed NIV or who did not tolerate or refused NIV/CPAP.<sup>7</sup> The decision to initiate or not long-term NIV in children has been shown to be stressful for parents.<sup>16</sup> Parents want comprehensive, balanced information on the impact of long-term NIV on their child but also on the family. When the child has a profoundly serious and life-limiting condition, they should have the option not to initiate long-term NIV.<sup>16</sup> A shared decision with a NIV trial during a determined period allowing the evaluation of PROMs, may be an option in this situation. The benefit of NIV/CPAP was assessed by PROMs. And finally, information on the parent (mother or father) who responded to the questions, as well as their age, education and income level, was not available.

In conclusion, this study shows the high adherence and major subjective benefit of long-term NIV/CPAP in selected children and young adults with life-limiting disorders. This supports the inclusion of pulmonary paediatricians in the management of these patients and the usefulness to screen and treat dyspnoea, by means of autoassessment and heteroassessment tools for patients who are unable to communicate, and SDB with minimally invasive tools in these patients in order to minimise burden on patients and families.

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## **BF is responsible for the entire content of the manuscript.**

BF made the design of the study, and wrote the first and last versions of the manuscript. All the coauthors approved and validated the information sheet, collected the data of their patients, and contributed to and approved the final version of the manuscript.

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## **Competing interests**

None declared.

## **Patient consent for publication**

Not applicable.

## **Ethics approval**

This study involves human participants and was approved by the Comité d’Ethique de Necker Enfants Malades (CENEM, n° CENEM19-5-BF) on 7 March 2019. Participants gave informed consent to participate in the study before taking part.

## **Provenance and peer review**

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## **Data availability statement**

Data are available upon reasonable request. Data are available on request.

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