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Interfaces for long-term noninvasive positive pressure ventilation in children

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Abstract *Objective:* The aim of the study was to report the type and tolerance of the interface chosen for long-term noninvasive positive pressure ventilation (NPPV) in children. *Methods:* This was a descriptive study carried out in the clinical setting of a pediatric university hospital in which all children started on long-term NPPV over a 18-month period were included. *Results:* NPPV was started in 97 children with neuromuscular disease or thoracic scoliosis ($n = 35$), obstructive sleep apnea with ($n = 32$) or without ($n = 21$) maxillofacial deformity, or lung disease ($n = 9$). All 25 children $\leq$2 years of age, as well as four older children, were fitted with custom-made nasal masks; all other children were fitted with an industrial nasal mask (50%), a facial mask (16%), or nasal prongs (2%). Industrial masks with and without manufactured leaks were used in 33 (34%) and 35 (36%) children, respectively. All patients with obstructive sleep apnea used interfaces with manufactured leaks, whereas all patients with neuromuscular disease or thoracic scoliosis used interfaces without manufactured leaks. Both types of interfaces were used in patients with lung disease. The interface had to be changed in 20 patients because of discomfort ($n = 16$), leaks ($n = 4$), facial growth ($n = 3$), skin injury ($n = 2$), or change of the ventilatory mode ($n = 2$). A second or third mask change was necessary in nine and four patients, respectively. *Conclusion:* The choice of the interface for NPPV in children is determined by the patient's age and the underlying disease. Discomfort is the main reason for mask change.

Keywords Noninvasive positive pressure ventilation · Interface · Nasal mask · Facial mask · Infant · Child

Introduction

Noninvasive positive pressure ventilation (NPPV) is increasingly used in children with severe obstructive sleep apnea syndrome (OSAS) and chronic respiratory failure due to neuromuscular disorders or lung disease. While it has been used in children and adolescents suffering from these various disorders for many years, more recently it is also being used in younger patients and patients with various diseases associated to severe bilateral facial deformities, such as Goldenhar syndrome, Treacher Collins syndrome, achondroplasia, craniostenosis, and Down syndrome [1–3]. In these patients, NPPV represents an interesting noninvasive alternative to tracheostomy,

which is associated with significant morbidity and may impair normal development and, particularly, language development [4, 5]. Discomfort and disruption of social and family life are common consequences of patients with a tracheostomy. Although tracheotomized children may be safely discharged home after careful family education and training, home treatment may be difficult or even unfeasible for some families [6]. In contrast, home treatment is easier with NPPV, which has the main advantage of being noninvasive with the possibility of an "on demand" use.

An increasing number of infants may benefit from NPPV in the newborn period, such as infants with Pierre Robin syndrome [7, 8]. However, the extended use of NPPV is limited by the paucity of well-adapted industrial masks for these young children. Indeed, the choice of the optimal interface for NPPV is of paramount importance for the success of NPPV, but is also challenging, especially in young children and those with facial deformity or asymmetry. The patient will not accept or tolerate NPPV in the case of skin injury, pain, discomfort, or air leaks around the mask.

The choice of the interface is determined not only by the age and the facial morphology of the patient but also by the ventilatory mode. NPPV can be classified as in "leak" ventilation [such as continuous positive airway pressure (CPAP) or bilevel positive pressure ventilation (BiPAP)] or "non-leak" ventilation (such as pressure support or volume-targeted ventilation with a valve or a double circuit). Interfaces with manufactured leaks are used for "leak" ventilation, and interfaces without manufactured leaks are used for "non-leak" ventilation. The choice between these two ventilatory modes is mainly determined by the type of underlying disease. Patients with upper airway obstruction or intrinsic positive end-expiratory pressure (PEEPI) will need a continuous positive airway pressure to maintain the patency of the upper airway or to counteract PEEPI [9]. "Leak" ventilation with CPAP or BiPAP by means of an interface with a manufactured leak is simple and perfectly appropriate for these patients [10, 11]. Patients with neuromuscular or lung disease who have no PEEPI may be ventilated with a pressure- or volume-targeted mode to ensure adequate alveolar ventilation, with the choice depending mainly on the habits of the prescriber. However, a PEEP is not necessary for these patients, and BiPAP devices generally perform not as well as classical ventilators [12]. In addition, volume-targeted ventilation may be required for patients with advanced restrictive disease, such as neuromuscular disease, in whom daytime mouthpiece ventilation may be necessary. Indeed, mouthpiece ventilation can only be performed with a volume-targeted mode, and the initial choice of a volume-targeted mode avoids any subsequent need for a change of the ventilatory mode.

To provide clinicians with information that will facilitate the choice of NPPV interface, we report our experience in a large group of infants and children who were recently started on long-term NPPV. The initial choice according to the age of the patient and the underlying disease is reported, as well as the eventual interface changes.

Materials and methods

Patients

All consecutive pediatric patients who were started on long-term NPPV in our unit between January 2010 and July 2011 were included in the study. NPPV was started in the presence of the parents in a specific pediatric NPPV in-patient unit by members of a well-trained and experienced staff. The patient was discharged home when he/she was able to sleep at least 6 h with NPPV during the night and presented normal nocturnal gas exchange with NPPV. A home visit was performed by the home care organization on the day of discharge, after 1 week, and then every 1–3 months or upon request. A systematic sleep study with NPPV was performed in the hospital 1 month after the start of NPPV. The parents were instructed to contact the NPPV unit in case of any problem with the interface and/or the ventilator.

Patients were classified into four groups according to the underlying disease: neuromuscular disease or thoracic scoliosis (NM/SC group), OSAS with maxillofacial deformity (MF group), OSAS without facial deformity (OSAS group), and lung disease (Lung group).

The study was approved by our institutional board, and written consent was obtained from all the parents and patients aged $\geq$6–8 years.

Interfaces

On admission, the most appropriate interface with regard to the patient's underlying disease and ventilatory mode, but also tolerance and comfort was selected (Fig. 1). The patient tried the interface with NPPV for repeated short periods during daytime. Nasal masks were preferred over facial masks, which were only used in the case of serious mouth leaks and/or the impossibility to close the mouth during sleep. In adolescents, nasal prongs were proposed as the first choice in the case of CPAP or BiPAP ventilation. The interface associated with the best tolerance and comfort, defined by the absence of any skin injury, pain, discomfort, and leaks, was selected. Custom-made masks were composed of a thermoformable plastic frame (VT Plastics, Gennevilliers, France) with an interior coverage of either self-sticking foam (Adhesia Laboratoire, Mulhouse, France) or a protection and comfort gel

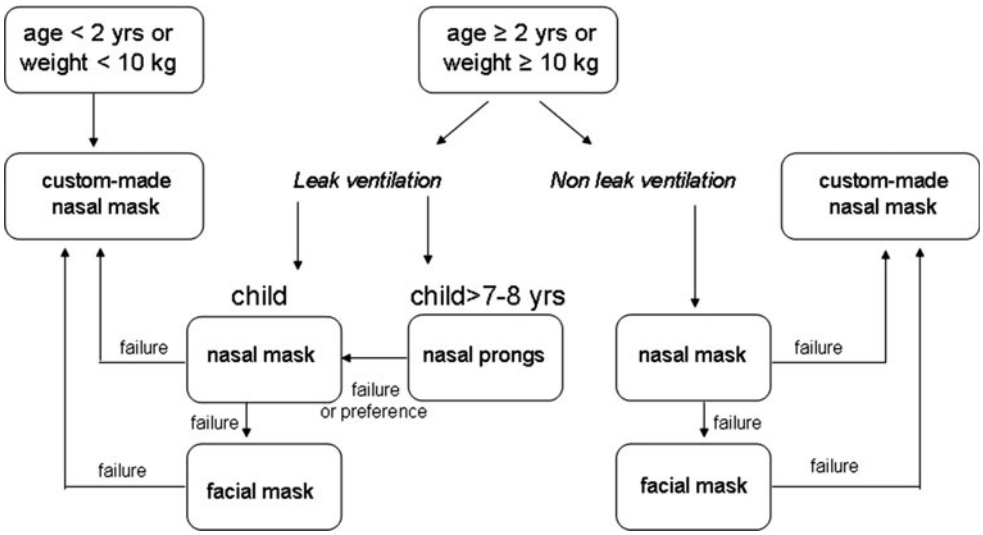

Fig. 1 The algorithm chart used to choose the interface for long-term noninvasive ventilation for children

(Adhesia Laboratoire, Mulhouse, France) [13]. The masks were modeled on plaster phantoms corresponding to the age and the physiognomy of the patient. Bedside adjustments were then realized, if necessary, by thermoforming the plastic frame to obtain the best comfort with minimal leaks. A plastic tube (inner diameter 22 mm) was fixed onto the mask by an autopolymerizable resin (Orthoresin, Dentsply, Weybridge, UK) and connected to a double circuit or an expiratory valve in the case of "non-leak" ventilation, or to a plateau exhalation valve (Respironics, Murrysville, PA) and a simple tube in the case of a "leak" ventilation. The masks were maintained by means of an industrial head gear (Respironics head gear "Child").

In the case of intolerance of the interface after the initial discharge [13], other interfaces were tried and the most appropriate and comfortable interface was selected by trained nurses with systematic medical supervision. The cause(s) of the intolerance and the type of the new interface were recorded.

Results

Patients

During the study period, 97 children were started on long-term NPPV (Table 1). The majority of the patients belonged to the NM/SC group and the MF group. Ages ranged from 1 month to 18 years. The patients of the MF group were younger than the patients belonging to the three other groups. Of note, 18 patients were younger than 1 year and 25 patients were $\leq$2 years. The clinical evolution of the 97 patients during the study period was excellent with no death or life-threatening event and no need for a tracheotomy.

Table 1 Characteristics of the patients

Characteristics	NM/SC group	MF group	OSAS group	Lung group
Number of patients	35	32	21	9
Age (years)	12.0 ± 4.5	4.3 ± 4.9	6.1 ± 4.4	10.0 ± 5.0
Weight (kg)	35.4 ± 18.8	29.2 ± 27.7	25.6 ± 13.5	15.2 ± 14.5
Pathology	Duchenne muscular dystrophy (11 patients) Spinal muscular atrophy (6) Congenital myopathy (10) Thoracic scoliosis (8)	Pierre Robin syndrome (7) Mucopolysaccharidosis (2) Goldenhar syndrome (2) Franceschetti syndrome (4) Idiopathic maxillofacial deformity (8) Achondroplasia (3) Craniostenosis (3) Down syndrome (1) Charge syndrome (1) Pycnodysostosis (1)	OSAS (10) Obesity (3) Turner sd (1) Noonan syndrome (1) Laryngomalacia (4) Laryngeal paralysis (1) Tracheomalacia (1)	COPD (4) BPD (1) Cystic fibrosis (4)

NM/SC Neuromuscular/scoliosis, *MF* maxillofacial, *OSAS* obstructive sleep apnea syndrome, *COPD* chronic obstructive pulmonary disease, *BPD* bronchopulmonary dysplasia

Table 2 Types of industrial interfaces used in the study population for long-term noninvasive positive pressure ventilation

Interfaces with manufactured leaks		Interfaces without manufactured leaks	
Manufacturer	Model	Manufacturer	Model
Nasal masks		Nasal masks	
Fisher and Paykel	Zest, 406, 407	Fisher and Paykel	406, 407
ResMed	Mirage FX	ResMed	Ultra Mirage
	Ultra Mirage		
	Pixi		
Respironics	Comfort classic	Respironics	Comfort gel
	Comfort gel		
	ComfortLite 2		
	Optilife		
Sleepnet	MiniMe		
Weinmann	Joyce Silkgel	Weinmann	Joyce Silkgel
Facial masks		Facial masks	
ResMed	Ultra Mirage facial	ResMed	Ultra Mirage Facial
	Quatro FX		
	Mirage quatro		
Respironics	Comfort Full		
Weinmann	Joyce Silk Gel	Weinmann	Joyce Silk Gel
	Joyce Full Face		Joyce Full Face
Nasal prongs			
ResMed	Swift LT-F, Swift FX		
Respironics	Opus		
Weinmann	NP15		

Interfaces

All patients ≤2 years old and four of the older children (two with maxillofacial deformity, one with OSAS, and one with spinal muscular atrophy) were fitted with custom-made nasal masks. The different industrial interfaces used in the remaining children are listed in Table 2. The interfaces were classified as nasal masks, facial masks, and nasal prongs and were separated into those interfaces with and without manufactured leaks, respectively. Figure 2 shows the first-choice interface for the patient population. Of the 67 children who received an industrial interface: 49 used a nasal mask, 16 a facial mask, and two nasal prongs. Industrial masks with and without manufactured leaks were used in 33 (34%) and 35 (36%) children, respectively.

Figure 3 shows the distribution of the different types of interfaces according to the underlying disease. All patients older than 2 years with OSAS with or without facial deformity used interfaces with manufactured leaks, with the exception of one patient in the MF group who had a severe maxillary retrusion and was fitted with modified nasal prongs (occlusion of the manufactured leaks) in order to be effectively ventilated with a volume-targeted mode. All patients with neuromuscular disease or thoracic scoliosis, with the exception of one patient, used interfaces without manufactured leaks. Interfaces with and without manufactured leaks were used equally in patients older than 2 years with lung disease. In 80% of the patients, the most appropriate interface could be determined after the first night.

The interface had to be changed in 20 patients because of discomfort ($n = 16$) and/or leaks ($n = 4$), facial growth ($n = 3$), skin injury ($n = 2$), and/or change of the ventilatory mode ($n = 2$) (Table 3). Nine patients needed a second mask change and among these nine patients, four with maxillofacial deformity required a third mask change. Changes were done within the same interface category or by switching to another trademark or using a different interface category.

Examples of custom-made masks, a nasal mask, and nasal prongs are shown in Figs. 4, 5, 6, 7.

Discussion

This study is the first to report the selection of the interface for long-term NPPV in a large cohort of children. The main advantages of the study are the large number of children treated in a recent and short time interval, ranging in age from <1 year up to 18 years, and the variety of underlying diseases. Despite the large experience of our NPPV team and close collaboration with a pediatric plastic surgery and maxillofacial team, the initial interface had to be changed during the study period in 20 (21%) children, mainly due to facial discomfort.

A large number of industrial interfaces are available, although the range is lower for children than for adults. It may thus be difficult for the physician to select the optimal interface for an individual patient. The choice will be

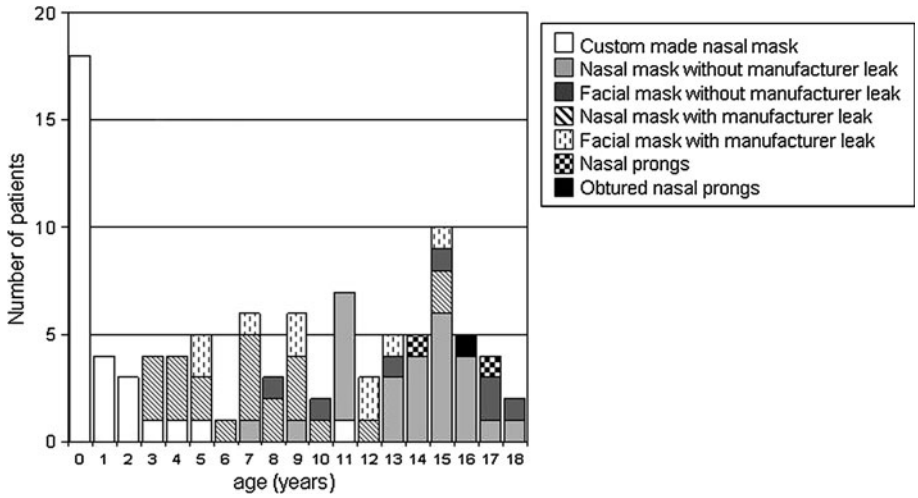

Fig. 2 First mask choice in the total population

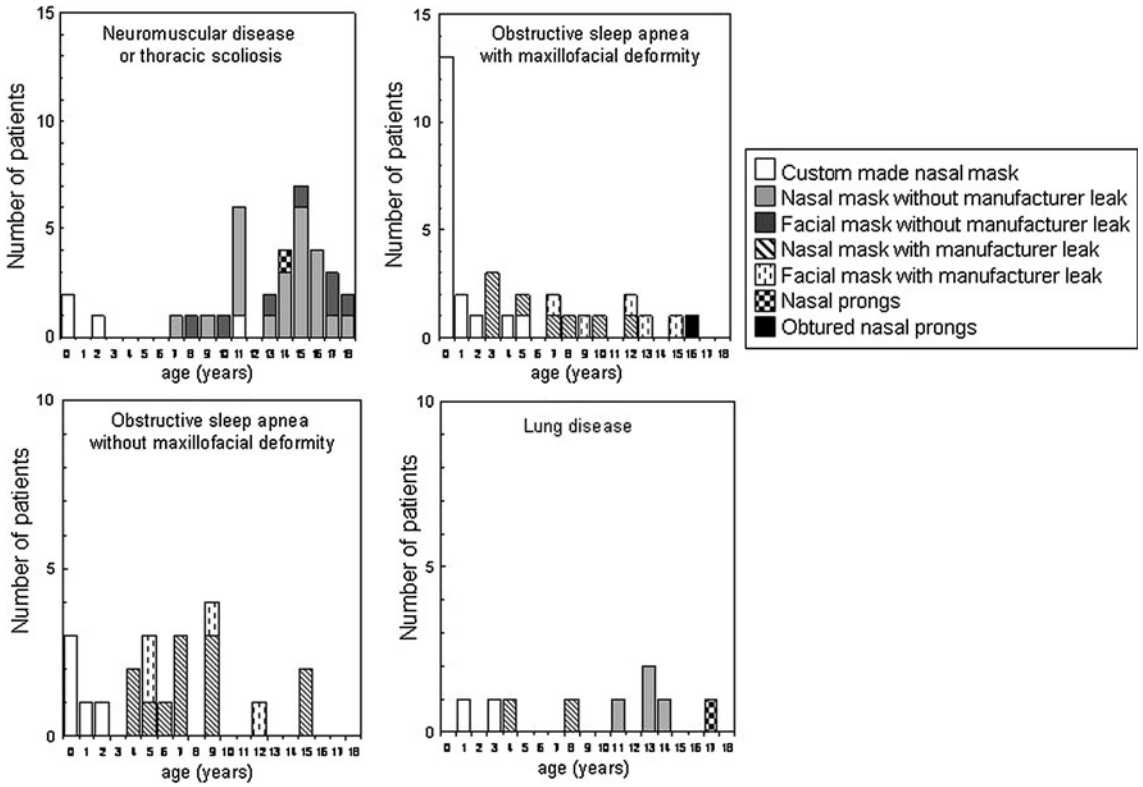

Fig. 3 Types of masks used according to the underlying disease

guided by the age of the patient, the underlying disease, the ventilatory mode (allowing an interface with or without a manufactured leak) and, most importantly, the facial physiognomy of the child. The larger use of "leak" ventilation in clinical practice, due to the large number of children having obstructive airway disorders, explains the greater number of interfaces with manufactured leaks in all age categories. However, in clinical practice, we have fitted the same proportion of patients with interfaces with and without manufacturer leaks, possibly due to the large number of patients with restrictive lung disease in our cohort. The ability of the child to keep his/her mouth closed during sleep will determine the choice of a nasal or a facial mask. However, we are reluctant to use a facial mask in a child who is not able to remove the mask by him/herself. This explains why facial masks were only used in children $\geq$5 years of age having no neuromuscular disease or a reduced mobility of the upper limbs.

Table 3 Masks changes and reasons for change during the study period

Age (years)	Pathology	First mask	Second mask	Third mask	Fourth mask
Neuromuscular group					
7	Congenital myopathy	N Discomfort	NP NPPV mode change		
9	Congenital myopathy	N Discomfort	NL NPPV mode change		
11	Spinal muscular atrophy	CM Discomfort + growth	NP		
11	Congenital myopathy	NL Discomfort + skin injury	NL Facial deformity	NP	
11	Congenital myopathy	N Discomfort + facial deformity	NPmod		
15	Congenital myopathy	N Discomfort + NPPV mode change	N		
16	Spinal muscular atrophy	N Discomfort	NPmod		
17	Muscular dystrophy	N Discomfort	N Discomfort + skin injury	NPmod	
Maxillofacial group					
3	Treacher-Collins	NL Discomfort	NL Discomfort + leaks	FL Discomfort + leaks	NL
4	Down syndrome + macroglossia	CM Growth	NL Discomfort	NL Discomfort	NL
5	OSAS + facial deformity	CM Discomfort	NL Facial deformity	NP leaks	NL
8	Maxillofacial deformity	NL Discomfort + leaks	NP Leaks	NL Leaks	FL
OSAS group					
4	OSAS	NL Discomfort + leaks	NL		
7	OSAS	NL Discomfort + leaks	NL		
7	Obesity	NL Leaks	FL Discomfort	F	
12	OSAS	FL Discomfort	FL		
Lung group					
1	BPD	CM Growth	NL		
11	Cystic fibrosis	N Discomfort	F Discomfort	F	
13	COPD	N Discomfort + NPPV mode change	NL Discomfort	NP	

CM custom made, *NL* nasal with manufacturer leak, *FL* facial with manufacturer leak, *NP* nasal prongs, *N* nasal without manufacturer leak, *F* facial without manufacturer leak, *NPmod* nasal prongs with obstruction of manufacturer leak, *NPPV* noninvasive positive pressure ventilation, *OSAS* obstructive sleep apnea syndrome, *BPD* bronchopulmonary dysplasia, *COPD* chronic obstructive pulmonary disease

Nasal prongs were successfully used in two adolescents of 14 or 17 years of age, respectively. This more recent type of interface is very well tolerated by patients because of the absence of a frontal support, which allows the patient to continue without much hindrance normal daily activities, such as reading, writing, and watching television. The exchange of a nasal mask for nasal prongs was associated with a marked reduction in maxillary retrusion in an adolescent who developed severe facial deformity within a few months after the start of NPPV. However, nasal prongs have manufactured leaks and can thus only be used with CPAP or BiPAP ventilation. Another adolescent with advanced neuromuscular disease developed such a severe maxillary retrusion that we decided to modify the nasal prongs by occluding the manufactured leaks in order to be able to deliver effective volume-targeted ventilation. Indeed, none of the BiPAP settings was able to correct his nocturnal hypoventilation and he did not tolerate high inspiratory pressures. We had thus to make the difficult choice between a tracheostomy, which the patient and his family refused, and the not recommended, but effective, modification of an industrial interface. This exceptional case underlines the legal and ethical problems associated with long-term NPPV in selected patients.

Infants with maxillofacial malformations, such as those with Pierre Robin sequence, represent an increasing

Fig. 4 A newborn baby with facial hypoplasia ventilated with a custom-made mask. Note the use of a pacifier

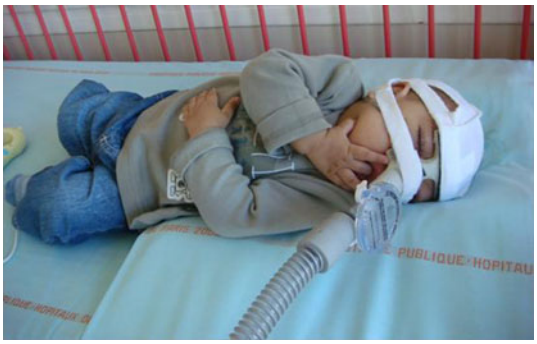

Fig. 5 An infant with a Pierre Robin sequence and a custom-made nasal mask

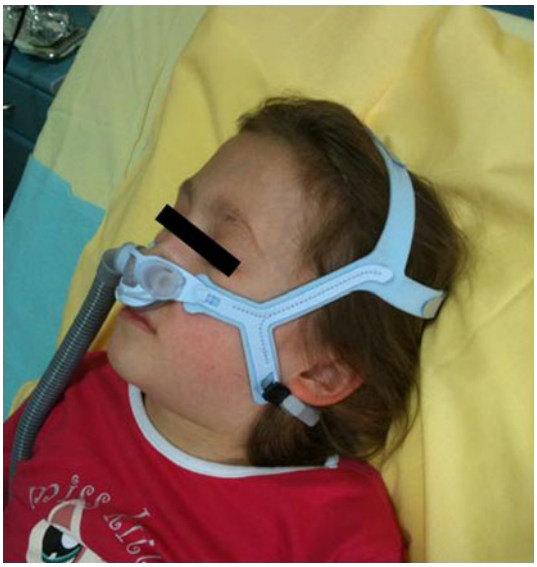

Fig. 6 A 7-year-old girl with Recklinghausen disease and tracheal compression ventilated with a nasal Pixi mask (Resmed, St Priest, France) without a frontal support

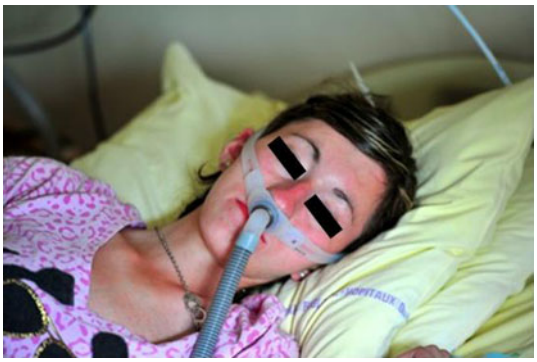

Fig. 7 An adolescent with nasal prongs (Swift LT-F Resmed, St Priest, France)

group of young children who may benefit from NPPV as an alternative to the more invasive tracheostomy [7] (Fig. 5).

It has to be noted that the interfaces listed in Table 2 reflect the experience and approaches of our unit and not the total number of interfaces available for children. However, we do note that there is still a lack of industrial interfaces for young patients, which limits the use of NPPV in this age group.

The main reason for mask change was discomfort (Table 3). Leaks as justification for the change of interface occurred in only four patients, and mainly in patients with a maxillofacial deformity due to the difficulty in adapting the interface. This low number may be explained by the careful selection of the most appropriate interface during the initiation period in the hospital. The rapid growth of the facial structures in young children explains why the interface had to be changed in three patients. The switch of a "non-leak" for a "leak" ventilation was also a reason for exchanging an interface without a manufactured leak for another interface with a manufactured leak. Of note, all patients who required a third mask change belonged to the MF group, which may be explained by the difficulty in finding an appropriate mask for children with facial deformity. The long-term side effects of NPPV in children, such as facial flattening or maxillary retrusion, should not be underestimated [13–16]. A systematic close surveillance of the tolerance of the interface is thus mandatory in children treated with long-term NPPV. The interface should be changed or modified at

the first sign of intolerance, discomfort, inefficacy of NPPV because of leaks, or facial deformity.

In conclusion, despite an increase in the number of interfaces available for children, there is still a lack of appropriate interfaces for young children and of interfaces without manufactured leaks. There is also a shortage of nasal prongs and facial masks for young children. In our study, even after a careful selection of the most appropriate interface by an experienced NPPV and maxillofacial team, discomfort and side effects occurred in as many as 21% of the patients, justifying a systematic and close monitoring of the NPPV interface.

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Conflicts of interest None.

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