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UPDATE

Non-invasive positive-pressure ventilation in children in otolaryngology

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Summary
Introduction: Obstructive diseases of the upper airways are common in children and sometimes difficult to manage. Non-invasive positive-pressure ventilation (NPPV) consists of delivering continuous positive pressure during all or part of the respiratory cycle via a non-invasive interface (face mask or nasal mask, or nasal prongs). NPPV is the treatment of choice for severe obstructive sleep apnoea in children and should be considered prior to tracheotomy and is also indicated in the case of persistent sleep-disordered breathing following surgical treatment, a frequent situation in children with a malformation of the head and neck or upper airways.
Discussion: A simple ventilator, able to deliver continuous positive airway pressure, is sufficient is most cases in otolaryngology. The interface represents the major technical limitation of NPPV, especially in infants for whom no appropriate commercial interface is available. A sleep study before and after initiation of NPPV, followed by regular follow-up examinations, is essential to confirm correction of gas exchanges and sleep quality in response to NPPV.
Conclusion: Finally, NPPV must be performed in a specialized paediatric centre with specific expertise in this field.

Introduction

Upper airway obstruction is very common in children. The main aetiology is hypertrophied tonsils and adenoids, which are simple to diagnose and treat in the vast majority of cases, as, in the absence of concomitant disease or comorbidity such as obesity, tonsillectomy and adenoidectomy ensure complete correction of airway obstruction. In the case of residual obstructive syndrome after tonsillectomy and adenoidectomy, the combination of a leukotriene receptor antagonist and topical nasal corticosteroids has been demonstrated to be effective [1]. However, upper airway obstruction can also be due to rarer diseases such as malformations of the head and neck (Pierre Robin sequence, craniostenosis, pycnodysostosis, Franceschetti syndrome, achondroplasia, storage diseases)

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Figure 1 Infant with Pierre Robin sequence ventilated via nasal mask.

Figure 3 Patient with craniostenosis and severe obstructive sleep apnoea syndrome despite several neurosurgical and head and neck operations: NPPV via face mask.

and upper airways (congenital or acquired laryngotracheal stenosis, laryngomalacia, tracheomalacia, lymphangioma) (Figs. 1 and 2). In these diseases, surgical management is often partially effective to enlarge the airway, but in some cases surgery may be impossible or even contraindicated. Non-invasive positive-pressure ventilation (NPPV) is particularly useful in these situations by increasing the calibre of the upper airways throughout the respiratory cycle by maintaining continuous positive airway pressure (CPAP). NPPV is currently recommended before tracheotomy, which, even only a few years ago, remained the only solution after failure of surgery. This non-invasive ventilatory assistance technique is associated with much lower morbidity and mortality and a much better quality of life than tracheotomy [2—4].

NPPV is a ventilatory assistance technique that preserves the patient's upper airways in contrast with endotracheal intubation, laryngeal mask, and tracheotomy. It consists of delivering positive pressure during all or part of the respiratory cycle via a nasal mask, face mask or nasal prongs, depending on the patient's age, tolerance, or facial morphology (Fig. 3). NPPV has been extensively developed in infants over recent years and now occupies an important place in the management of upper airway obstructive disease, as CPAP maintains or increases the calibre of the upper airways throughout the respiratory cycle. The major advantage of NPPV is maintenance of a minimum positive pressure throughout the respiratory cycle, during both inspiration and expiration.

Figure 2 Obese patient ventilated via nasal prongs.

The efficacy of NPPV is due to the fact that resistance to flow of a fluid in a tube, like air in the upper airways, is proportional to the inverse of the radius to the power of four [5]. A moderate increase of airway diameter, maintained patent by CPAP, therefore results in a marked reduction of resistance. As in adults, NPPV is preferably used in infants during sleep, associated with more severe airway obstruction, as sleep is associated with alteration of respiratory mechanics resulting in abnormalities of the ventilation-perfusion ratio, increased airway resistance and decreased residual functional capacity. Although diaphragmatic muscle activity is preserved, the activity of intercostal muscles and upper airway muscles is significantly decreased during sleep. Finally, ventilatory control and the activity of chemoreceptors are less effective during sleep than during waking. All of these changes explain the physiological reduction of alveolar ventilation during sleep. This hypoventilation is most marked during REM sleep, which explains why infants with upper airway obstruction are particularly vulnerable during this phase of sleep.

Due to its non-invasive nature, its relative ease of use, and the fact that it can be used on demand, NPPV is the preferred technique for home ventilatory assistance. Education of the parents and the child takes several days in a specialized centre, even for young infants. NPPV therefore now constitutes the first-line treatment for severe airway obstruction in children and a mandatory alternative to tracheotomy (Fig. 4).

Discussion

Why treat upper airway obstruction in children?

Many studies have demonstrated the harmful consequences of obstructive sleep apnoea syndrome in children. The correlation between apnoeas and sleep fragmentation related

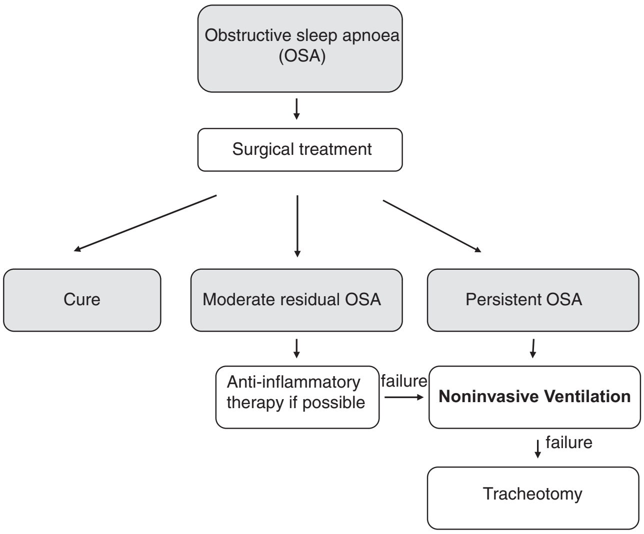

Figure 4 Place of NPPV in upper airway obstructive diseases in children.

to arousals (real or cortical) has been clearly established. Sleep fragmentation has many consequences, sometimes permanent, on the child's neurocognitive development (memory and attention disorders), behaviour (hyperactivity, irritability, agitation, or even depression), and cardiovascular (hypertension) and metabolic parameters (systemic inflammation and glucose intolerance) [6—10]. A number of these disorders observed in adults could be due to undiagnosed and untreated obstructive sleep apnoea during childhood.

Indications for NPPV

No validated criteria have been defined for initiation of NPPV in children with upper airway obstructive disease. The indication for NPPV is generally based on the presence of obstructive sleep apnoea on polysomnography (PSG) associated with suggestive clinical signs such as apnoeas observed by the family, persistent snoring, poor quality sleep with frequent arousals, agitated sleep, tiredness on awakening and during the day, enuresis, and especially irritability and concentration and memory difficulties (Fig. 5). Nocturnal blood gas abnormalities with hypoxaemia and nocturnal hypercapnia are a sign of severity and justify initiation of NPPV. A trial of NPPV can also be considered when PSG shows only moderate abnormalities, with poor sleep efficacy and fragmented sleep, but with the presence of very suggestive clinical signs such as those indicated above. The decision to stop or continue NPPV is then based on the results of another PSG and clinical assessment performed after one month of NPPV, depending on the benefit observed. Sleep quality questionnaires validated in children are very useful in these situations.

NPPV requires a certain degree of autonomous ventilation and this treatment cannot be indicated in the most severe patients who are unable to breathe unassisted for at least 15 to 30 min. However, NPPV has a residual effect, with persistence of the beneficial effects during spontaneous breathing between periods of NPPV after several days of regular use.

The efficacy of NPPV has been demonstrated in newborns and young infants with marked upper airway obstruction in the context of Pierre Robin sequence [11]. After initiating NPPV in the intensive care unit, these infants who initially had less than 30 min of ventilatory autonomy, were able to return home with NPPV at night and while sleeping during the day, without the need for tracheotomy (Fig. 1).

NPPV can also be indicated to facilitate extubation, which would otherwise be impossible due to persistent obstruction during sleep [12] (Fig. 6).

However, it is difficult to maintain NPPV 24 h a day in the long term and a minimum degree of cooperation is also essential. Nevertheless, except in the case of severe psychomotor retardation, the child's refusal of NPPV is an exceptional cause of failure. Other techniques such as hypnosis or sophrology can be particularly useful in these children who experience expectant anxiety due to their history of painful or traumatic surgery. In every case, NPPV constitutes an alternative to tracheotomy and must always be discussed and attempted before considering tracheotomy.

Whenever possible, initiation of NPPV must be planned ahead and explained to the child and the family. Initiation of NPPV in a specialized centre is particularly impor-

Figure 5 Patient with Willi-Prader syndrome: NPPV via nasal mask.

Figure 6 Patient tracheotomised at the age of 1 month due to congenital laryngeal palsy and extubated as a result of NPPV via nasal mask.

tant in order to choose the most appropriate ventilator interface, but also for therapeutic education of the child and the family. In the very great majority of cases, no major problems are encountered during this initiation and adaptation phase, especially when NPPV is presented as an alternative to tracheotomy.

Modalities of NPPV

The objective of NPPV in the context of upper airway obstruction is to maintain sufficient opening of the upper airways throughout the respiratory cycle during sleep. This requires selection of the most appropriate ventilator and interface. The ventilators used for NPPV are CPAP ventilators with a single-limb circuit and clearance of carbon dioxide during expiration via a calibrated leak system, either integrated into the interface or into the ventilator circuit. Many small, lightweight, and inexpensive ventilators are available for home NPPV. These ventilators are not battery-equipped and have limited alarms, but as this type of ventilation is not generally ''survival ventilation'', these options are not essential. Bilevel positive airway pressure (BiPAP) consists of delivering a higher pressure during inspiration. The advantage of BiPAP ventilation compared to CPAP in terms of efficacy or comfort in children with upper airway obstruction has not been demonstrated [13]. Delivery of this higher pressure during inspiration also implies that the ventilator is able to detect the patient's inspiratory trigger, which can be very difficult in young infants due to the very low flows and volumes involved, that cannot be detected by the machine [14,15]. The air delivered by the ventilator is humidified and reheated by the nasal fossae, but in the youngest infants, addition of a warm humidifier can improve tolerability and comfort of NPPV, especially during prolonged use during the night and daytime sleeping.

The major technical limitation of NPPV in young infants concerns the choice of interface [16]. Although many nasal and face masks are available for older children, very few commercial masks are available for infants and young children (Fig. 1). No satisfactory commercial mask is currently adapted to infants weighing less than 10 kg. Custom-made masks that can only be made in specialized centres are therefore necessary for these patients [16,17]. Nasal prongs are useful interfaces for children unable to support a face mask or nasal mask or with facial deformities (Fig. 3). Active research is currently underway to develop adapted and easy-to-use interfaces for routine NPPV in young children [16,18].

In practice, NPPV in children must be performed in a specialized multidisciplinary centre with specific expertise. It is initiated during the day, preferably while napping, and then at night. In young infants, NPPV is usually initiated in the arms of one of the parents. A nasal mask is preferably used, as it is smaller than the face mask, allowing use of a teat in infants, which, apart from its soothing properties, also promotes mouth closure and avoids leakages. The NPPV adjustment period, comprising selection of the most suitable equipment, correct settings, and familiarization of the child and the parents with the equipment, generally takes 2 days to 2 weeks with a mean of 3 to 5 days depending on the child's age and acceptance. A minimum of 6 h of continuous use is required to allow the child's return home. Sleep recordings are performed during the initial adjustment phase and arterial blood gases and sleep quality must be normal prior to discharge. Home surveillance is performed by a homecare technician and/or nurse, who must have paediatric training. The homecare provider must be contacted while the child is still in hospital and must be informed about the date of discharge so that the first home visit can be held on the day of discharge. In every case, parents are given written documents providing comprehensive information on NPPV and the names of people to be contacted in the event of problem 24 h a day, 7 days a week. Close and coordinated collaboration between the sleep centre and the homecare provider is essential for follow-up of home NPPV in children.

Limitations, complications

Complications of NPPV in children are rare and minor. The main problems encountered are technical: poorly adapted or incorrectly assembled circuit, ventilator dysfunction, defective interface. These incidents are rare with CPAP ventilators. Complications of NPPV are essentially related to the interface and consist of skin lesions and facial deformities [17]. Skin complications can be prevented or treated by regularly changing the interface and/or the use of a custom-made mask with regular review of acceptability. Custom-made masks induce fewer skin complications than commercial interfaces [17]. Facial deformities such as flattening or retrognathia are more difficult to prevent or treat, but NPPV often constitutes only temporary treatment for upper airway obstruction, and patients can be usually weaned after several months or several years. Facial deformities can resolve after stopping NPPV. Paediatric maxillofacial follow-up is therefore systematically recommended before and during NPPV. When NPPV needs to be continued, various alternative interfaces can be proposed, including nasal prongs, which have the advantage of not requiring any forehead support [16]. New nasal prongs for very young infants should be available in the near future, which should considerably reduce the rate of skin complications and facial deformities.

Follow-up

Surveillance of NPPV must be conducted in a centre specialized in NPPV in children, ideally attached to a paediatric maxillofacial unit in order to prevent and detect facial complications related to the interface [17]. This surveillance must be clinical, respiratory, ENT and maxillofacial, technical, or even nutritional. Overnight hospitalisations for sleep recording on NPPV are performed regularly at a frequency determined by the child's clinical status and age. The child is generally reviewed about one month after discharge, then every 2 to 6 months depending on the child's age and clinical status. The interface, ventilator and circuit, ventilator settings and at least nocturnal arterial blood gases and sleep quality with NPPV and the facial and skin tolerance of the mask are verified during overnight hospitalisations. Any signs of skin irritation require a change or adjustment of the interface. The impact of the interface on facial growth is determined by clinical examination [17]. Finally, a complete annual assessment is recommended, comprising polysomnography on NPPV, waking arterial blood gases, and a facial assessment.

Regular assessments evaluate the course of the obstructive syndrome in order to possibly propose discontinuation of NPPV. NPPV must be withdrawn progressively, first during the young child's afternoon nap and then every second night or for half of the night, under clinical and polysomnographic surveillance. NPPV can be stopped when clinical examination and a sleep study on spontaneous breathing are normal, but surveillance must be continued, as, due to the residual effect of NPPV, the obstructive syndrome can recur after several weeks or months, constituting an indication for resumption of NPPV [12].

Conclusion

NPPV is the treatment of choice for severe upper airway obstruction in children after failure or impossibility of surgical treatment. It is indicated in a minority of children with upper airway obstructive disease, but it can be highly beneficial in these children by reducing the frequency and/or duration of tracheotomy. Although the adverse effects of NPPV are minor and despite its relative simplicity, this treatment must be conducted and monitored by a specialized paediatric centre.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

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