Specific Equipment Required for Home Mechanical Ventilation in Children

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Educational Aims

- No ventilator presently available is perfect and is able to adequately ventilate all types of paediatric patients requiring noninvasive positive pressure ventilation (NPPV).
- Numerous ventilators are proposed for paediatric NPPV, but their performances are highly variable and depend on the ventilatory mode, type of trigger and circuit, but, most importantly, the type of patient.
- The inspiratory triggers of most of the ventilators are insufficiently sensitive for infants.
- A systematic paediatric bench and clinical evaluation is recommended before the use of a ventilator in clinical practice.
- Few industrial interfaces are available for children, and this shortage is even more important for infants.
- A systematic paediatric maxillofacial evaluation and follow-up is recommended in children before, during, and after the use of NPPV because of the risk of facial deformity.

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Noninvasive positive pressure ventilation (NPPV) may be used in a growing number of conditions and diseases in children, both in the acute and the chronic setting. However, respiratory mechanics and maxillofacial development are different in children as compared to adults, which justify age-adapted ventilators and interfaces. Children requiring NPPV represent a heterogeneous group, not only with regard to the underlying disease but also with regard to age, weight and maxillofacial physiognomy. A paucity of interfaces is available for young children. Individually adapted interfaces may be mandatory for these patients. The ventilators available for home NPPV have been shown to have severe limitations, especially with regard to the detection of the patient's inspiratory effort and their ability to deliver the required volume or pressure within an acceptable time frame for the patient. A close collaboration of the industry with paediatric experts in NPPV should be able to overcome these technical challenges.

10.1 Principles

The aim of noninvasive positive pressure ventilation (NPPV) is to improve or normalise alveolar hypoventilation by means of a ventilator and a noninvasive interface. Both equipments, the ventilator and the interface, should be adapted to the young child, which constitutes a real challenge for the youngest patients. Indeed, breathing pattern differs in children as compared to adults.

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Tidal volume is smaller and respiratory rate is higher in children. In children, normal tidal volume is approximately 10 ml/kg, with a respiratory rate of 40 breaths/min at birth and 20 breaths/min at the age of 2 years. Respiratory failure is associated with a decrease in tidal volume and an increase in respiratory rate. A ventilator should thus be able to deliver small tidal volumes with a relatively high frequency. Also, when a spontaneous mode is used, the ventilator should be able to detect the onset of the patient's inspiratory effort (by means of a change in pressure or flow) and deliver a preset pressure or volume within a time delay compatible with the patient's respiratory rate. As such, trigger time delays exceeding 100 ms for young children are too long and inadequate because the patient may have finished his inspiration before the delivery of the pressure or volume by the ventilator (Fauroux et al. 2008a).

The respiratory effort, i.e. the negative intrathoracic pressure that the patient has to generate during inspiration, varies according to the underlying condition. This inspiratory effort may be extremely high in case of upper airway obstruction or lung disease such as cystic fibrosis (Fauroux et al. 2001a, b, 2004a; Essouri et al. 2005), but very low in case of neuromuscular disease, because of the weakness of the respiratory muscles. It may be difficult for a ventilator to detect the onset of the inspiration in a patient who has a very low inspiratory effort because the change in airway pressure or flow will be too small. NPPV in young children with an "extreme" breathing pattern may thus be very challenging, requiring a ventilator able to detect minor changes in airway pressure or flow and capable of an adapted response within a tight time frame, i.e. 100–150 ms (Fauroux et al. 2001a; Essouri et al. 2005). Such requirements are further challenged by leaks, which are unavoidable during NPPV. Leaks are the main cause of ineffective ventilation with persistent hypercapnia, patient-ventilator dyssynchrony and NPPV failure (Gonzalez et al. 2003). The detrimental effects of leaks are more pronounced in young patients in whom the volume of leaks may represent a greater percentage of the tidal volume. The ratio between the tidal volume and the volume of the interface is important with regard to rebreathing. Indeed, a large interface, with regard to the patient's tidal volume, will increase the risk of rebreathing.

The anatomy of the facial bones and the proportions between the facial elements are different in children as compared to adults. The anatomy of the maxillofacial structures changes continuously during growth, which is particularly rapid during the two first years of life. Interfaces for NPPV need thus to be adapted specifically to the facial anatomy and physiognomy of children (Fauroux et al. 2005). They need to be changed frequently, especially within the first months of life. Skin injury occurs as a consequence of pressure sores which are defined as a lesion on any skin surface that occurs as a result of pressure. The principal causative factor is the application of localised pressure to an area of skin not adapted to the magnitude and duration of such external forces. Tissue damage will occur if both a critical pressure threshold and a critical time are exceeded. The soft tissue beneath the skin is thinner in children as compared to adults. They are thus at greater risk of skin injury during NPPV. The risk of skin injury is also higher in young children because they need NPPV during extended periods including nocturnal sleep and daytime naps (Fauroux et al. 2005). The repetitive pressure applied by NPPV on the facial structures may hinder the normal development of the facial bones. Facial growth occurs predominantly in an anterior and sagittal axis in children. NPPV hinders this normal facial growth causing facial deformity. Facial flattening and maxilla retrusion are commonly observed in children receiving long-term NPPV and justify a systematic evaluation and follow-up by a paediatric maxillofacial surgeon before and during NPPV (Fauroux et al. 2005).

10.2 Home Ventilators Adapted to Paediatric Needs

Numerous ventilators are proposed for paediatric NPPV, but their performances are highly variable and vary according to the ventilatory mode, type

of trigger and circuit, but, most importantly, the type of patient (Fauroux et al. 2008a, 2004b). Indeed, the same ventilator may be able to correctly ventilate an adolescent with end-stage cystic fibrosis lung disease but not an infant with congenital myopathy.

The choice of the best ventilator for a particular patient is a real challenge for the physician, not so much in the paediatric intensive care unit (PICU) where most of the ventilators have a NPPV mode, as in the home setting. Indeed, home ventilators become more sophisticated and tend to integrate continuously new options and measures. A large number of ventilators are now able to deliver different ventilatory modes, such as PS, with or without positive end-expiratory pressure (PEEP), as well as volume-targeted ventilation. Different circuits (simple, double or leak circuit) and triggers (pressure or flow triggers) may be available on the same ventilator. We have recently shown that the performance of a ventilator may vary according to the ventilatory mode or the type of trigger and circuit but also according to the patient profile (Fauroux et al. 2008a). Indeed, when we evaluated the performance of 17 home ventilators with six different paediatric patient profiles, our conclusions were (1) no ventilator is perfect and was able to adequately ventilate all the six different patient profiles, (2) the performance of the ventilators was very heterogeneous and depended on the type trigger and circuit and, most importantly, on the characteristics of the patient, and (3) the sensitivity of the inspiratory triggers of most of the ventilators was insufficient for infants. A systematic paediatric bench and clinical evaluation is thus recommended before use of a ventilator in clinical practice.

The first ventilators available for home NPPV were either volume- or pressure-targeted devices. Presently, all the home ventilators are able to deliver different, and sometimes combined, ventilatory modes. The simplest devices are pressure-targeted devices that deliver either a continuous positive airway pressure (CPAP) or a bilevel positive airway pressure, i.e. a higher positive pressure during inspiration and a lower positive pressure during expiration. These devices are used with a simple circuit and a leak, generally inserted in the mask, to allow carbon dioxide (CO_2) clearance. During CPAP, the device should be able to maintain the most constant positive airway pressure during the whole breathing cycle in patients with different diseases or conditions despite changes in respiratory mechanics (such as an increase in airway resistance or a decrease in lung compliance) or leaks. We have shown in a bench study using an adult profile that devices are able to measure the pressure loss in the circuitry and are able to adjust the pressure under dynamic conditions outline the other devices (Louis et al. 2010a). However, these observations have to be validated in the paediatric population. During a bilevel positive pressure ventilation, the ventilator should be able to detect the onset and the termination of the patient's inspiratory effort in order to optimise the synchronisation of the ventilator to the patient. The inspiratory and expiratory triggers of these devices may be fixed or adjustable with generally different levels of sensitivity. The principles and functioning of these triggers is often not very clear and, most importantly, not sufficiently sensitive for young children. Indeed, we have shown that the triggers of these devices are not able to detect the inspiratory effort of infants <18 months of age (Essouri et al. 2005). In these young infants, the use of a bilevel mode was associated with an important patient-ventilator dyssynchrony which resulted in an increase in the work of breathing. In this age group, CPAP is thus preferred to bilevel positive airway pressure. The lack of guarantee of minimal tidal volume or minute ventilation is a limitation of these devices. As such, some recent models have a "volume-guarantee" module, i.e. the possibility to compensate, by increasing the airway pressure and sometimes the inspiratory time, for an insufficient tidal volume. However, we have shown that these modules are not very efficient, especially during air leaks (Fauroux et al. 2010).

Other devices are more sophisticated and are able to deliver a positive pressure or volume with or without a positive airway pressure during expiration. These devices may be used with a simple or a double circuit. In case of a simple circuit, the CO_2 is cleared during the expiration by means of an expiratory valve. This expiratory valve should

Table 10.1 Interfaces for noninvasive positive pressure ventilation in children >8–10 kg

	Nasal mask	Facial mask	Nasal prongs	Mouthpiece
Age	Every age	Every age	Adolescent	Child and adolescent
Contraindications		Gastroesophageal reflux, patient without autonomy		
Advantages	Small volume, comfortable, allows eating and speaking, allows the use of a pacifier in infants	No mouth leaks	No facial contact	Use at libitum (neuromuscular patients)
Limitations	Mouth leaks	Large volume (risk of rebreathing)	Mouth leaks	Ability to seal lips around the mouthpiece (leaks)

have a low resistance to avoid a supplementary expiratory effort (Lofaso et al. 1996). When a spontaneous mode is used, the quality of the inspiratory and expiratory triggers is of major importance. We have shown that the quality of the triggers varies greatly among the different ventilators but also for a specific ventilator, according to the type of circuit, the interface and the patient profile (Fauroux et al. 2008a). Another importance technical requisite is the ability of the ventilator to reach the preset pressure or volume within a time frame and to maintain a constant airway pressure during the whole duration of the inspiration for pressure support (PS) (Fauroux et al. 2008a). This also varies significantly between the different ventilators (Fauroux et al. 2008a, 2004b).

In conclusion, numerous ventilators are proposed for home ventilation in children. In clinical practice, it is recommended to use the simplest ventilator to which the team is familiar, the most important issue being to check the correction of the nocturnal hypoventilation and the adaptation of the patient to the ventilator.

10.3 Machine Patient Interfaces

The interface is a major determinant of the success of NPPV. Different types of industrial interfaces can be used for home NPPV (Table 10.1). Unfortunately, there are no satisfactory interfaces for infants. These patients can thus only be ventilated with custom-made masks. The choice of the appropriate interface will depend on the ventilatory mode, the age of the patient, his autonomy with regard to the use of the interface, the necessity of daytime ventilation and the skin and facial tolerance. In children, a small interface is preferred because the risk of rebreathing increases with the increase of the interface's size. Moreover, a smaller interface will be less claustrophobic and generally better accepted and tolerated than a larger one. As such, nasal masks are preferred over facial masks. The latter are also strictly forbidden in children who do not have the ability to remove the masks by themselves, such as young children and those with neuromuscular disease, because of the potential risk of inhalation in case of reflux.

Air leaks are the main limitation of NPPV, especially with nasal masks. Leaks are the main cause of persistent hypercapnia (Gonzalez et al. 2003; Paiva et al. 2009). Simple practical measures, such as changing the mask, using a chin strap, increasing minute ventilation and changing the type of the ventilator, may be able to reduce the volume of air leaks and improve the efficacy of ventilation (Gonzalez et al. 2003; Paiva et al. 2009).

Skin erythema or irritation is also a major side effect of the interface (Fauroux et al. 2005). This complication has to be detected and prevented as early as possible by changing the interface or making a custom-made mask. The pressure applied by the interface on the growing facial structures of a child can induce facial deformities. A systematic maxillofacial evaluation

before, during and after the eventual withdrawal of NPPV is mandatory. The interface needs also to be changed frequently in young children, because of their rapid facial growth. It may be recommended to use and alternate different (types of) masks to minimise the risk of skin injury and facial deformity. Systematic humidification of the ventilator gas is not necessary for NPPV because of the respect of the upper airway. However, nasal intolerance due to excessive dryness can resolve after humidification of the ventilator gas. Most importantly, the evaluation of the efficacy of NPPV should consider the "couple" interface + ventilator because the change of the interface or the ventilator may modify the effectiveness of NPPV (Louis et al. 2010b).

10.4 Practical Organisation and Backup Systems

The major advantage of NPPV is that it can be applied at home, combining greater potential for psychosocial development and family function, at lesser cost. The use of home NPPV requires appropriate diagnostic procedures, appropriate titration of the ventilator, cooperative and educated families and a careful, well-organised follow-up. Prior to discharge, the patient's respiratory status should be stable on the actual ventilator and circuit the child will use at home, at least for several days. Settings on a home ventilator do not provide the same ventilation in the child as the same settings on a hospital ventilator, and the efficacy of home equipment must be evaluated in each child prior to discharge. Once the child is at home, and as the child grows, ventilator settings must be evaluated to ensure adequate gas exchange on a regular basis. Although the optimal frequency for these evaluations has not been determined, these evaluations should be performed more frequently in infants and small children because of their rapid growth. Sleep evaluations are recommended as a diagnostic tool before the initiating of NPPV, then as a control test of the efficacy of NPPV before discharge with the ventilator, and as a surveillance test during an overnight hospital admission during follow-up (Paiva et al. 2009). Careful extrapolation should be made from a sleep evaluation performed during daytime naps because this does not always reflect what happens during the night. Routine and emergency service must be available. Providers/home care equipment technicians and nurses should visit the patient at home at least every month to perform preventive maintenance and check of the interface and the ventilator. Evaluation of compliance should be systematically checked by counters on the equipment determining the amount of time the ventilator is effectively used and not only turned on.

Patients requiring home NPPV may need other supportive therapies. Cough-assisted techniques, by means of the mechanical insufflator-exsufflator, for example, are recommended in every patient with neuromuscular disease who is started on NPPV. The mechanical insufflator-exsufflator is very efficient and well-tolerated in helping the patient clear his respiratory secretions (Fauroux et al. 2008b). Oxygen therapy at home must be justified on the basis of an individual-based medical necessity, as determined by appropriate physiological monitoring, with pulse oximetry monitoring during periods of sleep, wakefulness, feeding and physical activity and arterial blood gases. CO_2 levels should be minimised first by ventilator use before considering oxygen therapy, especially for patients with neuromuscular disorders and obstructive sleep apnoea. It is important to remember that supplemental oxygen is not a replacement for assisted ventilation in patients who hypoventilate.

Children are frequently undernourished when starting NPPV. Chronic respiratory insufficiency is frequently associated with an increase in energy consumption, anorexia and malabsorption. Adequate nutrition is critical for growth and development of the lungs and the chest wall. Nutritional support, via a nasogastric tube, is frequently necessary during the first weeks or months. This can be performed by fashioning a port in a custom-made nasal mask, if gastrostomy feeding is not planned. In infants, discoordination of swallowing mechanisms is frequent, and

swallowing function should be evaluated to assess pulmonary aspiration risks. Many patients have also associated gastroesophageal reflux, which may require surgical correction. This can be combined with a gastrostomy if necessary.

It is essential that the child, if the age permits it, and the parents should have the opportunity to discuss the NPPV therapy in advance. Discussion should start long enough before the anticipated need to allow the child and the family to evaluate options thoroughly and to discuss their feelings. NPPV has here an essential first place as a noninvasive therapy but still represents an objective element reflecting a further step in the severity of a disease. It is crucial to determine short-term and intermediate-term goals of NPPV with the child and the family and to explain the principles of NPPV. A wide range of ventilators and masks are available, and great care will be taken to choose the most appropriate equipment and settings. The final objective is that NPPV translates into well-being and a better quality of life, with a total adherence of the child and his family. In progressive diseases such as some neuromuscular diseases, ventilatory failure may progress at daytime. These patients are generally equipped with a second ventilator. Diurnal NPPV, by means of the patient's usual nocturnal interface or a mouthpiece in older children, may be effective and can delay the discussion for a tracheotomy (Toussaint et al. 2006). However, in some patients, a tracheotomy will become necessary at a certain moment. Close monitoring of the patient's physiological status and disease progression, together with clear information of the family, is essential.

Future Perspectives

- Develop adequate and well-tolerated interfaces for young children.
- Develop home ventilator specifically adapted for young children.
- Develop and validate alternatives to polysomnography for the evaluation of the efficacy of NPPV at home in children.

Essentials to Remember

- No ventilator is perfect and is able to adequately ventilate all the different types of paediatric patients who may justify NPPV.
- The performance of ventilators is very heterogeneous and depends on the type trigger and circuit and, most importantly, on the characteristics of the patient.
- The sensitivity of the inspiratory triggers of most of the ventilators is insufficient for infants.
- The evaluation of the efficacy of NPPV should consider the "couple" interface + ventilator because the change of the interface or the ventilator may modify the effectiveness of NPPV.
- Patients should be regularly and closely monitored after NPPV initiation, to ensure a perfect correction of alveolar hypoventilation and to prevent and detect side effects of NPPV, such as skin injury or facial deformity.

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