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Non-invasive positive pressure ventilation in children ☆

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ABSTRACT

Non-invasive positive pressure ventilation is increasingly used in children both in acute and in chronic setting. Clinical data supporting safety, efficacy and limitations in children are growing. Technical problems related to the ventilators performance and interfaces selection have not been fully resolved, especially for younger children. Non-invasive ventilation can be applied at home. Its use at home requires appropriate diagnostic procedures, accurate titration of the ventilators, cooperative and educated families and careful, well-organized follow-up programs.

1. Introduction

An increasing number of children with chronic hypercapnic respiratory failure are currently treated with non-invasive ventilation [1]. Acute accesses to the emergency department or in-hospital follow-up controls of patients undergoing home long-term non-invasive ventilation are increasingly common occurrences [2–4].

Non-invasive ventilation has some undoubted peculiarities. By definition, it is a non-invasive technique, which can be applied on demand and preferably at night, causing less morbidity and discomfort [1,2]. It also allows preserving important functions such as swallowing, feeding, speaking, and coughing. Heating and humidification of the inspired air are greatly respected [5].

The introduction of non-invasive ventilation in pediatric practice has allowed the reduction of the number of children destined to tracheostomy, the limitation and/or the delay of the intubation of children with acute respiratory failure and/or exacerbation of chronic respiratory failure [5]; and also helped reduce the length of stay in the pediatric intensive care units of children who have been extubated and weaned from invasive ventilation in a shorter time [6]. Avoiding intubation prevents vocal cords or trachea damages, and reduces the risk of lower respiratory tract infections [7].

Non-invasive ventilation has also a better impact on quality of life of the patient with respect to the tracheostomy [5,10].

2. Definition, indications and objectives on non-invasive ventilation

There are two types of respiratory support: “invasive” and “non-invasive”. The distinction depends on the type of interface used for patient-ventilator connection. For non-invasive ventilation, gases are conducted into the airways via an external interface, in the case of invasive ventilation through an endotracheal tube or tracheostomy [2,10].

Non-invasive ventilation in children is indicated essentially for: 1) Diseases due to increased respiratory load (intrinsic cardiopulmonary disorders, abnormalities of the upper airways, especially skeletal deformities of the chest wall); 2) Disorders characterized by weakness of the respiratory muscles (neuromuscular diseases, spinal cord injuries); 3) Abnormal neurological control of ventilation (congenital or acquired alveolar hypoventilation syndrome) [2,5]. Main indications for non-invasive ventilation in children are summarized in Table 1. Authors' experience on non-invasive ventilation is shown in Fig. 1. Fig. 2 shows the increasing number of patient treated by non-invasive ventilation in the authors' institution since 1993.

Non-invasive ventilation can alleviate chronic respiratory failure through the correction of hypoventilation, the improvement of respiratory muscles function and reducing the workload of the respiratory system. An effective reduction of nocturnal hypercapnia by mechanical ventilation leads to an improvement of the daytime carbon-dioxide (CO_2), during spontaneous breathing [1]. According to Mehta et al. [10], objectives of non-invasive ventilation can be summarized into short and long-term goals. Short-term (including acute) goals are: relief from symptoms; reducing the work of breathing; improvement and stabilization of gas exchanges; optimization of the level of comfort; good patient-ventilator synchrony; minimization of risks; and avoiding intubation. Long-term goals are: improvement of the duration and quality of sleep, maximizing the quality of life, improvement of the functional status, and prolongation of survival.

3. Patient selection

Long-term non-invasive ventilation is not applicable to all children, as it requires a degree of cooperation, being more difficult to use in younger patients [2,5,10,11]. The ideal candidate for long term non-invasive ventilation should be a cooperative and in stable clinical condition patient [5,11]. Nocturnal or constant hypercapnia ($PaCO_2 > 55$–

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Table 1
Indications for NIV in children (modified from reference [11]).

Neuromuscular disorders
Duchenne muscular dystrophy
Spinal Muscular Atrophy
Nemaline myopathy
Abnormalities of the rib cage and chest wall
Progressive juvenile idiopathic scoliosis
Asphyxiating thoracic dystrophy (mild)
Advanced Cystic Fibrosis complicated by hypercapnia
Obesity-related disorders of ventilation
Prader–Willi Syndrome
Morbid obesity associated with obstructive sleep apnea
Overlap syndromes (obstruction of the upper airways and restrictive pulmonary dysfunction)
Spina bifida (pulmonary complications, Arnold–Chiari malformation, restrictive pulmonary dysfunction, obstruction of the upper airways)
Cerebral palsy (laryngeal dystonia and restrictive pulmonary dysfunctions)
Chronic obstruction of the upper airways
Obstructive sleep apnea complicated by hypercapnia
Down syndrome (maxillary hypoplasia, large tongue)
Craniofacial syndromes with midface or mandibular hypoplasia
Laringotracheomalacia
Chronic obstructive airways disease
Advanced Cystic Fibrosis
Disorders with central alveolar hypoventilation
Central Congenital Alveolar Hypoventilation Syndrome (CCHS)
Central Secondary Alveolar Hypoventilation
Rapid Onset Obesity with Hypoventilation, Hypothalamic, Autonomic Dysregulation Syndrome (ROHHAD)

60 mm Hg) should be present [11]. Indispensable prerequisite to its use is the presence of a certain degree of respiratory autonomy. Usually, it is applied at night and/or during daytime sleep (especially in younger children) [5]. Patients requiring ventilation throughout 24 h/day are usually not candidates for non-invasive ventilation [10,11].

4. Interfaces selection

The interface choice depends on the characteristics of the patient (age, facial characteristics, degree of cooperation, and severity of respiratory impairment). Regardless of the interface used, it is fundamental to limit the air leaks that may reduce the effectiveness of the ventilation [2].

In children, the interface acceptance is the first step for a successful non-invasive ventilation program. Interfaces should have good adhesion, a low resistance to airflow and should be light. The interface should exert less pressure on the skin that can be compatible with effective ventilation and the dead space volume should be minimized [1,11].

Many commercial interfaces are actually available for children. Nasal masks are the most often used interfaces, but there is getting a growing experience with oro-nasal and full-face masks [5,11]. Nasal pillows and mouthpieces are alternative options. In the authors' experience 93% of children use nasal-masks; 6% of children use oro-nasal or full-face masks; and the lasting 1% use other interfaces.

Transparent and with different form interfaces should be always used for each patient. The transparency facilitates a more easy and immediate inspection of the correct positioning [2,11]. The different forms reduce the risk of patient's discomfort, the development of side effects and facial deformities [5,11].

Accessories that allow keeping the interfaces in place are available for each type of them. A comfortable sealing, reducing the risk of air leakage, make them easier to use.

Helmet ventilation is a method that uses an interface consisting of a transparent helmet placed around the patient's head through which is possible to realize positive pressure ventilation. The helmet can be attached to the patient by axillary shoulder straps, waist belt and in the younger children through a "diaper" fixing system. Helmets are equipped with a protection system antisuffocation with automatic opening and a porthole airtight access to the patient [12].

5. Ventilators

Non-invasive ventilation in children can be performed with volume or pressure-targeted ventilators, according to the control variable through which the ventilator produces the inspiration [2,10]. A ventilator is in volume-controlled mode (volume-targeted ventilation) if the wave profile of the flow of gas delivered during inspiration does not change with changes in the compliance and resistance of the respiratory system [2,10]. The mechanical act generates a predefined volume of air or mixture of air/oxygen. A ventilator is pressure-controlled (pressure-targeted ventilation) if the wave profile of the pressure delivered during inspiration is not affected by changes in compliance or resistance of the respiratory system. The mechanical act generates a predefined pressure [2,10].

Some modern home ventilators provide both systems, and hybrid ventilations can be performed. Pressure-targeted ventilation is the most often used non-invasive ventilation modality [2,5,11]. It can be administered in various ways that can be characterized by being bound to strict parameters set by the physician, or flexible and modifiable by the patient himself by reason of its variable requests [10].

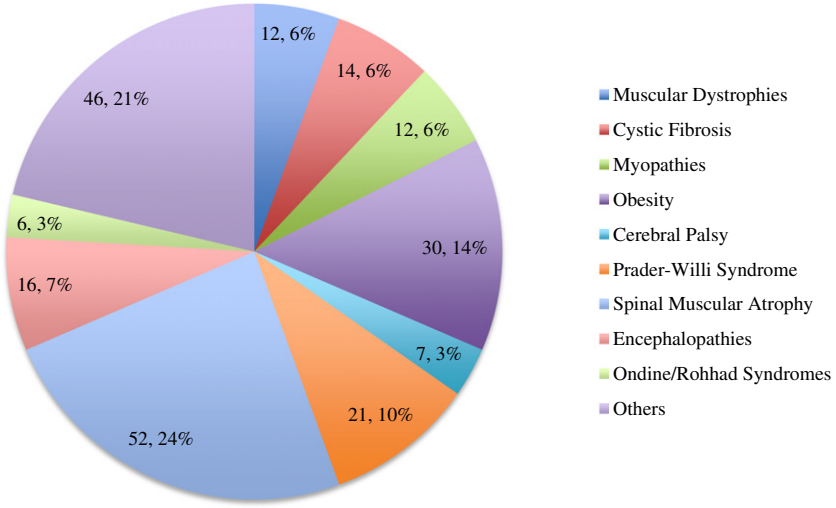

Fig. 1. Indications for non-invasive ventilation: authors' experience at Bambino Gesù Children's Research Institute (Rome, Italy).