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REVIEW ARTICLE

Noninvasive ventilation in pediatrics

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Abstract

Objective: To introduce the notion of noninvasive mechanical ventilation as an alternative for invasive ventilation in children, describing advantages and disadvantages, indications, and the process of equipment installation.

Sources of data: Literature review through PubMed/Medline, using as source articles focusing on noninvasive ventilation in pediatric populations, as well as consensus statements and metanalyses concerning noninvasive ventilation in adults.

Summary of the findings: The main indication for noninvasive ventilation is respiratory failure due to hypercapnia. It is contra-indicated in the presence of hemodynamic instability. The advantages of noninvasive ventilation include: the equipment is easy to install; it is not invasive and involves less discomfort; there is a lower incidence of complications associated with the endotracheal tube; lower cost. The disadvantages are: gastric distention; transient hypoxemia; facial skin necrosis. The equipment required for noninvasive ventilation includes an interface (mask) and a respirator. The removal of noninvasive ventilation equipment is relatively simple, but chronic patients may require assistance in the home.

Conclusions: Noninvasive ventilation is a less costly, efficient and simple to perform alternative in cases of respiratory failure without hemodynamic instability.

J Pediatr (Rio J) 2003;79(Suppl 2):S161-S68: Mechanical ventilation, noninvasive ventilation, pediatrics, respiratory failure.

Introduction

Since invasive mechanical ventilation was first used as a therapy for respiratory failure, complications have come to light associated with oral endotracheal intubation: ulceration or edema of the mucosa, hemorrhage, stenosis, pneumonia or sinusitis associated with invasive ventilation.[1]

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Noninvasive ventilation (NIV) is a therapeutic alternative in this context. This provides the liberation of mechanical pulmonary ventilation without the use of an artificial airway such as an endotracheal tube or tracheostomy cannula. The theoretical advantages of increasing alveolar ventilation without an artificial airway include: avoid complications associated with endotracheal tubes, improve patient comfort, preserve airway defense mechanisms, the tongue and deglutition. In addition, NIV offers great flexibility in terms of applying and withdrawing mechanical ventilation.[1]

Available methods for noninvasive mechanical ventilation include: negative external pressure, chest wall oscillation and mechanical ventilation by positive pressure via mask, which will be the subject of this article.

During the 1970s and 80s, two methods for noninvasive ventilation with positive pressure, using a facial or nasal mask were introduced into clinical practice:

– CPAP (Continuous Positive Airway Pressure), to improve oxygenation in patients with acute respiratory insufficiency with hypoxemia.
– IPPV (Intermittent Positive Pressure Ventilation), in order to increase ventilation and rest respiratory musculature in patients with chronic respiratory insufficiency, due to neuromuscular diseases and/or chronic obstructive pulmonary disease (COPD).

NIV objectives

Sufficient ventilation depends on the capacity of respiratory musculature to contract and the individual's demand, in addition to an efficient central respiratory drive. Any pathology which compromises this balance can lead to ventilation failure and prejudice gas exchange. Contraction abnormalities may surface as a result of intrinsic muscle weakness, as happens with patients with neuromuscular disease, or when muscles are forced to work at a mechanical disadvantage, as happens with patients with chest deformities. Increased demand may occur because of airway obstruction, either upper or lower, and because of alterations to pulmonary compliance. Alterations to respiratory drive may occur as a result of sedative drugs or congenital anomalies.[2]

In this context noninvasive mechanical ventilation is used with the objective of reducing areas of atelectasis and improve gas exchange[2] (Figures 1 and 2).

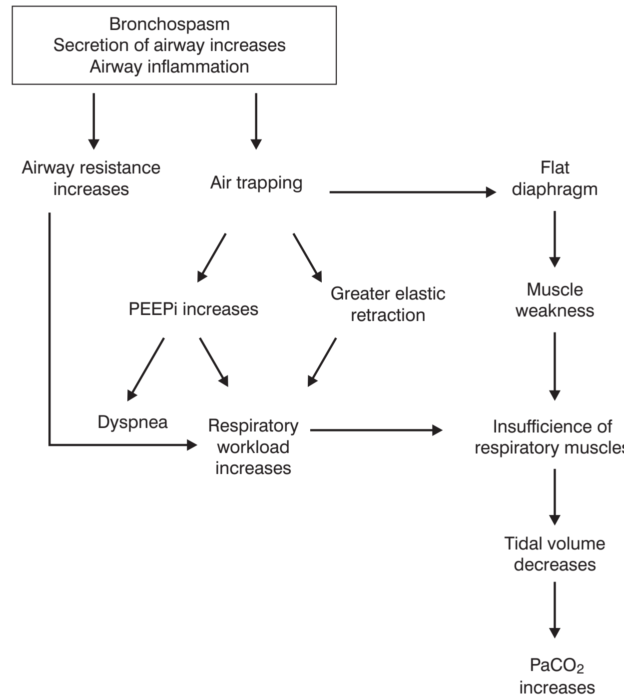

Figure 1 - Hypercapnic respiratory failure can occur due to increased respiratory workload (COPD, asthma) or normal respiratory workload (neuromuscular diseases). Its treatment involves increase of the alveolar ventilation (increase of the tidal volume and/or RR) and decrease of the respiratory effort

Indications and contraindications of NIV

Indications

The main indication for Noninvasive ventilation is respiratory insufficiency, primarily when there is hypercapnia, although it can also be used for some other hypoxemic pathologies, such as, for example, for weaning Table 1).

Childhood chronic lung disease: there are no studies of the pediatric age group. In adults with acute phase COPD the use of noninvasive ventilation reduces the need for intubation and also mortality. Furthermore it rapidly improves acidosis and reduces respiratory frequency. Complications are fewer and length of hospital stay is shorter with patients who have had noninvasive ventilation.[8]

Chest wall alterations (anatomic and functional): Kyphoscoliosis can distort the whole of the rib cage and interfere with inspiration, impeding deep breathing and coughing. Because of difficulties clearing secretions and achieving adequate ventilation, these patients can develop infections and alterations to gas exchange. Obese and ascitic patients, when in the supine position, have their diaphragms forced downwards by their abdominal organs, reducing residual functional capacity and compromising diaphragmatic contraction during inspiration.

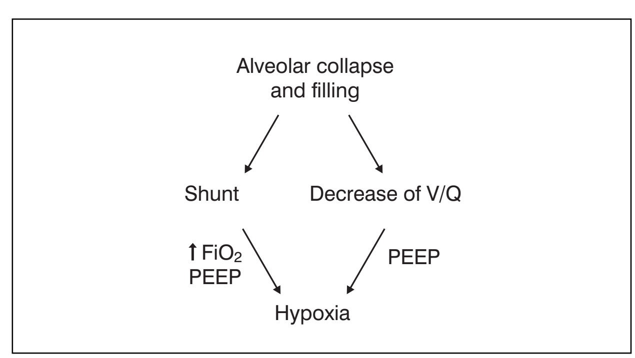

Figure 2 - Hypoxemia can occur due to hypoventilation (when there is elevation of $PaCO_2$) and due to the alteration of the ventilation/perfusion (V/Q) ratio. It is treated with increase of FiO_2 and alveolar recruitment when applying PEEP

Table 1 - Indications for non-invasive ventilation

Respiratory insufficiency with hypercapnia	**Respiratory insufficiency with hypoxemic pathologies**	**Other indications***
Aacute phase COPD (pH < 7.35)	Pneumonia	IMV weaning
Chest wall alterations	ARDS	Patients who do not need intubation
Neuromuscular diseases	Postoperative period	Thoracic trauma without pneumothorax
Central hypoventilation Obstructive sleep apnea	Acute pulmonary edema	

* Regardless the type of respiratory insufficiency.

Neuromuscular diseases: in a series of acquired or congenital neuromuscular diseases, respiratory failure is inevitable and is very often the terminal event. Some of these conditions are listed in Table 2.[3]

Support with noninvasive ventilation should be begun as soon as the first episode of hyperventilation is observed. Noninvasive ventilation is best indicated in patients with hypoventilation who have normal or near-normal bulbar function and who can breath without the apparatus for a certain amount of time. For children who are totally dependent on the apparatus, with deglutition difficulties or hyper secretion, a tracheostomy should be considered.[3] Despite the existence of evidence that efficient ventilatory support can improve the quality of life of these children and reduce the number of hospitalizations,[5,6] a study carried out with Duchenne muscular dystrophy, failed to demonstrate improvements in the incidence of hypercapnia or the number of episodes in which forced vital capacity fell 20% below initial values due to the preventative use of noninvasive ventilation during an eight-hour nocturnal period. Furthermore, the group on prophylactic noninvasive ventilation had increased mortality.[7] Therefore, the use of this method for prophylaxis is not indicated whereas for patients with symptoms of hypoventilation and hypercapnia it is.

Central Hypoventilation: central hypoventilation syndrome is defined as the absence of automatic respiratory control - drive. The majority of children are more seriously

Table 2 - Neuromuscular diseases associated to the ventilatory insufficiency

Congenital	
Muscular dystrophy	*Duchenne, Becker, congenital*
Congenital myopathies	
Congenital neuropathies	*Congenital motor-sensory neuropathies*
Glycogen storage diseases	*Acid maltase deficiency*
Spinal muscular atrophy	*Types I, II, III*
Myotonic dystrophy	*Congenital*
Mitochondrial myopathies	*Kearns-Sayre, Leigh, MELAS syndrome*
Acquired	
Polyneuropathies	*Guillain Barre syndrome*
Polymyositis	
Poliomyelitis	
Endocrinopathies	
Myastenia gravis	*Juvenile, infantile*
Nonprogressive chronic encephalopathy	

affected during their sleep. The treatment is to offer ventilatory support during sleep to overcome the absence of central drive. Traditionally this was carried out by means of a tracheostomy and positive pressure ventilation, however, we can also ventilate via the mask with positive pressure and avoid subjecting the child to a tracheostomy, since during the day breathing is voluntary and so without difficulties.[3]

Obstructive sleep apnea: during active stages of sleep (REM), there is a loss of pharyngeal muscle tone. In children with a predisposition to narrowing of this region during to anatomical alterations (Pierre-Robin, achondroplasia, craniofacial abnormalities or obesity), this loss of tone can cause a significant obstruction, with gas exchange affected. In these situations, the use of CPAP via mask may be solution until it is possible to plan surgery with more time available, meaning that a child can be allowed time to grow before being subjected to surgical stress.[3,4] Guilleminalt et al. evaluated 74 children with sleep disorders due to upper airway obstruction, 74 of whom were successfully treated with CPAP.[4]

Asthma or bronchiolitis: we did not find any controlled and randomized trials showing the use of NIV to be of benefit in children with asthmatic crises. In adults, Meduri et al.[9] demonstrated benefits from the use of noninvasive ventilation with 17 patients with asthmatic conditions, only two of whom required intubation. In a series of pediatric cases, Padman et al. demonstrated reductions in respiratory frequency, heart rate, dyspnea and improved oxygenation in more than 90% of the patients studied.[10] While NIV has been used with success in cases of asthmatic crises, both with adults and children there is yet insufficient evidence to recommend its routine employment.[11,12]

Pneumonia: there are few studies of pediatric patients. Fortenberry et al.[22] used NIV with 28 patients with hypoxemic respiratory failure and concluded that NIV can be used with safety in children with requiring better oxygenation due to mild to moderate hypoxemic respiratory failure. There are no controlled and randomized trials comparing noninvasive, with invasive or conventional treatment in children with hypoxemic respiratory failure.

Acute pulmonary edema: there are at least two controlled and randomized trials on adults which demonstrate that CPAP via facial mask rapidly improves vital signs and reduces the need for intubation.[13] We do not know of any controlled and randomized studies of the pediatric age group.

Weaning: prolonged weaning times are associated with increased risks of complications associated with ventilation, while, on the other hand, early weaning and later reintubation is a relatively common problem in ICUs. The introduction of noninvasive ventilation at an early stage for such patients appears highly attractive. The objective would be to reduce the time taken to weans and, therefore, complications associated with ventilation and so reduce the need for reintubation in borderline patients.[13] We do not know of any controlled and randomized studies that have been carried out on pediatric populations.

Contraindications for NIV

Contraindications are:

- Surgery, trauma or deformity of the face (impeding the use of a mask).
- Total obstruction of the upper airways.
- Failure of airway protective reflex.
- Respiratory hypersecretion.
- High risk of bronchoaspiration (vomit or upper digestive hemorrhage).
- Undrained pneumothorax
- Non respiratory organ failure: severe encephalopathy (Glasgow < 10), digestive hemorrhage, cardiac arrhythmia and hemodynamic instability (including SHOCK).

Remember that these contraindications may be relative, especially in the absence of personnel trained in intubation and in those patients who are not intubation candidates.[12,13]

Advantages and disadvantages of NIV

Advantages of NIV

Noninvasive nature: ventilation via mask is easy to start and stop. In an acute situation, it takes less time than intubation and avoids complications associated with the insertion of an endotracheal tube. During mechanical ventilation, Noninvasive positive-pressure ventilation eliminates the respiratory work imposed by the endotracheal tube. During weaning NIV eliminates the necessity of reintubation associated with early weaning extubation.

The invasive nature of endotracheal intubation is the primary motive used to justify the postponement of mechanical ventilation until more advanced stages of acute respiratory insufficiency. However the failure to rest respiratory muscles can lead to necrosis of musculature and, eventually, prolong the duration of mechanical ventilation.[14] Furthermore, mortality rates are higher among patients with respiratory insufficiency who are not intubated and develop respiratory arrest..

Weaning and extubation in conventional mechanical ventilation is made more difficult by concern about the need to reintubate. With NIV it is simpler, in the event that its removal is not effective, the mask is just refitted.

The average duration of NIV is shorter than with conventional intubation with an endotracheal tube. Factors which may be involved in this include: earlier intervention, less sedation and pain relief, reduced incidence of respiratory muscular atrophy (generally induced by conventional mechanical ventilation with an endotracheal tube), the elimination of respiratory work imposed by the endotracheal tube and a lower incidence of complications (particularly infections).[15,16]

Reduced discomfort: the pain resulting from the presence of the endotracheal tube in the oral cavity is the main source of discomfort among intubated patients. Furthermore, endotracheal intubation prevents the patient from speaking. Communication with relatives and health professionals is frustrating because of the inability to verbalize, which interferes with the ability to cooperate. The patient becomes agitated and is treated with sedatives, further compromising communication. The majority of patients with a facial or nasal mask tolerate them relatively well and present a gradual reduction in dyspnea.[17]

Reduced incidence of complications: nosocomial pneumonia is a frequent complication of mechanical ventilation and an important factor in the prognosis of mortality. Endotracheal intubation short circuits the airway barrier defenses, compromising mucociliary clearance, resulting in epithelial cell peeling, leading to greater bacterial adhesion and tracheal colonization. In addition it serves as a route by which microorganisms can enter the tracheobronchial tree.

A number of different laryngeal, pharyngeal and tracheal complications are caused by endotracheal tubes. These complications may occur at the moment of intubation (prolonged intubation attempt, right main stem bronchus intubation, arterial hypotension, airway damage), during the intubation period (mechanical malfunction of the endotracheal tube, leaking cuff, laryngeal ulceration), and after removal (stridor due to obstruction of the upper airways, hoarseness, difficulties with deglutition, stenosis of the trachea).[1,18]

Sinusitis is a common cause of fever with no obvious focus and of bacteremia in patients on mechanical ventilation. The risk is related to the presence of tubes in the nasopharynx and the duration of ventilation, and can, therefore, be minimized by NIV.

Disadvantages of NIV

Gastric distension: gastric distension is a rare occurrence among patients treated with NIV. It happens because in order to open the lower esophageal sphincter a pressure of more than 33 ± 12 mmHg is needed. Children are theoretically safe from significant gastric distension at pressures of up to 25 mmHg. When air enters the stomach during NIV a fluttering sound can be heard in the epigastrium and palpation reveals vibration. Respiratory physiotherapists and nurses should be trained to observe signs of abdominal distension.

Transitory hypoxemia: hypoxemia may result from the removal of the mask when supplementary oxygen is not offered. The use of continuous oximetry and suitable ventilator alarm settings are crucial to rapid intervention.

Necrosis of facial skin: the most common complication of NIV is the development of necrosis of the skin where the mask comes into contact, with an approximate incidence of 10%. Of the factors that contribute to skin necrosis, prominence should be given to tissue hypoxia due to the pressure of the mask. After removal of the mask, the lesion heals. The use of protection where the mask is fitted can avert necrosis.

NIV fitting

Material necessary

In addition to a mechanical ventilator, an interface is necessary to apply NIV: the mask. The mask may be facial (covering nose and mouth) or nasal, of a number of different shapes and sizes, including some with protection for areas of pressure (e.g. nasal protectors). They are attached to the face with elastic straps in the manner of a bridle and its suitability to the patient defines success or failure of the procedure, from which comes the importance of having a variety of masks available. Normally a facial mask is used for the first 24 hours and after the patient improves it is changed for a nasal one[12] (Figure 3).

Different types of masks have advantages and disadvantages. The main differences are summed up in Table 3.

Table 3 - Advantages and disadvantages of different types of mask

Type of mask	Advantages	Disadvantages
Facial mask	Better ventilation	Claustrophobia
	Less leakage	Speaking is not possible
	Skin lesion	Gastric insufflation
Nasal mask	Feeding and speaking	Less efficient
	Easy to adjust	Leakage through mouth
Double nasal tube	Less pressure on skin	Difficult to adjust
	No claustrophobia	Leakage through mouth

Types of ventilator

While there are many ventilators designed specifically for NIV (bi-level), in principle any ventilator is capable of performing noninvasive ventilation. Conventional respirators separate the inspiratory and expiratory gas mixtures, which prevents reinhalation and permits monitoring of inspiratory pressure and exhaled ventilation by minute, parameters on which the alarms are based. Ventilators of the bi-level type have only one circuit for gas and exhalation can be active (the ventilator opens an

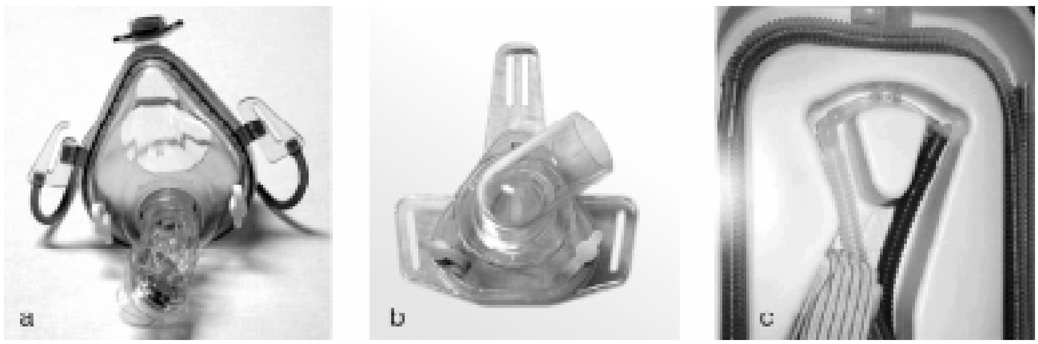

Figure 3 - Types of mask: **a)** facial mask, **b)** nasal mask, **c)** double nasal cannula

exhalation valve) or passive (expiratory positive airway pressure - EPAP- directs the expired air to leave via an exhaust valve). They are cheaper, easier to use and portable, they have an input valve for oxygen and do not need humidification[12,13] (Figure 4).

Ventilation modes

Controlled mechanical ventilation (CMV): total respiratory support, with no need for effort on the part of the patient. With respirators for NIV ventilation cycled by time is indicated for patients with ineffective respiratory drive. These respirators have an adjustment for the time in which a defined pressure will be achieved: if short pressure increase will be rapid, if longer the increase will be more progressive, a ramp, and more comfortable for the patient.

Figure 4 - Equipment used for noninvasive ventilation - BIPAP vision

Controlled assisted ventilation: a certain number of breaths is defined to be offered to the patient in the absence of effort. As the respirator delays its cycle when the patient exhibits respiratory effort, this mode is known as synchronized: SIMV, on conventional respirators or S/T (spontaneous/timed) on respirators for NIV.

Spontaneous assisted ventilation: better known as pressure supported ventilation (PSV). Inhalation by the patient triggers the respirator which offers a set pressure. It is important that the NIV apparatus has adjustable minimum respiratory frequency, for cases of respiratory pause (back up for apnea).

Continuous positive airway pressure (CPAP): is a way of offering oxygen with a little pressure, generally average airway pressure at around 5 cm H_2O. It improves ventilation in collapsed areas, and is much used via nasal in neonatology and via mask for patients with carcinogenic pulmonary edema and on respiratory physiotherapy.

Bi-level support pressure: the mode in which NIV respirators normally function: there is a support pressure (as with PSV), called IPAP (inspiratory positive airway pressure), and also an EPAP (expiratory positive airway pressure), which works as a CPAP, as it is on the same circuit.

Initial parameters

Below are some suggestions for initial parameters[12] (Table 4), which should be customized to the individual patient. Not all apparatus has all of the settings although that would be the ideal.

Table 4 - Suggestions for initial parameters for noninvasive ventilation

Mode	IPAP	EPAP	RR	Triggers	I:E ratio back up
S/T	12-15 cmH_2O (up to 20)	3-5 cmH_2O	5 ipm	Maximum sensitivity	1:3

IPAP: inspiratory positive airway pressure, EPAP: expiratory positive airway pressure, RR: respiratory rate.

Monitoring and control nosocomial infections

Monitoring

Minimum clinical assessment: level of consciousness, movement of the chest wall, use of accessory muscles for breathing, patient comfort, synchronization of respiratory effort with respirator cycle, respiratory frequency and heart beat. Should be sufficiently frequent to evaluate response to treatment and adjust respirator parameters.

Assessment with gasometry: after 1-2 hours of NIV and then after 4-6 hours, if the first showed little improvement. If there is no improvement in pH and $PaCO_2$ after this period, invasive ventilation should be considered.[12]

Oxygen saturation: should be monitored continuously, at least for the first 24 hours and supplementary oxygen should be administered to maintain $SatO_2 > 90\%$.

Control of nosocomial infection

While disposable masks and exhalation valves do exist in order to reduce the risk of cross-infection they increase costs considerably (especially since more than one mask is tried per patient before the one that fits best is chosen). Generally we use masks and circuits that are re-sterilizable and which pass through a process of washing, disinfection and sterilization (e.g. with heat or ethylene oxide). The elastic straps also have to be washed at sterilizing temperatures. There is also a bacterial filter which can be coupled to the exit of the respirator when in use. Also remember to wash the surface of the respirator between uses.[12,13]

Treatment failure

Before deciding that there has been a failure of treatment, it is a good idea to certify that treatment was adequate.

Was treatment of the primary disease suitable?

- Has medication prescribed been administered?
- Consider respiratory physiotherapy.

Were there complications?

- Pneumothorax, bronchoaspiration.

$PaCO_2$ remains elevated?

- Is the patient receiving too much O_2? Adjust FiO_2 to maintain $SatO_2 > 90\%$.
- Is much air leaking? Adjust the mask. If a nasal mask is being used check if a facial mask reduces leakage.
- Check if connections are correct and there are no holes in the circuit.
- Has re-inhalation occurred? Check that the exhalation valve (when present) is pervious (it may be blocked by secretions) and consider increasing EPAP
- Is the patient synchronized with the respirator? Observe the FR adjustment and I:E ratio (if assisted controlled). Check inspiratory and expiratory triggers (if adjustable). Consider increasing EPAP (if a COPD patient).
- Is ventilation adequate? Observe chest expansion. Increase IPAP or volume. Consider increasing inspiratory period or FR. Consider other ventilatory modes, if available.

$PaCO_2$ reduces, but PaO_2 remains low.

- Increase FiO_2.
- Consider increasing EPAP.

In the event that all this has been checked and the patient has not presented improvements, consider therapeutic failure - invasive ventilation is indicated.[12,13]

NIV Withdrawal

All patients put on NIV for acute hypoxemic respiratory failure should be subjected to spirometry and arterial gasometry in room air before discharge.[9]

All patients with spinal marrow damage, neuromuscular disease, chest wall deformities or morbid obesity should be assessed for home ventilation requirements.[12]

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