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Mouthpiece ventilation and complementary techniques in patients with neuromuscular disease: A brief clinical review and update

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Abstract

Noninvasive ventilatory support (NVS) is sometimes reported as suboptimal in patients with neuromuscular disease (NMD). The reasons for this include inadequate ventilator settings and/or lack of interface tolerance. NVS has been used for many years in patients with NMD disorders as a viable alternative to continuous ventilatory support via a tracheostomy tube. The mouthpiece ventilation (MPV) is a ventilatory mode that is used as daytime ventilatory support in combination with other ventilatory modalities and interfaces for nocturnal NVS. However, there is still a poor understanding of this method's benefits compared with other modalities. This review aims to highlight the indications and advantages along with the disadvantages of MPV.

Keywords

Neuromuscular disease, noninvasive ventilatory support, mouthpiece ventilation, daytime ventilatory support, complementary techniques

Introduction

The respiratory muscles are rarely spared in neuromuscular diseases (NMDs) even if the type of muscle involvement, severity, and time course greatly varies among the different diseases.[1] The most common NMDs in childhood are Duchenne muscular dystrophy, spinal muscular atrophy, and congenital muscular disorders, which include a large group of congenital muscular dystrophies. In adults, amyotrophic lateral sclerosis, myotonic dystrophy, and limb-girdle muscular dystrophy are the most common NMDs, which benefit from noninvasive ventilatory support (NVS). Each diagnosis has a different rate of respiratory decline which is summarized in Table 1.[2]

Before 1953, non-invasive ventilation (NIV) was practiced with the use of negative pressure ventilators: the "iron lung." Most people under the age of 40 are unlikely ever to have seen an iron lung, yet 40 years ago, these devices would have been a common sight in most hospitals throughout the world. Through the late 1920s and into the 1950s, the iron lung was considered to be the state of the art, high tech, and life support technology. Indeed, medical students of the time would have learnt about such devices as a recommended treatment for respiratory paralysis, used to maintain life for those whose breathing capabilities have been impaired or destroyed by poliomyelitis. They were noninvasive in the sense that no part of the device invaded the patient. The whole body was enclosed within the airtight chamber of the device, apart from the head, which protruded through a tight seal around the neck. This

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Table 1. Neuromuscular diseases benefiting from NVS due the progressiveness of respiratory impairment.

SMA type 1	Rapid worsening (0–3 years duration)
SMA type 2	Slow worsening (>15 years)
SMA type 3	Slow worsening (>15 years)
Acid maltase deficit	Slow worsening (>15 years)
DMD	Intermediate worsening (5–15 years)
Myotonic dystrophy (Steinert disease)	Intermediate worsening (5–15 years)
LGMD	Intermediate worsening (5–15 years)
ALS	Rapid worsening (0–3 years duration)

SMA: spinal muscular atrophy; DMD: Duchenne muscular dystrophy; LGMD: limb-girdle muscular dystrophy; ALS: Amyotrophic lateral sclerosis; NVS: noninvasive ventilatory support.

method of artificial respiration became known as external negative pressure ventilation. Despite their great success as a continuous ventilatory support, ventilation through tracheostomy became the standard since the epidemic Danish polio in 1952, because it was possible to mobilize patients and secretions could be easily managed.[3]

In 1956, the Harris Thompson portable 28-lb Bantam, positive pressure ventilator, was another turning point. Mouthpiece use slowly grew in patients with severe ventilatory impairment (vital capacity less than 500 ml) and with inefficient cough, resulting in a reduced ventilator dependence and hypersecretion.[4,5] NVS via a mouthpiece was used in 257 patients at the Goldwater Memorial Hospital from 1968 to 1987 with excellent results.[6] Despite the remarkable results,[4–6] few centers in the United States and around the world used the NVS via a mouthpiece, and until 10 years ago, there were only sporadic reports of NVS mouthpiece use in this patient group whose numbers have been increasing with time.[7–9] Intermittent NVS through a mouthpiece has been increasing for patients requiring continuous ventilatory support, with more favorable outcomes and better survival, as an alternative to mechanical ventilation via a tracheostomy.[10] Some of the reasons for its underutilization include clinicians being cautious about a technique which looks unsecure, lack of knowledge on how to set it up; advances in mask technology (including nasal pillow systems); the high prevalence of NVS centers using only pressure cycled ventilators,[11] previous lack of commercially available equipment to secure the mouthpiece to the wheelchair or bed and financial disincentives for noninvasive management.[12]

The fundamentals

NVS leads to improvements in arterial blood gas tensions, relieves shortness of breath, rests the inspiratory muscles, reduces the incidence of nosocomial infections, decreases hospitalizations for respiratory failure, and decreases mortality.[13] The greatest limitation of this technique is that it is impossible to implement continuously if the interface is uncomfortable,[14,15] but fortunately, now there are over 100 types of interfaces. The appropriate interface is crucial for NVS success with flexibility to switch between different types of interfaces in order to change the pressure points of the mask. The ability to do this can improve patient adherence to NVS.[16] However, few clinical trials have compared the effects produced by different types of interfaces on clinical outcomes and none has evaluated the impact of interfaces on the respiratory workload.[17,18]

The most frequent causes of NVS failure with consequent intubation of NMD patients are due to inappropriate settings of the ventilator[19,20] and misuse of mechanically assisted cough (MAC) to eliminate airway secretions.[21–24] Failure of NVS may also occur due to serious bulbar dysfunction[25] with decreased upper airway patency limiting the efficacy of NVS, secretions management or to inappropriate administration of sedative drugs, and/or additional oxygen therapy.[13,26]

Currently, tracheostomy is offered to patients affected by NMD who need around the clock NVS even though they would prefer to continue to use NVS.[27] Tracheostomy may lead to an increase in care costs, complications, and social isolation.[6] Tracheostomy is considered suitable for patients with severe dysfunction of the glottis as these patients have higher risk of aspiration pneumonia. Its requirement for NMD patients without bulbar impairment is unproven regardless of severity of ventilatory failure.[28–31]

Rationale for the use of ventilatory support with a mouthpiece

Nasal and oronasal interfaces are the most commonly used interfaces for NVS.[14–16] They allow ventilation through the nose and/or nose and mouth, and they are suitable interfaces if the patient is not claustrophobic

Table 2. Beneficial effects of NVS for neuromuscular patients.

Improves patency of the upper airways
Normalizes gas exchange
Improves quality of sleep and maintains gas exchange improvement during the day (nocturnal NVS)
Reduces symptoms related to chronic hypoventilation
Favors the rest of respiratory muscles
Resets the sensitivity of central chemoreceptors
Improves lung compliance
Reduces complications secondary to intercurrent infections
Slows thoracic deformity and decline in lung function
Improves quality of life
Decreases morbidity and reduces mortality

NVS: noninvasive ventilatory support.

or has any facial pressure sores.[32] Nasal interfaces also include nasal pillows, which have the advantage of minimal contact with the face. Therefore, there is a lower chance of developing interface-related pressure sores and there is often the mask of choice in the claustrophobic patient.

However, they have the disadvantage of higher air leak when higher inspiratory pressure is administered (>15 cmH_2O).[14,16] NMD patients, who use nocturnal NVS with nasal or oronasal interfaces, who start requiring daytime ventilation due to the progression of inspiratory muscle weakness will benefit from the effects of NVS reported in Table 2. However, using a nasal or oronasal interface 24 hour/day can decrease social interaction, impair eating, drinking, and speech and changes a patient's perception of themselves. The latter may have psychologically damaging consequences.

The use of angled mouthpieces supported by a metal flexible arm (if the patient has no strength to keep the mouthpiece near to the mouth) is an ideal solution for daytime ventilation in patients with functioning mouth muscles and some preserved neck movements. In the selected patient, it is easy to apply and easy to use even during daily living activities such as eating and talking.[33] Despite these obvious advantages, this modality is not commonly used. However, its effectiveness in improving long-term survival has been documented in a series of more than 700 NMD patients who required continuous ventilatory support.[29,34]

There are no published evidence-based guidelines concerning mouthpiece ventilation (MPV). Its application is mainly based on the experience of few centers.[9,34,35,36] However, these centers are matching the technology to the patients ever changing clinical condition. Randomized controlled studies cannot be easily performed in this area.[37]

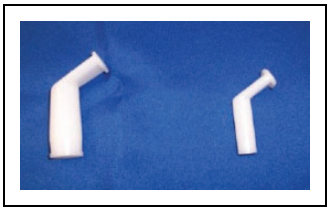

Figure 1. The 15- or 22-mm-angled mouthpieces.

MPV is considered by some patients to be more comfortable compared with nasal or facial interfaces, but it requires a more active participation of the patient and an initial training period for the staff to teach the patient how to use it. However, in the long term, it has the following significant advantages: less negative psychosocial impact on patient, no risk of facial pressure sores, enhanced speech and swallowing, and improved self-imagery. MPV allows the patient to be able to gloss oropharyngeal breathe (GPB) in the case of sudden failure of the ventilator or accidental disconnection from the ventilator,[38,39] which is not possible with tracheostomy.

There are various types of mouthpieces available for MPV.[16] Angled mouthpieces are the most commonly used, because they are the easiest for the patient to grip. There are different types and sizes of angled mouthpieces commonly 15 and 22 mm (Figure 1). In the 24 hour/day ventilator-dependent patient, daytime ventilation with angled mouthpiece in association with nocturnal NVS nasal, oral, or oronasal interface (or in selected patients, the use of a standard nozzle or of an orthodontic bite with a custom-molded flange covering the lips for the use of the mouthpiece, rather than an interface) has been reported to offer a better quality of life.[27]

With the MPV, any mode of ventilation including the pressure assisted, pressure support or bilevel positive airway, ventilation mode can be used with several ventilator circuits (double-limb or single-limb circuit with expiratory valve up to single limb with an intentional leak).[40,41] However, the latter does not permit air stacking. The volume-cycled ventilation with single-limb circuit appears the most suitable in allowing patient to air stack.[42–44]

Recently, a new ventilatory mode specially dedicated for MPV has been developed (MPV – Trilogy – Philips Respironics®, Pittsburgh, Pennsylvania, USA), with a dedicated arm and circuit without expiratory valve

Figure 2. Mouthpiece ventilation equipment. Source: reproduced with kind permission of K. Philips N.V.

allowing the patient to exhale outside the mouthpiece (Figure 2). This form of MPV has been reported to be safe and comfortable. The mode has a system of triggering dedicated for its purpose, which improves its use, by delivering air only when the patients touch the mouthpiece with their lips. This ventilatory mode has been tested with good results in selected patients and is commonly known as "kiss trigger."[40,41]

Advantages, disadvantages, and side effects of ventilation with mouthpiece

The most significant advantage compared with a nasal or oronasal interface is that mouthpiece is intermittently applied producing less interference with speech, better appearance, and absence of claustrophobia. A major disadvantage is the difficulty (but not the impossibility) of use at night[45] and the air leak from the mouth or nose.[46] In some patients, the mouthpiece may cause gastric distension (no more than nasal interface), increased salivation, and sometimes vomiting.[16]

Choice of ventilator, modes, and settings for MPV

MPV is usually performed using portable ventilators in volume-assisted/controlled mode.[47] Pressure modes are usually not used because of the high airflow that the devices continue to deliver when the patient is disconnected from the circuit and do not permit air stacking.[41,43,48,49] Volume-cycled modes allow the patient to choose at every inspiration the amount of air, which they want to inhale, adjusting the seal with the lips on the mouthpiece. A tidal volume between 700 and 1500 ml for adult patients ensures adequate ventilation and permits the patient to speak, shout, or cough.[15,49] The low pressure alarm will need to be set to minimum or where possible turned off altogether and the apnea alarm.[15,41,49]

In newer ventilators, it is possible to set a positive expiratory pressure (expiratory positive airway pressure (EPAP) or positive end expiratory pressure (PEEP)) to 0 cmH_2O. In other home volumetric ventilators, the minimum pressure alarm cannot be excluded; therefore, it is necessary to set up a PEEP (often 2 cmH_2O) which, due to the resistance to the airflow created from the angle of the mouthpiece, assures a pressure that prevents the continuous activation of the alarms.[40,41]

The patient activates the breath by putting the mouth on the mouthpiece and creating a small negative pressure in the circuit by sipping or inhaling from the mouthpiece. The negative pressure generated by a sip is much higher than that generated by a maximum static inspiratory pressure and can explain why a patient with advanced NMD can activate trigger without any inspiratory effort after a sip maneuver.[35]

MPV is available on both volume and pressure control mode using a single-limb circuit without expiratory valve connected to the ventilator.[40,50] A recent study has investigated the technical aspects that can influence the ventilator during MPV and provides

a practical setting strategy to avoid alarm activation of disconnection and low pressure (that represent major limitations for MPV use). A correct combination of tidal volume and inspiratory time avoided the activation of alarm for most ventilators. Only one of the tested ventilators did not allow MPV independently from the setting used.[50]

Complementary techniques

The air stacking maneuver is performed by the delivery of a series of deep breaths in via a resuscitation bag or MPV, and the patient does not exhale between the breaths. The number of breaths that are delivered via the resuscitation bag is the amount required to approach the total lung capacity. By increasing the inspiratory volume, the expiratory volume and flow will be greater and this will increase cough efficacy.[34] In this way, a patient who has an ineffective cough can often produce a peak cough flow (PCF) sufficient to eliminate secretions through an ''air-stacked cough.''[29,30,51] When the air stacking can no longer achieve PCF above 270 l/min[52], an abdominal thrust to assist cough can be added; this routine should be included in patient's daily schedule. The MAC can increase PCF from ineffective to above 270 l/min and enable the patient to clear secretions from the airways, maintain oxygen saturation above 95%, and prevent acute respiratory failure and intubation. Nevertheless, if NMD patients who are intubated or who have tracheostomy tubes with little or no free breathing ability, a protocol based on full NVS and MAC can help these patients to be extubated and decannulated.[53,54]

If the combination of NVS and MAC fails to maintain oxygen saturation above 95%, tracheostomy should be considered.[26]

Conclusion

Some authors still acknowledge tracheostomy as the most effective and secure form of continuous ventilatory support. However, there are studies showing that the survival is significantly longer and there are fewer reported complications with NVS[28] compared with a variety of other strategies. As noted earlier, a randomized controlled trial with NVS and tracheostomy unlikely to be conducted. NVS is a safe and acceptable alternative to ventilation by tracheostomy.[48] There is a widespread consensus that the NVS is preferable to tracheostomy during the early stages of early ventilatory failure in NMD patients,[1,7,24,37] but there remains controversy about long-term effectiveness. The side effects of tracheostomy are well known: dysphagia, decreased or loss of vocalization, the inability to perform GPB, and so on. Patients suffering from severe NMD, in whom nocturnal NVS alone becomes insufficient, should have a trial of NVS with a mouthpiece during the day. We hope that this review will encourage more centers to use this less invasive technique.

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