# We are IntechOpen, the world's leading publisher of Open Access books Built by scientists, for scientists

6,900
Open access books available

185,000
International authors and editors

200M
Downloads

Our authors are among the

154
Countries delivered to

TOP 1%
most cited scientists

12.2%
Contributors from top 500 universities




WEB OF SCIENCE™

Selection of our books indexed in the Book Citation Index in Web of Science™ Core Collection (BKCI)

## Interested in publishing with us?
## Contact book.department@intechopen.com

Numbers displayed above are based on latest data collected.
For more information visit www.intechopen.com



Chapter 6

# Open-Circuit Mouthpiece Ventilation: Indications, Evidence and Practicalities

Michalis Agrafiotis, Konstantina Nikolaou, Dimitra Siopi and Diamantis Chloros

Additional information is available at the end of the chapter

http://dx.doi.org/10.5772/intechopen.73243

**Abstract**

Open-circuit mouthpiece ventilation (MPV) is a method of noninvasive ventilation, which can be used to provide full-time support, induce lung volume recruitment, increase cough efficacy, defer tracheostomy and possibly improve survival and quality of life in advanced-stage neuromuscular patients. MPV might also be applicable to other chronic respiratory diseases as well as in acute exacerbations of chronic obstructive pulmonary disease and can also be employed for the extubation of unweanable neuromuscular patients. A candidate for MPV should be able to rotate his neck adequately, grab the mouthpiece with his lips and maintain sufficient control of the upper airway muscles. MPV is usually provided in the volume assisted-controlled mode with a tidal volume between 0.7 and 1.5 L, zero PEEP and backup rate set to the lower allowed value, allowing the patient to define his own ventilatory pattern. The "low pressure" and "apnea" alarm should be switched off, if possible, or special setting adjustments should be used to prevent their activation. Comprehensive patient training and dedicated nursing time are important for the application of MPV. MPV is considered a safe method for the majority of the patients, but accidental mouthpiece loss is an important concern.

**Keywords:** noninvasive ventilation, tracheostomy, Duchenne muscular dystrophy, amyotrophic lateral sclerosis, home ventilation

## 1. Introduction: rationale for mouthpiece ventilation

Mouthpiece ventilation (MPV) is a unique method of respiratory support specifically intended to provide full-time ventilatory assistance mainly to patients with chronic ventilatory failure and limited or no ventilator-free breathing time [1–3]. Together with negative pressure ventilation, MPV is probably one of the oldest methods of noninvasive ventilatory support since it

IntechOpen

© 2018 The Author(s). Licensee IntechOpen. This chapter is distributed under the terms of the Creative Commons Attribution License (http://creativecommons.org/licenses/by/3.0), which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited. CC BY

was initially employed as an alternative to tracheostomy by poliomyelitis survivors 60 years ago [3]. However, it was the pioneering work of a few investigators over the last three decades, which popularized the use of MPV for the management of chronic ventilatory failure, and since 2013, several MPV modes have been incorporated into modern home ventilators [3, 4–10].

The main indication of MPV is the provision of full-time ventilatory support for patients with chronic progressive neuromuscular diseases (NMD). Many NMDs cause respiratory disease by involving inspiratory, expiratory and upper airway muscles, leading to sleep disordered breathing, reduced respiratory pump efficiency and weak cough [11, 12]. From the respiratory physician's point of view, the most characteristic pathophysiologic trait of these NMDs is chronic alveolar hypoventilation which appears initially during the rapid eye movement sleep stage and extends eventually throughout all sleep stages before manifesting during daytime [13]. At advanced disease stages cough function is also severely impaired predisposing to respiratory infections and atelectasis [11, 12]. The institution of noninvasive positive pressure ventilation (NIV) for the support of the feeble respiratory muscles at the early stage of nocturnal hypoventilation is the mainstay of management of these patients and has been shown to improve survival, quality of sleep and quality of life [14–19]. However, as disease progresses and respiratory muscles continue to weaken, ventilatory requirements extend into the daytime. For patients with limited or no ventilator-free breathing time (e.g. ventilator use for > 16 or 20 hours per day), many practitioners would suggest transition to invasive ventilation via tracheostomy [20]. Importantly, an additional role of tracheostomy is the facilitation of secretion clearance given that at advanced disease stages cough flows are invariably reduced [11, 12]. In a European survey involving patients managed with home ventilation, patients with NMDs were the most likely to receive ventilatory support for a prolonged period (>6 years) and to have undergone tracheostomy procedures [21]. Nevertheless, long-term tracheostomies have been associated with several disadvantages including loss of voice, tube-related injuries, increased care-giver burden and disturbed self-image [2, 3, 9]. Although most patients would show preference toward a continued noninvasive mode of management [6, 22], standard nasal or oronasal masks are not suitable for this task since they commonly cause difficulties in eating, drinking and speech, sense of claustrophobia, limited field of vision, impaired social interaction and pressure lesions [1–3].

An alternative method for continuous noninvasive ventilator support is based on the 24 hour use of MPV or the combination of MPV during the daytime with nocturnal nasal or oronasal mask ventilation (MV). This method is complemented by ancillary strategies of which the most important are the "air-stacking" maneuver and the glossopharyngeal breathing (GPB) technique [1–3]. A good candidate for MPV should be able to rotate his neck and grab the mouthpiece with his lips and also maintain good control of his upper airway muscles (**Figure 1**) [10]. MPV is usually delivered via a home ventilator in the volume-assist-control mode (VAC) with the tidal volume (Vt) commonly set between 0.7 and 1.5 L (to correct for air leaks), zero PEEP and a back-up respiratory rate ideally set to zero [1–3]. Therefore, the patient has the ability to define his pattern of breathing according to his own ventilatory needs by taking as many breaths as he requires and by modifying the quantity of leak [8].

Volume-target MPV facilitates the application of the "air-stacking" maneuver, which is performed by teaching the patient to stack consecutive volumes of air delivered from the

http://dx.doi.org/10.5772/intechopen.73243



**Figure 1.** A patient with non-bulbar amyotrophic lateral sclerosis making use of mouthpiece ventilation (from Agrafiotis et al. [30], after permission).

ventilator until his lungs are maximally expanded. The maximum volume of air that can be held with a closed glottis is the patient's maximum insufflation capacity (MIC) [23]. The role of air-stacking is to preserve lung function by effecting lung volume recruitment and to avert chest wall strictures and contractures in patients with NMDs who experience a progressive decline in vital capacity (VC) [24, 25]. The difference MIC-VC depends directly on the integrity of glottic function and represents the amount of recruitable lung volume [24]. In a recent retrospective study of 151 patients with Duchenne muscular dystrophy (DMD), lung volume recruitment by air-stacking delayed the maximal VC decline by at least 5 years [26]. Other researchers have also reported a decrease in the rate of VC decline in patients with DMD [27] and a progressive increase in the MIC-VC difference, indicating a higher number of recruitable lung units [28]. In addition, lung expansion by air-stacking takes advantage of the increased respiratory system recoil pressure at high lung volumes to increase peak cough flow (PCF) and facilitate secretion clearance. In a group of 61 DMD patients, mean PCF increased from 137 to 236 L/min with the application of the air-stacking maneuver [29]. As an example, the application of air-stacking in a patient with amyotrophic lateral sclerosis (ALS) receiving MPV was associated with a MIC-VC difference of 0.4 L and an increase of PCF from 50 to 200 L/min [30]. Therefore, the use of MPV in combination with the air-stacking maneuver supports daytime ventilatory function and improves cough efficacy deferring tracheostomy.

GPB is another strategy commonly taught to MPV users. This technique is based on the use of the tongue to gulp consecutive boluses of air, while the glottis remains closed after each gulp to retain the inhaled air. By imitating the effects of a deep breath, GPB induces lung volume

recruitment and improves cough efficacy. In addition, GPB provides ventilator independency for patients with limited off-ventilator time and can be used as a rescue strategy in case of an accidental mouthpiece disconnection or ventilator failure [5, 6, 31].

During the last years, the use of MPV has also expanded to patients with other chronic respiratory diseases such as chest wall diseases [32] as well as to acute exacerbations of chronic obstructive pulmonary disease (COPD) [33, 34]. Nevertheless, despite the increasing interest in MPV, its application is limited to few centers specializing in noninvasive respiratory management [35]. Many practitioners are unfamiliar with its use, and several authors still consider tracheostomy as the most effective and secure method of ventilation for patients with advanced diseases [1].

In the rest of the chapter, we will present the evidence supporting the application of MPV for the management of respiratory disease and address several practical issues related to its use.

## 2. Evidence

### 2.1. Mouthpiece ventilation for full-time ventilatory support in neuromuscular patients

The application of MPV ventilation to 75 post-polio survivors with chronic respiratory failure was reported by Bach et al. [5]. All patients were using MPV as a major part of their respiratory support, but some (31%) were also using body respirators and the majority (88%) required full-time ventilator assistance. On average MPV was used for 1028 patient-years (14.8 years per person) for a total mortality of 1 death per 60.5 patient-years. Several of these MPV users married and some also joined the workforce.

Toussaint et al. [8] reported on 42 patients with DMD treated with MPV. Before introduced to MPV, all patients had diurnal hypercapnia by the end of the day while being treated optimally with nocturnal nasal ventilation. Survival at 1, 3, 5 and 7 years was 88, 66, 58 and 51%, respectively. Importantly, the use of MPV was associated with stabilization of VC, despite a deterioration in respiratory muscle strength, while transcutaneous $CO_2$ values improved. Some patients also reported improvements in dyspnea, appetite and swallowing. No important accidents or complications were observed over the 7-year follow-up period. VC was also preserved in a small cohort of 12 DMD patients who were prescribed MPV, a fact attributed to the concurrent use of lung volume recruitment strategies [36].

MPV was also compared to tracheostomy ventilation (TV) in 42 patients with DMD [9]. All tracheostomized patients (n = 16) used cuffless tubes and speaking valves connected in line with the ventilator circuit. While TV was associated with a higher incidence of tracheal injuries, mucous hypersecretion and lung infections, MPV users had a slightly higher incidence of weight loss and need for enteral feeding. Causes of death did not roughly differ between the two groups, however one MPV user died as a result of loss of mouthpiece during wheelchair malfunction, while a TV patient died during an endoscopic procedure.

Bedard and McKim [10] reported retrospectively on the use of MPV in patients with ALS. Of 37 patients in total, 27 were considered to be successful MPV users (consistent use > 1 month). The majority of the successful patients had less severe bulbar symptoms and demonstrated recruitable lung volume with MIC > VC. Importantly, all successful users experienced improved dyspnea scores and normalization of $CO_2$ values. For this group of patients, FVC decreased as disease progressed, but MIC was relatively preserved and MIC-VC difference increased. The majority of them could effect a PCF > 180 L/min with lung volume recruitment throughout disease course. In addition, a PCF > 180 L/min at initiation of MPV in successful users was associated with significantly better mean survival (637 vs. 240 days).

In addition, Khirani et al. [37] obtained questionnaires on quality of life issues from 30 neuromuscular MPV users. The majority of the patients reported reductions in dyspnea (73%) and fatigue (93%), and some of them also improvements in the ease of speech (43%) and swallowing (27%).

Overall, the above evidence indicates that MPV may defer tracheostomy, improve or stabilize clinically relevant lung function variables and possibly improve quality of life and confer a survival benefit to neuromuscular patients. The application of MPV seems relatively safe, although the possibility of mouthpiece loss is not a negligible concern. In addition, the concurrent use of other noninvasive aids, e.g. mechanical insufflation-exsufflation devices, might bias the interpretation of these studies. From this point of view, it is worth noting that in the study by Toussaint et al. [8], mechanical insufflation-exsufflation was available for only 7% of the patients. Nevertheless, further prospective studies are warranted to explore the impact of MPV on the outcomes of neuromuscular patients.

### 2.2. Mouthpiece ventilation for "unweanable" ventilator-dependent neuromuscular patients

MPV, sometimes combined with mechanical insufflation-exsufflation, has been successfully employed as a noninvasive method of weaning or removal of artificial tubes for patients with acute or chronic ventilatory failure, the majority of which had various NMDs. Many of these patients continued using MPV for several years [5, 6, 38]. A recent study [7] evaluated a simplified protocol for the extubation of neuromuscular patients with no ventilator-free breathing time. All patients (n = 157) who were normocapnic on invasive mechanical ventilation could maintain an $SpO_2$ > 95% for 12 hours on room air and could reverse desaturations with the use of mechanical insufflation-exsufflation device were extubated to noninvasive nasal, oronasal or mouthpiece volume assisted-controlled ventilation. Intensive use of mechanical insufflation-exsufflation was provided after extubation. Patients using MPV could determine the amount of volume they required and, when possible, could wean themselves *after* extubation by taking gradually fewer breaths from the mouthpiece. This protocol effected extubation success (defined as discharge without reintubation) in 157 (98%) of the patients, of whom 46% remained full-time ventilator-dependent. Although this study does not provide specific data with respect to each interface, it does exemplify the usefulness of mouthpiece for providing full-time ventilatory support without the requirement of an invasive tube.

### 2.3. Mouthpiece ventilation in other chronic respiratory diseases

Nicolini et al. [32] recruited 18 mechanical ventilation-naive patients with severe kyphoscoliosis in a prospective 4-year study, which evaluated the impact of combined diurnal MPV and nocturnal MV on lung function, clinical outcomes and health-related quality of life. They observed significant improvements in spirometric indices, blood gases, static mouth pressures, ventilatory drive and polygraphic variables at 6 months. In addition, patients reported improvements in quality of life aspects as sleep, physical well-being, eating, leisure, self-confidence and mood. Mortality at the end of the study period was 22.2%. When compared to a historical group of kyphoscoliotic patients who received only nocturnal MV, survival was better for the combined group at 180, 360 and 720 days.

### 2.4. Mouthpiece ventilation in acute respiratory exacerbations

There is limited evidence on the effectiveness, safety and tolerance of MPV in the setting of acute respiratory failure. A randomized cross-over prospective physiologic study compared four different interfaces with various internal volumes including a mouthpiece with minimal internal volume in critically ill patients. No difference was noted in gas change and respiratory effort variables, but the mouthpiece was associated with more leaks and asynchronies and a significantly less comfort on a visual analogue scale [39]. In a cross-over study which compared short-term mouthpiece and face mask tolerance in a cohort of 27 intensive care unit (ICU) patients treated for acute respiratory failure, five patients were withdrawn due to poor tolerance of the mouthpiece. For the remaining subjects and their nurses, facemask was associated with a nonsignificant better comfort, but mouthpiece required a significantly higher nursing time. While oxygenation and blood gases significantly improved with both interfaces, only face masks were associated with a significantly lower respiratory rate [40].

Some of the above findings were challenged by more recent clinical studies. Glerant et al. [33] conducted a retrospective matched case-control study in which MPV was compared to nasal MV and standard medical care in 87 COPD patients admitted to a respiratory ICU due to acute hypercapnic exacerbation of mild severity (average pH 7.3). In both groups, assist-control or pressure support modes were used. MPV was applied for 20 minutes every hour during the day and at less frequent intervals during the night. All MPV patients used a nose clips and had to hold the mouthpiece firmly and keep their mouth closed to avoid leaks. This study observed a nonsignificant lower intubation rate for MPV as compared to MV users (7 vs. 14%) and similar improvements in blood gases although these changes occurred much later in the MPV group, a fact attributed to a longer learning period for these patients. Overall, the duration of NIV and ICU stay did not differ between these two groups.

The same question was revisited by a recent randomized controlled trial. Nicolini et al. [34] randomized 50 COPD patients presenting with acute exacerbation of mild-moderate severity (pH 7.25–7.30) to receive either nasal MV or MPV in the pressure support mode. No case of NIV failure was observed, and blood gas values showed similar trends while the duration of NIV and hospital stay did not differ between the two groups. Common complications included skin breakdown for the MV group and gastric distention for the MPV group. However, tolerance and device acceptability was better for the MPV group.

http://dx.doi.org/10.5772/intechopen.73243

Criticism for both the abovementioned studies focused on the use of nasal masks in the setting of an acute exacerbation and the absence of long-term results. In addition, the study by Nicolini et al. [33] was underpowered to assess changes in blood gases, which was the primary outcome [41]. Pending the results of further investigations, MPV might be considered for COPD patients with a mild-moderate acute exacerbation who are intolerant of nasal or oronasal masks but retain a good level of consciousness and are not severely distressed in order to understand and apply this technique.

## 3. Practicalities in the application of mouthpiece ventilation

MPV is mainly indicated for neuromuscular patients with chronic ventilator failure when they develop daytime hypercapnia despite optimized nocturnal ventilatory support [8] or when they manifest deteriorating daytime breathlessness with increasing ventilator dependence [10]. MPV can be performed with (1) a home life-support ventilator, (2) a single or a double-limb circuit, (3) various types of mouthpieces, and (4) adjustable support arm or custom-made straps to mount the mouthpiece close to the head for patients with advanced motor disabilities (**Figure 2**). The presence of an exhalation valve in the circuit is not a prerequisite for the delivery of MPV; however, it might be necessary for switching to nocturnal MV for patients using non-vented circuits. The ideal candidate should be able to grab the mouthpiece with his lips and adequately rotate his neck [10]. MPV can be combined with MV during sleep or applied 24 hours per day using specifically designed interfaces [6, 31].

### 3.1. Patient education

Most of the patients considered for MPV have already been using MV for several years. Nevertheless, the experience of MPV is quite different and some patients may feel uncomfortable and express reluctance to continue. The application of MPV requires active participation

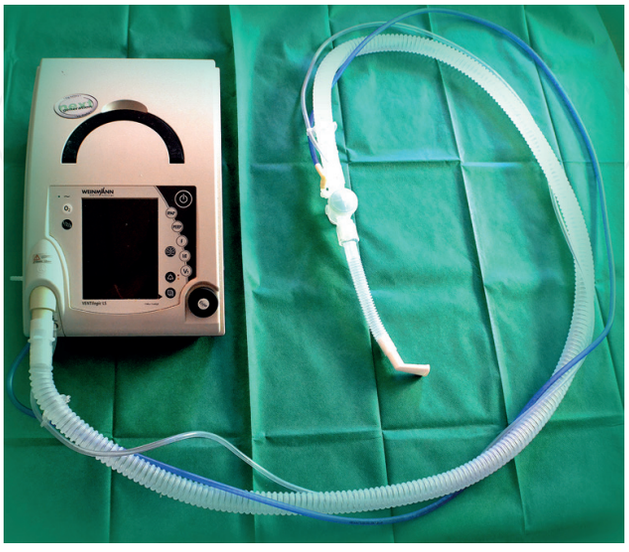


**Figure 2.** Setup for mouthpiece ventilation.

from the patient, increased nursing time and longer periods of training. We generally instruct patients to "sip" from the mouthpiece, in a manner similar to drinking a beverage using a straw (**Figure 1**). When this maneuver is applied, the soft palate moves posterocaudally sealing off the nasopharynx and minimizing nasal leaks. Importantly, the "sip" maneuver generates a higher negative pressure than maximum static inspiratory pressure, a fact that makes triggering easier for the frail neuromuscular patients [1].

### 3.2. Ventilator settings

The ventilator is usually set to the volume assisted-controlled mode with a Vt between 0.7 and 1.5 L, while PEEP (or EPAP) and backup rate are set to zero or to the lowest manufacturer-defined value. Recommendations on how to choose Vt and inspiratory time (Ti) are generally scarce [3]. If the patient is breathing comfortably while using MV, we begin with a similar Vt, albeit increased by 0.1–0.3 L to account for leaks and we commonly use a Ti at least as high as 1 s. Then we gradually increase Vt and/or Ti over several hours or days as much as tolerated. Many patients might have been using bilevel MV for many years before being introduced to volume-targeted MPV. While in bilevel ventilation, peak inspiratory flow is determined by the preset pressure, respiratory resistance and patient effort [42] in traditional volume-targeted ventilation inspiratory flow is ventilator-defined and is commonly delivered using a square waveform shape. Nevertheless, if the patient becomes severely breathless when switched to volume-targeted MPV, the use of a decelerating flow shape which delivers higher peak inspiratory flows at the start of the breath might be considered [43] . Generally, the patient should be able to define his own ventilatory pattern by determining the number of breaths and the quantity of leak [8]. Some experts would choose to use a pressure, rather than a flow-regulated, inspiratory trigger to avoid autotriggering [44]. However, despite the fact that in many new generation home ventilators only flow triggering is available, autotriggering with MPV seems to be less common than initially thought [37, 45]. Inspiratory trigger should be sensitive enough to reduce the work of breathing. However, since MPV users commonly fail to trigger the ventilator [45], a number of backup breaths could be set to ensure adequate ventilation and avert fatigue. On the other hand, machine-triggered breaths during patient disconnection from the mouthpiece might be a source of discomfort as a result of high flow on the user's face [45]. Nevertheless, in some newer generation ventilators, triggering can be simply effected by creating a small negative pressure at the mouthpiece ("kiss-trigger") [3, 37]. It must be noted that standard turbine ventilators are not designed to perform under conditions of rapidly changing load. A bench study which evaluated five different modern home ventilators observed significant swings in Vt when conditions of disconnection and reconnection were experimentally reproduced [46].

### 3.3. Alarms

One of the major problems in open-circuit systems is the prevention of "low pressure" and "apnea" alarm activation. As a general rule, these alarms should be switched off when possible or set to the lowest sensitive value allowed by the manufacturer. Several new generation home ventilators are more versatile in alarm customization with some even incorporating specifically designed software for MPV [37, 45]. Nevertheless, the use of high resistance mouthpieces together with smart combinations of Vt and Ti may create a sufficient back pressure which will

http://dx.doi.org/10.5772/intechopen.73243

prevent "low pressure" alarm activation, when the last cannot be switched off [37, 44]; practical recommendations for alarm customization for several home life-support ventilators have become recently available [45]. When 'apnea' alarming cannot be deactivated, a minimum of backup breaths should be set corresponding to the maximum allowed apnea time [44]. It should be noted, however, that at least in some types of ventilators, backup rate manipulation might also influence Ti, making alarm customization even more complicated [45].

### 3.4. Mouthpieces

There are a few types of mouthpieces available in the market nowadays, including angled 15 and 22 mm mouthpieces, straw-like as well as lip-sealing interfaces or orthodontic bite plates (**Figure 3**). The performance of these interfaces in the delivery of MPV has been assessed in a limited number of studies. Khirani et al. [37] compared three different angled mouthpiece configurations, a large mouthpiece (22 mm) and a small mouthpiece (15 mm) with and without a filter in a bench study which validated six different types of life-support home ventilators. The resistance was higher with the 15 mm mouthpiece with a filter, and this configuration was also

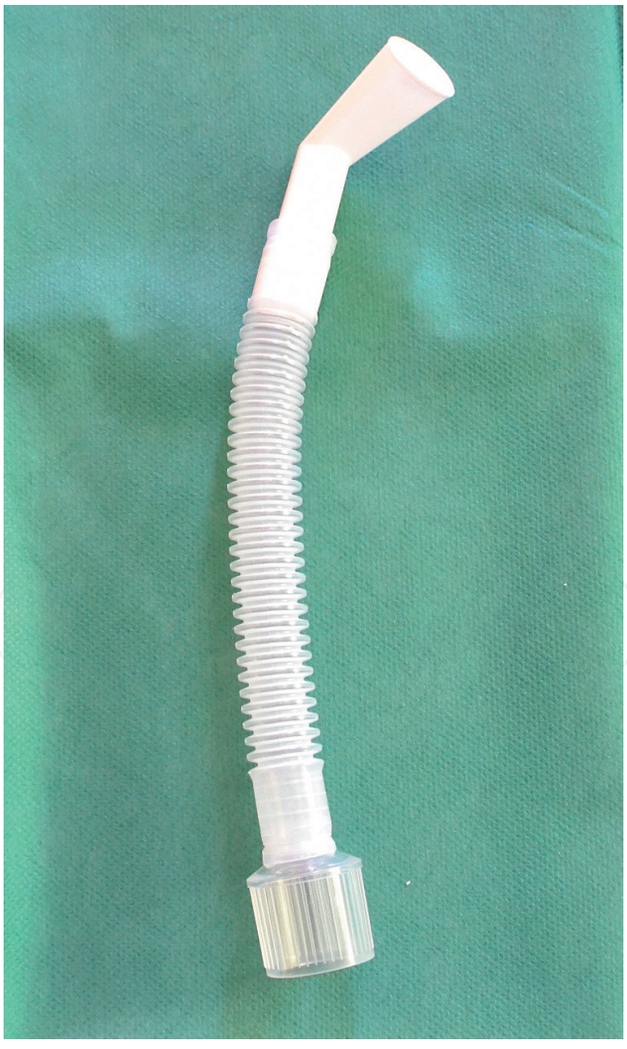


**Figure 3.** A 15 mm mouthpiece.

associated with a lower incidence of "low pressure" alarm activation. In another bench study, Ogna et al. [47] used four different home ventilators to compare the mechanical properties of various mouthpieces including a newly designed one. With respect to the most commonly used interfaces, respiratory resistance was lower with the 15 mm rigid-angled mouthpiece and higher with the straw mouthpiece. In volume-targeted modes with a Vt set to 1 L, both interfaces performed equally well across the different ventilators in delivering a volume close to the predetermined, while in pressure-targeted modes the effective pressure with the straw mouthpiece was slighter lower as a result of the increased resistance. The clinical implications of these findings remain unclear. For the time being, the choice of the interface should be tailored to the individual needs of each patient. An angled 15 mm mouthpiece seems to be a rational choice because its configuration increases resistance to the airflow preventing the activation of the "low pressure" alarm and in addition it is easier for the patient to grab [1]. If "low pressure" alarming persists, the addition of a filter to the circuit might be a simple and practical solution to the problem [37]. In addition, MPV can also be delivered during sleep with the use of specifically designed orthodontic bite plates or lip-sealing retention systems with attached Velcro straps to avoid disconnection. The use of these interfaces might cause desaturation, fragmented sleep and repeated arousals in a minority of the patients due to nasal leakage. Nasal pledges or clips can be applied to patients with significant nasal leaks [31].

### 3.5. Speaking and deglutition

Speaking is commonly problematic in patients with advanced respiratory disease as it requires higher than tidal inspiratory volumes and may slow the breathing rate causing breathlessness and fatigue [48]. If a patient on MPV needs to speak, he must take a large breath from the mouthpiece and then speak while expiration is driven by the expiratory muscles and the respiratory system recoil pressure. Nevertheless, speaking with the use of the mouthpiece might be associated with longer pauses (in order to breath in) and difficulties in choosing the right strategy for mouthpiece positioning and use [48].

Breathing and swallowing are normally competitive procedures and their coordination is disrupted in respiratory disease with respiration taking precedence over swallowing. Swallowing in neuromuscular patients is characterized by piecemeal deglutition, increased time to swallow a bolus and an increased number of swallows during inspiration. The institution of positive pressure ventilation stabilizes breathing and improves swallowing performance [49]. The practice of MPV should theoretically contribute to the restoration of swallowing and breathing coordination as users have to alternate between taking deep breaths from the mouthpiece and swallowing. In addition, MPV should also maintain the supraglottic pressure required for effective swallowing, while the high inspired volumes improve cough efficacy providing protection against aspiration [48]. Nevertheless, as disease progresses and ventilator-free time is reduced, less time is available for swallowing, while the presence of food in the mouth and in the pharynx does not allow patients to breath in safely. To deal with this problem, some patients use the ventilator to perform air-stacking in order to increase lung volume and afford longer periods of apnea without breathlessness [9]. Although weight loss and feeding problems in MPV users have been reported [9], it is not clear whether these should be attributed to the interface *per se* or to disease progression and increasing breathlessness.

It should be noted that a disconnection from the mouthpiece for eating and speaking might compromise effective gas exchange. In a small study including eight MPV users who were monitored with polygraghy during daily activities, most of the patients could speak and eat without ventilator assistance; however, prolonged disconnections (>3 min) (e.g. during meals) were associated with significant drops in $SpO_2$ and increases in transcutaneous $PCO_2$ [50].

### 3.6. Complications

A few complications associated with MPV have been so far reported, of which mouthpiece loss is the most important [9]. This complication can be avoided if the mouthpiece's position is secured using specifically designed support arms or customary-made straps. Fixation of the mouthpiece on the shoulders allows the interface to follow the patient's movements [8]. In addition, MPV users could be taught the technique of GPB to maintain ventilator independency in case of accidental mouthpiece loss [6, 31]. Other complications include salivation, aerophagia, abdominal distention and orthodontic problems [1, 2, 6, 51]. There are no available data on the management of excessive salivation in mouthpiece users. For more severe cases the administration of an anticholinergic agent such as amitriptyline might be considered [9, 52]. Aerophagia and abdominal distention are common complications of noninvasive ventilatory support and have been associated with respiratory distress and ventilator dependence [51]. For patients with gastrostomies, unclamping the gastric tube to "burp out" the air is a quick method to effect symptom relief. Sometimes a nasogastric or a rectal rube (for patients with colonic distention) might also be helpful [51]. For patients with persistent symptoms switching to pressure-targeted ventilation could be an option, although this mode is not suitable for the application of lung volume recruitment and air-stacking maneuvers. If the patient is maintained on a volume-targeted mode, setting a lower pressure limit to effect secondary pressure cycling is an alternative option. Vomiting and aspiration as a result of gastric distention as well as pneumothorax have been so far theoretical concerns, but they represent potentially life-threatening events [4, 53]. Orthodontic complications are not uncommon in long-term users; however, they pose mostly an esthetic rather than a functional concern and specifically designed orthodontic interfaces have become available in the market [6, 31, 53]. Patients on MPV and full-time ventilator dependence can safely undergo dental procedures using nasal interfaces as long as oxygen saturation is monitored and oxygen or sedatives are avoided [54].

## 4. Conclusion

MPV is a "re-discovered" method of noninvasive ventilation that can be used to provide full-time ventilatory support, recruit lung volume, improve cough efficacy, defer tracheostomy and possibly improve survival and quality of life in neuromuscular patients. MPV might also be beneficial for patients with other chronic respiratory diseases or in acute COPD hypercapnic exacerbation. The successful application of MPV requires careful selection of patient, interface, ventilator and alarm settings, increased nursing time and comprehensive patient training.

## Conflict of interest

None for all authors.

## Funding

This work was supported by the Hellenic Society of Respiratory and Occupational Lung Diseases

## Author details

Michalis Agrafiotis*, Konstantina Nikolaou, Dimitra Siopi and Diamantis Chloros

*Address all correspondence to: m.agrafiotis@gmail.com

Department of Pulmonary Medicine, Respiratory Neuromuscular Outpatient Clinic, "Georgios Papanikolaou" General Hospital of Thessaloniki, Exohi, Greece

## References

[1] Pinto T, Chatwin M, Banfi P, Winck JC, Nicolini A. Mouthpiece ventilation and complementary techniques in patients with neuromuscular disease: A brief clinical review and update. Chronic Respiratory Disease. May 2017;**14**(2):187-193

[2] Benditt JO. Full-time noninvasive ventilation: possible and desirable. Respiratory Care. Sep 2006;**51**(9):1005-1012

[3] Garuti G, Nicolini A, Grecchi B, Lusuardi M, Winck JC, Bach JR. Open circuit mouthpiece ventilation: Concise clinical review. Revista Portuguesa de Pneumologia. Jul–Aug 2014;**20**(4): 211-218

[4] Bach JR, Alba AS, Bohatiuk G, Saporito L, Lee M. Mouth intermittent positive pressure ventilation in the management of postpolio respiratory insufficiency. Chest. Jun 1987;**91**(6): 859-864

[5] Bach JR, Alba AS. Noninvasive options for ventilatory support of the traumatic high level quadriplegic patient. Chest. Sep 1990;**98**(3):613-619

[6] Bach JR, Alba AS, Saporito LR. Intermittent positive pressure ventilation via the mouth as an alternative to tracheostomy for 257 ventilator users. Chest. Jan 1993;**103**(1):174-182

[7] Bach JR, Gonçalves MR, Hamdani I, Winck JC. Extubation of patients with neuromuscular weakness: A new management paradigm. Chest. May 2010;**137**(5):1033-1039

[8] Toussaint M, Steens M, Wasteels G, Soudon P. Diurnal ventilation via mouthpiece: Survival in end-stage Duchenne patients. The European Respiratory Journal. Sep 2006;**28**(3):549-555

[9] Soudon P, Steens M, Toussaint M. A comparison of invasive versus noninvasive full-time mechanical ventilation in Duchenne muscular dystrophy. Chronic Respiratory Disease. 2008;**5**(2):87-93

[10] Bédard ME, McKim DA. Daytime mouthpiece for continuous noninvasive ventilation in individuals with amyotrophic lateral sclerosis. Respiratory Care. Oct 2016;**61**(10): 1341-1348

[11] Benditt JO. The neuromuscular respiratory system: Physiology, pathophysiology, and a respiratory care approach to patients. Respiratory Care. Aug 2006;**51**(8):829-837

[12] Benditt JO, Boitano LJ. Pulmonary issues in patients with chronicneuromuscular disease. American Journal of Respiratory and Critical Care Medicine. May 15, 2013;**187**(10):1046-1055

[13] Ragette R, Mellies U, Schwake C, Voit T, Teschler H. Patterns and predictors of sleep disordered breathing in primary myopathies. Thorax. Aug 2002;**57**(8):724-728

[14] Simonds AK, Muntoni F, Heather S, Fielding S. Impact of nasal ventilation on survival in hypercapnic Duchenne muscular dystrophy. Thorax. Nov 1998;**53**(11):949-952

[15] Jeppesen J, Green A, Steffensen BF, Rahbek J. The Duchenne muscular dystrophy population in Denmark, 1977-2001: Prevalence, incidence and survival in relation to the introduction of ventilator use. Neuromuscular Disorders. Dec 2003;**13**(10):804-812

[16] Ward S, Chatwin M, Heather S, Simonds AK. Randomised controlled trial of non-invasive ventilation (NIV) for nocturnal hypoventilation in neuromuscular and chest wall disease patients with daytime normocapnia. Thorax. Dec 2005;**60**(12):1019-1024

[17] Bourke SC, Tomlinson M, Williams TL, Bullock RE, Shaw PJ, Gibson GJ. Effects of non-invasive ventilation on survival and quality of life in patients with amyotrophic lateral sclerosis: A randomised controlled trial. Lancet Neurology. Feb 2006;**5**(2):140-147

[18] Vrijsen B, Buyse B, Belge C, Robberecht W, Van Damme P, Decramer M, Testelmans D. Noninvasive ventilation improves sleep in amyotrophic lateral sclerosis: A prospective polysomnographic study. Journal of Clinical Sleep Medicine. Apr 15, 2015;**11**(5):559-566

[19] Boentert M, Brenscheidt I, Glatz C, Young P. Effects of non-invasive ventilation on objective sleep and nocturnal respiration in patients with amyotrophic lateral sclerosis. Journal of Neurology. Sep 2015;**262**(9):2073-2082

[20] King AC. Long-term home mechanical ventilation in the United States. Respiratory Care. Jun 2012;**57**(6):921-930

[21] Lloyd-Owen SJ, Donaldson GC, Ambrosino N, Escarabill J, Farre R, Fauroux B, Robert D, Schoenhofer B, Simonds AK, Wedzicha JA. Patterns of home mechanical ventilation use in Europe: Results from the Eurovent survey. The European Respiratory Journal. Jun 2005;**25**(6):1025-1031

[22] Bach JRA. Comparison of long-term ventilatory support alternatives from the perspective of the patient and care giver. Chest. Dec 1993;**104**(6):1702-1706

[23] Kang SW, Bach JR. Maximum insufflation capacity. Chest. 2000 Jul;**118**(1):61-65

[24] Ishikawa Y, Bach JR. Physical medicine respiratory muscle aids to avert respiratory complications of pediatric chest wall and vertebral deformity and muscle dysfunction. European Journal of Physical and Rehabilitation Medicine. 2010 Dec;**46**(4):581-597

[25] Molgat-Seon Y, Hannan LM, Dominelli PB, Peters CM, Fougere RJ, McKim DA, Sheel AW, Road JD. Lung volume recruitment acutely increases respiratory system compliance in individuals with severe respiratory muscle weakness. ERJ Open Research. Mar 14, 2017;**3**(1):00135-2016

[26] Chiou M, Bach JR, Jethani L, Gallagher MF. Active lung volume recruitment to preserve vital capacity in Duchenne muscular dystrophy. Journal of Rehabilitation Medicine. Jan 19, 2017;**49**(1):49-53

[27] Katz SL, Barrowman N, Monsour A, Su S, Hoey L, McKim D. Long-term effects of lung volume recruitment on maximal inspiratory capacity and vital capacity in Duchenne muscular dystrophy. Annals of the American Thoracic Society. Feb 2016;**13**(2):217-222

[28] Bach JR, Mahajan K, Lipa B, Saporito L, Goncalves M, Komaroff E. Lung insufflation capacity in neuromuscular disease. American Journal of Physical Medicine & Rehabilitation. Sep 2008;**87**(9):720-725

[29] Ishikawa Y, Bach JR, Komaroff E, Miura T, Jackson-Parekh R. Cough augmentation in Duchenne muscular dystrophy. American Journal of Physical Medicine & Rehabilitation. Sep 2008;**87**(9):726-730

[30] Agrafiotis M, Renessis V, Kousta A, Athanassiadou A, Tryfon S, Chloros D. Noninvasive ventilation via a mouthpiece in a patient with amyotrophic lateral sclerosis. Pneumon. Jan–Mar 2017;**30**(1) (in press)

[31] Bach JR. Continuous noninvasive ventilation for patients with neuromuscular disease and spinal cord injury. Seminars in Respiratory and Critical Care Medicine. Jun 2002;**23**(3): 283-292

[32] Nicolini A, Barlascini C, Piroddi IM, Garuti G, Banfi PI. Effectiveness and safety of mouthpiece ventilation and nocturnal non-invasive ventilation in patients with kyphoscoliosis: Short and long-term outcomes after an episode of acute respiratory failure. Revista Portuguesa de Pneumologia (2006). Mar–Apr 2016;**22**(2):75-81

[33] Glerant JC, Rose D, Oltean V, Dayen C, Mayeux I, Jounieaux V. Noninvasive ventilation using a mouthpiece in patients with chronic obstructive pulmonary disease and acute respiratory failure. Respiration. 2007;**74**(6):632-639

[34] Nicolini A, Santo M, Ferrari-Bravo M, Barlascini C. Open-mouthpiece ventilation versus nasal mask ventilation in subjects with COPD exacerbation and mild to moderate acidosis: A randomized trial. Respiratory Care. Dec 2014;**59**(12):1825-1831

[35] Bach JR, Gonçalves MR, Hon A, Ishikawa Y, De Vito EL, Prado F, Dominguez ME. Changing trends in the management of end-stage neuromuscular respiratory muscle failure: Recommendations of an international consensus. American Journal of Physical Medicine & Rehabilitation. Mar 2013;**92**(3):267-277

[36] McKim DA, Griller N, LeBlanc C, Woolnough A, King J. Twenty-four hour noninvasive ventilation in Duchenne muscular dystrophy: A safe alternative to tracheostomy. Canadian Respiratory Journal. Jan–Feb 2013;**20**(1):e5-e9

[37] Khirani S, Ramirez A, Delord V, Leroux K, Lofaso F, Hautot S, Toussaint M, Orlikowski D, Louis B, Fauroux B. Evaluation of ventilators for mouthpiece ventilation in neuromuscular disease. Respiratory Care. Sep 2014;**59**(9):1329-1337

[38] Bach JR, Saporito LR. Criteria for extubation and tracheostomy tube removal for patients with ventilatory failure. A different approach to weaning. Chest. Dec 1996;**110**(6):1566-1571

[39] Fraticelli AT, Lellouche F, L'her E, Taillé S, Mancebo J, Brochard L. Physiological effects of different interfaces during noninvasive ventilation for acute respiratory failure. Critical Care Medicine. Mar 2009;**37**(3):939-945

[40] Schneider E, Dualé C, Vaille JL, Ouchchane L, Gillart T, Guélon D, Schoeffler P. Comparison of tolerance of facemask vs. mouthpiece for non-invasive ventilation. Anaesthesia. Jan 2006;**61**(1):20-23

[41] Carlucci A, Gregoretti C. Mouthpiece ventilation: Just a home-care support? Respiratory Care. Dec 2014;**59**(12):1951-1953

[42] Hess DR. Ventilator waveforms and the physiology of pressure support ventilation. Respiratory Care. Feb 2005;**50**(2):166-186

[43] Agrafiotis M, Kotsifou E, Renessis V, Athanassiadou A, Kousta A, Chloros D. Decelerating flow shape for volume-targeted mouthpiece ventilation. The Clinical Respiratory Journal. (in press)

[44] Boitano LJ, Benditt JO. An evaluation of home volume ventilators that support open-circuit mouthpiece ventilation. Respiratory Care. Nov 2005;**50**(11):1457-1461

[45] Carlucci A, Mattei A, Rossi V, Paracchini E, Raineri SM, Gregoretti C. Ventilator settings to avoid nuisance alarms during mouthpiece ventilation. Respiratory Care. Apr 2016;**61**(4): 462-467

[46] Ogna A, Prigent H, Falaize L, Leroux K, Santos D, Vaugier I, Orlikowski D, Lofaso F. Accuracy of tidal volume delivered by home mechanical ventilation during mouthpiece ventilation: A bench evaluation. Chronic Respiratory Disease. May 3, 2016. pii: 1479972316647177

[47] Ogna A, Prigent H, Falaize L, Leroux K, Santos D, Vaugier I, Orlikowski D, Lofaso F. Bench evaluation of commercially available and newly developed interfaces for mouthpiece ventilation. The Clinical Respiratory Journal. Dec 27, 2016 (in press)

[48] Britton D, Benditt JO, Hoit JD. Beyond tracheostomy: Noninvasive ventilation and potential positive implications for speaking and swallowing. Seminars in Speech and Language. Aug 2016;**37**(3):173-184

[49] Terzi N, Orlikowski D,Prigent H, Denise P,Normand H,Lofaso F. Noninvasive monitoring of breathing and swallowing interaction. In: Mark Schwartz EMG Methods for Evaluating Muscle and Nerve Function. In-tech; 2012. ISBN: 978-953-307-793-2. Available from: http://www.intechopen.com/books/emgmethods-or-evaluating-muscle-and-nerve-function/noninvasive-monitoring-of-breathing-and-swallowinginteraction

[50] Nardi J, Leroux K, Orlikowski D, Prigent H, Lofaso F. Home monitoring of daytime mouthpiece ventilation effectiveness in patients with neuromuscular disease. Chronic Respiratory Disease. Feb 2016;**13**(1):67-74

[51] Bach JR, Mehta AD. Respiratory muscle aids to avert respiratory failure and tracheostomy: A new patient management paradigm. Journal of Neurorestoratology. 2014;**2**:25-35

[52] Banfi P, Ticozzi N, Lax A, Guidugli GA, Nicolini A, Silani V. A review of options for treating sialorrhea in amyotrophic lateral sclerosis. Respiratory Care. Mar 2015;**60**(3):446-454

[53] Nava S, Navalesi P, Gregoretti C. Interfaces and humidification for noninvasive mechanical ventilation. Respiratory Care. Jan 2009;**54**(1):71-84

[54] Tran J, Bach JR, Gonçalves MR. Alternatives to mouthpiece noninvasive ventilatory support to permit dental care. American Journal of Physical Medicine & Rehabilitation. Feb 2014;**93**(2):182-185

IntechOpen