Physical Medicine and Rehabilitation - International

Austin Publishing Group

Open Access Full Text Article

Review Article

Mouthpiece Ventilation in Patients with Neuromuscular Disease: A Brief Clinical Review

Nicolini A, Russo D , Barlascini CO*, Sclifò F§, Grecchi B^, Garuti GC°, Banfi PI°°

Respiratory Rehabilitation Unit and ALS Centre, Italy
*Public Health Department, Italy
§ Allergy & Respiratory Diseases Clinic, University of Genoa, Italy
^ Physical Medicine and Rehabilitation, Italy
° Pulmonary Department Santa Maria Bianca, Italy
°°Don Gnocchi Foundation IRCSS, Italy

***Corresponding author:** Nicolini Antonello, Respiratory Rehabilitation Unit and ALS Centre, Italy

Received: July 31, 2014; **Accepted:** September 22, 2014; **Published:** September 25, 2014

Abstract

The Open-circuit Mouthpiece Ventilation (MPV) is a mode of ventilation that uses a mouthpiece interface which the patient holds with the lips when he wants to be supported during inspiration. There exists a poor understanding of this method's benefits compared to other modalities. Non-invasive ventilation (NIV) is sometimes reported as suboptimal in neuromuscular disease patients due to excessive secretions in the airways , hypercapnia due to inadequate ventilator settings , or because of a lack of tolerance of the interface. Interfaces that cover the nose and / or mouth and nose are the most commonly used, but may cause skin lesions and claustrophobia. Many of these drawbacks can be avoided by using a mouthpiece to administer the NIV. The MPV is used by many patients as daytime ventilatory support in combination with other modes of ventilation interface effective for night ventilation. There are two models mouthpiece of different sizes 15 and 22 mm .NIV has been used for years in patients with neuromuscular disorders as a viable alternative to continuous ventilatory support via tracheotomy tube. NIV is associated with a reduced risk of pneumonia and other respiratory complications. Its use in the volumetric mode allows air-stacking to improve cough. The mouthpiece interface facilitates speech and swallowing, factors that lead to a better quality of life for patients. This review aims to highlight the indications, along with the advantages and disadvantages of MPV.

Keywords: Non-invasive ventilation; Mouthpiece; Open-circuit mouthpiece ventilation (MPV); Neuromuscular disease; Ventilator settings

Introduction

Neuromuscular diseases represent a heterogeneous group of disorders of the muscle, nerve, and/or some neuromuscular junction. The respiratory muscles are rarely spared in neuromuscular diseases even if the type of muscle involvement, severity, and time course greatly varies among the different diseases [1]. The most common neuromuscular diseases in childhood are Duchenne muscular dystrophy (DMD), spinal muscular atrophy (SMA) and congenital myophathy, congenital muscular dystrophy (CMD) . In adults, amyotrophic lateral sclerosis, myotonic myopathy (Steiner's disease) and limb-girdle muscular dystrophy (LGMD) are the most common neuromuscular diseases which can benefit from NIV treatment according their progressiveness are reported in the Table 1[2].Before 1953, the non-invasive ventilation (NIV) was practiced through the use of negative pressure ventilators : "iron lung ","armor". Despite of their great success as a continuous ventilator support, ventilation through tracheotomy became the standard since the epidemic Danish polio in 1952, because it was possible to move patients and secretions could be easily managed [3]. In 1953 patients who used lungs steel or armor began to use the ventilation mouthpiece during the day. In the United States intermittent positive pressure ventilation (IPPV) through a mouthpiece has been the accepted practice for patients requiring continuous ventilatory support as an alternative to mechanical ventilation via a tracheotomy tube [3]. The portable ventilator manufactured by Bantam Harris Thompson in 1956 was another turning point. Mouthpiece use slowly grew in patients with severe ventilatory defect due to restrictive illnesses (vital capacity lower than 500 ml) and with inefficient cough [3]. The result was a reduced dependence on the ventilator with less hyper secretion [3, 4]. Non invasive ventilation via mouthpiece was used in 257 patients requiring continuous ventilatory support at the Goldwater Memorial Hospital from 1968 to 1987 with excellent results. [5]. Despite remarkable results by Dr. JR. Bach's group [3,4,5], few centers in the United States used the ventilation via mouthpiece in neuromuscular patients .Until ten years ago there were only sporadic reports of NIV mouthpiece in this patient group whose numbers have continued to increase with time. Server a et al [6] and Toussaint et al. [7, 8] changed this.

The Fundamentals

NIV improves gas exchange, relieves shortness of breath, allows inspiratory muscles to rest, reduces the incidence of nosocomial infections. Mortality from and hospitalizations for respiratory failure decrease. [9] The great limitation of this technique is that it is impossible to implement on a chronic basis if the interface is uncomfortable [10,11]. Fortunately, now there are over 100 types of interfaces. The most frequent causes of NIV failure with consequent intubation of patients are due to inappropriate settings of the ventilator. The group treating these patients has to be experienced. Skin breakdowns due to pressure points of interfaces must be prevented. Strict skin care protocols are essential .Misuse of mechanically assisted coughing (MAC) to eliminate airway secretions [10, 11, 12] must be prevented. However, the failure of NIV may also occur due to a

Citation: Nicolini A, Russo D , Barlascini CO, Sclifò F, Grecchi B, Garuti GC, et al. Mouthpiece Ventilation in Patients with Neuromuscular Disease: A Brief Clinical Review. Phys Med Rehabil Int. 2014;1(3): 4.

Table 1: Neuromuscular diseases can benefit fron NIV according the progressiveness of respiratory impairment.

Spinal muscular atrophy (SMA) Type 1	Rapid worsening (0-3 years)
Spinal muscular atrophy Type 2	Slow worsening (>15 years)
Spinal muscular atrophy Type 3	Slow worsening (>15 years)
Acid maltase deficit	Slow worsening (>15 years)
Duchenne dystrophy (DMD)	Intermediate worsening (5-15 years)
Myotonic dystrophy (Steinert's disease)	Intermediate worsening (5-15 years)
Limb girdle muscular dystrophy	Intermediate worsening (5-15 years)
Amyotrophic lateral sclerosis (ALS)	Rapid worsening (0-3 years)

serious bulbar dysfunction, or a severe cognitive deficit involving a lack of cooperation in the maneuvers of assisted cough, or due to the inappropriate administration of sedative drugs or additional oxygen [12,13]. These latter problems are harder to overcome. For these reasons the choice of the appropriate interface is crucial for the NIV success [14]. Being able to switch between different types of interfaces to change the pressure points of the mask on the skin can help to increase patient adherence to NIV [14]. However, few clinical trials have compared effects produced by different types of interfaces on clinical outcomes; none has evaluated the impact of interfaces on the respiratory workload [15, 16].

Rationale for the Use of Ventilation with Mouthpiece (MPV)

Nasal and oro-nasal masks are the most practical as well as the most commonly used for the administration of NIV, especially during sleep [14, 17, 18]. They permit ventilation through the nose or nose and mouth: they would be perfect interfaces if their use was not limited by claustrophobia, discomfort and skin lesions [14, 18, 19]. Nasal interfaces also include nasal pillows: they have the advantage of producing no skin lesions and claustrophobia, but they have the disadvantage of higher air leaks when high inspiratory pressure is administered[14,18]. Currently tracheotomy is widely offered to patients affected by neuromuscular diseases who need 24 hour assistance; mouthpiece NIV has been the alternative. Tracheotomy may increase costs, complications, and social isolation [20]. When the choice is given, to them, patients usually prefer non-invasive ventilation [21]. Patients with neuromuscular diseases who are usually ventilated during the night with nasal or oronasal masks, but they may have a level of inspiratory muscle weakness that requires continuous ventilation. The application of an oro- nasal interface can interfere with social interaction; impair eating, drinking, and talking. This mask changes a patient's perception of him/herself. This last may have drastic consequences psychologically. The use of angled mouthpieces wired by a metal flexible arm support (if the patient has no strength to keep the mouthpiece near to the mouth) is the ideal solution for daytime ventilation in patients who can still grab mouthpiece with preserved neck movement and hold it in mouth . The open-circuit mouthpiece ventilation, has been reported to be safe and comfortable in patients confined to wheelchairs. In the selected patient, it's easy to apply and to use even during daily living activities such as eating and talking [22, 23]. 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 500 neuromuscular disease patients who required continuous ventilatory support [23]. Tracheotomy is considered mandatory for the survival of patients who have severe bulbar dysfunction and high risk of recurrent and massive aspiration pneumonia. Its utility for neuromuscular patients without bulbar impairment is not conclusively proved regardless of severity of ventilator failure [24, 25, 26]. It must be stated that there are no published evidence-based guidelines concerning mouthpiece ventilation. Its application is mainly based on the experience of few centers [20. 21, 22, 23]. Patient selection is of paramount importance. Match the technology with the patient; be ready to change the technology as is required by the patient's changing condition. Obviously, randomized controlled studies pose enormous practical and ethical problems. Mouthpiece ventilation is more comfortable compared to nasal or facial masks, but it requires a more active participation of the patient and a longer 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 [1, 3]:

1) Less negative psychosocial impact on patient

2) no risk of pressure breakdown on the face

3) Better speech than with the oro-nasal and nasal mask

4) Better ability to eat and drink

5) Improved image of the self

6) Improved security (compared to tracheotomy)

It permits the use of glosso-pharyngeal breathing in case of sudden failure of the ventilator or accidental disconnection from the ventilator [20, 21, 22, 27]. This last advantage can save lives. There are various types of mouthpiece for non invasive ventilation [14]. The angled mouthpiece is more commonly used, because they are easier for the mouth to grip. There are 2 types of angled mouthpiece, one of 15 and one of 22 mm (Figure 1). In full-time ventilator users, daytime ventilation with angled mouthpiece in combination with nasal or oro-nasal mask ventilation during the night (or in selected patients, the use of a standard nozzle or of an orthodontic bite with a custom-molded flange covering the lips for use of the mouthpiece, rather than a mask) during the night offers a better life [22].With the mouthpiece any mode of ventilation including the pressure assisted ventilation mode can be used (e.g., closed system with the continued support of the interface on the mouth). During the day, particularly when the patient is sitting, a system in which the mouthpiece should be placed to close to the mouth by adjustable arm and the patient can grab the mouthpiece as desired, close to the mouth by an adjustable arm. It should be removable for talking, eating, or breathing independently. In this case the volumetric ventilation appears the most suitable

Figure 1: 22 and 15 mm angled mouthpieces with adaptors.

because the flows provided are slow and steady; it also allows air-stacking maneuver [28, 29, 30]. Recently special software for the open mouthpiece ventilation that facilitates the setting of alarms with a system of triggering dedicated to this mode which facilitates its use, by activating the emission of air only by the positioning of the patient's lips on the mouth piece has come on the market; this software has been tested with good results in selected patients and is commonly known as "kiss the trigger ". [17, 31, 32, 33] .

Advantages, Disadvantages and Side Effects of Ventilation with Mouthpiece

The most significant advantage compared to a nasal or oronasal mask is that mouthpiece produces less interference with speech, better appearance, and absence of claustrophobia. The greatest disadvantage is the difficulty of use at night [22, 34]. Other disadvantages are air leaks from the mouth or nose [1, 14, 32, 34]. Moreover, the mouthpiece may cause gastric distension , even salivation and even more rarely vomiting [14]. Often these "non-nocturnal problems can be fixed. The failure of the MPV and /or NIV is seen: a) if patients are not cooperative, or b) more often, in the presence of a severe bulbar dysfunction, unable to cooperate. If the combination of NIV, interface, and MAC fails to maintain a constant oxygen saturation between 94 - 95%, tracheotomy should be considered.

Ventilator Types, Settings, and Settings for the Ventilation Open-Circuit Mouthpiece

The mouthpiece ventilation is usually performed using portable ventilator in volume assisted - controlled mode (ACV) [32] to provide adequate ventilatory support and allow the patient to perform air -stacking maneuvers. Volumetric mode allows 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 700-1500 ml for adult patients (depending on the amount of air the patient wants to let enter the mouth) , ensures proper ventilation: so that he/she can even talk, shout or cough [17,31,32] . Moreover, the maneuver of air -stacking is done by taking a series of breaths at high tidal volume without exhaling , trying to get an air volume approaching the total lung capacity to make cough as effectively as possible [23,31,32] .In this way , a patient who has a ineffective cough can often produce a peak flow sufficient to eliminate secretions through an "air-stacked cough" [26] . Some home ventilators was tested for delivering MPV [31,36]: among them Trilogy (Philips Respironics) has a dedicated particular function called MPV "Kiss -trigger " (see above) as well as an arm dedicated to support the mouthpiece to facilitate its use [36]. The pressure modes are usually shut off because of the high flow that the devices continue to deliver when the patient is disconnected to the mouthpiece and because they do not permit air –stacking [28,31,32,37,38].In the new generation ventilators manufactured by Philips - Respironics one must not set a positive expiratory pressure (EPAP or PEEP) (set it to Zero PEEP or ZEEP) .The alarms for apnea, minimum pressure, and minimum volumes can easily be shut off in order to avoid their unnecessary activation. In most home volumetric ventilators the minimum pressure alarm cannot be excluded; therefore it is necessary to set up a PEEP (often 2 cm H_2O) , which, thanks to the resistance to the flow of air created from the angle of the mouthpiece creates a pressure that prevents the continuous activation of the alarms. The most common mode of ventilation is ACV with a tidal volume between 0.7 and 1.5 L without PEEP (EPAP) , alarm with low set pressure to minimum possible or excluded , and the maximum apnea time [17,32] (see Table 1) [38]. The patient activates the breath by putting the mouth on the mouthpiece and creating a small negative pressure in the circuit as drinking or inhaling from the mouthpiece (Figure 2). With the "Kiss trigger", ventilation is activated when the patient simply leans the mouth on the mouthpiece; no other effort is required for the patient [36, 38] . Sometimes volume cycling ventilation produces gastric distention; in these cases, one switches to pressure cycling ventilation [32, 33, 37, 38].

Conclusion

Some authors still think that the tracheotomy is the most effective and secure form of continuous ventilatory support. However, we believe there is a better way. There are studies showing that the survival is significantly longer and has fewer complications with NIV [25] compared to a variety of other strategies. As noted above, a direct randomized comparison with tracheotomy is neither feasible or nor ethical. NIV is a safe and acceptable alternative to ventilation by tracheotomy [39]. There is now a widespread consensus that the NIV is preferable to tracheotomy IPPV during the early stages of early ventilatory failure in patients neuromuscular disorders, but there continues to be widespread doubt about its long term effectiveness.

Figure 2: Patient activate the breath putting the mouth in the mouthpiece creating a small negative pressure in the circuit.

The problems of tracheotomy are well- known: dysphagia, difficult with speech, the impossibility of glosso-pharyngeal breathing. Patients suffering from severe neuromuscular diseases, in whom only nocturnal NIV becomes insufficient, should have a trial of non-invasive ventilation with a mouthpiece. We hope that this review will encourage many centers to use this less invasive technique.

References

1. Faroux B, Khirani S. Neuromuscular disease and respiratory physiology in children: putting lung function into perspective. Respirol 2014; 19: 782-791
2. Robert D,Argaud L. Non-invasive positive ventilation in the treatment of sleep-related breathing disorders Sleep Med 2007; 8: 441-452
3. Bach JR, Goncalves MR, Hon A, Ishikawa Y, De Vito EL, Prado F , Dominguez ME .Changing trends in the management of end-stage neuromuscular respiratory muscle failure. Am J Phys Med Rehab 2012; 91: 1 -1
4. Bach JR, Alba AS. Sleep and nocturnal mouthpiece IPPV efficiency in postpoliomyelitis ventilator users . Chest 1994, 106, 1705-1710
5. Bach JR, Alba AS, Saporito LR. Intermittent positive pressure ventilation via the mouth as an alternative to tracheostomy for 257 ventilator users. Chest 1993, 103:174-182
6. Servera E, Sancho J, Zafra MJ et al. Alternatives to endotracheal intubation for patients with neuromuscular diseases. Am J Phys Med Rehabil. 2005; 84: 549-555.
7. Toussaint M, Steens M, G Wasteets, Soudon P. Diurnal ventilation via mouthpiece : survival in end-stage Duchenne patients.Eur Resp J 2006; 28: 549-555
8. Toussaint M, Chatwin M, Soudon P. Mechanical ventilation in Duchenne patients with chronic respiratory insufficiency : clinical implications of 20 years published experience.Chronic Resp Dis. 2007 ; 4: 167-177
9. Bach JR, Rajaraman R, Ballanger F, Tzeng AC, Ishikawa Y, Kulessa R, et al. Neuromuscular ventilator insufficiency: effect of home mechanical ventilator. Use vs oxygen therapy on pneumonia and hospitalization rate. Am J Phys Rehabil. 1998; 77: 8-19
10. Bansal T. Neuromuscular ventilatory insufficiency: effect of home mechanical ventilator use vs oxygen therapy on pneumonia and hospitalization rates.Am J Phys Med Rehabil. 1998 ; 77: 8-19
11. Meduri GU, Turner RE, Abou-Shala N, Wunderink R,Tolley E. Noninvasive positive pressure ventilation via facemask. First line intervention in patients with acute hypercapnicand hypoxemic respiratory failure. Chest 1996; 109: 179–193
12. Gomez-Merino E, Bach JR. Duchenne muscular dystrophy: prolongation of life by noninvasive ventilation and mechanically assisted coughing. Am J Phys Med Rehabil 2002; 81: 411–415.
13. Bach JR, Bianchi C, Aufiero E. Oximetry and indications for tracheotomy in amyotrophic lateral sclerosis. Chest 2004; 126: 1502-1507
14. Nava S, Navalesi P, Gregoretti C. Interfaces and humidification for noninvasive mechanical ventilation.Respir Care 2009; 54: 71-82
15. Anton A, Tarrega J, Giner J, et al: Acute physiologic effects of nasal and full-face masks during noninvasive positive-pressure ventilation in patients with acute exacerbations of chronic obstructive pulmonary disease. Respir Care 2003; 48: 922–925
16. Kwok H, McCormack J, Cece R, et al: Controlled trial of oronasal versus nasal mask ventilation in the treatment of acute respiratory failure. Crit Care Med 2003; 31: 468–473
17. Hess DR Noninvasive ventilation in neuromuscular disease: equipment and application. Respir Care 2006; 51: 896-911
18. Pisani L,Carlucci A,Nava S. Interfaces for noninvasive ventilation: technical aspects and efficiency. Min Anest 2012; 78: 1154-1161
19. Sferrazza Papa GF, Di Marco F, Akoumianaki E ,Brochard L. Recent advances in interfaces for non-invasive ventilation: from bench studies to practical issues. Min Anestesiol 2012; 78:1146-1153
20. Bach JR, Alba AS, Saporito LR. Intermittent positive pressure ventilation via the mouth as an alternative to tracheostomy for 257 ventilators users. Chest 1993; 103: 174–182
21. Bach JR. A comparison of long-term ventilatory support alternatives from the perspective of the patient and caregiver. Chest 1993; 104: 1702–1706
22. Bach JR, O'Brien J, Krotenberg R, Alba AS. Management of end stage respiratory failure in Duchenne muscular dystrophy. Muscle Nerve 1987; 10: 177–182
23. Dean S, Bach JR. The use of noninvasive respiratory muscle aids in the management of patients with progressive neuromuscular diseases. Respir Care Clin N Am 1996; 2: 223–240
24. Simonds AK, Muntoni F, Heather S, Fielding S. Impact of nasal ventilation on survival in hypercapnic Duchenne muscular dystrophy. Thorax 1998; 53: 949–952
25. McKim D, Griller N, LeBlanc C, Woolnough A, King J. Twenty-four hour noninvasive ventilation in Duchenne muscular dystrophy: a safe alternative to tracheostomy. Can Resp J 2013; 20: 5- 9
26. Benditt JO, Boitano L. Pulmonary issue in patients with chronic neuromuscular disease. Am Rev Resp Crit Care Med 2013;187:1046-1055
27. Bach JR, McDermott IG Strapless oral-nasal interface for positive-pressure ventilation. Arch Phys Med Rehabil. 1990; 71: 910-913
28. Ambrosino N,Carpene N,Ghirardi M. Chronic respiratory care for neuromuscular disease in adults. Eur Resp J 2009; 34: 444-451
29. Boitano L. Equipment options for cough augmentation,ventilation,and noninvasive interfaces in neuromuscular respiratory management. Pediatrics 2009;123: 5226-5230
30. Soudon P,Steens M,Toussaint M.A comparison of invasive versus non invasive full-time mechanical ventilation in Duchenne muscular dystrophy. Chron Respir Dis 2008; 5: 87-93
31. Boitano LJ, Benditt JO. An evaluation of home ventilators to support open-circuit mouthpiece ventilation. Respir care 2005; 50:1457-1461
32. Hess DR. The growing role of noninvasive ventilation in patients requiring prolonged mechanical ventilation. Respir Care 2012; 57: 900-918
33. Benditt JO,Boitano LJ. Pulmonary issues in patients with chronic neuromuscular diseases. Am J Respir Crit Care Med 2013; 187; 1046-1055
34. Benditt JO. Full-time noninvasive ventilation :possible and desiderable. Respir Care 2006; 51: 1005-1012
35. Kirby NA, Barnerias MJ, Siebens AA. An evaluation of assisted cough in quadriparetic patients. Arch Phys Med Rehabil 1966; 47: 705-710
36. Khirani S,Ramirez A,Delord V,Leroux K,lofaso F,Eng SH,Toussaint M et al. Evaluation of ventilators for mouthpiece ventilation in neuromuscular disease. Respir care 2014; 59: 1-9
37. Bach JR, Kang SW. Disorders of ventilation: weakness, stiffness and mobilization. Chest 2000; 117: 301-303
38. Garuti G, Nicolini A, Grecchi B, Lusuardi M, Winck JC, Bach JR. Open circuit mouthpiece ventilation: concise clinical review. Rev Port Pneumol 2014; 20: 211-218
39. Heritier Barras AC, Adler D, Ferfoglia RI, Ricou B, Gasche Y, Leuchter I, et al. Is tracheostomy still an option in amyotrohic lateral sclerosis? SMW 2013; 143: 1-9

Phys Med Rehabil Int - Volume 1 Issue 3 - 2014
ISSN : 2471-0377 | www.austinpublishinggroup.com

Citation: Nicolini A, Russo D , Barlascini CO, Sclifò F, Grecchi B, Garuti GC, et al. Mouthpiece Ventilation in Patients with Neuromuscular Disease: A Brief Clinical Review. Phys Med Rehabil Int. 2014;1(3): 4.