Review Article

Mouthpiece Ventilation in Neuromuscular Disease

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Abstract

Mouthpiece non invasive mechanical ventilation is more frequently used in recent years because mouthpiece ventilation modes are being introduced to available portable ventilators. Often, a patient affected by neuromuscular disease needs noninvasive mechanical ventilation. Sometimes the patient does not accept the treatment, due to excessive air leakage, claustrophobia, anxiety and conjunctivitis. The mouthpiece makes less interference with speech, better appearance, less negative impact on the patient, no risk of skin breakdown and absence of claustrophobia. The mpv has been used to treat various neuromuscular diseases such as amyotrophic lateral sclerosis, Duchenne dystrophy, Steinert dystrophy; data in literature documenting that the mouthpiece ventilation resulted an effective, preferable and comfortable alternative.

INTRODUCTION

Ventilation with mouthpiece is used since 1990,[1] and there are already evidence in literature documenting effectiveness of treatment and increased compliance by the patient. Despite this, there is little knowledge and practicality of the use of noninvasive mechanical ventilation (NIV) via a simple mouthpiece [2]. Only in recent years, 2013, mouthpiece ventilation modes are being introduced to commercially available portable ventilators, increasing the interest for this interface.

Two sides of the same coin

In the natural history of many neuromuscular diseases noninvasive mechanical ventilation is required. The choice of a proper interface can play a decisive role in therapy but often complications such as skin lesions (Figure 1), excessive pressure, and claustrophobia interfere with appropriate treatment. Actually mouthpiece ventilation (MPV) interferes less with speaking, coughing and patient’s appearance is improved. The patient can independently disconnect from the mouthpiece to speak, eat, cough or call a family member (Figure 2). It is clear that it present no risk of skin breakdown, conjunctivitis, absence of claustrophobia and lower probability of gastric distension. It is safer by permitting use of glossopharyngeal breathing in the event of sudden ventilator failure or accidental disconnection from the ventilator. 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 neuromuscular disease patients who required continuous ventilatory support [3].

Mouthpieces for daytime use may elicit salivation and more rarely vomiting [4] and long-term use can cause orthodontic deformities after 20 years use [5,6]. Nasal pledges or nose clips can be used to avoid air leak through the nares for patients using lip covering interfaces for mouthpiece NIV during sleep [4-6]. However, although rarely, air may also be swallowed and cause gastric distension. Mouthpiece and nasal NIV are open systems of ventilator support, the low pressure alarms of ventilators not having mouthpiece NIV modes can often be sound. Back pressure from a 15 mm angled mouthpiece is sufficient to prevent a low-pressure alarm set at 2 cmH₂O.

Carlucci et al. recently studied how to set different type of ventilator when using the mouthpiece [4]. They found that an appropriate alarm setting and combination of VT and TI

Figure 1 Skin lesion during mechanical ventilation with nasal mask in patient with amyotrophic lateral sclerosis.
would allow the majority of the tested ventilators to be used for mouthpiece ventilation without alarm activation [4]. The patient triggers the breath by placing the mouth on the mouthpiece and creating a small negative pressure in the circuit by sipping or inhaling.

Failure of the MPV and /or NIV is seen if patients are not cooperative, or in the presence of a severe bulbar dysfunction, unable to cooperate [5-7].

Mouthpieces are very useful in adjunct daytime ventilation for patients suffering from neuromuscular diseases who do not have the ability to maintain acceptable diurnal arterial blood gases without frequent intermittent periods of assistance [7,8]. Khirani et al., report their study of ventilators for mouthpiece ventilation in patients with neuromuscular disease, and confirm the usefulness of this interface. The authors find subjects are satisfied with mouthpiece ventilation [9] Nardi et al, also describes that patients were satisfied with MPV and preferred the mouthpiece to the nasal mask, this aspect can favours adherence to NIV but the risk of the use of MPV is that the patient may unknowingly under ventilate themselves because of the frequent disconnection from mouthpiece [10]. The time of disconnection is probably the major limit of this approach to NIV. The authors documented that the periods of disconnection were associated with > 5 mmHg pCO2 increases and > 2 % spO2 decreases but no clinical complication occurred before or after the monitoring period. Few patients tolerated prolonged disconnections without developing hypercapnia. The most common type of asynchrony was ineffective effort, also suggesting a need for improved trigger sensitivity. Recently introduced MPV software allowing in sufflation to be triggered only by the positioning of the patient’s lips seems to hold pro. For this reason we have seen in recent years growing interest and use of the MPV in patients with neuromuscular disease. Moreover, the software of many new ventilators are adding the mouthpiece mode. During the night while sleeping, most patients use a mask because a mouthpiece requires collaboration and is uncomfortable. However in our experience, ventilation with mouthpiece can promote adherence to night ventilation.

In what diseases?

Sclerosis lateral amyotrophic: There are few data in literature. Bédard and McKim recently studied utilization of daytime mouthpiece ventilation in an ALS population using 24-h NIV. Results confirm the effectiveness of mouthpiece ventilation as well as the importance of preserved bulbar function and ability to generate an adequate peak cough flow with lung-volume recruitment for survival. The authors point out that despite its effectiveness and convenience, mouthpiece ventilation is rarely used in individuals with ALS needing continuous ventilatory support [11] Full-time NIV using different interfaces has been previously reported, including in ALS. Bach et al reported NIV via the mouth in 257 subjects with neuromuscular diseases (5 with ALS), of whom 144 used 24-h NIV (2 with ALS). With adequate bulbar muscle function, mouthpiece ventilation was shown to be an effective alternative to tracheostomy. Mouthpiece ventilation was also shown to be useful in weaning of 155 of 157 “unweanable” patients with neuromuscular diseases [12,13].

Duchenne Distrophy

The use of mouthpiece ventilation in patients with Duchenne dystrophy is documented in literature [14-16]. Some authors examine the impact of diurnal mouthpiece intermittent positive pressure ventilation and conclude that daytime mouthpiece ventilation is safe, prolongs survival and stabilises vital capacity in Duchenne muscular dystrophy patients [14]. McKim et al. argue that twenty-four hours NIV should be considered a safe alternative for patients with DMD because its use may obviate the need for tracheostomy in patients with chronic respiratory failure requiring more than nocturnal ventilation alone [15]. In this case mouthpiece ventilation improves patient and caregiver’s quality of life. [16]. In our opinion, this interface should always be considered for patients with Duchenne muscular dystrophy that have to start NIV. As time passes, patients with DMD develop a constant hypoventilation and need respiratory support 24 h a day; then, mouthpiece can be very valuable, particularly in patients who use the NIV many hours a day and presenting skin
lesions, gastric distension or eye irritation, sometimes alternating nasal and full face masks.

Other Neuromuscular Disease

Neuromuscular diseases, such as polio, have been managed with positive pressure non invasive ventilation. During the polio era, it was well recognized that patients otherwise supported by iron lungs could be liberated from their tanks with positive pressure mouthpieces [17]. Bach and others have reported a large number of patients with neuromuscular diseases managed long beyond the point of respiratory failure with 24 h NIV. Even patients previously ventilated 24 h per day via a tracheostomy have been converted to non invasive mechanical ventilation with MPV [18]. Bach also describes noninvasive acute and long-term management of quadraplegia due to high spinal cord lesions. This includes full-setting, continuous ventilatory support by noninvasive intermittent positive pressure ventilation to support inspiratory muscles and mechanically assisted coughing to support inspiratory and expiratory muscles. The author explains how it can also be used to extubate or decannulate 'unweanable' patients with spinal cord injury, to prevent intercurrent respiratory tract infections from developing into pneumonia and acute respiratory failure [19]. There are limited data on the use of MPV in patient with Steinert dystrophy. In our experience (Figure 3) it can be useful for Steinert patients who previously rejected the application of NIV for tightness, claustrophobia and poor compliance interface. The use of MPV has allowed us to treat patients who had previously refused nasal, oral or oro-nasal interface.

CONCLUSION

The mouthpiece should always be considered for patients with neuromuscular disease that has to start NIV; it is useful to promote a positive approach and rapid acceptance of the new condition.

The mouthpiece ventilation should be considered for patients poorly tolerant to NIV and particularly in patients who use the NIV many hours a day and presenting skin lesion, gastric distension or eye irritation, sometimes alternating with the nasal or full face mask; it is used also in weaning from tracheostomy tube. Mouthpiece ventilation must be always considered as a possible option. The clinician needs to know in deep details the patient’s ventilator to properly set the parameters. The existence of a dedicated mode simplifies the setting but in fact, the interface can be used with most of the ventilators. The use of MPV, alone or combined with other interfaces improves the quality of neuromuscular patient’s life and promotes greater adherence to mechanical ventilation.

REFERENCES