Case Report

Noninvasive ventilation via mouthpiece in a patient with amyotrophic lateral sclerosis

A method to avoid tracheostomy and improve cough efficacy

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Key words:
- Noninvasive ventilation;
- Mouthpiece;
- Cough augmentation;
- Air-stacking;
- Tracheostomy

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ABSTRACT

A 62 year old gentleman, recently diagnosed with amyotrophic lateral sclerosis, was admitted due to acute-on-chronic hypercapnic failure. He was successfully managed with noninvasive bilevel ventilation and was discharged home under the recommendation to continue the use of noninvasive ventilation during sleep. Three months later however, his condition had deteriorated and he was using mechanical ventilation for >18 hours/day. His vital capacity had dropped from 1.3 to 0.7 L and he had a peak cough flow (PCF) of 50 L/min. To defer tracheostomy the patient was started on mouthpiece ventilation in the assist volume control mode, with a tidal volume of 0.9 L, a zero PEEP and a back-up rate of 14/breaths/min. The patient was taught how to utilize mouthpiece ventilation in order to perform the "air-stacking" maneuver, which increased his PCF to 200 L/min. The management of this case suggests that the use of noninvasive respiratory support strategies can obviate tracheostomy and improve cough efficacy in selected patients with neuromuscular diseases.

Pneumon 2017, 30(2):97-101.

1. INTRODUCTION

Pulmonary complications are very common in neuromuscular diseases (NMDs) and a significant cause of morbidity and mortality[1]. The introduction of noninvasive mechanical ventilation at the early stages of symptomatic sleep hypoventilation and the effective use of various cough augmentation methods has been shown to improve mortality and quality of life[1-3]. However due to the progressive character of many of NMDs, ventilatory requirements increase with time and eventually the majority of these patients will become ventilator dependent for a great part of the day. At this point many practitioners would consider transition to invasive ventilatory management via tracheostomy[4-6]. On the other hand, some experts assert

that noninvasive management can still be continued at advanced disease stages without risk for the patient, with a lower rate of complications and with similar or probably even improved mortality as compared to invasive management. The combination of mouthpiece ventilation with cough augmentation techniques such as the "air-stacking" maneuver is a noninvasive strategy that has become very popular the recent years[3,5,6].

We describe here a case of advanced amyotrophic lateral sclerosis (ALS) with rapid respiratory deterioration and we explain how the effective application of the abovementioned noninvasive strategy obviated tracheostomy and improved the patient's cough efficacy.

2. CASE REPORT

A 62 year old gentleman with a recent diagnosis of ALS was admitted to our department due to severe respiratory distress and hypercapnic acidosis. For the past 5 months he had had progressive lower limb weakness and deteriorating dyspnea on exertion. On admission the patient was obviously distressed, unable to lie flat and was using his axillary respiratory muscles to breathe. The revised Amyotrophic Lateral Sclerosis Functional Rating Scale (ALSFRS-R) score[3,5,6] was equal to 28 (out of 48 points) with a bulbar subscore of 12 (out of 12 points). His chest radiograph was unremarkable but his arterial blood gases on room air revealed acute-on-chronic hypercapnic acidosis: pH 7.346, PCO_2 54.5 mmHg, PO_2 57.9 mmHg, HCO_3 29 mmol/L. Spirometry revealed a moderately severe restrictive deficit: FEV_1 1.09 L (40%), FVC 1.29 L (38%) and FEV_1/FVC 84%, however, his FVC decreased by 0.35 L (27%) on the supine position. His maximum inspiratory mouth pressure (MIP) was -47 cm H_2O, his maximum expiratory mouth pressure (MEP) was +51cm H_2O and his sniff nasal inspiratory pressure (SNIP) was -22 cm H_2O. Moreover the patient's peak cough flow (PCF) was also reduced to 160 L/min. The patient was treated with antibiotics and bilevel noninvasive ventilation via an oronasal mask and displayed significant clinical and functional improvement. He was discharged 10 days later on a prescription of a BPAP ST device with an inspiratory positive airway pressure (IPAP) of 16 cm H_2O, an expiratory positive airway pressure (EPAP) of 6 cm H_2O and a back-up rate of 16 breaths/min. The patient was breathing comfortably without the assistance of his axillary inspiratory muscles and was using mechanical ventilation only during sleep. His arterial blood gases on room air had improved to near normal values: pH 7.403, PCO_2 44 mmHg, PO_2 68 mmHg, HCO_3 27 mmol/L. Overnight oximetry under ventilatory support revealed an oxygen desaturation index (ODI) of 5/hour, while the time with a hemoglobin saturation (SaO_2) <90% (T90) was 4% of the total recording time.

On his follow-up assessment 3 months later, the patient displayed however further deterioration. His lower limb weakness had progressed and he was confined on a wheelchair, but he retained significant functionality of both upper arms and he was able to speak and swallow without problems. His ALSFRS-R score had decreased to 24 points, but his bulbar subscore had not changed. Importantly, although his blood gases did not show any change (pH 7.370, PCO_2 41 mmHg, PO_2 65 mmHg, HCO_3 23 mmol/L), the patient was using his BPAP device for >18 hours per day. His FVC was reduced by 0.58 L (45%) to a value of 0.71 L, his SNIP had decreased to -16 cm H_2O and his new PCF value was 50 L/min. The patient was severely breathless and was using again his axillary muscles during his off-ventilator time; he also persistently complained of difficulty in bringing up sputum. Moreover, he had developed a pressure ulcer on his nose as a result of the prolonged use of the oronasal mask.

Although tracheostomy is thought to be the standard management option for neuromuscular patients with chronic respiratory failure and almost full-time ventilator dependence, the patient was amenable to consider an alternative, noninvasive management strategy. The oronasal interface was substituted with an angled 15 mm mouthpiece and the noninvasive respiratory support was continued throughout the day with the ventilator set at the assist volume control (AVC) mode to deliver a tidal volume of 0.9 L with an inspiratory time of 1.3 seconds and a square flow waveform, a zero PEEP, and a back-up rate of 14 breaths/min. The patient was able to bring the mouthpiece to his mouth with his hands and manage his ventilation according to his own needs by controlling the leak and choosing the number of breaths he required. Some "obtrusive" alarms, including the "low pressure", the "apnea response" alarm were deactivated. The patient learned relatively easily to use the mouthpiece and experienced a significant improvement in his symptoms (Figure 1).

In addition, the patient was taught to perform the "air-stacking" maneuver. This maneuver is performed by asking the patient to take a series of consecutive breaths from a volume ventilator (or a manual resuscitator), without exhaling in the intervening period, in order to inflate his lungs as much as possible. The maximum inspiratory volume that the patient can hold in his lungs with his

FIGURE 1. Use of noninvasive mouthpiece ventilation by a patient with advanced amyotrophic lateral sclerosis.

glottis closed using this maneuver is called maximum insufflation capacity (MIC)[8]. By stacking air from his ventilator, our patient achieved a MIC of 1.12 L, while his PCF improved to 200 L/min, when the patient coughed from MIC. Bilevel ventilation via an alternative oronasal interface was continued for nocturnal support in the BPAP ST mode with IPAP increased to 18 cm H_2O; for night use all relevant alarms remained active. Oximetry was used to assess the safety and efficacy of both nocturnal and diurnal settings and arterial blood gas sampling also verified sufficient gas exchange function during diurnal mouthpiece ventilation (e.g. pH 7.424, PCO_2 33 mmHg, PO_2 85 mmHg, HCO_3 21 mmol/L).

3. DISCUSSION

ALS is a devastating neurodegenerative disease causing death usually within 3 years, most commonly due to respiratory failure[9]. We report here a case of a non-bulbar ALS patient who displayed a very rapid decline of motor and respiratory function (>0.5 L drop in VC within the 3-month follow-up period) resulting in complete loss of ambulation, but most importantly almost full-time ventilator dependence and weak cough. The combination of mouthpiece ventilation with the technique of air-stacking allowed the patient to avoid tracheostomy and to make use of full-time noninvasive ventilatory support.

Alveolar hypoventilation is a major pathophysiologic trait of many NMDs, appearing initially during sleep and extending subsequently into the daytime[1]. Nocturnal noninvasive positive pressure ventilation is the standard mode of initial management of alveolar hypoventilation in NMDs[2], however as respiratory muscles weakness progresses, the ventilator-free breathing time is reduced significantly. When the number of hours of ventilator use per day exceed an arbitrarily defined threshold (e.g. >16 or 20 hours), many practitioners would consider transition to invasive ventilatory support via tracheostomy[4]. At this point, tracheostomy also serves an additional role by facilitating secretion clearance, given that in advanced disease stages, cough flows are invariably severely reduced[1]. Generally, a PCF <160 L/min is associated with impaired ability to clear secretions and remove debris from the airways and a PCF <270 L/min identifies a patient at increased risk of developing respiratory failure during a trivial respiratory tract infection[1,6]. Nevertheless, there are many disadvantages in long-term tracheostomy ventilation including tube related complications, loss of voice and disturbed self-image[6]. On the other hand, while many patients would prefer to continue noninvasive ventilatory support on a 24-hour basis[10], conventional nasal or oronasal masks are not suitable for this option because prolonged use is associated with difficulties in eating, drinking and talking, claustrophobia, limited field of vision, impaired social interaction and pressure lesions[6].

An alternative method for providing full-time noninvasive ventilatory support is based on the use of a mouthpiece interface for the daytime combined with mask ventilation during sleep. This strategy has been advocated as early as 1993[11], but lately it has witnessed increasing popularity for the management of advanced NMDs such as Duchenne's muscular dystrophy (DMD) and ALS[5,10,12,13]. To apply this method, the patient should be able to rotate his head and grab the mouthpiece with his lips, therefore it might not be feasible for patients with facial muscle weakness or advanced bulbar symptoms[13]. Specially designed support arms can be used to mount the mouthpiece close to the mouth for patients with motor disabilities. Mouthpiece ventilation is provided usually in the AVC mode with a tidal volume between 0.7 and 1.5 L, a zero PEEP, a back-up respiratory rate set

to the minimum allowed, while the inspiratory time titrated to patient comfort[6,14]. The patient has the ability to define his ventilatory pattern by triggering the ventilator as many times as required and by modifying the quantity of leak[12]. A number of back-up breaths can be set for patients who are too frail to trigger the ventilator, however in some modern ventilators triggering can be simply effected by creating a small negative pressure at the mouthpiece ("kiss-trigger")[15]. The angled configuration of the mouthpiece creates a "back-pressure" that "tricks" the "low-pressure" alarm of the ventilator. As general rule the "low pressure" and the "apnea response" alarm should be deactivated[6,14].

Mouthpiece ventilation is ideal for the application of the air-stacking maneuver. In fact, the application of air-stacking maneuver to improve cough flows and minimize atelectasis was popularized by Kang and Bach and has been characterized as one of "the most important complementary interventions that permits the long term use of noninvasive ventilation"[16]. The maneuver can be performed with a volume ventilator, a resuscitation bag or the technique of glossopharyngeal breathing. Air-stacking requires an adequate glottic closure and is based on achieving maximum lung insufflation by delivering consecutive volumes of air[8,16]. The higher the difference between MIC and VC, the higher the possibility that the patient can be managed successfully with noninvasive strategies[16]. In a cohort of 61 DMD patients, mean PCF increased from 138 to 236 L/min with the application of air-stacking maneuver[17]. The effectiveness of the air-stacking maneuver was also obvious in the case of our patient: by stacking air, he achieved MIC-VC difference of 0.4 L and his PCF improved from 50 to 200 L/min.

Based on data from retrospective studies, the non-invasive management of DMD patients with strategies that include mouthpiece ventilation and air stacking was associated with improved mortality[3], whereas tracheostomy ventilation was associated with a higher rate of complications, mainly tracheal injury[5]. On the other hand, there is limited information on the use of mouthpiece ventilation in patients with ALS. Bach suggested that ALS patients who can air-stack to volumes higher than their VC and can produce a PCF >180 L/min are most likely to avoid tracheostomy for a longer time[18]. In a recent study, Bedard and Mckim reported retrospectively on 39 ALS patients who were treated with mouthpiece ventilation over a 17 year period. A low bulbar ALSFRS-R subscore (<6) was a poor predictor of success; among mouthpiece users those who could achieve a PCF >180 L/min with air-stacking had a better survival as compared to those with a PCF <180 L/min (637 vs. 240 days)[13]. In a another study involving 30 mouthpiece ventilation users, the majority of the subjects reported lesser dyspnea (73%) and fatigue (97%) and, to a lesser extent, improvements in speech and eating[15]. Nevertheless, problems with mouthpiece ventilation have also been reported and include increased sialorrhoea, orthodontic deformities, and nasal leaks[14]. Moreover, alarm and setting customization is still problematic in many home ven tilators even when specifically designed software for mouthpiece ventilation is available[15]. On the other hand, for patients with advanced bulbar involvement, air-stacking is hardly effective[13]. Strategies for cough augmentation in these cases include the use of the mechanical insufflator-exsufflator or the delivery of inspiratory volumes as high as the patient's reference inspiratory capacity[8].

4. CONCLUSION

Mouthpiece noninvasive ventilation is an attractive strategy to deliver 24 hour noninvasive respiratory support to patients with advanced NMDs who retain the ability to grab the mouthpiece with their lips and can perform air-stacking maneuvers. This technique remains largely underutilized despite its many obvious advantages. More studies are required to explore the potential of mouthpiece ventilation in the vast field of respiratory care.

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