Open-circuit ventilation with nasal pillows for daytime ventilatory support in a patient with advanced bulbar amyotrophic lateral sclerosis: A case report

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ABSTRACT

Full-time non-invasive ventilation (NIV) is an essential treatment for patients with advanced neuromuscular diseases. This case report discusses a patient aged 77 years with bulbar amyotrophic lateral sclerosis (ALS) who transitioned to daytime NIV due to daytime respiratory failure and interface-related discomfort. Conventional open-circuit, mouthpiece ventilation was unsuitable due to advanced bulbar dysfunction. An alternative approach involved a nasal pillows interface with an open-circuit setup, utilizing the assisted pressure-controlled mode (22 cm H_2O, 15 breaths/min, inspiratory time 1.5 s, 0 cm H_2O PEEP). This intervention mitigated the patient's dyspnea, sustaining SpO_2 levels between 94% and 95% without supplemental oxygen, facilitated activities such as reading and social interaction, and was well tolerated. Although a family member contacted by phone confirmed that the patient was comfortable with using this method of daytime ventilatory assistance at home, follow-up was limited as the patient succumbed to a respiratory infection a month later. This report highlights the feasibility of nasal pillows as an alternative interface for daytime NIV in neuromuscular patients, particularly when other options are unfeasible. While not originally designed for high-demand ventilatory assistance, the nasal pillow interface was effective in supporting the patient's daytime ventilatory needs with good tolerance. This approach underscores the importance of personalized solutions for the management of the complex respiratory needs of patients with ALS.

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INTRODUCTION

Full-time non-invasive ventilation (NIV) is increasingly being used for the management of advanced-stage neuromuscular disease[1-3]. Several new interfaces are now available in the market and can improve NIV compliance and reduce shortcomings associated with long-term use, including skin lesions and claustrophobia[1,2]. Open-circuit mouthpiece ventilation (MPV) is becoming increasingly popular for the management of daytime ventilatory dependence. However, individualization of care with respect to interface, ventilatory settings, and support arm selection is essential for successful patient management. Furthermore, patients with weak mouth muscles or advanced bulbar disease may not be able to use MPV[1-5]. In this following case, nasal pillows, traditionally used for respiratory sleep disorders, were employed for daytime ventilatory support in a patient with advanced bulbar amyotrophic lateral sclerosis (ALS)[2].

CASE PRESENTATION

We report the case of a man aged 77 years with bulbar (ALS), diagnosed two years ago, who had been using nocturnal NIV via a vented oronasal mask in the Intelligent Volume-Assured Pressure-Support mode (iVAPS; Astral 100, ResMed, San Diego, California, USA) for the previous year, targeting a tidal volume of 10 mL/kg. His previous history was significant for hypertension and hyperlipidemia and he quit smoking 12 years ago. The patient complained of deteriorating shortness of breath during the previous 2 months and he had extended his ventilator use into daytime for an average of 15 h/day. Over this period, his diurnal PCO_2 increased from 40 to 44 mmHg, his vital capacity dropped from 0.7 L (20% predicted, 4 months ago) to a non-measurable value, and his sniff inspiratory pressure (SNIP) was reduced from -22 to -5 cm H_2O. His ALFRS-R score was 25, with a bulbar sub-score of 6 (speech: 0, salivation: 4, swallowing: 2). The patient had retained adequate upper and lower limb mobility, but prolonged use of his oronasal mask resulted in increasing discomfort and prevented him from putting on his glasses to read the newspaper and watch TV; his nose bridge showed a reddening although no pressure sores were

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Figure 1. Assembly for the provision of daytime ventilation by combining nasal pillows and open-circuit setup in the Astral 100 ventilator

ID: internal diameter.

noted. Based on the above findings (interface complications, daytime ventilatory dependence, increased diurnal PCO_2, severely impaired respiratory function tests)[4,6] full-time ventilatory support was considered, and, given his advanced bulbar disease bulbar disease, tracheostomy was extensively discussed as an option. However, the patient adamantly denied this option, pointing to the invasive nature of the procedure, distortion of his self-image, and increased burden of care for his family. As an alternative solution daytime non-invasive ventilatory support was considered and a trial of MPV was discussed. The patient could close his mouth; however, because of his advancing bulbar disease we were concerned about his ability to keep the mouthpiece in place safely. Therefore, we opted to use an opencircuit setup with a single-limb circuit, an active exhalation valve, and a nasal pillows interface (AirFit P10, ResMed, San Diego, California, USA) which was distally attached to the exhalation valve via a 22 mm adapter (Figure 1). Daytime ventilation was delivered in the assisted pressure-controlled mode (aPC), with a pressure level of 22 cm H_2O, a backup respiratory rate (RR) of 15 breaths/min, an inspiratory time of 1.5 s, and a PEEP of 0 cm H_2O. To mitigate the patient's breathlessness without causing intolerance, the inspiratory pressure level was adjusted gradually. Subsequently, the patient's breathlessness was palliated and he consistently maintained a SpO_2 of 94–95%, while using the ventilator without supplementary oxygen. The patient was comfortable with this combination and, despite the high inspiratory pressure level, did not complain of pressure intolerance, significant leaks, nasal irritation, or dry mouth. His nighttime ventilatory settings were not changed. The family was contacted by phone one week following hospital discharge and they confirmed that he continued using the nasal pillows in the open-circuit setup during daytime, reporting no problems, but unfortunately, he succumbed to a respiratory infection a month later. Ventilator usage data were not available.

DISCUSSION

In our case, the open-circuit ventilatory setup was coupled with the nasal pillows, an interface primarily designed for the management of respiratory sleep disorders, especially obstructive sleep apnea. The minimalistic design of the pillows allowed the patient to read, watch TV, and socialize. Although nasal pillows can be considered for the provision of daytime ventilation in advanced neuromuscular disease, high inspiratory pressures (>15 cm H_2O) are likely to induce significant leaks and impair adhesion[2]. When open-circuit ventilation is implemented, volume-preset is favored over

pressure-preset modes, because only the former permits air-stacking, while in the latter leaks may induce acute pressure drops and ventilation overshoot, which is usually not well tolerated[4,6]. Although we used a pressure-preset mode, we believe that the narrow diameter and tapered configuration of the pillows may have helped to maintain pressure within the circuit and mitigate this effect, which could likely explain why the high inspiratory pressure (22 cm H_2O) was well tolerated. According to standard practice in MPV implementation, PEEP was set to zero[4]. Additionally, given that the interface was always in place, a positive end-expiratory pressure might have caused discomfort during exhalation. Notably, the nasal pillows are a vented interface and thus, when used together with the leak-tolerant, open-circuit setup, it must be assumed that the leakage from the vent consists of a part of the random total system leakage[4]. This feature along with the flexibility in alarm adjustment in MPV modes makes the nasal pillows an attractive alternative for patients who cannot achieve a proper seal with the mouthpiece.

Initially pioneered by John Affeldt for poliomyelitis survivors[7], open-circuit MPV fell into obscurity as a result of the widespread use of tracheostomy ventilation but was subsequently revived by Bach and other investigators[1,2,8-14]. The growing interest in non-invasive respiratory support for chronically ventilator-dependent patients has led many manufacturers to incorporate MPV modes into their home ventilators[11] and expert guidance on MPV implementation has recently been published[4,6]. MPV is also increasingly being used in advanced-stage ALS patients. Bedard and McKim[8] reported retrospectively on MPV implementation on 39 non-bulbar ALS patients and showed improved survival for those who attained a peak cough flow >180 L/min using lung volume recruitment maneuvers. More recently Nicolini et al.[9] demonstrated equal survival rates for MPV and tracheostomy ventilation in patients with ALS; however, quality of life was better for MPV users[9]. To our knowledge, no study has reported the use of nasal pillows coupled with the open-circuit ventilatory setup. In a study of spinal ALS patients, reported in abstract form, daytime ventilation was provided with nasal pillows, while the ventilator was set to the standard aPC mode and PEEP to the minimally allowed value[15].

Open-circuit ventilation is commonly delivered with the aid of a mouthpiece in the assisted volume-controlled mode to permit air-stacking. Air-stacking requires adequate glottic function and consists of delivering consecutive lung volumes until the patient attains the maximum inspiratory volume air that he/she can hold with a closed glottis (maximum insufflation capacity). This maneuver facilitates lung volume recruitment and augments cough efficacy by allowing the patient to cough from a higher end-inspiratory volume, after disconnecting from the mouthpiece[5]. Although, theoretically a patient will also be able to air-stack while using the nasal pillows, it is possible that he/she may not be able to cough efficiently with the interface in place, if at the same time, the ventilator delivers a programmed breath. However, this problem can be circumvented by having a caregiver disconnect temporarily the pillows to allow the patient to cough or by setting the ventilator back-up RR to zero, if tolerated. Additionally, our patient could not air-stack because of advanced bulbar disease, and therefore the use of nasal pillows coupled with the open-circuit setup served solely to palliate his breathlessness and provide daytime non-invasive ventilatory support. Therefore, given that our patient was comfortable with the aPC mode, we opted to maintain him in this setting.

Strengths and limitations

A main strength of this report is that it details the specifics of combining the open circuit setup with the nasal pillow interface. Importantly, the patient showed good adjustment to the new mode and the high inspiratory pressure, despite the presence of advanced bulbar disease. Last, our case indicates that, although the nasal pillows were not originally designed to sustain high ventilatory requirements, daytime ventilation with nasal pillows is a feasible option for ventilator-dependent neuromuscular patients who cannot achieve a good mouthpiece seal or for those who cannot tolerate other interfaces. The main limitation of this report is the short follow-up interval of the patient, including limited follow-up data. However, with these settings the patient achieved a SpO_2 of 94–95% while using the ventilator without supplemental oxygen, which is an acceptable titration end-point[16]. Blood gases were not drawn as our main focus was symptom palliation, rather than optimization of ventilatory settings. Despite lacking ventilator data, consistent ventilator usage was verified by reliable and dedicated caregivers.

CONCLUSION

In this report, we explore the practicality of nasal pillows as a daytime NIV interface for neuromuscular patients in situations where other methods are infeasible. Despite not being designed for high-volume ventilation, the nasal pillows proved effective and comfortable in supporting the patient's daytime ventilatory needs. It should be emphasized that, because MPV is not commonly practiced in Greece[17], mouthpiece support arms are generally not available in the market, a fact necessitating the use of makeshift arms and rendering the implementation of MPV even more problematic. From this point of view, the combination of nasal pillows with the open-circuit setup could be a viable alternative. Physician improvisation should be encouraged for the management of the complex problems of these patients.

CONFLICTS OF INTEREST

The authors have completed and submitted the ICMJE Form for disclosure of Potential Conflicts of Interest and none was reported.

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DATA AVAILABILITY

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