Clinical Case Report

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Application of different ventilator modes in patients with amyotrophic lateral sclerosis according to certain clinical situations

A Case Report

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Abstract

Rationale: Amyotrophic lateral sclerosis (ALS) is a rapidly progressing neurodegenerative disease that involves limb, axial, bulbar, and respiratory muscles. Fatigue and weakness of the respiratory muscles eventually induce respiratory insufficiency, which is one of the main causes of death in patients with ALS. In ALS patients with respiratory insufficiency, application of a ventilator is indispensable. Although there are various modes of ventilation, these modes are classified roughly into volume-controlled ventilation (VCV) and pressure-controlled ventilation (PCV). There have been several reports that VCV is preferable to PCV in neuromuscular disorder patients, such as ALS patients, but there is still debate on which ventilator mode is better.

Patient concerns: Respiratory difficulty despite ventilator application.

Diagnosis: Three ALS patients with respiratory difficulty.

Intervention: Changing ventilator mode to improve symptoms of respiratory difficulty.

Outcomes: Considering case 1 shows that the VCV mode may have an advantage in managing respiratory insufficiency of patients in situations where the inner diameter of the airway decreases because of increased sputum. In contrast, cases 2 and 3, it is shown that changing to the PCV mode may be one of the treatment options if not enough tidal volume can be supplied to resolve respiratory insufficiency because of an increase in leakage volume.

Lessons: Therefore, in this study, through considering several cases of ALS patients whose clinical symptoms were improved by changing ventilation mode, we tried to investigate the adequacy of each ventilation mode under certain clinical situations.

Abbreviations: AC = Assist control, ALS = amyotrophic lateral sclerosis, ALSFRS-R = revised amyotrophic lateral sclerosis functional rating scale, EKG = electrocardiography, EPAP = expiratory positive airway pressure, IPAP = inspiratory positive airway pressure, NIPPV = non-invasive positive pressure ventilation, PCV = pressure-controlled ventilation, PEEP = positive end-expiratory pressure, PEG = percutaneous endoscopic gastrostomy, PIP = peak inspiratory pressure, S/T = Spontaneous/Timed, TPPV = tracheostomy positive pressure ventilation, VCV = volume-controlled ventilation, VTe = expired tidal volume.

Keywords: amyotrophic lateral sclerosis, pressure-controlled ventilation, respiratory insufficiency, volume-controlled ventilation

1. Introduction

Amyotrophic lateral sclerosis (ALS) is a rapidly progressing neurodegenerative disease that involves limb, axial, bulbar, and respiratory muscles.[1] Fatigue and weakness of the respiratory muscles eventually induce respiratory insufficiency, which is one of the main causes of death in patients with ALS.[1] In ALS patients with respiratory insufficiency, application of a ventilator is indispensable. Among them, non-invasive positive pressure ventilation (NIPPV) is known to increase life span and quality of life in patients with ALS. However, because of bulbar palsy, tracheostomy positive pressure ventilation (TPPV) is also widely used in clinical practice.[2]

Although there are various modes of ventilation, these modes are classified roughly into volume-controlled ventilation (VCV) and pressure-controlled ventilation (PCV). There have been several reports that VCV is preferable to PCV in neuromuscular disorder patients, such as ALS patients,[2,3] but there is still debate on which ventilator mode is better. In actual clinical practice, the ventilation mode often tends to be determined according to the patient's disease, the clinical situation, or the physician's familiarity with a particular ventilation mode. The absence of the principle of ventilator mode adjustment in rare diseases, such as ALS, may cause difficulties for physicians who do not have much clinical experience of managing mechanical ventilators.

Therefore, in this study, through considering several cases of ALS patients whose clinical symptoms were improved by changing ventilation mode, we tried to investigate the adequacy of each ventilation mode under certain clinical situations. The

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family of patient was informed that data concerning the case would be submitted for publication, and they provided consent. This case series was approved by ethics committee of our hospital (Institutional Review Board of Daegu Fatima Hospital).

2. Case 1

A 64-year-old male limb-onset ALS patient was admitted to the department of pulmonary medicine with respiratory insufficiency. At admission, his revised amyotrophic lateral sclerosis functional rating scale (ALSFRS-R) was 24 of 48, [4] and he was being fed via a percutaneous endoscopic gastrostomy (PEG) tube. After admission to the respiratory medicine department, his respiratory insufficiency was improved after applying TPPV with PCV mode (Spontaneous/Timed [S/T] mode, inspiratory positive airway pressure [IPAP] 13 cmH_2O, expiratory positive airway pressure [EPAP] 5 cmH_2O inspiratory time 1.2 s, frequency 12 times/minute) via a home ventilator Triology 100. After 2 weeks of treatment, he was transferred to the department of rehabilitation for pulmonary rehabilitation. Several days after being transferred to the department of rehabilitation, he began to complain of discomfort in breathing, with mildly increased saliva and sputum. There was no significant interval change in electrocardiography (EKG), and the serum level of creatine phosphokinase-MB (CPK-MB) and Troponin I was also within normal range. In transcutaneous CO_2 monitoring, the transcutaneous CO_2 level and the oxygen saturation were slightly changed from 42 to 45 mmHg (normal range; 35–45 mmHg), from 98% to 93% (normal range; 94%–100%), but no significant abnormalities were observed elsewhere. During application of ventilation with PCV mode, expired tidal volume (VTe) showed a finding that decreased to about 400 mL of 290 mL. Then, the cough assist machine was applied 3 times daily, but his discomfort in breathing did not improve. Two days after the onset of symptoms, the ventilation mode was changed to VCV mode (assist control [AC] mode, tidal volume 400 mL, positive end-expiratory pressure [PEEP] 5 cmH_2O, inspiratory time 1.2 s, frequency 12 times/minute). After changing the ventilator mode to VCV, a constant tidal volume was maintained regardless of peak inspiratory pressure (PIP), and his discomfort, O_2 saturation, and transcutaneous CO_2 level improved slightly was significantly improved (transcutaneous CO_2 level 40–41 mmHg; O_2 saturation 98%–99%).

3. Case 2

Case 2 is a 54-year-old male bulbar-onset ALS patient. For 6 months, he had been undergoing NIPPV via a home ventilator Triology 100 (Philips Respironics, Murrysville, PA) with active circuit. Several days before the hospital admission, he started to complain of chest discomfort, so he was admitted to our rehabilitation department for ventilation adjustment. His ALSFRS-R was 36 of 48, and his ventilation mode was VCV mode (AC mode, tidal volume 600 mL, PEEP 0 cmH_2O, inspiratory time 1.2 s, frequency 12 times/minute).

In the neurological examination at the time of admission, the speech subscore of the ALSFRS-R scale decreased from 3 to 2, indicating that bulbar palsy had progressed more than before. In addition, the transcutaneous CO_2 level was increased from 43 to 50 mmHg, and the PIP was reduced from 10 to 7 cmH_2O. However, sputum was not increased and there was no significant interval change in EKG, and the serum level of CPK-MB and Troponin I was also within normal range. The inspiratory tidal volume was increased to improve the patient's clinical symptoms while maintaining VCV mode. After increasing the tidal volume to 800 mL, his discomfort and transcutaneous CO_2 level improved slightly (44 mmHg); however, he continued to complain of intermittent chest discomfort. One week after admission, the patient's ventilation mode was changed to PCV (S/T mode, IPAP 10 cmH_2O, EPAP 2 cmH_2O, inspiratory time 1.2 s, frequency 12 times/minute) to maintain a constant PIP. Even after changing the ventilator mode, the leakage increased significantly between the facial mask and the face because of the bulbar palsy, but the PIP remained constant and his clinical symptoms improved significantly. He refused TPPV, and was discharged home a few days later.

4. Case 3

Case 3 is an 83-year-old male limb-onset ALS patient. He had been undergoing TPPV via a home ventilator Triology 100 (Philips Respironics, Murrysville, PA) with a passive circuit for 1 month. His ALSFRS-R was 21 of 48.[4] His ventilation mode was initially VCV mode (AC mode, tidal volume 360 mL, PEEP 4 cmH_2O, inspiratory time 1.5 s, frequency 14 times/minute). During application of the above ventilator mode, PIP was well maintained at 13~14 cmH_2O, and both pCO_2 and SaO_2 were well maintained. Then, he began to complain about difficulty in breathing despite the constant application of the ventilator. Interestingly, his discomfort in breathing was worse in the supine position rather than in the lateral decubitus position. Thus, he could not keep lying in supine position for a long time and spent more time lying in the lateral decubitus position. He did not complain that sputum quantity had increased, and there was no evidence of pneumonia on chest radiography and blood tests. In transcutaneous CO_2 monitoring (Sentec AG, Therwil, Switzerland), the transcutaneous CO_2 level and the oxygen saturation were slightly changed from 37 to 41 mmHg, from 98% to 96%, but no significant abnormalities were observed except for increase in respiratory rate (RR; from 13~14 to 24~26 breaths/min) (Fig. 1A and B).

In the supine position, VTe and PIP were decreased compared to the lateral decubitus position (Fig. 1A and B). In addition, he also complained that he had a lot of air leakage through his mouth in his supine position when the ventilator supplied air. It is thought that the leakage volume increased between the tracheal tube and the trachea in the supine position, so the balloon of the tracheal tube was inflated more. After inflating the balloon of the tracheal tube, he showed a slight improvement in his breathing difficulty, but still complained of difficulty in breathing when in the supine position. Then, the patient's ventilation mode was changed to PCV (S/T mode, IPAP 13 cmH_2O, EPAP 5 cmH_2O, inspiratory time 1.2 s, frequency 14 times/minute) to maintain a constant PIP. After applying PCV mode, he was able to maintain a constant PIP regardless of the position he lay in (Fig. 1C and D), and his uncomfortable symptoms and RR were significantly improved (14~16 breaths/min) (Fig. 1C and D). In addition, transcutaneous CO_2 level and O_2 saturation also improved slightly (transcutaneous CO_2 level 36 mmHg; O_2 saturation 99%).

5. Discussion

As mentioned before, there is still debate on which ventilator mode is better, and the ventilation mode often tends to be determined according to the patient's disease, the clinical situation, or the physician's familiarity with particular ventilation mode in clinical practice. Moreover, for ALS patients, unlike

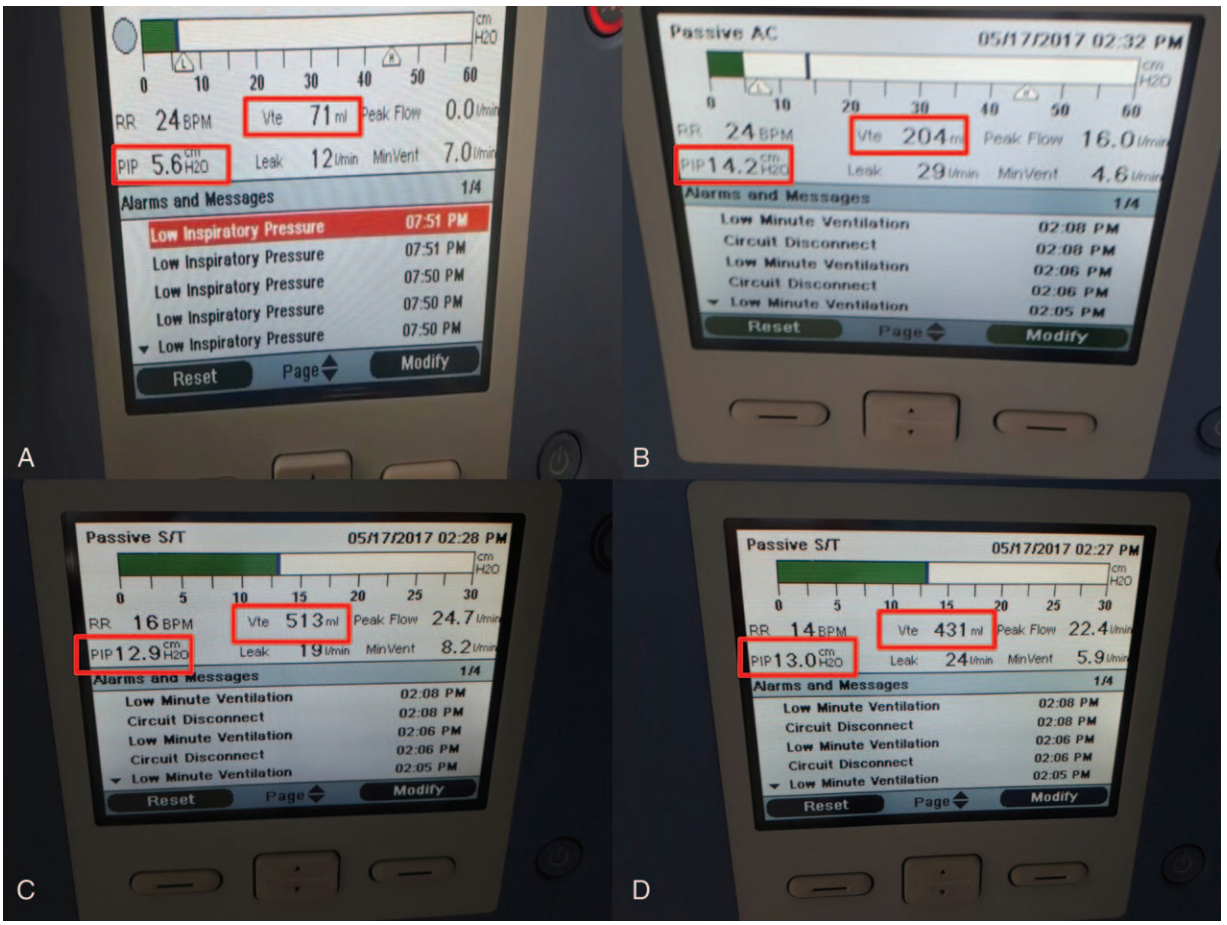

Figure 1. Ventilator monitor in the case 1 patient with amyotrophic lateral sclerosis. (A) Ventilator monitor when the volume-controlled ventilation (VCV) mode was applied in the supine position. (B) Ventilator monitor when the VCV mode was applied in the decubitus position. (C) Ventilator monitor when the pressure-controlled ventilation (PCV) mode was applied in the supine position. (D) Ventilator monitor when the PCV mode was applied in the decubitus position.

acute respiratory distress syndrome or chronic obstructive pulmonary disease, there are few studies on what type of respiratory mode is better for patient's respiratory insufficiency improvement. Therefore, through case series of ALS patients whose condition had been improved by changing the ventilation mode, we tried to investigate the adequacy of each ventilation mode under certain clinical situations.

In case 1, the patient's respiratory insufficiency was controlled well by the PCV mode at first. However, as the amount of sputum increased, he started to complain of discomfort in breathing. Then, the patient's discomfort in breathing improved after changing from the PCV mode to the VCV mode. The reason the patient began to complain about the PCV mode, which was previously well controlled, is thought to have been that the inner diameter of the airway narrowed because of increased sputum. Thus, patient discomfort may have occurred because of the reduced internal diameter of the airway, resulting in the PIP reaching the target pressure (IPAP) with less tidal volume than before. It is thought that the patient's discomfort improved after changing to the VCV mode because it supplies a constant tidal volume to the lung, regardless of PIP.

In case 2, the patient was previously well controlled in the VCV mode, but as the bulbar palsy progressed, the amount of leakage between the face mask and the face increased and the patient began to complain of discomfort in breathing. With increased leakage owing to the progressive bulbar palsy, it seems that the tidal volume, which was supplied in the previous VCV mode and which was effective in resolving the patient's respiratory insufficiency, could not maintain an adequate PIP to resolve this patient's respiratory insufficiency. Also in case 1, it seems that the greater leakage volume between the tracheal tube and the trachea in the supine position compared to the lateral decubitus position seems to have caused the patient's discomfort in breathing. Thus, it is thought that adequate PIP could not be maintained in the supine posture for tidal volume, which was well controlled in the lateral decubitus position. Therefore, in case 3, after changing to the PCV mode to maintain a constant PIP regardless of posture, the patient's discomfort in breathing seemed to be improved. Considering cases 2 and 3, it is shown that changing to the PCV mode may be one of the treatment options if not enough tidal volume can be supplied to resolve respiratory insufficiency because of an increase in leakage volume. In contrast, case 1 shows that the VCV mode may have an advantage in managing respiratory insufficiency of patients in situations wherein the inner diameter of the airway decreases because of increased sputum.

Of course, it may be unreasonable to generalize only with these cases; however, considering the fact that ALS is a rare disease, this case series seems to be sufficiently helpful for physicians who are not familiar with ventilator application to determine the ventilation mode.

Moreover, in the future, it may be more useful to investigate the adequacy of the ventilation mode in certain clinical situations,

rather than simply investigating the adequacy of the ventilation mode according to the disease.

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